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UNITED STATES
SECURITIES AND EXCHANGE COMMISSION

Washington, D.C. 20549

FORM 10-Q

(Mark One)

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

For the quarterly period ended June 30, 2023
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 July 28, 2023, there were 70,167,898 common shares outstanding in aggregate, comprised of:
35,238,254 Class A common shares, par value $0.000273235 per share
1,795,158 Class B common shares, par value $0.000273235 per share
17,075,868 Class A1 common shares, par value $0.000273235 per share
16,057,618 Class B1 common shares, par value $0.000273235 per share

Kiniksa Pharmaceuticals, Ltd.

FORM 10-Q

FOR THE THREE MONTHS ENDED JUNE 30, 2023

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SPECIAL NOTE REGARDING FORWARD-LOOKING STATEMENTS

This Quarterly Report on Form 10-Q (this "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, clinical and 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 "Summary Risk Factors," "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 a small commercial stage biopharmaceutical company and our expectation to incur losses for the foreseeable future;
- our future capital needs and our need to raise additional funds;
- our ability to manufacture sufficient quantities of our products and product candidates to meet patient and partner demand;
- our ability to successfully complete the technology transfer of the manufacturing process for ARCALYST drug substance;
- 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;
- the decision by any applicable regulatory authority to clear our current or future product candidates for clinical development and, ultimately, 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, out-licensing activities, collaborations or other strategic transactions and our ability to realize value therefrom;
- 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;
- 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 began generating product revenue in 2021, have incurred operating losses in the past, expect to incur operating losses for the foreseeable future and may never achieve corporate profitability on a sustained basis;
- we depend heavily on the commercial success of ARCALYST and may be unsuccessful in our efforts to commercialize ARCALYST on a sustained basis, support our sales, marketing, and distribution activities and maintain applicable infrastructure for these activities either directly and/or through agreements with third parties;
- our success depends heavily on the sustained commercial execution of ARCALYST and the future success of one or more of our product candidates, which are in various stages of development; for ARCALYST, our success is dependent on growing and sustaining market acceptance by prescribers, patients and payors; for our product candidates, for our product candidates in clinical development, our success is dependent upon us obtaining regulatory approval and ultimately commercializing one or more of our product candidates on a timely basis;
- 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 market opportunities for our products and product candidates, if approved, may be smaller than we estimate, or any approval that we obtain may be based on a narrower definition of our targeted patient population, either of which may materially adversely affect our revenue and ability to achieve profitability;
- we may 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; 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;
- 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 studies and/or clinical trials, including as a result of delays in obtaining regulatory approvals to conduct clinical trials, activating sites, enrolling participants, and conducting trials, which could delay or prevent our product development activities;
- we rely on third parties, including contract research organizations (“CROs”) to activate our clinical trial sites and 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 development and manufacturing organizations (“CDMOs”) 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

ARCALYST and our product candidates; and if these third parties do not have sufficient manufacturing capacity at our desired times or otherwise fail to perform satisfactorily, including by producing insufficient supply of commercial and clinical stock to meet patient demand or clinical trial requirements, or are impacted by delays or supply shortages, our product development activities, regulatory approval, and commercialization efforts may be delayed, prevented or impaired;

- we are conducting a technology transfer of the manufacturing process for ARCALYST drug substance from Regeneron Pharmaceuticals, Inc. ("Regeneron") to a new CDMO, and the process to complete the technology transfer and qualify a new CDMO may be subject to significant risks and uncertainties;
- for our products and product candidates that 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 such 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;
- we have entered into and may seek to enter into collaboration, licensing or other transactions to further develop, commercialize or otherwise realize value from one or more of our product candidates, and the expected value we hope to realize, including through milestone, royalty or other payments, may be less than we expect; 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. 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)

	June 30, 2023	December 31, 2022
Assets		
Current assets:		
Cash and cash equivalents	$ 112,567	$ 122,715
Short-term investments	72,425	67,893
Accounts receivable, net	24,650	12,660
Contract asset	—	7,656
Inventory	23,958	21,599
Prepaid expenses and other current assets	12,440	10,537
Total current assets	246,040	243,060
Property and equipment, net	1,136	1,658
Operating lease right-of-use assets	13,191	5,385
Other long-term assets	2,353	5,824
Intangible asset, net	17,750	18,250
Deferred tax assets	203,862	185,495
Total assets	$ 484,332	$ 459,672
Liabilities and Shareholders' Equity		
Current liabilities:		
Accounts payable	$ 648	$ 7,899
Accrued expenses	35,316	30,112
Deferred revenue	4,674	—
Operating lease liabilities	2,238	3,301
Other current liabilities	4,596	5,754
Total current liabilities	47,472	47,066
Non-current liabilities:		
Non-current deferred revenue	12,006	12,000
Non-current operating lease liabilities	11,283	2,618
Other long-term liabilities	1,915	1,839
Total liabilities	72,676	63,523
Commitments and contingencies (Note 14)		
Shareholders' equity:		
Class A common shares, par value of $0.000273235 per share; 35,074,019 shares and 34,750,560 shares issued and outstanding as of June 30, 2023 and December 31, 2022, respectively	9	9
Class B common shares, par value of $0.000273235 per share; 1,795,158 shares and 1,813,457 shares issued and outstanding as of June 30, 2023 and December 31, 2022, respectively	1	1
Class A1 common shares, $0.000273235 par value; 17,075,868 shares issued and outstanding as of June 30, 2023 and December 31, 2022	5	5
Class B1 common shares, $0.000273235 par value; 16,057,618 shares issued and outstanding as of June 30, 2023 and December 31, 2022	4	4
Additional paid-in capital	900,956	888,120
Accumulated other comprehensive income	13	44
Accumulated deficit	(489,332)	(492,034)
Total shareholders' equity	411,656	396,149
Total liabilities and shareholders' equity	$ 484,332	$ 459,672

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 June 30,		Six Months Ended June 30,	
	2023	2022	2023	2022
Revenue:				
Product revenue, net	$ 54,495	$ 26,972	$ 97,154	$ 49,161
License and collaboration revenue	16,978	—	22,664	10,000
Total revenue	71,473	26,972	119,818	59,161
Costs and operating expenses:				
Cost of goods sold	7,699	5,029	14,735	9,248
Collaboration expenses	13,986	3,672	22,274	11,926
Research and development	23,767	13,798	38,939	34,615
Selling, general and administrative	29,175	23,841	58,220	46,059
Total operating expenses	74,627	46,340	134,168	101,848
Loss from operations	(3,154)	(19,368)	(14,350)	(42,687)
Other income	1,915	103	3,747	137
Loss before income taxes	(1,239)	(19,265)	(10,603)	(42,550)
Benefit (provision) for income taxes	16,211	(716)	13,305	(2,641)
Net income (loss)	$ 14,972	$ (19,981)	$ 2,702	$ (45,191)
Net income (loss) per share attributable to common shareholders—basic	$ 0.21	$ (0.29)	$ 0.04	$ (0.65)
Net income (loss) per share attributable to common shareholders—diluted	$ 0.21	$ (0.29)	$ 0.04	$ (0.65)
Weighted average common shares outstanding—basic	69,918,287	69,289,972	69,835,452	69,213,860
Weighted average common shares outstanding—diluted	71,634,729	69,289,972	71,420,026	69,213,860
Comprehensive income (loss):				
Net income (loss)	$ 14,972	$ (19,981)	$ 2,702	$ (45,191)
Other comprehensive income (loss):				
Unrealized gain (loss) on short-term investments and currency translation adjustments, net of tax	(42)	13	(31)	(24)
Total other comprehensive income (loss)	(42)	13	(31)	(24)
Total comprehensive income (loss)	$ 14,930	$ (19,968)	$ 2,671	$ (45,215)

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

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

	Common Shares (Class A, B, A1 and B1)		Additional Paid-In Capital	Accumulated Other Comprehensive Income	Accumulated Deficit	Total Shareholders' Equity
	Shares	Amount				
Balances at December 31, 2022	69,697,503	$ 19	$ 888,120	$ 44	$ (492,034)	$ 396,149
Issuance of Class A common shares under incentive award plans	135,576	—	90	—	—	90
Share-based compensation expense	—	—	6,115	—	—	6,115
Unrealized gain on short-term investments and currency translation adjustments	—	—	—	11	—	11
Net loss	—	—	—	—	(12,270)	(12,270)
Balances at March 31, 2023	69,833,079	$ 19	$ 894,325	$ 55	$ (504,304)	$ 390,095
Issuance of Class A common shares under incentive award plans	169,584	—	158	—	—	158
Share-based compensation expense	—	—	6,473	—	—	6,473
Unrealized loss on short-term investments and currency translation adjustments	—	—	—	(42)	—	(42)
Net Income	—	—	—	—	14,972	14,972
Balances at June 30, 2023	70,002,663	$ 19	$ 900,956	$ 13	$ (489,332)	$ 411,656

	Common Shares (Class A, B, A1 and B1)		Additional Paid-In Capital	Accumulated Other Comprehensive Loss	Accumulated Deficit	Total Shareholders' Equity
	Shares	Amount				
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

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

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

	Six Months Ended June 30,	
	2023	**2022**
Cash flows from operating activities:		
Net income (loss)	$ 2,702	$ (45,191)
Adjustments to reconcile net income (loss) to net cash used in operating activities:		
Depreciation and amortization expense	1,198	1,307
Share-based compensation expense	12,588	12,707
Non-cash lease expense	1,610	1,481
Amortization of premiums and accretion of discounts on short-term investments	(2,213)	240
Loss on disposal of property and equipment	175	34
Deferred income taxes	(18,367)	—
Changes in operating assets and liabilities:		
Prepaid expenses and other current assets	(1,925)	(7,421)
Accounts receivable, net	(11,990)	(3,823)
Inventory	(2,359)	(16,241)
Contract asset	7,656	—
Other long-term assets	3,178	2,918
Accounts payable	(7,251)	654
Accrued expenses and other current liabilities	4,046	(1,889)
Operating lease liabilities	(1,814)	(1,436)
Deferred revenue	4,680	12,000
Other long-term liabilities	76	(10)
Net cash used in operating activities	(8,010)	(44,670)
Cash flows from investing activities:		
Proceeds from sale of property and equipment	—	91
Purchases of property and equipment	(58)	(115)
Purchases of short-term investments	(91,028)	(55,734)
Proceeds from the maturities of short-term investments	88,700	71,300
Net cash provided (used) in investing activities	(2,386)	15,542
Cash flows from financing activities:		
Proceeds from issuance of Class A common shares under incentive award plans and employee share purchase plan	1,087	1,558
Payments in connection with Common Stock tendered for employee tax obligations	(839)	(593)
Net cash provided by financing activities	248	965
Net decrease in cash and cash equivalents	(10,148)	(28,163)
Cash and cash equivalents at beginning of period	122,715	122,470
Cash and cash equivalents at end of period	$ 112,567	$ 94,307
Supplemental information:		
Cash paid for income taxes	$ 6,716	$ 3,293
Supplemental disclosure of non-cash investing and financing activities:		
Change in right-of-use asset as a result of new, modified, and terminated leases	$ 9,416	$ —

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 immune-modulating assets, ARCALYST, KPL-404 and mavrilimumab, are based on strong biologic rationale or validated mechanisms, target a spectrum of underserved cardiovascular and autoimmune conditions and offer the potential for differentiation.

The Company is subject to risks and uncertainties common to small 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 and whether any of the Company's current or future product candidates, if approved, will be commercially viable. Such risks and uncertainties 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 accrual for research and development expenses, the valuation of our deferred tax assets and the valuation of share-based awards. Estimates are periodically reviewed in light of changes in circumstances, facts and experience. Changes in estimates are recorded in the period in which they become known. Actual results could differ from those estimates.

Unaudited Interim Consolidated Financial Information

The accompanying unaudited consolidated financial statements have been prepared in accordance with GAAP for interim financial information. The accompanying unaudited consolidated financial statements do not include all of the information and footnotes required by GAAP for complete consolidated financial statements. The 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 ended December 31, 2022 (the "2022 Form 10-K"). The Company's accounting policies are described in the Notes to

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

Consolidated Financial Statements included in the Company's 2022 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 June 30, 2023 and the results of its operations for the three and six months ended June 30, 2023 and 2022, the changes in its shareholders' equity for the three and six months ended June 30, 2023 and 2022 and its cash flows for the six months ended June 30, 2023 and 2022. The results for the three and six months ended June 30, 2023 are not necessarily indicative of results to be expected for the year ending December 31, 2023, 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 June 30, 2023, the Company had an accumulated deficit of $489,332. During the six months ended June 30, 2023, the Company reported net income of $2,702 and used $8,010 cash in operating activities. As of June 30, 2023, the Company had cash, cash equivalents and short-term investments of $184,992.

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 or sustain ARCALYST commercial revenue in future periods, the Company would need to seek additional financing through public or private securities offerings, debt financings, 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.

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.

2. 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.

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- 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 June 30, 2023 Using:			
	Level 1	Level 2	Level 3	Total
Assets:				
Cash equivalents — money market funds	$ 60,746	$ —	$ —	$ 60,746
Cash equivalents — U.S. Treasury notes	—	16,245	—	16,245
Short-term investments — U.S. Treasury notes	—	72,425	—	72,425
	$ 60,746	$ 88,670	$ —	$ 149,416

	Fair Value Measurements as of December 31, 2022 Using:			
	Level 1	Level 2	Level 3	Total
Assets:				
Cash equivalents — money market funds	$ 20,929	$ —	$ —	$ 20,929
Cash equivalents — U.S. Treasury notes	—	15,009	—	15,009
Short-term investments — U.S. Treasury notes	—	67,893	—	67,893
	$ 20,929	$ 82,902	$ —	$ 103,831

During the six months ended June 30, 2023 and the year ended December 31, 2022, there were no transfers between Level 1, Level 2 and Level 3. The money market funds were valued using quoted prices in active markets, which represent a Level 1 measurement in the fair value hierarchy. The Company's cash equivalents and short-term investments as of June 30, 2023 and December 31, 2022 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
June 30, 2023					
Cash equivalents — U.S. Treasury notes	$ 16,241	$ 4	$ —	$ —	$ 16,245
Short-term investments — U.S. Treasury notes	72,433	7	(15)	—	72,425
	$ 88,674	$ 11	$ (15)	$ —	$ 88,670

	Amortized Cost	Gross Unrealized Gains	Gross Unrealized Losses	Credit Losses	Fair Value
December 31, 2022					
Cash equivalents — U.S. Treasury notes	$ 15,006	$ 3	$ —	$ —	$ 15,009
Short-term investments — U.S. Treasury notes	67,891	6	(4)	—	67,893
	$ 82,897	$ 9	$ (4)	$ —	$ 82,902

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As of June 30, 2023, we consider the unrealized losses in our investment portfolio to be temporary in nature and not due to credit losses. We have the ability to hold such investments until recovery of the fair value. We utilize the specific identification method in computing realized gains and losses. We had no realized gains and losses on our available-for-sale securities for the three and six months ended June 30, 2023 or 2022.

3. Product Revenue, Net

Product revenue, net, from sales of ARCALYST was as follows:

	Three Months Ended June 30,		Six Months Ended June 30,	
	2023	2022	2023	2022
Product revenue, net	$ 54,495	$ 26,972	$ 97,154	$ 49,161

The following table summarizes balances and activity in each of the product revenue allowance and reserve categories for the six months ended June 30, 2023:

	Contractual Adjustments	Government Rebates	Returns	Total
Balance at December 31, 2022	$ 1,464	$ 2,084	$ 351	$ 3,899
Current provisions relating to sales in the current year	7,247	3,891	163	11,301
Adjustments relating to prior years	(72)	(32)	(78)	(182)
Payments/returns relating to sales in the current year	(6,043)	(1,666)	—	(7,709)
Payments/returns relating to sales in the prior years	(1,392)	(1,987)	(39)	(3,418)
Balance at June 30, 2023	$ 1,204	$ 2,290	$ 397	$ 3,891

Total revenue-related reserves as of June 30, 2023 and December 31, 2022, included in our consolidated balance sheets, are summarized as follows:

	June 30, 2023	December 31, 2022
Components of accounts receivable	$ (290)	$ (304)
Components of other current liabilities	4,181	4,203
Total revenue-related reserves	$ 3,891	$ 3,899

Primarily all of the Company's trade accounts receivable arise from product revenue in the United States due from the Company's third party logistics provider.

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4. Inventory

Inventory consisted of the following:

	June 30, 2023	December 31, 2022
Raw materials	$ —	$ —
Work-in-process	6,623	6,312
Finished Goods	17,335	15,287
	$ 23,958	$ 21,599

5. Property and Equipment, Net

Property and equipment, net consisted of the following:

	June 30, 2023	December 31, 2022
Furniture, fixtures and vehicles	$ 224	$ 224
Computer hardware and software	345	345
Leasehold improvements	3,931	3,931
Lab equipment	4,076	4,017
Construction in progress	—	—
Total property and equipment	8,576	8,517
Less: Accumulated depreciation	(7,440)	(6,859)
Total property and equipment, net	$ 1,136	$ 1,658

Depreciation expense was $292 and $350 during the three months ended June 30, 2023 and 2022, respectively, and $581 and $709 during the six months ended June 30, 2023 and 2022, respectively.

As of June 30, 2023 and December 31, 2022, $170 and $226, respectively, of our property and equipment, net was in the United Kingdom.

6. Leases

Kiniksa US and Kiniksa UK leases office and laboratory space under operating leases. Additionally, Kiniksa US leases vehicles under an operating lease. In May 2023, the Company entered into a lease amendment to extend the term of the Lexington, Massachusetts headquarters lease by forty-eighty months to August 31, 2028. The Company accounted for the lease amendment as a modification and recorded increases in the right-of-use-assets and lease liability of $8,515.

The components of lease cost for three and six months ended June 30, 2023 and 2022:

	Three Months Ended June 30,		Six Months Ended June 30,	
	2023	2022	2023	2022
Operating lease cost	$ 871	$ 806	$ 1,748	$ 1,612
Variable lease cost	177	90	397	170
Total lease cost	$ 1,048	$ 896	$ 2,145	$ 1,782

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	June 30, 2023
Weighted-average remaining lease term (years)	3.97
Weighted-average discount rate	6.50%

Maturities of operating leases liabilities were as follows:

As of June 30, 2023	
2023 (remaining six months)	$ 1,871
2024	2,866
2025	3,098
2026	2,954
2027	2,996
2028 and thereafter	2,037
Total future minimum lease payments	$ 15,822
Less imputed interest	(2,301)
Present value of lease liabilities	$ 13,521

7. Intangible Assets

Intangible assets, net of accumulated amortization as of June 30, 2023 and December 31, 2022 are summarized in the following table.

	Estimated life	As of June 30, 2023			As of December 31, 2022		
		Cost	Accumulated Amortization	Net	Cost	Accumulated Amortization	Net
Regulatory milestone	20 years	$ 20,000	$ 2,250	$ 17,750	$ 20,000	$ 1,750	$ 18,250
		$ 20,000	$ 2,250	$ 17,750	$ 20,000	$ 1,750	$ 18,250

8. Accrued Expenses

Accrued expenses consisted of the following:

	June 30, 2023	December 31, 2022
Accrued research and development expenses	$ 7,713	$ 8,378
Accrued employee compensation and benefits	9,242	11,213
Accrued collaboration expenses	13,986	7,522
Accrued legal, commercial and professional fees	4,125	2,866
Other	250	133
	$ 35,316	$ 30,112

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”).

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2015 Plan

As of June 30, 2023, there were 1,950,667 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, 2023, increased by 2,787,900 Class A common shares. As of June 30, 2023, 5,542,432 shares remained available for future grant under the 2018 Plan.

2018 ESPP

In December 2022, the Company's board of directors resolved not to increase the number of Class A common shares reserved for issuance under the 2018 ESPP. As of June 30, 2023, 562,890 Class A common shares were available for future issuance under the 2018 ESPP.

Options

Share option activity under the Plans is summarized as follows:

	Number of Shares	Weighted Average Exercise Price
Outstanding as of December 31, 2022	10,144,618	$ 13.36
Granted	1,080,434	$ 11.80
Exercised	(66,639)	$ 10.35
Forfeited	(194,635)	$ 14.91
Outstanding as of June 30, 2023	10,963,778	$ 13.20
Share options exercisable as of June 30, 2023	6,533,441	$ 13.49
Share options unvested as of June 30, 2023	4,430,337	$ 12.77

As of June 30, 2023, total unrecognized compensation expense related to the unvested share option awards was $34,670 which is expected to be recognized over a weighted average remaining period of 2.52 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 in March 2022, 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 vested in one installment in March 2023.

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During the three months ended June 30, 2023 and 2022, the Company recognized compensation expense of $1,753 and $932, respectively, related to RSUs including those granted in connection with the RLTIP. During the six months ended June 30, 2023 and 2022, the Company recognized compensation expense of $3,282 and $1,551, respectively, related to RSUs including those granted in connection with the RLTIP.

The following table summarizes RSU activity, including the RSUs issued under the RLTIP for the six months ended June 30, 2023:

	Number of Shares	Weighted Average Grant Date Fair Value
Unvested RSUs as of December 31, 2022	1,742,401	$ 12.76
Granted	566,188	$ 11.54
Vested	(271,925)	$ 14.64
Forfeited	(93,668)	$ 11.93
Unvested RSUs as of June 30, 2023	1,942,996	$ 12.18

As of June 30, 2023, total unrecognized compensation cost related to the RSU awards was $20,162 which is expected to be recognized over a weighted average remaining period of 3.05 years.

Share-Based Compensation

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

	Three Months Ended June 30,		Six Months Ended June 30,	
	2023	2022	2023	2022
Cost of goods sold	$ 311	$ 112	$ 597	$ 292
Research and development expenses	1,395	1,608	2,839	3,584
Selling, general and administrative expenses	4,767	4,956	9,152	8,831
	$ 6,473	$ 6,676	$ 12,588	$ 12,707

10. Out-Licensing Agreements

Genentech License Agreement

In August 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”).

Under the Genentech License Agreement, the Company received an upfront payment of $80,000 for the license. In the first quarter of 2023, following the Company’s last delivery of certain drug supplies to Genentech, the Company received an additional $20,000 payment. In the second quarter of 2023, following the achievement of a development milestone related to a new indication under the Genentech License Agreement, Genentech became obligated to make an additional cash payment of $15,000. Under the terms of the Genentech License Agreement, the Company is eligible to receive a total of approximately $600,000 in contingent payments, including specified development, regulatory and

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sales-based milestones, before fulfilling the Company's upstream financial obligations, of which approximately $585,000 remain as of June 30, 2023. 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 completing its Phase 2b clinical trial assessing the efficacy, safety and tolerability of vixarelimab in reducing pruritis in prurigo nodularis.

Accounting for the Genentech License Agreement

As of the Genentech Effective Date, the Company identified the following 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 Company determined the transaction price of the Genentech License Agreement consisted of the $80,000 upfront payment and the $20,000 variable consideration related to the delivery of the initial drug supply and drug product resupply which was added to the transaction price in the fourth quarter of 2022. In the second quarter of 2023, the Company added $15,000 to the transaction price following the achievement of a development milestone related to a new indication under the Genentech License Agreement.

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 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 each of the four performance obligations noted above.

Performance Obligation	Method of Recognition
Exclusive license for vixarelimab	Point in time; 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 on that date.
Initial drug supply delivery	Point in time upon delivery.
Drug product resupply delivery	Point in time upon delivery.
Completion of the phase 2b clinical trial for vixarelimab	Over time; using the cost-to-cost input method, which is believed to best depict the transfer of control to the customer. Under the cost-to-cost input method, the percent of completion is based on the ratio of actual costs incurred as of the period end to the total estimated costs. Revenue is recorded as a percentage of the allocated transaction price times the percent of completion.

The Company recognized $16,978 and $22,664 of collaboration revenue under the Genentech License Agreement during the three and six months ended June 30, 2023, respectively. As a result of the $15,000 development milestone the Company recognized revenue of $14,001 and $13,148, during the three and six months ended June 30, 2023, respectively, related to performance obligations satisfied in prior periods. The remaining revenue was recognized as a result of further progress towards completion of the of the phase 2b clinical trial for vixarelimab, which is being

recognized over time using the cost-to-cost input method. The Company expects to recognize the remaining deferred revenue associated with the Genentech License Agreement over the remaining portion of the Phase 2b clinical trial for vixarelimab.

Huadong Collaboration Agreements

In February 2022 (the "Effective Date"), the Company entered into two collaboration and license agreements (each, a "Huadong Collaboration Agreement" and together, the "Huadong 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 "Huadong Licensed Product" and together, the "Huadong 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 "Huadong Territory"). The Company otherwise retained its current rights to the Huadong Licensed Products outside the Huadong Territory.

Under the Huadong Collaboration Agreements, the Company received a total upfront cash payment of $22,000, which includes $12,000 for the Huadong Territory license of rilonacept and $10,000 for the Huadong 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 Huadong Licensed Product-by-Huadong Licensed Product basis ranging from the low-teens to low-twenties on annual net sales of each Huadong Licensed Product in the Huadong 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 Huadong Licensed Product-by-Huadong 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 Huadong Licensed Product in such country or region in the Huadong 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 Huadong Licensed Product in such country or region in the Huadong Territory, and (iii) the expiration of the last regulatory exclusivity for the applicable Huadong Licensed Product in such country or region in the Huadong Territory.

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

Accounting for the Mavrilimumab Huadong Collaboration Agreement

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

The Company determined the transaction price at the inception of the mavrilimumab Huadong 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.

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 year ended December 31, 2022 under the mavrilimumab Huadong 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 the Rilonacept Huadong Collaboration Agreement

As of the Effective Date, the Company identified one performance obligation in the rilonacept Huadong Collaboration Agreement: the exclusive license for rilonacept and clinical and commercial manufacturing obligations for rilonacept products in the Huadong Territory. Huadong cannot exploit the value of the exclusive license for rilonacept products in the Huadong Territory without receipt of supply as the exclusive license for rilonacept products in the Huadong Territory does not convey to Huadong the right to manufacture and therefore the Company has combined the exclusive license for rilonacept products in the Huadong Territory and the manufacturing obligations into one performance obligation.

The Company determined the transaction price at the inception of the rilonacept Huadong Collaboration Agreement which includes $12,000, consisting of the upfront payment. The Company also includes an estimate of variable consideration associated with the clinical and commercial 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.

The Company recognizes revenue for the single performance obligation in the rilonacept Huadong Collaboration Agreement consisting of the exclusive license for rilonacept and clinical and commercial manufacturing obligations for rilonacept products in the Huadong 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 Huadong 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 Huadong Collaboration Agreement for the three and six months ended June 30, 2023 as there has been no delivery of materials under the rilonacept Huadong Collaboration Agreement to date. The transaction price of $11,895 is recorded in non-current deferred revenue and $105 is recorded in current deferred revenue, based upon timing of anticipated future shipments.

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The following table summarizes the Company's contract assets and contract liabilities in connection with license and collaboration agreements for the six months ended June 30, 2023:

	Balance at Beginning of Period	Additions	Revenue Recognized	Reclassification	Balance at End of Period
Six Months Ended June 30, 2023					
Contract Assets:					
Genentech vixarelimab	$ 7,656	$ —	$ —	$ (7,656)	$ —
Contract Liabilities:					
Genentech vixarelimab	$ —	$ 35,000	$ (22,664)	$ (7,656)	$ 4,680
Huadong rilonacept	12,000	—	—	—	12,000
Total Contract Liabilities	$ 12,000	$ 35,000	$ (22,664)	$ (7,656)	$ 16,680

11. License and Acquisition Agreements

Biogen Asset Purchase Agreement

In September 2016, the Company entered into an asset purchase agreement (the "Biogen Agreement") with Biogen MA Inc. ("Biogen") to acquire all of Biogen's right, title and interest in and to certain assets used in or relating to 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 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 clinical, regulatory and net sales milestone payments to Biogen of up to $329,000, of which $315,000 remains as of June 30, 2023. Additionally, the Company could be obligated to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the high single-digit percentages and ending below the teens.

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).

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.

In August 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.

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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 six months ended June 30, 2023 and 2022, the Company recorded research and development expense of $34, related to the annual maintenance in connection with the Biogen Agreement.

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 six months ended June 30, 2023 and 2022, 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 ("BLA") 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 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. 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) the technology transfer of the manufacturing process for ARCALYST drug substance will 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 six months ended June 30, 2023, the Company recognized $13,986 and $22,274 respectively, of expenses related to the profit sharing agreement presented within collaboration expenses. For the three and six months ended June 30, 2022, the Company recognized $3,672 and $11,926 respectively, of expenses related to the profit sharing

agreement presented within collaboration expenses. For the six months ended June 30, 2022, expenses related to the profit sharing agreement presented within collaboration expenses included $6,000 of profit sharing expense related to the rilonacept Huadong Collaboration Agreement.

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 clinical and commercial use. 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 six months ended June 30, 2023 and 2022, the Company did not incur any research and development expense related to the purchase of drug materials under the clinical supply agreement. The Company's inventory balance as of June 30, 2023 and December 31, 2022, of $23,958 and $21,599, respectively, were purchased under the commercial supply agreement. As of June 30, 2023, 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.

The Company is obligated to make clinical regulatory milestone payments for the first two indications and sales milestone payments of up to $1,257,500 in the aggregate, of which $1,242,500 remain as of June 30, 2023. 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 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.

KINIKSA PHARMACEUTICALS, LTD.
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(Unaudited)

During the three and six months ended June 30, 2023 and 2022, the Company did not record research and development expense in connection with milestone payments due under the MedImmune Agreement.

12. Net Income (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 included in our Form 10-K). As the liquidation and dividend rights are identical, losses are allocated on a proportionate basis and the resulting net income (loss) per share attributed to common shareholders will, therefore, be the same for both Class A and Class B common shares on an individual or combined basis.

Basic and diluted net income (loss) attributable to common shareholders was calculated as follows:

	Three Months Ended June 30,		Six Months Ended June 30,	
	2023	2022	2023	2022
Numerator:				
Net income (loss) attributable to common shareholders	$ 14,972	$ (19,981)	$ 2,702	$ (45,191)
Denominator:				
Weighted-average basic shares outstanding	69,918,287	69,289,972	69,835,452	69,213,860
Effect of dilutive securities				
Options to purchase common shares	1,206,683	—	1,165,632	—
Unvested RSUs	509,759	—	418,942	—
Weighted-average diluted shares	71,634,729	69,289,972	71,420,026	69,213,860
Basic EPS	$ 0.21	$ (0.29)	$ 0.04	$ (0.65)
Diluted EPS	$ 0.21	$ (0.29)	$ 0.04	$ (0.65)

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 six months ended June 30, 2022 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:

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	Three Months Ended June 30,		Six Months Ended June 30,	
	2023	2022	2023	2022
Share options to purchase common shares	8,204,258	9,573,543	8,219,041	9,573,543
Unvested RSUs	255,769	1,158,744	690,695	1,158,744
Total anti-dilutive shares	8,460,027	10,732,287	8,909,736	10,732,287

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 six months ended June 30, 2023 is due to Kiniksa UK's and Kiniksa US's income subject to taxation in each of their respective countries. Income tax benefit for the three and six months ended June 30, 2023 was $16,211 and $13,305, respectively. The benefit for income taxes is primarily driven by the release of the valuation allowance on the Company's U.S. deferred tax assets. This is partially offset by provision for income taxes driven by income earned in the UK and U.S. reduced by tax benefits from Foreign Derived Intangible Income ("FDII") deduction and U.S. federal and state research and development credits.

Management regularly assesses the need for a 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 second quarter of 2023, the Company assessed the valuation allowance on its U.S. deferred tax assets and considered positive evidence, including cumulative U.S. income in recent years, primarily related to cost plus arrangements and expectations regarding future profitability. The Company determined it was more likely than not that its U.S. deferred tax assets are realizable in the future and released the associated valuation allowance as of June 30, 2023. This resulted in a benefit of $19,185. There are no material deferred tax assets in the jurisdictions outside Kiniksa US and Kiniksa UK.

A number of countries have begun to enact legislation to implement the Organization for Economic Cooperation and Development's ("OECD") international tax framework. The Company is currently monitoring these developments and is in the process of evaluating the potential impact on its results of operations.

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).

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Manufacturing Commitments

The Company entered into supply agreements with Regeneron to provide both clinical supply and commercial product (see Note 11). In May 2023, the Company signed a letter of intent with a contract development and manufacturing organization (a "CDMO") related to its technology transfer of the manufacturing process for ARCALYST drug substance. The Company has additionally entered into agreements with several CDMOs to provide the Company with preclinical and clinical trial materials for its non-ARCALYST assets. As of June 30, 2023, the Company had committed to minimum payments under all of these agreements totaling $139,417, of which $62,496 is 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 June 30, 2023 or December 31, 2022.

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, and our audited consolidated financial statements and related notes for the year ended December 31, 2022 included in our Annual Report on Form 10-K (our "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 (the "SEC") our actual results could differ materially from the results, performance or achievements expressed in or implied by these forward-looking statements.

Overview

We are a biopharmaceutical company focused on discovering, acquiring, developing and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. Our immune-modulating assets, ARCALYST, KPL-404 and mavrilimumab, are based on strong biologic rationale or validated mechanisms, target a spectrum of underserved cardiovascular and autoimmune conditions, and offer the potential for differentiation.

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 ("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 across the United States through a network of distributors. ARCALYST is also approved in the United States for the treatment of Cryopyrin-Associated Periodic Syndromes ("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 ("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, as well as third party proceeds, with Regeneron. In March 2023, Regeneron initiated a technology transfer of the manufacturing process for ARCALYST drug substance and we are working with Regeneron to qualify a new CDMO.

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. ("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. ("BIDMC"). 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 ("RA"), Sjogren's syndrome, Graves' disease and systemic lupus erythematosus. In May 2021, we announced positive final data from our Phase 1 single-ascending-dose 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 clinical trial of KPL-404 in RA, which is designed to evaluate pharmacokinetics, safety and efficacy with subcutaneous administration. In January 2023, we announced that we had completed enrollment of the multiple-ascending-dose portion of such trial. We are currently enrolling the proof-of-concept portion of the trial. 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 ("GM-CSFRα"). In 2017, we licensed exclusive worldwide rights in all indications to mavrilimumab from MedImmune, Limited ("MedImmune"). We are pursuing collaborative study agreements to evaluate the potential of mavrilimumab in rare cardiovascular diseases where the GM-CSF mechanism has been

implicated. We previously evaluated mavrilimumab in giant cell arteritis ("GCA"), a chronic inflammatory disease of the medium-to-large arteries, and COVID-19-related acute respiratory distress syndrome ("ARDS").

Vixarelimab is an investigational monoclonal antibody inhibitor of signaling through oncostatin M receptor beta ("OSMRβ"), which was previously part of our portfolio of immune-modulating assets. In September 2022, we closed an agreement granting Genentech, Inc. and F. Hoffmann-La Roche Ltd (collectively, "Genentech") an exclusive worldwide license to develop and commercialize vixarelimab. 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 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 three and six months ended June 30, 2023, our net income was $15.0 million and $2.7 million, respectively, primarily as a result of the release of our deferred US tax asset valuation allowance; however, as of June 30, 2023, we had an accumulated deficit of $489.3 million. Notwithstanding the foregoing, we expect to incur operating losses for the foreseeable future 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 expenses related to product manufacturing, including technology transfer costs, marketing, sales and distribution of ARCALYST. We may also incur expenses in connection with the in-licensing or acquisition of additional product candidates.

As of June 30, 2023, we had cash, cash equivalents and short-term investments of $185.0 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.

In February 2022, we entered into two collaboration and license agreements (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 year ended December 31, 2022. We deferred the $12.0 million related to the rilonacept license agreement as of June 30, 2023, and will recognize revenue as materials are shipped.

In August 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. In the first quarter of 2023, following delivery of certain drug supplies to Genentech, we received an additional $20.0 million payment. In the second quarter of 2023, following the achievement of a development milestone related to a new indication under the Genentech License Agreement, Genentech became obligated to make an additional cash payment of $15.0 million. We will be eligible to receive up to a total of approximately $600,000 in contingent payments, including specified development, regulatory and sales-based milestones, of which approximately $585,000 remain as of June 30, 2023, 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 have recognized $110.3 million of revenue of the $115.0 million transaction price under the Genentech License Agreement and will recognize the remaining revenue over the remaining duration of the in-progress Phase 2b prurigo nodularis clinical trial.

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, supply chain activities and the technology transfer of the manufacturing process for drug substance.

Collaboration expenses

Collaboration expenses consist of Regeneron's share of the profit related to ARCALYST sales under the license agreement with Regeneron (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. With respect to the technology transfer of the manufacturing process for ARCALYST drug substance initiated by Regeneron in March 2023, to the extent permitted by the Regeneron Agreement, the fully-burdened costs of each of us and Regeneron incurred in performing 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 CROs that are primarily engaged in the oversight and conduct of our clinical trials and CDMOs 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, CDMOs 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 external commercialization, marketing, and professional fees for legal, patent, and accounting services.

We have been commercializing ARCALYST since April 2021 and expect that our selling, general and administrative expenses will continue to increase in the future as we continue to perform commercialization and sales activities.

Other Income

Other income consists of interest income recognized from investments in money market funds, U.S. Treasury notes and other miscellaneous income 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 primarily relates to the release of the valuation allowance on our U.S. 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 June 30, 2023 and 2022

The following table summarizes our results of operations for the three months ended June 30, 2023 and 2022:

	Three Months Ended June 30,		
	2023	2022	Change
		(in thousands)	
Revenue:			
Product revenue, net	$ 54,495	$ 26,972	$ 27,523
License and collaboration revenue	16,978	—	16,978
Total revenue	71,473	26,972	44,501
Costs and Operating expenses:			
Cost of goods sold	7,699	5,029	2,670
Collaboration expenses	13,986	3,672	10,314
Research and development	23,767	13,798	9,969
Selling, general and administrative	29,175	23,841	5,334
Total operating expenses	74,627	46,340	28,287
Loss from operations	(3,154)	(19,368)	16,214
Other income	1,915	103	1,812
Loss before benefit (provision) for income taxes	(1,239)	(19,265)	18,026
Benefit (provision) for income taxes	16,211	(716)	16,927
Net income (loss)	$ 14,972	$ (19,981)	$ 34,953

Product Revenue, Net

We recognized net revenue from the sale of ARCALYST of $54.5 million for the three months ended June 30, 2023, compared to $27.0 million for the three months ended June 30, 2022, an increase of $27.5 million. The increase in product revenue was primarily driven by an increase in patients.

License and Collaboration Revenue

We reported $17.0 million of license and collaboration revenue for the three months ended June 30, 2023, related to the Genentech License Agreement. This license and collaboration revenue was primarily driven by the achievement of a $15.0 million development milestone related to a new indication under the Genentech License Agreement. We expect to recognize the remaining $4.7 million of the transaction price still to be recognized related to the Genentech License Agreement over the life of the in-progress Phase 2b prurigo nodularis clinical trial. We did not record any license and collaboration revenue during the three months ended June 30, 2022.

Cost of Goods Sold

We recognized cost of goods sold of $7.7 million for the three months ended June 30, 2023, compared to $5.0 million for the three months ended June 30, 2022, an increase of $2.7 million. The increase in cost of goods sold relates primarily to the increase in sales of ARCALYST.

Collaboration Expenses

Collaboration expenses were $14.0 million for the three months ended June 30, 2023, compared to $3.7 million for the three months ended June 30, 2022, an increase of $10.3 million. Collaboration expenses increased due to increased revenue from sales of ARCALYST.

Research and Development Expenses

	Three Months Ended June 30,		
	2023	2022	Change
	(in thousands)		
Rilonacept (ARCALYST)	$ 450	$ (614)	$ 1,064
KPL-404	11,649	1,712	9,937
Mavrilimumab	308	977	(669)
Vixarelimab	2,278	3,634	(1,356)
Unallocated research and development expenses:			
Personnel related (including share-based compensation)	5,393	5,209	184
Other	3,689	2,880	809
Total research and development expenses	$ 23,767	$ 13,798	$ 9,969

Research and development expenses were $23.8 million for the three months ended June 30, 2023, compared to $13.8 million for the three months ended June 30, 2022, an increase of $10.0 million.

The direct costs for our ARCALYST program were $0.5 million during the three months ended June 30, 2023, compared to ($0.6) million during the three months ended June 30, 2022, an increase of $1.1 million. The expense increase for the three months ended June 30, 2023, is due to the limited remaining close-out costs of the RHAPSODY trial, our global, pivotal Phase 3 clinical trial in recurrent pericarditis. The reduction in expenses during the three months ended June 30, 2022 related primarily to lower than expected RHAPSODY trial cost, identified during the close-out activities offset by the limited period remaining costs.

The direct costs for our KPL-404 program were $11.6 million during the three months ended June 30, 2023, compared to $1.7 million during the three months ended June 30, 2022, an increase of $9.9 million. The increase in expenses incurred primarily related to the manufacturing of clinical material, initiation of cohort three and continuation of the first two cohorts of our Phase 2 trial in RA during the three months ended June 30, 2023, as compared to initial cost of the first two cohorts of our Phase 2 trial in RA, for the three months ended June 30, 2022.

The direct costs for our mavrilimumab program were $0.3 million during the three months ended June 30, 2023, compared to $1.0 million during the three months ended June 30, 2022, or a decrease of $0.7 million. The decrease in expenses incurred is primarily related to the continued wind-down of the Phase 3 clinical trial in COVID-19 related ARDS.

The direct costs for our vixarelimab program were $2.3 million during the three months ended June 30, 2023, compared to $3.6 million during the three months ended June 30, 2022, a decrease of $1.4 million. The decrease in expenses was primarily related to a decrease in active patients in our ongoing Phase 2b clinical trial in prurigo nodularis.

Unallocated research and development expenses were $9.1 million for the three months ended June 30, 2023, compared to $8.1 million for the three months ended June 30, 2022, an increase of $1.0 million. The increase of $1.0 million in unallocated research and development expenses was primarily due to an increase in headcount. Personnel-related costs for the three months ended June 30, 2023 and 2022 included share-based compensation of $1.4 million and $1.6 million, respectively.

Selling, General and Administrative Expenses

Selling, general and administrative expenses were $29.2 million for the three months ended June 30, 2023, compared to $23.8 million for the three months ended June 30, 2022. The increase of $5.3 million was primarily due to an increase of $3.6 million in personnel-related costs related to the expansion of the salesforce and an increase in professional fees of $0.8 million. Personnel-related costs for the three months ended June 30, 2023 and 2022 included share-based compensation of $4.8 million and $5.0 million, respectively.

Other Income

Other income was $1.9 million for the three months ended June 30, 2023 compared to other income of $0.1 million for the three months ended June 30, 2022. The increase of $1.8 million was primarily due to higher interest rates on U.S. Treasury notes and a larger average balance in short term investments.

Provision for Income Taxes

For the three months ended June 30, 2023, we recorded an income tax benefit of $16.2 million relating primarily to the release of the valuation allowance on our U.S. deferred tax assets offset by income earned in the UK and the U.S., net of the Foreign Derived Intangible Income ("FDII") deduction and U.S. federal and state research and development credits ("R&D Credits") utilized. For the three months ended June 30, 2022, we recorded an income tax provision of $0.7 million relating primarily to the current tax expense due to income from our cost plus arrangements in the United States, net of R&D Credits utilized. During the three months ended June 30, 2022, there was no UK tax provision because Kiniksa UK was in a net loss position and had a full valuation allowance against its deferred tax asset. We expect our UK current tax provision to be partially reduced by net operating losses to the extent available. We expect that our reported income tax expense will increase in future periods.

Comparison of the Six Months Ended June 30, 2023 and 2022

	Six Months Ended June 30,		
	2023	2022	Change
		(in thousands)	
Revenue:			
Product revenue, net	$ 97,154	$ 49,161	$ 47,993
License and collaboration revenue	22,664	10,000	12,664
Total revenue	119,818	59,161	60,657
Operating expenses:			
Cost of goods sold	14,735	9,248	5,487
Collaboration expenses	22,274	11,926	10,348
Research and development	38,939	34,615	4,324
Selling, general and administrative	58,220	46,059	12,161
Total operating expenses	134,168	101,848	32,320
Loss from operations	(14,350)	(42,687)	28,337
Other income	3,747	137	3,610
Loss before benefit (provision) for income taxes	(10,603)	(42,550)	31,947
Benefit (provision) for income taxes	13,305	(2,641)	15,946
Net income (loss)	$ 2,702	$ (45,191)	$ 47,893

Product Revenue, Net

We recognized net revenue from the sale of ARCALYST of $97.2 million for the six months ended June 30, 2023, compared to $49.2 million for the six months ended June 30, 2022, an increase of $48.0 million. The increase in product revenue was primarily driven by an increase in patients.

License and Collaboration Revenue

We reported $22.7 million of license and collaboration revenue for the six months ended June 30, 2023, related to the Genentech License Agreement, compared to $10.0 million for the six months ended June 30, 2022, an increase of $12.7 million. This increase in license and collaboration revenue was primarily driven by the achievement of a $15.0 million development milestone related to a new indication under the Genentech License Agreement, offset by the $10.0 million upfront payment recognized during the six months ended June 30, 2022 upon the signing of the mavrilimumab Collaboration Agreement in February of 2022.

Cost of Goods Sold

We recognized cost of goods sold of $14.7 million for the six months ended June 30, 2023, compared to $9.2 million for the six months ended June 30, 2022, an increase of $5.5 million. The increase in cost of goods sold relates primarily to the increase in sales of ARCALYST.

Collaboration Expenses

Collaboration expenses were $22.3 million for the six months ended June 30, 2023, compared to $11.9 million for the six months ended June 30, 2022, an increase of $10.3 million. Collaboration expenses increased due to increased revenue from sales of ARCALYST.

Research and Development Expenses

	Six Months Ended June 30,		
	2023	2022	Change
	(in thousands)		
Rilonacept (ARCALYST)	$ 1,008	$ 1,242	$ (234)
KPL-404	15,095	3,918	11,177
Mavrilimumab	36	4,489	(4,453)
Vixarelimab	4,325	6,439	(2,114)
Unallocated research and development expenses:			—
Personnel related (including share-based compensation)	11,806	12,179	(373)
Other	6,669	6,348	321
Total research and development expenses	$ 38,939	$ 34,615	$ 4,324

Research and development expenses were $38.9 million for the six months ended June 30, 2023, compared to $34.6 million for the six months ended June 30, 2022, an increase of $4.3 million.

The direct costs for our ARCALYST program were $1.0 million during the six months ended June 30, 2023, compared to $1.2 million during the six months ended June 30, 2022, a decrease of $0.2 million. The expense reduction for the six months ended June 30, 2023, is due to the limited remaining close-out costs of the RHAPSODY trial, our global, pivotal Phase 3 clinical trial in recurrent pericarditis. The expenses for the six months ended June 30, 2022, related primarily to the long-term extension portion of the RHAPSODY trial.

The direct costs for our KPL-404 program were $15.1 million during the six months ended June 30, 2023, compared to $3.9 million during the six months ended June 30, 2022, an increase of $11.2 million. The increase in expenses incurred primarily related to the manufacturing of clinical material, initiation cost of cohort three and continuation of the first two cohorts of our Phase 2 trial in RA during the six months ended June 30, 2023, as compared to initial cost of the first two cohorts of our Phase 2 trial in RA, for the six months ended June 30, 2022.

The direct costs for our mavrilimumab program were less than $0.1 million during the six months ended June 30, 2023, compared to $4.5 million during the six months ended June 30, 2022, or a decrease of $4.5 million. The

decrease in expenses incurred is primarily related to the continued wind-down activities of the Phase 3 clinical trial in COVID-19 related ARDS.

The direct costs for our vixarelimab program were $4.3 million during the six months ended June 30, 2023, compared to $6.4 million during the six months ended June 30, 2022, a decrease of $2.1 million. The decrease in expenses was primarily related to a decrease in active patients in our ongoing Phase 2b clinical trial in prurigo nodularis.

Unallocated research and development expenses were $18.5 million for the six months ended June 30, 2023 and 2022. Personnel-related costs for the six months ended June 30, 2023 and 2022 included share-based compensation of $2.8 million and $3.6 million, respectively.

Selling, General and Administrative Expenses

Selling, general and administrative expenses were $58.2 million for the six months ended June 30, 2023, compared to $46.1 million for the six months ended June 30, 2022. The increase of $12.2 million was primarily due to an increase of $8.1 million in personnel-related costs and $2.1 million in sales and marketing costs related to the expansion of the salesforce. Personnel-related costs for the six months ended June 30, 2023 and 2022 included share-based compensation of $9.2 million and $8.8 million, respectively.

Other Income

Other income was $3.7 million for the six months ended June 30, 2023 compared to other income of $0.1 million for the six months ended June 30, 2022. The increase was primarily due to higher interest rates on U.S. Treasury notes and a larger average balance in short term investments.

Provision for Income Taxes

For the six months ended June 30, 2023, we recorded an income tax benefit of $13.3 million relating primarily to the release of the valuation allowance on our U.S. deferred tax assets offset by income earned in the UK and the U.S., net of the FDII deduction and R&D Credits utilized. For the six months ended June 30, 2022, we recorded an income tax provision of $2.6 million relating primarily to the current tax expense due to income from our cost plus arrangements in the United States, net of R&D Credits utilized. During the six months ended June 30, 2022, there was no UK tax provision because Kiniksa UK was in a net loss position and had a full valuation allowance against its deferred tax asset. We expect our UK current tax provision to be partially reduced by net operating losses to the extent available. We expect that our reported income tax expense will increase in future periods.

Liquidity and Capital Resources

As of June 30, 2023, our principal source of liquidity was cash, cash equivalents and short-term investments, which totaled $185.0 million. Net income (loss) was $2.7 million and ($45.2) million for the six months ended June 30, 2023 and 2022, 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, pay annual maintenance fees and to meet due diligence requirements, each based upon specified events. Pursuant to the Regeneron Agreement, we have entered into a supply agreement with Regeneron to purchase both clinical and commercial product. We have committed to minimum payments to Regeneron of $31.2 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 $15.8 million, of which $3.0 million are due within one year. In connection with our ongoing technology transfer of ARCALYST drug substance manufacturing, we have entered into a manufacturing commitment with a CDMO to establish a new manufacturing site for ARCALYST drug substance. Such commitment, which includes the purchase of raw materials and related service fees, will obligate us to minimum payments of $97.3 million, $20.4 million of which 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 first quarter of 2023, we received $20.0 million for delivery of certain drug supplies as part of the Genentech License Agreement. In the second quarter of 2023, following the achievement of a development milestone related to a new indication under the Genentech License Agreement, Genentech became obligated to make an additional cash payment to us of $15,000.

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:

	Six Months Ended June 30,	
	2023	2022
	(in thousands)	
Net cash used in operating activities	$ (8,010)	$ (44,670)
Net cash provided (used) in investing activities	(2,386)	15,542
Net cash provided by financing activities	248	965
Net decrease in cash and cash equivalents	$ (10,148)	$ (28,163)

Operating Activities

During the six months ended June 30, 2023, operating activities used $8.0 million of cash which primarily consisted of our net income of $2.7 million, offset by working capital increase of $5.7 million and non-cash items of $5.0 million. Working capital increased primarily due to a $12.0 million increase in accounts receivable and $7.3 million decrease in accounts payable attributable to timing of inventory purchases, offset by a $7.7 decrease in contract assets, a $4.7 million increase in deferred revenue both related to the $20.0 million received as part of the Genentech License Agreement and a $4.0 million decrease in accrued expenses attributable to a lower level of late stage clinical trials. Non-cash items were comprised primarily of an increase in deferred income tax assets of $18.4 million and $2.2 million of amortization of premiums and accretion of discounts on short-term investments, offset by $12.6 million of stock based compensation.

During the six months ended June 30, 2022, operating activities used $44.7 million of cash, primarily resulting from our net loss of $45.2 million as well as net cash used by our operating assets and liabilities of $15.2 million offset by non-cash charges of $15.7 million. Net cash used by our operating assets and liabilities for the six months ended June 30, 2022 consisted primarily of a $16.2 million increase in inventory, and a $7.4 million increase in prepaid expenses and other current assets, offset by an increase of $12.0 million in deferred revenue as a result of the rilonacept Huadong Collaboration Agreement, and a $3.0 million decrease in other long-term assets.

Investing Activities

During the six months ended June 30, 2023, investing activities used $2.4 million of cash, primarily consisting of $91.0 million of purchases of short-term investments, offset by $88.7 million of maturities of short-term investments.

During the six months ended June 30, 2022 investing activities provided $15.5 million of cash, primarily consisting of $71.3 million from proceeds of maturities of short-term investments, partially offset by $55.7 million of purchases of short-term investments.

Financing Activities

During the six months ended June 30, 2023 and 2022, net cash provided by financing activities was $0.2 million and $1.0 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, make arrangements with additional third party manufacturers to successfully manufacture our products and product candidates and perform activities related to our technology transfer of the manufacturing process for ARCALYST drug substance;
- 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.

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, “Financial Statements (Unaudited)” 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;
- 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 June 30, 2023, 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 and has been volatile over the past several fiscal quarters. 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 June 30, 2023.

Changes in Internal Control over Financial Reporting

There was no change in our internal control over financial reporting (as defined in Rules 13a-15(f) and 15d-15(f) under the Exchange Act) during the three months ended June 30, 2023 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 that began generating revenue from product sales in 2021. We have a history of operating losses 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 depends on the continued commercialization of ARCALYST and the development and eventual commercialization of one or more of our current or future product candidates.

We have incurred operating losses in the past and expect to incur such losses in the future. For the six months ended June 30, 2023, our net income was $2.7 million, primarily as a result of the release of our US deferred tax asset valuation allowance, however as of June 30, 2023, we had an accumulated deficit of $489.3 million. Notwithstanding the foregoing, 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 product candidates for which we may obtain marketing approval;
- our research and preclinical and clinical development of our product candidates, including our Phase 2 clinical trial for KPL-404 in RA as well as the completion of our in-progress Phase 2b dose-ranging clinical trial of vixarelimab for the treatment of prurigo nodularis as required by the Genentech Licensing Agreement;
- manufacturing our products and product candidates for clinical or commercial use, increasing our manufacturing capabilities, adding additional manufacturers or suppliers and performing activities related to our technology transfer of the process for manufacturing ARCALYST drug substance;
- 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 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, a pandemic or other outbreak of disease, or the global economic slowdown and rising inflation.

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. Corporate profitability, when and if achieved, may not be sustained 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.

To further our operational plans, we may require substantial additional financing, which we may not be able to obtain when needed or on acceptable term.

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.

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, if we are unable to grow or sustain ARCALYST commercial revenue, we may need to obtain substantial additional funding to progress our operating plans via accessing capital markets. If we are unable to raise capital when needed on acceptable terms, if at all, we may 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. 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 organizational 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, 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, 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 global political turmoil, including pandemics or other outbreaks of disease and the ongoing war in Ukraine;

- the number, size and type of our 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 (a "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, 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;
- litigation arising out of, but not limited to, product liability claims, intellectual property disputes, disputes arising from our collaboration and license agreements and employment-related disputes;
- 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.

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, if approved.

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.

In addition to ARCALYST commercial revenue, we expect to finance our cash needs through private or public securities offerings, debt financings, 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 may not be able to continue to commercialize ARCALYST or be successful in commercializing any future products, potentially impairing the commercial potential for our current and future products to generate any revenue.

We have been generating revenue from 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 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;
- an absence or reduction in 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 to compete with current or future competitor products and/or biosimilars;
- 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;
- any delays in the ongoing technology transfer of the process for manufacturing ARCALYST drug substance;
- 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 or sustain 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.

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.

Our current or future products may not gain or sustain 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 in the U.S., 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 its approved indications, or any of our future products 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 products 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 and our ability to maintain and expand favorable labeling when and if needed;
- limitations or warnings contained in the labeling approved by regulatory authorities, including any additions mandated by authorities after initial approval;
- 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;
- publications of scientific literature and consensus papers favorable to the administration of our products and product candidates; and
- other potential advantages over alternative treatment methods.

If ARCALYST or any of our future approved products, if any, fail to gain or sustain market acceptance, our ability to generate revenue will be adversely affected. Even if ARCALYST or any future products 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 current and future products, 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 current and future products, 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 its approved indications or any of our future products, 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 future product and alternative treatments from third party payors (e.g., governmental authorities, private health insurers and other organizations). We currently enjoy largely 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, our ability to successfully commercialize ARCALYST or any of the product candidates for which we obtain marketing approval may be adversely affected. Moreover, any coverage or reimbursement that may be obtained may be decreased or eliminated in the future. For example, in January 2023, one of the large private health insurers that currently covers ARCALYST placed 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 may 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 also revisit their previously established coverage policies from time to time and often introduce more challenging price negotiation methodologies under a number of circumstances, including, but not limited to the entrance of new competitors, including branded drugs, generics and biosimilars, into 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. 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.

We have and may continue periodically 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 a small, commercial stage biopharmaceutical company, our future success relies on the effective commercialization of one or more of our current and future 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 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 a small commercial stage company, the loss of any payor, especially a large payor, or limitations on 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.

Price increases that outpace inflation may also 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 significant and sustained 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.

For more information, see "*Risk Factors – General Risk Factors – Enacted and future healthcare legislation may have a material adverse effect on our business and results of operations*."

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 products we commercialize;
- injury to our reputation and significant negative media attention;
- regulatory investigations that could require costly recalls or product modifications;
- difficulty in enrolling participants in clinical trials or withdrawal of clinical trial participants;
- significant costs to defend the related litigation;
- substantial monetary awards to trial participants;
- 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, training, expanding and/or maintaining a sales force is expensive and time-consuming and could impede any ongoing commercialization efforts or delay a prospective 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. A failure to maintain or expand a commercial sales force, as needed, may also materially impact any ongoing commercialization efforts.

If we enter into arrangements with third parties to perform sales, marketing, distribution and other commercial support services, our product revenues or the profitability of these revenues to us are likely to be lower than if we were to market and sell any product ourselves. In addition, we may not be successful in entering into arrangements with third parties to market and sell our products, 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 products 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 new products or impact ongoing commercialization of currently marketed products. 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 commercially successful and our business, results of operations, financial condition and prospects will be materially adversely affected.

Any future growth outside of the United States 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 current and future products in markets outside of the United States either on our own or through 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 certain 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 potentially costly clinical trial that compares our product candidate to other available therapies. Failure to demonstrate sufficiently desirable results to such parties may result in adverse pricing or reimbursement decisions. 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 current and future products 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 more stringent than 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 similar foreign regulations ("cGMP"). As such, we and our CDMOs 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 ("BLAs") or Marketing Authorization Applications ("MAAs"). Accordingly, we and our CDMOs 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 marketing approval is obtained via the accelerated approval pathway, we could be required to conduct a successful confirmatory clinical trial to confirm clinical benefit for our products. An unsuccessful confirmatory 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 any of our current or future products, such as adverse events of unanticipated severity or frequency, or problems with the facility where a 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 CDMOs' 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 occur, 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 potentially significant enforcement actions.

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 our current and future products.

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 (the “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;

- the Health Insurance Portability and Accountability Act (“HIPAA”), which imposes criminal and civil liability for executing a scheme to defraud any healthcare benefit program or obtain, by means of false or fraudulent pretenses, representations or promises, any of the money or property owned by, or under the

custody or control of, any healthcare benefit program, regardless of the payor (e.g., public or private) and knowingly and willfully falsifying, concealing or covering up a material fact or making any materially false statement in connection with the delivery of or payment for healthcare benefits, items or services. Similar to the federal Anti-Kickback Statute, a person or entity does not need to have actual knowledge of the statute or specific intent to violate it in order to have committed a violation;

- the federal civil monetary penalties laws, which impose civil fines for, among other things, the offering or transfer of remuneration to a Medicare or state healthcare program beneficiary if the person knows or should know it is likely to influence the beneficiary's selection of a particular provider, practitioner, or supplier of services reimbursable by Medicare or a state healthcare program, unless an exception applies;

- 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 anti-kickback 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 European Union (the "EU") and other jurisdictions, including reporting requirements detailing interactions with and payments to healthcare professionals, which may be applicable even if we are not commercializing a product in such jurisdictions.

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.

Each of our product candidates requires substantial preclinical or clinical development and manufacturing support and, if approved, an organization to facilitate a successful product launch and commercialization, which may not be synergistic with our existing capabilities, before we will be able to generate any revenue from product sales. The success of our current and 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 (“INDs”), and clinical trial applications (“CTAs”) 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 regulations, or similar foreign standards (“GLP”);

- 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 to 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 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, efficacy and cGMP requirements 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, if approved;

- 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 post-marketing requirements imposed by regulatory authorities, including any required post-marketing 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 we could experience significant delays in, or an inability to, timely or successfully commercialize our product candidates. 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 – To further our operational plans, we may require substantial additional*

financing, which we may not be able to obtain when needed or on acceptable term" 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."*

Though we are currently commercializing ARCALYST, we evenly split profits on ARCALYST sales with Regeneron, and the relevant markets for its indications may prove not to be large enough to allow us to generate significant and sustained revenue from ARCALYST sales. Moreover, even if we successfully obtain regulatory approvals to 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. See, "*—Risks Related to Commercialization—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*."

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 future clinical trials will be conducted as initially planned or completed on our initial projected schedule, if at all. Further, even if conducted on time, a clinical trial may result in unfavorable or statistically insignificant results. 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 expenses 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 CTA under the EU Clinical Trials Regulation ("CTR") 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, manufacturing and controls ("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 an institutional review board (an "IRB") or ethics committee for each clinical trial site before a trial may be initiated, which approval could be delayed, rejected or suspended. Our planned clinical trials may also include other applicable committees intended to monitor the safety profile of our product candidates. IRBs, regulatory authorities or other applicable safety committees 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 certain supplemental BLAs ("sBLA") to the FDA, an MAA to the European Medicines Agency (the "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 participants 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, ethics committee approval or positive opinion 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 participants 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 manner or in compliance with all applicable laws and regulations, including the FCPA;
- failure to perform in accordance with the FDA's good clinical practices ("GCPs") or applicable comparable regulatory guidelines in other countries;

- participants not completing a clinical trial or not returning for post-treatment follow-up, including as a result of trial demands on participants;
- clinical trial sites withdrawing from or being unable to conduct activities, or participants withdrawing from clinical trials, including as a result of a pandemic or other outbreak of disease and the ongoing war in Ukraine;
- participants experiencing serious adverse events or undesirable side effects or being exposed to unacceptable health risks;
- participants 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 sufficient quantities of or 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 in 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.

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. 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 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 such 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 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 in which the clinical trial takes place, to both the competent national health authority and an independent ethics committee, the CTR introduced a centralized process and only requires the submission of a single application for multi-center trials. 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. Clinical trials for which an application was submitted (i) prior to January 31, 2022 under the Clinical Trials Directive, or (ii) between January 31, 2022 and January 31, 2023 and for which the sponsor has opted for the application of the Clinical Trials Directive remain governed by the said Directive until January 31, 2025. After this date, all clinical trials (including those which are ongoing) will become subject to the provisions of the CTR. Compliance with the CTR requirements by us and our third party service providers, such as our CROs, may impact our development plans.

The United Kingdom's (the "UK") regulatory framework in relation to clinical trials is derived from existing EU legislation (as implemented into UK law, through secondary legislation). However, on January 17, 2022, the Medicines and Healthcare products Regulatory Agency (the "MHRA") launched an eight-week consultation on

reframing the UK legislation for clinical trials with the aim 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 UK government published its response to the consultation in March 2023, confirming that it would bring forward changes to the legislation. These resulting legislative amendments will be closely watched and will determine how closely the UK regulations will be aligned with the CTR. A decision by the UK government not to closely align any new legislation with the new approach that has been adopted in the EU may have an effect on the cost of conducting clinical trials in the UK as opposed to other countries.

Under the terms of the Protocol on Ireland/Northern Ireland, provisions of the CTR which relate to the manufacture and import of investigational medicinal products and auxiliary medicinal products apply in Northern Ireland. In February 2023 the UK Government and the European Commission reached a political agreement on the "Windsor Framework" which will revise the Protocol on Ireland/Northern Ireland in order to address some of the perceived shortcomings in its operation. Under the proposed changes, Northern Ireland would be reintegrated under the regulatory authority of the MHRA with respect to medicinal products. The implementation of the Windsor Framework will occur in various stages, with new arrangements relating to the supply of medicines into Northern Ireland due to take effect in 2025.

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*."

We may find it difficult to enroll participants 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 participants for 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 participants to test 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 participants 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 participants available to us because some participants 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 participants 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 in the future experience delays, or we may be prevented from completing our clinical trials. Participant enrollment depends on many factors, including:

- the size and nature of the patient population;
- the severity of the disease being studied;

- participant referral practices of prescribers;
- participant eligibility criteria for the clinical trial and evolving standards of care;
- the proximity of participants 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 participant consents;
- our ability to recruit clinical trial investigators with applicable competencies and experience;
- the risk that participants 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 participants' 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 participants may prove costly, especially since we are looking to identify a subset of the participants 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 participant enrollment for any other reason, our costs may significantly increase and the timeline for recruiting participants, 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 ("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, including clinical trials evaluating our current products in new indications, 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. Further, third parties may have rights to independently develop and commercialize our current and future products and product candidates, which may increase the likelihood of adverse safety results. For example, Regeneron retains worldwide rights to develop and commercialize ARCALYST for local administration to the eye and ear and oncology, and Huadong holds rights to develop and commercialize ARCALYST and mavrilimumab in the Asia Pacific region, excluding Japan. The development of our product candidates and, if approved, commercialization of our products for new indications or new patient populations by these third parties may increase the possibility of uncovering adverse safety results not previously discovered during our own clinical development process or U.S. commercialization. Such effects, if uncovered by such third parties, may lead to regulatory authorities ordering 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 addition, the compassionate use of our products and product candidates, or evaluation of our products and 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 participant 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 are subject to the risk that one or more of the clinical observations may materially change as participant enrollment continues and more participant 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, 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. The European Commission's proposal for revision of several legislative instruments related to medicinal products (potentially reducing the duration of regulatory data protection, revising eligibility for expedited pathways, etc.) was published in April 2023. The proposed revisions remain to be agreed and adopted by the European Parliament and European Council (not expected before early 2025) and 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 products or product candidates to other therapies for the treatment of the same indication;
- regulatory authorities, following the discovery of adverse safety signals or side effects from approved therapeutics or therapeutics in development in the same or related class as our products or product candidates, could require us to collect additional data or conduct additional clinical trials;

- 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 participants required for clinical trials 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 participants for 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 our candidates 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 or payers may not approve the price we intend to charge, may grant approval contingent on the performance of costly postmarketing clinical trials, may impose certain postmarketing 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 exclusivity and designation in the United States for ARCALYST for the treatment of pericarditis and mavrilimumab for the treatment of GCA, respectively. In addition, we have received Orphan Drug designation in the EU for ARCALYST for the treatment of idiopathic pericarditis. In the future, we may seek Orphan Drug designation for certain of our other product candidates in the United States or 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 medicinal products that are intended for the diagnosis, prevention or treatment of life-threatening or chronically debilitating conditions where either (i) such conditions affect no more than five in 10,000 persons in the EU when the application is made or (ii) the product, without the benefits derived from Orphan Drug status, would not generate sufficient return in the EU to justify investment; and for which no satisfactory method of diagnosis, prevention, or treatment exists for marketing in the EU, or if such method exists, the product will be of significant benefit to those affected by such condition. In the EU, Orphan Drug designation entitles a party to financial incentives such as reduction of fees or fee waivers, and upon grant of an MA, to 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 medicinal product 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 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 EU, 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 may be unable to successfully obtain marketing approvals for any of our current or future product candidates. Failure to 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.

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, which may impair our ability to generate additional revenue. 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 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 recently hit its debt limit, which has caused certain regulatory agencies, such as the FDA, to furlough critical FDA employees and stop critical activities.

Separately, in response to the COVID-19 pandemic, the FDA postponed most inspections of domestic and foreign manufacturing facilities at various points. Even though the FDA has since resumed standard inspection operations of domestic facilities where feasible, the FDA has continued to monitor and implement changes to its inspectional activities to ensure the safety of its employees and those of the firms it regulates as it adapts to the evolving COVID-19 pandemic, and any resurgence of the virus or emergence of new variants may lead to further inspectional delays. Any resurgence of the virus may cause additional postponements, exacerbating the previously discussed risks. Regulatory authorities outside the United States were similarly impacted by the COVID-19 pandemic. If a prolonged government shutdown occurs, or if global health concerns 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; the commercial manufacture of our current and future products; and labeling and packaging activities for our current and future products. We rely on these third parties to produce our products and product candidates at sufficient quality and quantity to support our and our collaboration partners' 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, or not in a timely manner due to, for example, production interruptions caused by equipment failure and an inability to source adequate replacement parts and equipment, which could delay, prevent or impair our commercialization or research and development efforts. From time to time, we have identified events in the ARCALYST manufacturing process that prevented distribution of ARCALYST material as planned, though this has yet to impact our ability to source sufficient ARCALYST material to cover our needs. Further, equipment used in the ARCALYST manufacturing process may no longer be supported by vendors in the event of equipment failure. Such equipment may also not be repaired, replaced or qualified in a timely manner. These issues may be exacerbated by increased clinical or commercial demand by us or our collaboration partners, should we decide to develop ARCALYST in one or more additional indications or in additional territories. 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 future 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.

Regeneron and its CDMOs are the sole manufacturers of ARCALYST and will remain so until we complete the technology transfer of the manufacturing process for ARCALYST drug substance to a new CDMO. 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 CDMOs 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 CDMOs 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.

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

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 CDMO, 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.

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 existing commercial supply or 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 or new manufacturers, which could impact the timing and subsequent success of our planned commercial supply or clinical trials. In addition, as we plan to produce clinical trial and commercial material at a CDMO, the CDMO may be required to adopt different manufacturing protocols or processes. For example, in March 2023, Regeneron formally initiated a technology transfer with respect to the manufacturing process for ARCALYST drug substance. Any CDMO that we select as part of this process may find it necessary to utilize a different manufacturing process than that used by Regeneron, which could require lengthy development, regulatory review and approval. For more information see "*Risk Factors — Risks Related to Manufacturing and Our Reliance on Third Parties — We are conducting a technology transfer with respect to the manufacturing process of ARCALYST drug substance from Regeneron to a new CDMO and the analytical testing methods of ARCALYST drug substance and drug product to new CTLs. Such technology transfer will be subject to significant risks and uncertainties.*"

The facilities used by our CDMOs 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 CDMOs for compliance with cGMPs and other regulatory requirements in connection with the manufacture of current and future products and product candidates. If our CDMOs 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 CDMOs and have the ability to audit their compliance and performance, we have no direct control over the ability of our CDMOs to maintain adequate quality control, quality assurance and qualified personnel other than through quality monitoring in accordance with our agreements with the CDMOs. 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 or any replacement CDMO with respect to the commercial supply of ARCALYST, we must place purchase orders based on projected demand. 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. We may also be required to estimate and order safety stock as part of our planned technology transfer of the manufacturing process for ARCALYST drug substance, which will be subject to a number of the same risks and uncertainties. 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 our current and future products. If our current CDMOs 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 CDMOs 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.

We are conducting a technology transfer with respect to the manufacturing process of ARCALYST drug substance from Regeneron to a new CDMO and the analytical testing methods of ARCALYST drug substance and drug product to new CTLs. Such technology transfer will be subject to significant risks and uncertainties.

In March 2023, Regeneron, our sole supplier of ARCALYST drug substance, initiated a technology transfer related to the manufacturing process of ARCALYST drug substance and the analytical testing methods of ARCALYST drug substance and drug product. We plan to collaborate with Regeneron to qualify and contract with a new CDMO who will serve as the new manufacturer of ARCALYST drug substance and new contract testing labs ("CTLs") who will serve as the new testing labs of ARCALYST drug substance and drug product.

Pharmaceutical development, manufacture and analytical testing requires significant expertise and capital investment, and the manufacture and testing of biologics, in particular, can be complex and difficult. While we have selected a replacement CDMO and replacement CTLs, we are still in the early stages of the technology transfer process and still must determine whether such CDMO and CTLs can meet our requirements regarding production costs and yields, process controls, quality control, quality assurance, data integrity and cGMP compliance, among other factors. We would also need to source sufficient raw materials to facilitate new manufacturing and analytical testing, which may be affected by supply chain disruptions, materials shortages or an inability to negotiate satisfactory terms with suppliers.

The technology transfer process is a time-consuming and difficult task that may require significant time and focus from our management and technical teams. Further, because of the complexities of this process, the technology transfer may be subject to substantial delay, which could materially harm our business and operations.

Because such CDMO would be manufacturing ARCALYST drug substance at a new manufacturing site and with a potentially different manufacturing process, and such CTLs would be testing ARCALYST drug substance and drug product at new testing sites and potentially with different testing methods, we expect that the FDA will need to approve such changes before we are able to complete the technology transfer. The FDA generally requires that any new CDMO be able to manufacture drug substance at sufficient levels of comparability with the materials produced by the original manufacturer. Failure to provide sufficient evidence of comparability may result in the FDA requesting a bioequivalence or pharmacokinetic study, which would delay our expected technology transfer timeline. Even if such study were to be performed, there is no guarantee that the FDA would accept our findings and approve any new facilities for the manufacture of ARCALYST drug substance.

Regeneron is contractually obligated to continue manufacturing ARCALYST drug substance for at least a portion of the time that it will take to qualify a replacement CDMO. During such time, Regeneron will remain subject to many of the risks described elsewhere in this "*Risk Factors*" section, including the risk that it is unable to manufacture sufficient quantities of ARCALYST and at sufficient quality to meet ours and our patients' and collaborators' needs. Further, because we expect the timeline for any successful technology transfer to extend beyond Regeneron's contractual obligations, our ability to meet patient demand will depend significantly on whether we can secure sufficient safety stock from Regeneron, negotiate continued ARCALYST drug substance manufacture by Regeneron beyond its contractual obligations or some combination thereof. Purchasing significant amounts of safety stock would require substantial upfront capital investment and, if the technology transfer process is delayed beyond our expectation, such safety stock may expire or be depleted before a new CDMO can begin manufacturing ARCALYST drug substance. Regeneron may also disagree with our forecasted safety stock requirements and manufacture less ARCALYST drug substance than we request, exposing us to risks if the process is significantly delayed. Any arrangement that we negotiate with Regeneron to manufacture ARCALYST beyond their contractual obligations may not be on as favorable terms as our current relationship, which could materially increase our costs and as a result negatively impact our financial condition and results of operations. A failure to secure sufficient safety stock or negotiate satisfactory manufacturing terms with Regeneron could result in supply shortages for our patients and collaborators while we work to complete the technology transfer.

A failure to either complete our planned technology transfer on our expected timeline or at an acceptable cost and/or secure sufficient supply of ARCALYST through the technology transfer process would have a material impact on our business, financial condition and results of operations.

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 cleanup 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, if approved, and adversely affect our business.

The manufacture of our current and future products and 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, KPL-404 and mavrilimumab. Due to the highly technical requirements of manufacturing our current and future products and 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 our current and future products and 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 our current and future products and 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 and an epidemic or pandemic or other outbreak of disease. 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*."

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 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 our products and 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 is currently our sole source manufacturer, but with its initiation of a technology transfer of the manufacturing process for ARCALYST drug substance in March 2023, will cooperate with us to qualify a suitable replacement CDMO. For more information see "*Risk Factors — Risks Related to Manufacturing and Our Reliance on Third Parties — We are conducting a technology transfer with respect to the manufacturing process of ARCALYST drug substance from Regeneron to a new CDMO and the analytical testing methods of ARCALYST drug substance and drug product to new CTLs. Such technology transfer will be subject to significant risks and uncertainties.*" 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. Successful completion of a technology transfer of the manufacturing process for ARCALYST drug substance will be integral to our ability to meet such requirements. With respect to ARCALYST and mavrilimumab, 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, in the future, enter into collaboration and other agreements requiring us to provide commercial or clinical drug supply to third party partners. This includes additional vixarelimab drug substance that Genentech may request from us pursuant to the terms of the Genentech License Agreement. A failure by our CDMOs 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 CDMOs 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 CDMOs 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 studies and 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 participants 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 a pandemic or other outbreak of disease or as the result of war, conflict or terrorism as with the ongoing war in Ukraine;
- 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, 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 we 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; 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. Clinical stage development programs include: VTX2735 and VTX3232 (Ventyx Biosciences, Inc.), ZYIL-1 (Zydus Lifesciences Ltd.), HT-6184 (Halia Therapeutics, Inc.), OLT1177 (Olatec Therapeutics LLC), DFV-890 (Novartis A.G.), Selnoflast (Roche), NT-0167 and NT-0796 (NodThera Ltd.), Somalix and Onzomelid (Roche).

In addition, there are several molecules in development designed to inhibit the NLRP3 inflammasome, an intracellular sensor of a broad range of danger signals, that leads to the release of IL-1β and IL-18. Clinical stage development programs include: VTX2735 and VTX3232 (Ventyx Biosciences), ZYIL-1 (Zydus Lifesciences), HT-6184 (Halia), OLT1177 (Olatec Therapeutics), DFV-890 (Novartis), Selnoflast (Roche), NT-0167 and NT-0796 (NodThera), Somalix and Onzomelid (Roche). 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 various programs in clinical development antagonizing the CD40 / CD154 costimulatory pathway. Novartis A.G. is developing CFZ-533, or iscalimab (anti-CD40) for subcutaneous administration for the treatment of Sjögren's Syndrome. Certain other programs are designed for intravenous administration only. Horizon Therapeutics plc (in the process of being acquired by Amgen Inc.) is developing the Tn3 fusion protein, dazodalibep (anti-CD40L); Biogen, Inc. and UCB S.A. are developing dapirolizumab pegol (anti-CD40L) for the treatment of moderately to severely active Systemic Lupus Erythematosus; and Eledon Pharmaceuticals, Inc. is developing AT-1501 (anti-CD40L) for use by patients undergoing kidney transplantation. There are other programs that present the potential for subcutaneous administration. Sanofi S.A./ImmuNext Inc. are developing frexalimab (anti-CD40L) for the treatment of Multiple Sclerosis, Primary Sjögren's Syndrome and Systemic Lupus Erythematosus, Bristol Myers-Squibb is developing BMS-986325 (anti-CD40) for the treatment of Primary Sjögren's Syndrome; and H. Lundbeck A/S is developing Lu AG22515 (bi-specific, anti-CD40L & Albumin (scFv)2-Fab).

With respect to mavrilimumab, there are programs in clinical development in various indications that modulate GM-CSF signaling from I-MAB Biopharma Co. Ltd. (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.

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 participant 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. Further, a competitor conducting a clinical trial in a rare disease indication for which we market a product may reduce the number of patients on our commercial therapy by recruiting such patients to be trial participants. Our competitors also may obtain FDA or other regulatory approval and/or marketing exclusivity for their products more rapidly than we may obtain 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 certain of 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;
- we may incur substantial liabilities as part of an acquisition or merger that may not be offset by the benefits of the acquired assets or the synergies we hope to realize; 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 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, and any such transactions or arrangements that we 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 collaborations, 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 not achieve applicable development, regulatory, or commercial milestones, which may materially impact the collaboration revenue that we expect to realize from such relationship

- may delay, dispute or refuse to pay milestone and royalty payments, which may 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, KPL-404 and mavrilimumab. 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 BIDMC 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 we or our licensees may not pursue or maintain in the future, patent protection for our products or product candidates in every country or territory in which our products or product candidates may be sold, 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 (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 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 products and 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, KPL-404, mavrilimumab, 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 in 2023, not including any patent term extensions. A U.S. patent covering methods of using ARCALYST in the treatment of recurrent pericarditis was 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 owned by us have statutory expiration dates in 2036, not including any extensions. The issued composition of matter patents licensed from BIDMC 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 (supplementary protection certificate) 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 or our licensees 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 or our licensees are unable to obtain patent term extension or the term of any such extension is less than requested, the period during which our patent rights can be enforced for that product will be shortened and 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 or our licensees 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 adequate and continuing patent protection sufficient to exclude others from commercializing products similar or identical to our product candidates. In such cases, regulatory exclusivity is expected to be relied upon for our or our licensees' 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 or our licensees 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 even then, 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 or our licensees 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 the date our inventions were invented, or may file patent applications before we or our licensees do. In such case, we or our licensees 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 or our licensees 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, product candidates 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 or our licensees. 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, our licensees or our licensors were the first to file any patent application related to our product and 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, our licensees' 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, our licensees' 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 or our licensees' ability to stop others from using or commercializing similar or identical technology and products, without payment to us, could limit the duration of the patent protection covering our technology, product and product candidates, or could reduce the period of time during which our licensees are obligated to make royalty payments to us for the sale of licensed products. Such challenges may also result in our inability to manufacture or commercialize our product and 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 or our licensees 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 our product or 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 or 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 or our licensees' ability to successfully commercialize our product or 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 or product candidates, we could lose the ability to continue the development and commercialization of the related product or product candidate. 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, KPL-404 and mavrilimumab. 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 BIDMC 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 patents and other rights to third parties;
- the ownership of inventions, know-how and other intellectual property, including intellectual property rights resulting from the joint creation or use of intellectual property by us and our licensors, licensees, partners or collaborators;
- 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 in-licensed, out-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, or our sublicensees cause us to fail, to meet our obligations under our agreements in a material respect, the respective licensor/seller would have the right to terminate the respective agreement. We then not only would have to return the licensed technology, but we may also be required to grant the licensor rights to any intellectual property controlled by us and developed during the period the agreement was in force that relate to the applicable licensed technology. This means that the licensor/seller for each of these agreements could effectively take control of the

development and commercialization of our product and 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, or our sublicensees cause us to fail, to meet our obligations under these agreements in any material respect, including seeking to cure any breach by us or our sublicensees, 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 in-licenses could result in our loss of exclusive rights and may lead to a complete termination of our product development and any commercialization efforts for our product and 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 or product candidate could impair our ability to raise additional capital, generate revenue and may significantly harm our business, financial condition and prospects.

Additionally, under the Regeneron Agreement, Regeneron retains worldwide rights to develop and commercialize ARCALYST for local administration to the eye and ear and oncology and 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 identifying adverse safety results that may impact the commercialization of ARCALYST for the treatment of recurrent pericarditis in our territory.

We have also entered into agreements to grant to others licenses under our owned intellectual property and sublicenses under intellectual property that we license from others for those third parties to develop and commercialize ARCALYST, mavrilimumab and vixarelimab, including the Collaboration Agreements with Huadong and the Genentech License Agreement. Under each of these agreements, our licensees have certain responsibilities to develop and commercialize the applicable licensed drugs, make timely milestone and royalty payments, provide to us certain information regarding their activities and indemnify us with respect to their development and commercialization activities under the terms of the agreements. Additionally, under the Genentech License Agreement, we granted Genentech the first right to file, prosecute, maintain, defend, enforce and extend the life of the patents that we own and licensed to Genentech. Our licensees or collaborators may:

- have significant discretion in determining the efforts and resources that they will apply to our collaboration;
- not pursue development and commercialization of the applicable licensed drugs or may elect not to continue or renew development or commercialization programs based on clinical trial results, changes in their strategic focus due to their acquisition of competitive products or product candidates or their internal development of competitive products and product candidates, availability of funding or other external factors, such as a business combination that diverts resources or creates competing priorities;
- delay clinical trials, provide insufficient funding for a clinical trial program, stop a clinical trial, abandon a product candidate, repeat or conduct new clinical trials or require a new formulation of a product candidate for clinical testing;
- independently develop, or develop with third parties, products or product candidates that compete directly or indirectly with our product or product candidates;
- not commit sufficient resources to or otherwise not perform satisfactorily in carrying out their contractual obligations;
- not properly maintain or defend our intellectual property rights or may use our intellectual property or proprietary information in a way that gives rise to actual or threatened litigation that could jeopardize or invalidate our intellectual property or proprietary information or expose us to potential liability;
- raise disputes that cause the delay or termination of the research, development or commercialization of our current or future products and product candidates or that results in costly litigation or arbitration that diverts management attention and resources;

- own or co-own intellectual property covering products that result from our collaboration with them, and in such cases, we would not have the exclusive right to develop or commercialize products covered by such intellectual property;

- raise disputes with respect to the ownership or inventorship of any intellectual property developed pursuant to our collaborations or licenses;

- conduct sales and marketing activities or other operations that may not be in compliance with applicable laws resulting in civil or criminal proceedings;

- fail to pay us milestone and royalty payments when due;

- cause us to be named defendants in lawsuits due to their improper use of the licensed intellectual property and not indemnify us against losses in such lawsuits;

- enforce licensed intellectual property rights against third parties that lead such third parties to challenge the validity or enforceability of the licensed intellectual property and potentially cause the licensed intellectual property to become invalid or rendered unenforceable;

- fail to maintain issued licensed patents that are under their control, or prosecute licensed patent applications in ways that diminish their value, all of which actions may adversely affect our business if our agreements with them terminate and the rights to the licensed intellectual property return to us or an upstream licensor; and

- breach their agreements with us, which may lead us to breach our obligations with our upstream licensors, potentially causing us to lose our upstream licenses.

As such, the occurrence of any of these events with respect to our licensing and collaboration arrangements may have an adverse effect on our business.

Finally, 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 sublicensees 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 KPL-404 and mavrilimumab. If the claims of any of these patents are asserted against us, we do not believe our proposed activities related to KPL-404 and mavrilimumab 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 licensees 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 or our licensees 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 or licensees 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 or our licensees 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 our product or one of our product candidates, we or our licensees 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 or our licensees lose a patent lawsuit outside of the United States, alleging our infringement of a competitor’s patents, we or our licensees could be prevented from marketing our current or future products and product candidates 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 or our licensees 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 or our licensees may not be able to prevent third parties from infringing upon or misappropriating or from successfully challenging our intellectual property rights. Uncertainties resulting from the initiation and continuation of patent litigation or other proceedings could have a material adverse effect on our ability to compete in the marketplace.

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

The USPTO and various governmental patent agencies outside of the United States require compliance with a number of procedural, documentary, fee payment and other similar provisions during the patent application process. In addition, periodic maintenance fees on issued patents often must be paid to the USPTO and patent agencies outside of the United States over the lifetime of the patent. While an unintentional lapse can in many cases be cured by payment of a late fee or by other means in accordance with the applicable rules, there are situations in which noncompliance can result in abandonment or lapse of the patent or patent application, resulting in partial or complete loss of patent rights in the relevant jurisdiction. Non-compliance events that could result in abandonment or lapse of a patent or patent application include, but are not limited to, failure to respond to official actions within prescribed time limits, non-payment of fees and failure to properly legalize and submit formal documents. If we or our licensees fail to appropriately file and prosecute patent applications covering the licensed products, product candidate or technologies, and maintain any patent issuing from such patent applications, we or our licensees 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 and 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 or our licensees may not be able to prevent third parties from practicing inventions covered by our patents, whether owned or in-licensed, in all countries outside the United States. Competitors may use our or technologies in jurisdictions where we or they have not obtained patent protection, or where we or they 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 or our licensees' ability to enforce our patents to stop infringing activities is inadequate. These products may compete with our products and product candidates or the products and product candidates that we have out-licensed, and our or our licensees' 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 and product candidates, we cannot ensure that we or our licensees will be able to initiate or maintain similar efforts in all jurisdictions in which we or they may wish to market our or our out-licensed products and product candidates. Accordingly, our or our licensees' 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 the ability to obtain and enforce adequate intellectual property protection for our technology.

In addition, geo-political actions in the United States and in foreign countries could increase the uncertainties and costs surrounding the prosecution or maintenance of our patent applications or those of any current or future licensors and the maintenance, enforcement or defense of our issued patents or those of any current or future licensors. For example, a decree was adopted by the Russian government in March 2022, allowing Russian companies and individuals to exploit inventions owned by patentees from the United States without consent or compensation. Consequently, we would not be able to prevent third parties from practicing our inventions in Russia or from selling or importing products made using our inventions in and into Russia. Accordingly, our competitive position may be impaired, and our business, financial condition, results of operations and prospects may be adversely affected.

Changes to patent laws in the United States and other jurisdictions could diminish the value of patents in general, thereby impairing our ability to protect our product or our current or future 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 (the "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 or our licensees 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 or 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 or our licensees 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 or our licensees 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 or our licensees' 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 or our licensees' 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 licensees' 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 or our licensees' ability to obtain or maintain patent protection for our or our out-licensed proprietary technology or our or their ability to enforce our or our out-licensed proprietary technology, respectively.

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 or our licensees' existing patents and patents that we might obtain in the future; shorten the term that has been lengthened by patent term adjustment of our or our licensees' 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.

Finally, Europe's new Unitary Patent system and Unified Patent Court (the "UPC") may present uncertainties for our ability to protect and enforce our patent rights against competitors in Europe. In 2012, the European Patent Package (the "EU Patent Package"), regulations were passed with the goal of providing a single pan-European Unitary Patent system and a new UPC, for litigation involving European patents. Implementation of the EU Patent Package occurred in June 2023. Under the UPC, all European patents, including those issued prior to ratification of the European Patent Package, will by default automatically fall under the jurisdiction of the UPC. The UPC will provide our competitors with a new forum to centrally revoke our European patents and allow for the possibility of a competitor to obtain pan-European injunctions. It will be several years before we will understand the scope of patent rights that will be recognized and the strength of patent remedies that will be provided by the UPC. Under the EU Patent Package we will have the right to opt our patents out of the UPC over the first seven years of the court's existence, but doing so may preclude us from realizing the benefits of the new unified court.

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 or 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, leading to regional devastation and global disruptions. Additionally, in the wake of the invasion, the United States, the EU, the UK and other countries implemented sanctions, export controls and other measures against Russia and Russian-aligned parties. Such sanctions and other measures, as well as the existing and potential future 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 conflict in Ukraine has adversely affected 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 were able to secure alternative clinical trial sites in other European countries, there is no guarantee that such sites will be able to provide a sufficient number of participants to satisfy our clinical need and at a reasonable cost. Further, should the conflict or externalities related to the conflict extend beyond Ukraine and we are forced to terminate or suspend ongoing or planned clinical trial operations, 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 CDMOs, 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*."

A global pandemic, such as the COVID-19 pandemic, and measures taken in response to such pandemic, could have an adverse impact that is significant on our business and operations as well as the business or operations of the third parties with whom we conduct business or otherwise engage, which may have a material adverse effect on our business, operations and financial position.

Global pandemics, such as the COVID-19 pandemic, and measures taken in response to such pandemics, 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.

In the past, governmental authorities around the globe implemented 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, where we conduct our clinical trials or where we otherwise conduct business or engage with other third parties. In addition, the COVID-19 had a direct impact on our business and operations by, among other things:

- disrupting global supply chains for our products and product candidates, the raw materials required for manufacturing our products and product candidates and important ancillary products needed to administer our products and product candidates;

- causing disruptions, staffing shortages, production slowdowns, stoppages or reprioritizations at the third party CDMOs that we rely on to produce our products and product candidates;

- impeding clinical trial activities, including activities related to enrolling and monitoring our clinical participants;
- limiting our ability to access third party payors, prescribers and patient advocacy groups to build disease awareness;
- limiting our workforce's ability to collaborate in-person at our facilities; and
- causing disruption and volatility in U.S. and global capital markets.

Such impacts were also felt by a number of the third parties with whom we interact, which further affected our business and operations. In the event that a new global pandemic emerges, or a new variant of the COVID-19 pandemic emerges, we may be subject to the same or similar restrictions and adverse events. We cannot ultimately predict the scope and severity of any such future event; however, such events may be severe and have a material impact on our business, results of operations and financial condition.

The UK's withdrawal from the EU may have a negative effect on our business.

Since January 1, 2021 the UK has operated under a distinct regulatory regime to the EU, with EU pharmaceutical laws applying in respect of the UK only in Northern Ireland (as set out in the Protocol on Ireland/Northern Ireland). In February 2023, however, the UK Government and the European Commission reached a political agreement on the "Windsor Framework" which will revise the Protocol on Ireland/Northern Ireland in order to address some of the perceived shortcomings in its operation. Under the proposed changes, Northern Ireland would be reintegrated under the regulatory authority of the MHRA with respect to medicinal products. The implementation of the Windsor Framework will occur in various stages, with new arrangements relating to the supply of medicines into Northern Ireland due to take effect in 2025.

EU laws which have been transposed into UK law through secondary legislation continue to be applicable as "retained EU law". 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 ("TCA"), which put in place a framework for a future EU-UK relationship, 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, until new legislation has been published, it remains uncertain as to what extent the UK legislation will align with the 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 MHRA launched an eight-week consultation on reframing the UK legislation for clinical trials, with the aim 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 resulting new legislation will determine how aligned the UK clinical trials regime is compared to the CTR. A decision by the UK not to closely align any new legislation with the new approach that has been adopted in the EU may have an effect on the cost of conducting clinical trials in the UK as opposed to other countries.

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 new 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. Any of these factors could have a significant adverse effect on our business, financial condition, results of operations and prospects.

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.

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 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, introduced 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 2032 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. The impact of the IRA on our business and the pharmaceutical industry cannot yet be fully determined but 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. 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 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.

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 which may have a material impact on our business and operations.

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. 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 CDMOs, 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 CDMOs, 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. We may also 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 CDMO, 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 CDMOs, 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 cyberattacks 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, directly from individuals (or their healthcare providers) who enroll in our patient support program and directly from individuals who consent to be included in our marketing database. 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, the California Consumer Privacy Act (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 has increased the likelihood of, and the risks associated with data breach litigation. Further, the California Privacy Rights Act (the "CPRA") generally went into effect on January 1, 2023 and significantly amends the CCPA. It imposes 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 also creates a new California data protection agency authorized to issue substantive regulations and could result in increased privacy and information security enforcement. Similar laws have been passed in other states and are continuing to be proposed at the state and federal level, reflecting a trend toward more stringent privacy legislation in the United States. Additional compliance investment and potential business process changes may also be required.

Furthermore, the Federal Trade Commission ("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 GDPR, and legislation of EU member states and European Economic Area ("EEA") countries implementing it. The GDPR imposes strict requirements for processing the personal data of individuals within the EEA. In addition, some of the personal data we process in respect of clinical trial participants is special category or sensitive personal data under the GDPR, and subject to additional compliance obligations and to local law derogations. 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. In addition to fines, a breach of the GDPR may result in regulatory investigations, reputational damage, orders to cease/ change our data processing activities, enforcement notices, assessment notices (for a compulsory audit) and/ or civil claims (including class actions).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.

Case law from the Court of Justice of the European Union ("CJEU") states that reliance on the standard contractual clauses (a standard form of contract approved by the European Commission as an adequate personal data transfer mechanism) alone may not necessarily be sufficient in all circumstances and that transfers must be assessed on a case-by-case basis. We currently rely on the EU standard contractual clauses and the UK Addendum to the EU standard contractual clauses, as applicable, to transfer personal data outside the EEA and the UK, including to the United States, with respect to both intragroup and third party transfers. Following a period of legal complexity and uncertainty regarding international personal data transfers, particularly to the United States, we expect the regulatory guidance and enforcement landscape to continue to develop, in relation to transfers to the United States and elsewhere. As a result, we may have to make certain operational changes and we will have to implement revised standard contractual clauses and other relevant documentation for existing data transfers within required time frames.

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. As we continue to expand into other foreign countries and jurisdictions, we may be subject to additional laws and regulations that may affect how we conduct business.

Failure or perceived failure to comply with the GDPR, the UK GDPR 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 CDMOs, 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 CDMOs, 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, be incorrectly perceived as product promotion, lead to the loss of trade secrets or other intellectual property or result in public exposure of personal information of our employees, clinical trial participants, customers and others or information regarding our product candidates or clinical trials. Clinical trial participants 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; attract, retain and motivate qualified personnel; and implement succession planning efforts to ensure our long-term success.

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, sales, marketing and clinical personnel from other pharmaceutical companies, universities and research institutions, as applicable. 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.

Effective succession planning is also important to our long-term success and ability to operate as a generational company. As we encounter employee turnover, including turnover of key personnel, we may be unable to timely train or locate replacement personnel in a way that delays our strategic planning and clinical and commercial execution.

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 and evolving focus on environmental, social and governance ("ESG") matters could increase our costs, harm our reputation, adversely impact our access to capital and financial results or otherwise adversely impact our business.

There has been increasing and evolving public focus by investors, patients, environmental activists, the media and governmental and nongovernmental organizations on a variety of ESG matters, such as climate change and diversity, equity and inclusion matters. We may experience pressure from stakeholders, including our suppliers, employees, patients and shareholders, to set goals or make commitments relating to ESG matters that affect us, including the design and implementation of specific risk mitigation strategic initiatives relating to ESG topics. If we are not effective in addressing ESG matters affecting our business, or setting and meeting relevant ESG 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 ESG 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 ESG matters has resulted in the adoption of new laws and regulations, including new reporting requirements, and may result in the adoption of additional laws and regulations in the future. 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. Such ESG matters may also impact our suppliers or patients, which may adversely impact our business, financial condition and results of operations.

In addition, organizations that provide information to investors on corporate governance and related matters have developed ratings processes for evaluating companies on ESG matters. Such ratings are used by some investors to inform their investment or voting decisions. Unfavorable ESG ratings could lead to negative investor sentiment toward us and/or our industry, which could have a negative impact on our access to and costs of capital. To the extent ESG matters negatively impact our reputation, we may be affected in a number of ways, including an inability to recruit and retain personnel and a decrease in the trading price of our Class A common shares.

Climate change, and related regulation, may result in increased costs or otherwise negatively impact our operations and harm our business.

The impacts of climate change on the global economy and our industry are rapidly evolving. Physical impacts of climate change (including but not limited to floods, droughts, more frequent and/or intense storms and wildfires), could negatively impact our business and operations, as well as the business and operations of our third party CDMOs and CROs upon whom we rely. Such events may result in damage or loss of our products and product candidates during their manufacture and shipment, cause delays in clinical development due to trial site disasters or result in losses of critical data, any of which may adversely impact our operations. An evolving climate may also result in uncertain and potentially onerous regulatory requirements as agencies and governmental authorities adjust, such as new or changed emissions reporting and auditing requirements. Failure to comply with such requirements in a timely manner may adversely affect our reputation, business, or financial performance.

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 June 30, 2023, 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 June 30, 2023 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 June 30, 2023, 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 may be volatile and fluctuate substantially, which could result in substantial losses for holders of our Class A common shares.

Our share price may be subject to change as a result of volatility in the stock market driven by events often unrelated to our operating performance. 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 pandemics or other outbreaks of disease 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.

Market conditions are often difficult to predict and there can be no assurance as to the performance of our Class A common shares or 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 about us 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 June 30, 2023, approximately 2.0 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 (the "Securities Act"), and such rule, Rule 144. In addition, as of June 30, 2023, there were approximately 12.9 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.

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 June 30, 2023, on an as-converted to Class A common shares basis, these shareholders collectively held approximately 33.8 million of our Class A common shares. If any of these shareholders sell, convert or transfer, or indicate an intention to sell, convert or transfer, a substantial amount of their common shares (after certain restrictions on conversion or resale lapse), the market price of our Class A common shares could decline.

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 June 30, 2023, 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 1.8 million Class A common shares held by certain of our executive officers as of June 30, 2023. 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 currently benefit from the reduced disclosure requirements applicable to "smaller reporting companies", which may make our Class A common shares less attractive to investors.

As a result of our public float (the market value of our voting and non-voting common shares held by non-affiliates) as measured on the last business day of our second fiscal quarter, we will be required to comply with new, increased disclosure obligations beginning with our first quarterly report for the three months ended March 31, 2024. Until such time, we may avail ourselves of the reduced disclosure requirements available to "smaller reporting companies", as defined under the rules promulgated under the Securities Act.

For so long as we may utilize the reduced disclosure rules applicable to smaller reporting companies, we are permitted and intend to rely on exemptions from certain disclosure requirements that are applicable to other public companies that are 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 ("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 we expect to increase in connection with our exit from smaller reporting company status. 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, and we will be required to include an attestation report on internal control over financial reporting issued by our independent registered public accounting firm. To maintain compliance with Section 404 within the prescribed period, we engage in a process to document and evaluate our internal control over financial reporting, which is both costly and challenging. In this regard, we will need to continue to dedicate internal resources, engage outside consultants and refine and revise a detailed work plan to assess and document the adequacy of internal control over financial reporting, continue steps to improve control processes as appropriate, validate through testing that controls are functioning as documented and implement a continuous reporting and improvement process for internal control over financial reporting. Despite our efforts, there is a risk that neither we nor our independent registered public accounting firm will be able to conclude that our internal control over financial reporting is effective as required by Section 404. This could result in an adverse reaction in the financial markets due to a loss of confidence in the reliability of our financial statements.

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

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

- a classified board of directors with staggered three-year terms;
- directors only to be removed for cause;
- an affirmative vote of 66 2/3% of the voting power of our voting shares for certain "business combination" transactions that have not been approved by our board of directors;
- our 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 (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 in the future, it could adversely impact our tax position and our effective tax rate. 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.

We may be treated as a passive foreign investment company ("PFIC") for U.S. federal income tax purposes. 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 as a PFIC for the taxable year ending December 31, 2022. 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, 2023, and do not believe that the Company or its subsidiaries will be treated as a PFIC for the taxable year ending December 31, 2023. 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, 2023. 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" 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, Use of Proceeds and Issuer Purchases of Equity Securities

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	Sixth Amendment of Sublease, dated May 24, 2023, 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

SIGNATURES

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

KINIKSA PHARMACEUTICALS, LTD.

Date: August 1, 2023

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

Exhibit 10.1

SIXTH AMENDMENT OF SUBLEASE

SIXTH AMENDMENT OF SUBLEASE (the "Sixth Sublease Amendment") made as of the 24th day of May, 2023, 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, by Fifth Amendment of Sublease dated as of July 27, 2022 between Landlord and Kiniksa (the "Fifth Sublease Amendment"), Landlord and Kiniksa extended the Term of the Sublease, upon the terms set forth in such Fifth Sublease Amendment;

WHEREAS, the Term of the Sublease is scheduled to expire on August 31, 2024 (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 Sixth Sublease Amendment; and

WHEREAS, Landlord and Kiniksa are entering into this Sixth 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 Sixth Sublease Amendment is scheduled to expire on August 31, 2024, is hereby extended for the period commencing on September 1, 2024 and expiring on August 31, 2028 (the "Third 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 Sixth Sublease Amendment (the Sublease, as modified and extended by this Sixth Sublease Amendment is herein referred to as the "Amended Sublease").

2. Monthly Base Rent shall be payable by Kiniksa to Landlord during the Third Extended Term as follows:

   a. from September 1, 2024 through August 31, 2025 in the monthly amount of $233,016.67.
   b. from September 1, 2025 through August 31, 2026 in the monthly amount of $240,007.17.
   c. from September 1, 2026 through August 31, 2027 in the monthly amount of $247,230.68.
   d. from September 1, 2027 through August 31, 2028 in the monthly amount of $254,640.61.

3. For the sake of clarity, Kiniksa shall have no further option to extend the Term of the Sublease beyond the Third Extended Term.

4. (a) Landlord shall provide to Tenant a special allowance equal to the product of (i) $17.50 and (ii) the Rentable Floor Area of the Premises (the "Sixth Amendment Tenant Allowance"). The Sixth Amendment Tenant Allowance shall be used and applied by Tenant solely on account of the cost of work performed by Tenant in accordance with the terms of the Sublease as herein amended ("Tenant's Work"). Provided, in each instance of Tenant's Work, that the Tenant (i) has completed all of such Tenant's Work in accordance with the terms of the Amended Sublease, (ii) has paid for all of such Tenant's Work in full and has delivered to Landlord lien waivers from all persons who might have a lien as a result of the Tenant's Work, in the recordable forms acceptable to Landlord, (iii) has delivered to Landlord its certificate specifying the cost of such Tenant's Work and all contractors, subcontractors and suppliers involved with Tenant's Work, together with evidence of such cost in the form of paid invoices, receipts and the like, (iv) has delivered to Landlord a final set of record drawings for Tenant's Work, (v) has satisfied the requirements of (i) through (iv) above and made request for such payment on or before the date that is August 31, 2026, (vi) is not otherwise in default under the Amended Sublease, and (vii) there are no liens (unless bonded to the reasonable satisfaction of Landlord) against Tenant's interest in the Amended Sublease or against the Building arising out of Tenant's Work, then within thirty (30) days after the satisfaction of the foregoing conditions, the Landlord shall pay to the Tenant the lesser of the amount of such costs so certified or the amount of the Sixth Amendment Tenant Allowance. For the purposes hereof, the cost to be so reimbursed by Landlord shall include the cost of leasehold improvements but not the cost of any of Tenant's personal property, trade fixtures or trade equipment, moving expenses or any so-called soft costs except that Tenant may utilize up to a maximum of fifteen percent (15%) (the "Cap Amount") of the Sixth Amendment Tenant Allowance towards wiring and cabling in the Premises and architectural, engineering and design fees associated with Tenant's Work (the "Special Costs") so long as the conditions to application of the Sixth Amendment Tenant Allowance set forth above have been satisfied (including, without limitation, the requirement that Sixth Amendment Tenant Allowance be utilized on or before August 31, 2026. Landlord shall pay to Tenant the lesser of the amount of such costs so certified or the Cap Amount within thirty (30) days after the satisfaction of the foregoing conditions.

   (b) Notwithstanding the foregoing, Landlord shall be under no obligation to apply any portion of the Sixth Amendment Tenant Allowance for any purposes other than as provided in this Section 4 nor shall Landlord be deemed to have assumed any obligations, in whole or in part, of Tenant to any contractors, subcontractors, suppliers, workers or materialmen. Further, in no event shall Landlord be required to make application of any portion of the Sixth Amendment Tenant Allowance on account of any supervisory fees, overhead, management fees or other payments to Tenant, or any partner or affiliate of Tenant. In the event that such cost of Tenant's Work is less than the Sixth Amendment Tenant Allowance, Tenant shall not be entitled to any payment or credit nor shall there be any application of the same toward Monthly Base Rent or Additional Rent owed by Tenant under the Amended Sublease. Landlord shall be entitled to deduct from the Sixth Amendment Tenant Allowance any Landlord Plan Review Costs associated with Tenant's Work as defined and set forth in Section 14(l) of the Consent to Sublease dated March 13, 2018 among Landlord, Original Tenant and Kiniksa.

5. (a) Tenant warrants and represents that Tenant has not dealt with any broker in connection with the consummation of this Sixth 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.

(b) Landlord warrants and represents that Landlord has not dealt with any broker in connection with the consummation of this Sixth 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 Sixth Sublease Amendment it has no demands, causes of action, claims or other actions against the other under the Sublease.

7. This Sixth 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 Sixth 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 Sixth 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

By: /s/ Patrick 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: SVP, General Counsel

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
10. Recognition and Attornment Agreement and Amendment of Sublease between Landlord and Kiniksa dated as of November 6, 2020
11. Fifth Amendment of Sublease dated July 27, 2022 between Landlord and Kiniksa

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.

August 1, 2023

/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.

August 1, 2023

/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 June 30, 2023 (the "Report") fully complies with the requirements of Section 13(a) or 15(d) of the Securities Exchange Act of 1934, as amended; and

(2) The information contained in the Report fairly presents, in all material respects, the financial condition and results of operations of the Company.

August 1, 2023

/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 June 30, 2023 (the "Report") fully complies with the requirements of Section 13(a) or 15(d) of the Securities Exchange Act of 1934, as amended; and

(2) The information contained in the Report fairly presents, in all material respects, the financial condition and results of operations of the Company.

August 1, 2023

/s/ Mark Ragosa
Mark Ragosa
Chief Financial Officer
(Principal Financial Officer)