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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, 2025**

**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 International, plc

(Exact Name of Registrant as Specified in Its Charter)

| **England and Wales** | **98-1795578** |
|---|---|
| (State or Other Jurisdiction of Incorporation or Organization) | (I.R.S. Employer Identification No.) |

23 Old Bond Street, Floor 3
London, W1S 4PZ
England, United Kingdom
(781) 431-9100
**(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)**

N/A

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 Ordinary 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 25, 2025, there were 74,107,668 ordinary shares outstanding in aggregate, comprised of:

43,472,928 Class A ordinary shares, nominal value $0.000273235 per share

1,795,158 Class B ordinary shares, nominal value $0.000273235 per share

12,781,964 Class A1 ordinary shares, nominal value $0.000273235 per share

16,057,618 Class B1 ordinary shares, nominal value $0.000273235 per share

**Kiniksa Pharmaceuticals International, plc**

**FORM 10-Q**

**FOR THE THREE MONTHS ENDED JUNE 30, 2025**

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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 commercial strategy; potential value drivers; potential indications; potential market opportunities and competitive position; ongoing, planned and potential clinical trials and other studies; timing and potential impact of clinical data; future results of operations and financial position; expected timeline for our cash, cash equivalents and short-term investments; product development; prospective products and product candidates; supply of drug products at acceptable cost and quality; collaborators, license and other strategic arrangements; the expected timeline for achievement of our clinical milestones; potential marketing authorizations from the United States Food and Drug Administration (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;
- competitive and potentially competitive products and technologies;
- our ability to source sufficient quantities of our products and product candidates to meet patient and partner demand at acceptable cost and quality specifications;
- our ability to successfully complete the technology transfer of the manufacturing process for ARCALYST drug substance;
- 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;
- the decision of third party payors not to cover or maintain coverage of or to establish burdensome requirements prior to covering or maintaining coverage of ARCALYST or any of our current or future 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 permit clinical development of, to grant regulatory exclusivity for and to approve marketing and sale of our current and future product candidates;
- our ability to anticipate and prevent adverse events caused by our products and 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;
- potential product liability claims;
- changes to federal, state and foreign regulatory requirements applicable to our products and product candidates;
- the impact of current and future healthcare reforms, including those affecting the delivery of or payment for healthcare products and services;
- our ability to obtain, maintain, protect and enforce our intellectual property rights related to our products and product candidates;
- incurring losses in the future, potentially requiring us to raise additional funds;
- general economic, industry and market uncertainty, including due to tariff policy and geopolitical tensions; 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 place undue reliance on our forward-looking statements. Except as required by applicable law, we do not assume and specifically disclaim any obligation to update any forward-looking statements, whether as a result of any new information, future events, changed circumstances or otherwise.

## 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 ordinary shares. The principal risks and uncertainties affecting our business include the following:

- we may not be able to continue to commercialize ARCALYST or be successful in commercializing any future products, potentially impacting the commercial potential for our current and future products and our ability to generate revenue;
- successful commercialization of our products and product candidates, if approved, 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;
- current and future healthcare legislation or executive or administrative action may have a material adverse effect on our business and results of operations;
- if we are unable to advance our product candidates in clinical development, obtain regulatory approval and pursue commercialization, or experience significant delays in doing so, our business may be significantly harmed;
- the incidence and prevalence for target patient populations of our products and product candidates have not been established with precision; if the market opportunities for our products and product candidates are smaller than we estimate, or any approval that we obtain is based on a narrower definition of our targeted patient population, our revenue and ability to sustain profitability may be materially adversely affected;
- clinical development of our product candidates is a lengthy and expensive process with uncertain timelines, costs and outcomes;
- we may encounter substantial delays in our preclinical studies and/or 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;
- we contract with third parties, including independent contract development and manufacturing organizations ("CDMOs") to manufacture our commercial supply of ARCALYST and clinical supply of our product candidates and for certain research and development, which is highly regulated and complex, and we 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 are conducting a technology transfer of the manufacturing process for ARCALYST drug substance from Regeneron Pharmaceuticals, Inc. ("Regeneron") to Samsung Biologics Co., Ltd. ("Samsung"), and the analytical testing methods of ARCALYST drug substance and drug product to new contract testing labs ("CTLs"), which will be subject to significant risks and uncertainties;
- we rely, and expect to continue to rely, on third parties, including independent investigators and contract research organizations ("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 or commercialize our product candidates, and our business could be substantially harmed;

- if we are unable to adequately protect our proprietary technology or obtain and maintain patent protection for our technology and patents, if the scope of the patent protection obtained is not sufficiently broad, or if the terms of our patents are insufficient to protect 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;

- we have a history of operating losses and may require substantial additional financing in the future, which we may be unable to obtain when needed or on acceptable terms;

- we face significant competition, 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, in-license or acquire additional product candidates or technologies, 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 that 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 strategic transactions or arrangements to further develop, commercialize or otherwise attempt to realize value from one or more of our products and product candidates; such arrangements or transactions may not be successful or on favorable terms, which could adversely affect our ability to develop, commercialize or attempt to realize value from our products and product candidates;

- we may be adversely affected by continuing geopolitical tensions, including the introduction of tariffs or reciprocal tariffs that may be imposed by the United States and its global trading partners that could collectively cause economic uncertainty and increased costs to product development and manufacturing;

- the concentration of ownership of the voting power of our ordinary shares, including our Class B ordinary shares, and conversion rights of the holders of our Class A1 and Class B1 ordinary shares, which are held primarily by entities affiliated with certain of our directors, may prevent new investors from influencing significant corporate decisions and may have an adverse effect on the price of our Class A ordinary shares; and

- the rights afforded to our shareholders are governed by English law; not all rights available to shareholders under United States law will be available to holders of our ordinary 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 INTERNATIONAL, PLC**
**CONSOLIDATED BALANCE SHEETS**
**(In thousands, except share and per share amounts)**
**(Unaudited)**

| | June 30, 2025 | December 31, 2024 |
|---|---|---|
| **Assets** | | |
| Current assets: | | |
| Cash and cash equivalents | $ 192,037 | $ 183,581 |
| Short-term investments | 115,745 | 60,046 |
| Accounts receivable, net | 31,910 | 41,724 |
| Inventory | 48,181 | 26,364 |
| Prepaid expenses and other current assets | 32,438 | 20,084 |
| Total current assets | 420,311 | 331,799 |
| Property and equipment, net | 1,076 | 662 |
| Operating lease right-of-use assets | 10,304 | 10,376 |
| Other long-term assets | 8,195 | 10,315 |
| Intangible asset, net | 15,750 | 16,250 |
| Deferred tax assets | 205,514 | 211,151 |
| Total assets | $ 661,150 | $ 580,553 |
| | | |
| **Liabilities and Shareholders' Equity** | | |
| Current liabilities: | | |
| Accounts payable | $ 8,956 | $ 2,039 |
| Accrued collaboration expenses | 52,389 | 48,157 |
| Accrued expenses | 35,959 | 32,355 |
| Operating lease liabilities | 2,522 | 1,993 |
| Other current liabilities | 18,001 | 16,077 |
| Total current liabilities | 117,827 | 100,621 |
| Non-current liabilities: | | |
| Non-current deferred revenue | 31,811 | 31,811 |
| Non-current operating lease liabilities | 7,243 | 7,862 |
| Other long-term liabilities | 9,262 | 1,823 |
| Total liabilities | 166,143 | 142,117 |
| Commitments and contingencies (Note 13) | | |
| Shareholders' equity: | | |
| Class A ordinary shares, nominal value of $0.000273235 per share; 43,397,623 shares and 41,881,319 shares issued and outstanding as of June 30, 2025 and December 31, 2024, respectively | 12 | 11 |
| Class B ordinary shares, nominal value of $0.000273235 per share; 1,795,158 shares issued and outstanding as of June 30, 2025 and December 31, 2024 | 1 | 1 |
| Class A1 ordinary shares, nominal value of $0.000273235 per share; 12,781,964 shares issued and outstanding as of June 30, 2025 and December 31, 2024 | 4 | 4 |
| Class B1 ordinary shares, $0.000273235 nominal value; 16,057,618 shares issued and outstanding as of June 30, 2025 and December 31, 2024 | 4 | 4 |
| Additional paid-in capital | 989,811 | 959,722 |
| Accumulated other comprehensive loss | (53) | (163) |
| Accumulated deficit | (494,772) | (521,143) |
| Total shareholders' equity | 495,007 | 438,436 |
| Total liabilities and shareholders' equity | $ 661,150 | $ 580,553 |

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

**KINIKSA PHARMACEUTICALS INTERNATIONAL, PLC**
**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, | |
|---|---|---|---|---|
| | 2025 | 2024 | 2025 | 2024 |
| Revenue: | | | | |
| Product revenue, net | $ 156,797 | $ 103,394 | $ 294,582 | $ 182,279 |
| License and collaboration revenue | — | 5,237 | — | 6,210 |
| Total revenue | 156,797 | 108,631 | 294,582 | 188,489 |
| Costs and operating expenses: | | | | |
| Cost of goods sold | 18,603 | 12,322 | 36,471 | 22,905 |
| Collaboration expenses | 52,418 | 30,014 | 96,208 | 50,815 |
| Research and development | 18,753 | 24,017 | 38,078 | 50,351 |
| Selling, general and administrative | 46,863 | 42,395 | 90,393 | 81,077 |
| Total operating expenses | 136,637 | 108,748 | 261,150 | 205,148 |
| Income (loss) from operations | 20,160 | (117) | 33,432 | (16,659) |
| Other income | 2,717 | 2,421 | 5,010 | 4,687 |
| Income (loss) before income taxes | 22,877 | 2,304 | 38,442 | (11,972) |
| Provision for income taxes | (5,045) | (6,212) | (12,071) | (9,640) |
| Net income (loss) | $ 17,832 | $ (3,908) | $ 26,371 | $ (21,612) |
| Net income (loss) per share attributable to ordinary shareholders—basic | $ 0.24 | $ (0.06) | $ 0.36 | $ (0.31) |
| Net income (loss) per share attributable to ordinary shareholders—diluted | $ 0.23 | $ (0.06) | $ 0.34 | $ (0.31) |
| Weighted average ordinary shares outstanding—basic | 73,438,530 | 71,004,640 | 73,041,920 | 70,818,831 |
| Weighted average ordinary shares outstanding—diluted | 77,942,082 | 71,004,640 | 76,984,393 | 70,818,831 |
| Comprehensive income (loss) | | | | |
| Net income (loss) | $ 17,832 | $ (3,908) | $ 26,371 | $ (21,612) |
| Other comprehensive income (loss) | | | | |
| Unrealized gain (loss) on short-term investments and currency translation adjustments, net of tax | 112 | (77) | 110 | (136) |
| Total other comprehensive income (loss) | 112 | (77) | 110 | (136) |
| Total comprehensive income (loss) | $ 17,944 | $ (3,985) | $ 26,481 | $ (21,748) |

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

**KINIKSA PHARMACEUTICALS INTERNATIONAL, PLC**
**CONSOLIDATED STATEMENTS OF SHAREHOLDERS' EQUITY**
**(In thousands, except share amounts)**
**(Unaudited)**

| | Ordinary Shares (Class A, B, A1 and B1) | | Additional Paid-In | Accumulated Other Comprehensive | Accumulated | Total Shareholders' |
|---|---|---|---|---|---|---|
| | Shares | Amount | Capital | Loss | Deficit | Equity |
| **Balances at December 31, 2024** | 72,516,059 | $ 20 | $ 959,722 | $ (163) | $ (521,143) | $ 438,436 |
| Issuance of Class A ordinary shares under incentive award plans | 281,273 | — | 2,768 | — | — | 2,768 |
| Share-based compensation expense | — | — | 7,748 | — | — | 7,748 |
| Unrealized loss on short-term investments and currency translation adjustments, net of tax | — | — | — | (2) | — | (2) |
| Net income | — | — | — | — | 8,539 | 8,539 |
| **Balances at March 31, 2025** | 72,797,332 | $ 20 | $ 970,238 | $ (165) | $ (512,604) | $ 457,489 |
| Issuance of Class A ordinary shares under incentive award plans | 1,235,031 | 1 | 10,697 | — | — | 10,698 |
| Share-based compensation expense | — | — | 8,876 | — | — | 8,876 |
| Unrealized gain on short-term investments and currency translation adjustments, net of tax | — | — | — | 112 | — | 112 |
| Net income | — | — | — | — | 17,832 | 17,832 |
| **Balances at June 30, 2025** | 74,032,363 | $ 21 | $ 989,811 | $ (53) | $ (494,772) | $ 495,007 |

| | Ordinary Shares (Class A, B, A1 and B1) | | Additional Paid-In | Accumulated Other Comprehensive | Accumulated | Total Shareholders' |
|---|---|---|---|---|---|---|
| | Shares | Amount | Capital | Income (Loss) | Deficit | Equity |
| **Balances at December 31, 2023** | 70,460,617 | $ 20 | $ 916,763 | $ 6 | $ (477,950) | $ 438,839 |
| Issuance of Class A ordinary shares under incentive award plans | 358,479 | — | 3,613 | — | — | 3,613 |
| Share-based compensation expense | — | — | 7,206 | — | — | 7,206 |
| Unrealized loss on short-term investments and currency translation adjustments, net of tax | — | — | — | (59) | — | (59) |
| Net loss | — | — | — | — | (17,704) | (17,704) |
| **Balances at March 31, 2024** | 70,819,096 | $ 20 | $ 927,582 | $ (53) | $ (495,654) | $ 431,895 |
| Issuance of Class A ordinary shares under incentive award plans | 263,182 | — | (178) | — | — | (178) |
| Share-based compensation expense | — | — | 7,363 | — | — | 7,363 |
| Unrealized loss on short-term investments and currency translation adjustments, net of tax | — | — | — | (77) | — | (77) |
| Net loss | — | — | — | — | (3,908) | (3,908) |
| **Balances at June 30, 2024** | 71,082,278 | $ 20 | $ 934,767 | $ (130) | $ (499,562) | $ 435,095 |

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

**KINIKSA PHARMACEUTICALS INTERNATIONAL, PLC**
**CONSOLIDATED STATEMENTS OF CASH FLOWS**
**(In thousands)**
**(Unaudited)**

| | Six Months Ended June 30, | |
|---|---|---|
| | **2025** | **2024** |
| **Cash flows from operating activities:** | | |
| Net income (loss) | $ 26,371 | $ (21,612) |
| Adjustments to reconcile net income (loss) to net cash provided by operating activities | | |
| Depreciation and amortization expense | 703 | 896 |
| Share-based compensation expense | 16,624 | 14,569 |
| Non-cash lease expense | 1,759 | 1,615 |
| Net amortization of premiums and accretion of discounts on short-term investments | (589) | 109 |
| Net gain on disposal of property and equipment | (24) | (24) |
| Deferred income taxes | 5,637 | 14,934 |
| Changes in operating assets and liabilities: | | |
| Prepaid expenses and other current assets | (12,195) | (9,517) |
| Accounts receivable, net | 9,814 | 815 |
| Inventory | (21,817) | (3,736) |
| Other long-term assets | 1,975 | (7,201) |
| Accounts payable | 6,992 | (933) |
| Accrued expenses, accrued collaboration expenses and other current liabilities | 9,502 | 21,940 |
| Operating lease liabilities | (1,777) | (2,319) |
| Deferred revenue | — | (449) |
| Other long-term liabilities | 7,439 | 68 |
| Net cash provided by operating activities | 50,414 | 9,155 |
| **Cash flows from investing activities:** | | |
| Proceeds from sale of property and equipment | — | 25 |
| Purchases of property and equipment | (264) | (84) |
| Purchases of short-term investments | (129,793) | (125,539) |
| Proceeds from the maturities of short-term investments | 74,633 | 104,325 |
| Net cash used in investing activities | (55,424) | (21,273) |
| **Cash flows from financing activities:** | | |
| Proceeds from issuance of Class A ordinary shares under incentive award plans and employee share purchase plan | 15,925 | 5,129 |
| Payments in connection with ordinary shares tendered for employee tax obligations | (2,459) | (1,694) |
| Net cash provided by financing activities | 13,466 | 3,435 |
| **Net increase (decrease) in cash and cash equivalents** | 8,456 | (8,683) |
| Cash and cash equivalents at beginning of period | 183,581 | 107,954 |
| Cash and cash equivalents at end of period | $ 192,037 | $ 99,271 |
| | | |
| **Supplemental information:** | | |
| Cash paid for income taxes | $ 3,009 | $ 1,510 |
| | | |
| **Supplemental disclosure of non-cash investing and financing activities:** | | |
| Change in right-of-use asset as a result of new, modified, and terminated leases | $ 1,687 | $ 1,463 |
| Additions to property and equipment included in accounts payable and accrued expenses and other liabilities | 338 | 178 |

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

## 1. Nature of the Business and Basis of Presentation

Kiniksa Pharmaceuticals International, plc (the "Company" or "Kiniksa International") is a biopharmaceutical company developing and commercializing novel therapies for diseases with unmet medical need, with a focus on cardiovascular indications. The Company's portfolio of immune-modulating assets is based on strong biologic rationale or validated mechanisms, targets a spectrum of underserved cardiovascular and autoimmune conditions and offers the potential for differentiation.

The Company is the successor issuer to Kiniksa Pharmaceuticals, Ltd. ("Kiniksa Bermuda"). On June 27, 2024, the Company and Kiniksa Bermuda completed a transaction pursuant to a Bermuda court-approved scheme of arrangement (the "Scheme"), which had been previously approved by Kiniksa Bermuda's shareholders. Pursuant to the Scheme, the shareholders of Kiniksa Bermuda became the shareholders of the Company and the Company became the ultimate parent and holding company of the Kiniksa organization, thereby effecting a change of incorporation from Bermuda to the United Kingdom (the "UK) (the "Redomiciliation"). As used herein, and unless the context otherwise requires, references to the "Company" prior to the Redomiciliation shall refer to Kiniksa Bermuda and from and after the Redomiciliation, to Kiniksa International. In addition, references to "ordinary shares" prior to the Redomiciliation are to Kiniksa Bermuda's common shares and from and after the Redomiciliation are to Kiniksa International's ordinary shares.

The Company is subject to risks common to companies in the biopharmaceuticals industry including, but not limited to, commercialization of existing and new products, conducting clinical research and development, its current and future products and product candidates, risks from existing or new competition, protection of proprietary intellectual and other technology and compliance with United States and foreign regulations and approval requirements.

***Principles of Consolidation***

The Redomiciliation was accounted for as a change in the reporting entity between entities under common control and the historical basis of accounting was retained as if the entities had always been combined for financial reporting purposes. The consolidated financial statements for periods prior to the Redomiciliation are the consolidated statements of Kiniksa Bermuda as the predecessor to the Company for accounting and reporting purposes and, upon completion of the Redomiciliation, such historical consolidated financial statements became Kiniksa International's historical consolidated financial statements.

The accompanying consolidated financial statements have been prepared in accordance with accounting principles generally accepted in the United States of America ("GAAP") and include the accounts of the Company and its wholly owned subsidiaries, Kiniksa Pharmaceuticals Corp. ("Kiniksa US"), Kiniksa Bermuda and Kiniksa Pharmaceuticals (UK), Ltd. ("Kiniksa UK") as well as the subsidiary of Kiniksa US, Primatope Therapeutics, Inc. ("Primatope") and 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. Where the Kiniksa Pharmaceuticals International, plc entity is referred to in its single, unconsolidated form, it is referred to as "Kiniksa International".

***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, and the valuation of the Company's deferred tax assets. Estimates are periodically reviewed in light of changes in circumstances, facts and experience. Changes in estimates are recorded in the period in which they become known. Actual results could differ from those estimates.

***Unaudited Interim Consolidated Financial Information***

The accompanying unaudited consolidated financial statements have been prepared in accordance with GAAP for interim financial information. The accompanying unaudited consolidated financial statements do not include all of the information and footnotes required by GAAP for complete consolidated financial statements. The information included in this Quarterly Report on Form 10-Q should be read in conjunction with the Company's audited consolidated financial statements and the accompanying notes included in the Company's Annual Report on Form 10-K for the year ended December 31, 2024 (the "2024 Form 10-K"). The Company's accounting policies are described in the Notes to Consolidated Financial Statements included in the Company's 2024 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, 2025 and the results of its operations for the three and six months ended June 30, 2025 and 2024, the changes in its shareholders' equity for the three and six months ended June 30, 2025 and 2024 and its cash flows for the six months ended June 30, 2025 and 2024. The results for the three and six months ended June 30, 2025 are not necessarily indicative of results to be expected for the year ending December 31, 2025, 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, 2025, the Company had an accumulated deficit of $494,772. During the six months ended June 30, 2025, the Company reported a net income of $26,371 and had cash provided from operating activities of $50,414. As of June 30, 2025, the Company had cash, cash equivalents and short-term investments of $307,782. 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.

***Recently Issued Accounting Pronouncements***

In December 2023, the Financing Accounting Standards Board ("FASB") issued Accounting Standards Update ("ASU") No. 2023-09, *Income Taxes (Topic 740): Improvements to Income Tax Disclosures*. The amendments require (i) enhanced disclosures in connection with an entity's effective tax rate reconciliation and (ii) income taxes paid disaggregated by jurisdiction. The amendments are effective for annual periods beginning after December 15, 2024. The Company does not expect the adoption of the amendments to have a material impact on its financial statements.

In November 2024, the FASB issued ASU 2024-03, *Income Statement-Reporting Comprehensive Income-Expense Disaggregation Disclosures (Subtopic 220-40): Disaggregation of Income Statement Expenses*, requiring public entities to disclose additional information about specific expense categories in the notes to the financial statements on an interim and annual basis. ASU 2024-03 is effective for fiscal years beginning after December 15, 2026, and for interim periods beginning after December 15, 2027, with early adoption permitted. The Company is currently evaluating the impact of adopting ASU 2024-03.

## 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.

- 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, 2025 Using: | | | |
|---|---|---|---|---|
| | Level 1 | Level 2 | Level 3 | Total |
| Assets: | | | | |
| Cash equivalents — money market funds | $ 139,483 | $ — | $ — | $ 139,483 |
| Short-term investments — U.S. Treasury notes | — | 115,745 | — | 115,745 |
| | $ 139,483 | $ 115,745 | $ — | $ 255,228 |

| | Fair Value Measurements as of December 31, 2024 Using: | | | |
|---|---|---|---|---|
| | Level 1 | Level 2 | Level 3 | Total |
| Assets: | | | | |
| Cash equivalents — money market funds | $ 135,275 | $ — | $ — | $ 135,275 |
| Short-term investments — U.S. Treasury notes | — | 60,046 | — | 60,046 |
| | $ 135,275 | $ 60,046 | $ — | $ 195,321 |

During the six months ended June 30, 2025 and the year ended December 31, 2024, 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, 2025 and December 31, 2024 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, 2025 | | | | | |
| Short-term investments — U.S. Treasury notes | 115,771 | — | (26) | — | 115,745 |
| | $ 115,771 | $ — | $ (26) | $ — | $ 115,745 |

| | Amortized Cost | Gross Unrealized Gains | Gross Unrealized Losses | Credit Losses | Fair Value |
|---|---|---|---|---|---|
| December 31, 2024 | | | | | |
| Short-term investments — U.S. Treasury notes | 60,022 | 24 | — | — | 60,046 |
| | $ 60,022 | $ 24 | $ — | $ — | $ 60,046 |

As of June 30, 2025, the Company considers the unrealized losses in its investment portfolio to be temporary in nature and not due to credit losses. The Company has the ability to hold such investments until recovery of the fair value. The Company utilizes the specific identification method in computing realized gains and losses. The Company

had no realized gains and losses on its available-for-sale securities for the three and six months ended June 30, 2025 or 2024.

## 3. Product Revenue, Net

The Company derives substantially all of its product revenue, net from sales of ARCALYST in the United States, which was as follows:

| | Three Months Ended June 30, | | Six Months Ended June 30, | |
|---|---|---|---|---|
| | 2025 | 2024 | 2025 | 2024 |
| Product revenue, net | $ 156,797 | $ 103,394 | $ 294,582 | $ 182,279 |

The following table summarizes balances and activity in each of the product revenue allowance and reserve categories for the six months ended June 30, 2025:

| | Contractual Adjustments | Government Rebates | Returns | Total |
|---|---|---|---|---|
| Balance at December 31, 2024 | $ 3,495 | $ 8,640 | $ 2,294 | $ 14,429 |
| Current provisions relating to sales in the current year | 18,042 | 12,527 | 916 | 31,485 |
| Adjustments relating to prior years | — | (484) | — | (484) |
| Payments/returns relating to sales in the current year | (16,000) | (4,396) | — | (20,396) |
| Payments/returns relating to sales in the prior years | (3,495) | (5,201) | — | (8,696) |
| Balance at June 30, 2025 | $ 2,042 | $ 11,086 | $ 3,210 | $ 16,338 |

Total revenue-related reserves as of June 30, 2025 and December 31, 2024, included in the Company's consolidated balance sheets, are summarized as follows:

| | June 30, 2025 | December 31, 2024 |
|---|---|---|
| Components of accounts receivable | $ (670) | $ (444) |
| Components of other current liabilities | 17,008 | 14,873 |
| Total revenue-related reserves | $ 16,338 | $ 14,429 |

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.

## 4. Inventory

Inventory consisted of the following:

| | June 30, 2025 | December 31, 2024 |
|---|---|---|
| Raw materials | $ 9,743 | $ 9,972 |
| Semi-finished goods | 32,726 | — |
| Finished goods | 10,566 | 21,246 |
| Total inventory | $ 53,035 | $ 31,218 |
| Balance Sheet Classification: | | |
| Inventory | $ 48,181 | $ 26,364 |
| Other long-term assets | 4,854 | 4,854 |
| Total inventory | $ 53,035 | $ 31,218 |

As of June 30, 2025, $18,386 of semi-finished goods were associated with the Company's technology transfer of ARCALYST manufacturing to a new facility, which is pending regulatory approval. The Company believes it is probable that regulatory approval will be obtained, and all such inventory will be available for commercial distribution in the future.

## 5. Property and Equipment, Net

Property and equipment, net consisted of the following:

| | June 30, 2025 | December 31, 2024 |
|---|---|---|
| Furniture, fixtures and vehicles | $ 177 | $ 183 |
| Computer hardware and software | 379 | 379 |
| Leasehold improvements | 3,920 | 3,931 |
| Lab equipment | 3,743 | 4,207 |
| Construction in progress | 634 | 155 |
| Total property and equipment | 8,853 | 8,855 |
| Less: Accumulated depreciation | (7,777) | (8,193) |
| Total property and equipment, net | $ 1,076 | $ 662 |

Depreciation expense was $13 and $119 during the three months ended June 30, 2025 and 2024, respectively, and $48 and $273 during the six months ended June 30, 2025 and 2024, respectively.

## 6. Intangible Assets

Intangible assets, net of accumulated amortization as of June 30, 2025 and December 31, 2024 are summarized in the following table.

| | Estimated life | As of June 30, 2025 | | | As of December 31, 2024 | | |
|---|---|---|---|---|---|---|---|
| | | Cost | Accumulated Amortization | Net | Cost | Accumulated Amortization | Net |
| Regulatory milestone | 20 years | $ 20,000 | $ 4,250 | $ 15,750 | $ 20,000 | $ 3,750 | $ 16,250 |

Amortization expense was $250 during the three months ended June 30, 2025 and 2024 and $500 during the six months ended June 30, 2025 and 2024.

## 7. Accrued Expenses

Accrued expenses consisted of the following:

| | June 30, 2025 | December 31, 2024 |
|---|---|---|
| Accrued research and development expenses | $ 16,947 | $ 11,004 |
| Accrued employee compensation and benefits | 13,182 | 17,046 |
| Accrued legal, commercial and professional fees | 5,521 | 3,617 |
| Other | 309 | 688 |
| | $ 35,959 | $ 32,355 |

## 8. Share-Based Compensation

As part of the Redomiciliation, Kiniksa International assumed the sponsorship of, and all rights and obligations of Kiniksa Bermuda under Kiniksa Bermuda's equity compensation plans, which include the 2018 Incentive Award Plan (the "2018 Plan") and the 2018 Employee Share Purchase Plan (the "2018 ESPP"). Upon the effectiveness of the 2018 Plan, the Company ceased granting awards under its 2015 Equity Incentive Plan (as amended, the "2015 Plan" and together with the 2018 Plan, the "Plans").

***2015 Plan***

As of June 30, 2025, there were 1,126,908 Class A ordinary shares reserved for issuance pursuant to outstanding awards under the 2015 Plan that were granted prior to the effectiveness of the 2018 Plan.

***2018 Plan***

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"), PSUs (as defined below) 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, 2025, increased by 2,900,642 Class A ordinary shares. As of June 30, 2025, 6,132,156 shares remained available for future grant under the 2018 Plan.

***2018 ESPP***

In December 2024, the Company's board of directors approved an increase, as of January 1, 2025, of 90,000 Class A ordinary shares under the 2018 ESPP. As of June 30, 2025, 724,213 Class A ordinary shares were available for future issuance under the 2018 ESPP.

***Restricted Share Units***

The Company grants RSUs with service conditions to eligible employees as part of its equity incentive compensation. Such 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.

***Market and Performance-Based Share Units***

In the second quarter of 2024, the Company began periodically granting performance-based restricted share units to certain employees under the 2018 Plan that are earned based upon (i) the achievement of certain specified ARCALYST revenue targets ("Revenue PSUs") and (ii) the Company's total shareholder return ("TSR") relative to the TSR of each member of a specified peer group ("TSR PSUs"). The TSR PSUs and Revenue PSUs are subject to a three-year service period.

In addition, the Company from time-to-time grants performance-based restricted share units to certain eligible employees pursuant to 2018 Plan that are earned based upon certain development and regulatory milestones ("Development PSUs" and, together with the Revenue PSUs and TSR PSUs, "PSUs"). The Company's currently outstanding Development PSUs are subject to earnout percentages based upon the date of applicable milestone achievement.

***Performance Share Options***

Beginning in the second quarter of 2025, the Company began granting performance share options ("PSOs") to certain eligible employees pursuant to the 2018 Plan representing the right to purchase shares of the Company's Class A ordinary shares. Such PSOs vest, if at all, upon the achievement of certain specified development and regulatory milestones and are subject to earnout percentages based upon the date of applicable milestone achievement.

The following table summarizes RSU and PSU activity for the six months ended June 30, 2025:

| | RSUs | | PSUs | |
|---|---|---|---|---|
| | Number of Shares | Weighted Average Grant Date Fair Value | Number of Shares | Weighted Average Grant Date Fair Value |
| Unvested as of December 31, 2024 | 2,250,602 | $ 17.36 | 59,137 | $ 22.06 |
| Granted | 576,083 | $ 22.29 | 416,416 | $ 27.63 |
| Vested | (342,204) | $ 14.69 | — | $ — |
| Forfeited | (212,005) | $ 18.24 | (17,987) | $ 26.57 |
| Unvested as of June 30, 2025 | 2,272,476 | $ 18.93 | 457,566 | $ 26.96 |

The following table summarizes share option and PSO activity for the six months ended June 30, 2025:

| | Options | | PSOs | |
|---|---|---|---|---|
| | Number of Shares | Weighted Average Exercise Price | Number of Shares | Weighted Average Exercise Price |
| Outstanding as of December 31, 2024 | 11,286,994 | $ 15.25 | — | $ — |
| Granted | 1,074,941 | $ 23.31 | 359,995 | $ 27.36 |
| Exercised | (1,250,472) | $ 12.26 | — | $ — |
| Forfeited | (280,790) | $ 19.67 | (7,723) | $ 27.74 |
| Outstanding as of June 30, 2025 | 10,830,673 | $ 16.28 | 352,272 | $ 27.36 |
| Share options exercisable as of June 30, 2025 | 7,226,765 | $ 14.62 | — | $ — |
| Share options unvested as of June 30, 2025 | 10,830,673 | $ 16.28 | 352,272 | $ 27.36 |

As of June 30, 2025, total unrecognized compensation cost related to RSUs, Revenue PSUs, TSR PSUs, and share options was $82,058 which is expected to be recognized over a weighted average remaining period of 2.50 years. As of June 30, 2025, total unrecognized compensation cost related to outstanding Development PSUs and PSOs was $10,874 which will be recognized when the applicable milestones are deemed probable of achievement through the date the awards vests with a cumulative catch-up.

***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, | |
|---|---|---|---|---|
| | 2025 | 2024 | 2025 | 2024 |
| Cost of goods sold | $ 566 | $ 353 | $ 995 | $ 712 |
| Research and development expenses | 1,717 | 1,501 | 3,219 | 2,954 |
| Selling, general and administrative expenses | 6,593 | 5,509 | 12,410 | 10,903 |
| | $ 8,876 | $ 7,363 | $ 16,624 | $ 14,569 |

## 9. 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 in September 2022 (the "Genentech Effective Date").

Under the Genentech License Agreement, the Company received an upfront payment of $80,000 for the license. During the year ended December 31, 2023, the Company received cash payments of $20,000 following delivery of certain drug supplies to Genentech and $15,000 following Genentech's achievement of a development milestone related to a new indication under the Genentech License Agreement. In the fourth quarter of 2023, following the achievement of a development milestone related to a second indication under the Genentech License Agreement, Genentech became obligated to make an additional cash payment of $10,000 which the Company received in the first quarter of 2024. In the second quarter of 2024, the Company received a cash payment of $5,000 following the achievement of a development milestone related to the third indication under the Genentech License Agreement. 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 sales-based milestones, before fulfilling the Company's upstream financial obligations, of which approximately $570,000 remained as of June 30, 2025. 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. In 2024, the Company fulfilled its responsibility under the Genentech License Agreement with respect to 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 2022. In 2023 and 2024, the Company added $25,000 and $5,000, respectively, to the transaction price following Genentech's achievement of development milestones 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; that is upon transfer of the license to Genentech, as control of the license was transferred on the Genentech Effective Date and Genentech could begin to use and benefit from the license 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 did not recognize any collaboration revenue under the Genentech License Agreement during the three and six months ended June 30, 2025. The Company recognized $5,156 and $5,261 of collaboration revenue under the Genentech License Agreement during the three and six months ended June 30, 2024, respectively. As a result of the $5,000 development milestone the Company recognized revenue of $4,994 and $4,989 during the three and six months ended June 30, 2024, respectively, related to performance obligations satisfied in prior periods. As of June 30, 2025, the Company has recognized as revenue all of the transaction price associated with the Genentech License Agreement.

***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 ARCALYST and develop, manufacture and commercialize mavrilimumab 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 ARCALYST and mavrilimumab outside the Huadong Territory.

In April 2025, the Company and Huadong entered into a mutual termination agreement pursuant to which the parties agreed to terminate the mavrilimumab Huadong Collaboration Agreement and release all claims related thereto.

Under the Huadong Collaboration Agreements, the Company received a total upfront cash payment of $22,000, which included $12,000 for the Huadong Territory license of ARCALYST and $10,000 for the Huadong Territory license of mavrilimumab. In 2024, following the achievement of a regulatory milestone under the ARCALYST Huadong Collaboration Agreement, Huadong became obligated to make an additional cash payment of $20,000 to the Company which the Company received in the first quarter of 2025. The Company will be eligible to receive up to approximately $50,000 in contingent sales-based milestone payments for ARCALYST, all of which remain outstanding as of June 30, 2025. Due to its termination, the Company does not expect to receive any future payments under the mavrilimumab Huadong Collaboration Agreement. The Company was previously eligible to receive up to approximately

$576,000 in payments for mavrilimumab, including specified development, regulatory and sales-based milestones. Huadong was also obligated to pay the Company tiered percentage royalties ranging from the mid-teens to low twenties on annual net sales of mavrilimumab in the Huadong Territory. Huadong will also be obligated to pay the Company tiered percentage royalties ranging from the low-to-mid teens on annual net sales of ARCALYST in the Huadong Territory, subject to certain reductions tied to ARCALYST manufacturing costs and certain other customary reductions, with an aggregate minimum floor. Royalties will be payable on a country-by-country or region-by-region basis until the later of (i) 12 years after the first commercial sale of ARCALYST 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 ARCALYST in such country or region in the Huadong Territory, and (iii) the expiration of the last regulatory exclusivity for ARCALYST 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 included an estimate of variable consideration associated with the clinical manufacturing supply of certain materials when those materials were 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 recognized 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. Upon the termination of the mavrilimumab Huadong Collaboration Agreement in April 2025, the Company will not recognize any additional revenue from this agreement.

*Accounting for the ARCALYST Huadong Collaboration Agreement*

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

The Company determined the transaction price at the inception of the ARCALYST Huadong Collaboration Agreement, which includes $12,000, consisting of the upfront payment. In 2024, the Company added $20,000 to the transaction price following the achievement of a regulatory milestone. 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 ARCALYST Huadong Collaboration Agreement consisting of the exclusive license for ARCALYST and clinical and commercial manufacturing obligations for ARCALYST 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 ARCALYST Huadong Collaboration Agreement and may be reduced or increased based on changes in the various activities.

The Company did not recognize any revenue under the ARCALYST Huadong Collaboration Agreement during the three and six months ended June 30, 2025. The Company recognized $189 of the upfront payment in collaboration revenue during the six months ended June 30, 2024, under the ARCALYST Huadong Collaboration Agreement related to materials delivered. The Company did not recognize any revenue under the ARCALYST Huadong Collaboration Agreement during the three months ended June 30, 2024. As of June 30, 2025, $31,811 of the transaction price is recorded in non-current deferred revenue, based upon timing of anticipated future shipments.

The following table summarizes the Company's contract liabilities in connection with license and collaboration agreements for the six months ended June 30, 2025:

| | Balance at Beginning of Period | Additions | Revenue Recognized | Reclassification | Balance at End of Period |
|---|---|---|---|---|---|
| **Six Months Ended June 30, 2025** | | | | | |
| Contract Liabilities: | | | | | |
| Huadong ARCALYST | $ 31,811 | $ — | $ — | $ — | $ 31,811 |

## 10. License and Acquisition Agreements

***Biogen Asset Purchase Agreement***

In September 2016, the Company entered into an asset purchase agreement (the "Biogen Agreement") with Biogen MA Inc. ("Biogen") to acquire all of Biogen's right, title and interest in and to certain assets used in or relating to 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 payments to Biogen of up to $179,000 upon the achievement of specified clinical and regulatory milestones in multiple indications in various territories, of which $165,000 remains as of June 30, 2025. Additionally, the Company could be obligated to make up to an aggregate of $150,000 of payments upon the achievement of specified annual net sales milestones and to pay tiered royalties on escalating tiers of annual net sales of licensed products starting in the high single-digit percentages and ending below the teens.

The Company also agreed to pay certain obligations under third party contracts retained by Biogen that relate to the vixarelimab program. Under these retained contracts, the Company paid a one-time upfront sublicense fee and is

obligated to pay insignificant annual maintenance fees as well as clinical and regulatory milestone payments of up to an aggregate of $1,575.

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

The Company and Biogen have amended the Biogen Agreement twice following its initial effective date. The first amendment, in July 2017, clarified the scope of the antibodies subject to the Biogen Agreement. The second amendment, effective in August 2022, was entered into in connection with the Genentech License Agreement, and amended certain defined terms, including "Net Sales," "Indication," "Product," "Combination Product" and "Valid Claim." In addition, the tiered royalty rates to be paid by the Company to Biogen increased by an amount equal to less than one percent. Upon the termination or expiration of the Genentech License Agreement 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, 2025, the Company recorded expenses of $30 and $44, respectively, related to milestone payments and the annual maintenance fee in connection with the Biogen Agreement. During the three and six months ended June 30, 2024, the Company recorded expenses of $11 and $72, respectively, related to a milestone and the annual maintenance fee in connection with the Biogen Agreement.

***Beth Israel Deaconess Medical Center License Agreement***

In 2019, the Company acquired all of the outstanding securities of Primatope Therapeutics, Inc. ("Primatope"), the company that owned or controlled the intellectual property related to abiprubart. In connection with the Company's acquisition of Primatope, 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 abiprubart (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, 2025, the Company did not record any expenses in connection with the BIDMC Agreement. During the three and six months ended June 30, 2024 the Company recorded expenses of $8 and $35, respectively, 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, 2025, the Company recognized $52,389 and $96,164 respectively, of expenses related to the profit sharing agreement presented within collaboration expenses. For the three and six months ended June 30, 2024, the Company recognized $29,943 and $50,066 respectively, of expenses related to the profit sharing agreement presented within collaboration expenses.

The Company has a supply agreement with Regeneron pursuant to which the Company may order both clinical and commercial product. The 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, 2025 and 2024, the Company did not incur any research and development expense related to the purchase of drug materials under the supply agreement. As of June 30, 2025 and December 31, 2024, the Company recorded inventory of $24,906 and $21,246 respectively, related to the purchase of commercial product under the supply agreement. As of June 30, 2025, the Company had non-cancelable purchase commitments under the supply agreement (see Note 13).

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 (subsequently acquired by AstraZeneca PLC) ("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 was obligated to use commercially reasonable efforts to develop and commercialize the licensed products.

In February 2025, the Company delivered a notice of termination to MedImmune, notifying them of its intent to terminate the MedImmune Agreement, for convenience, effective May 22, 2025. Following such date, the exclusive worldwide sublicense rights to certain intellectual property rights to make, use, develop and commercialize mavrilimumab were returned to MedImmune. The Company did not record any significant charges related to the termination of the MedImmune Agreement.

During the three and six months ended June 30, 2025 and 2024, the Company did not record any expenses in connection with milestone payments due under the MedImmune Agreement.

## 11. 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 ordinary shares are identical, except with respect to voting, transferability and conversion (see Exhibit 4.2 to the Company's Current Report on Form 8-K12B, filed on June 28, 2024). 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 ordinary shareholders will, therefore, be the same for both Class A and Class B ordinary shares on an individual or combined basis.

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

| | Three Months Ended June 30, | | Six Months Ended June 30, | |
|---|---|---|---|---|
| | 2025 | 2024 | 2025 | 2024 |
| Numerator: | | | | |
| Net income (loss) attributable to ordinary shareholders | $ 17,832 | $ (3,908) | $ 26,371 | $ (21,612) |
| Denominator: | | | | |
| Weighted-average shares outstanding | 73,438,530 | 71,004,640 | 73,041,920 | 70,818,831 |
| Effect of dilutive securities | | | | |
| Options to purchase ordinary shares | 3,504,516 | — | 3,105,795 | — |
| Performance options to purchase ordinary shares | — | — | — | — |
| Unvested RSUs | 924,675 | — | 780,562 | — |
| Unvested PSUs | 74,362 | — | 56,116 | — |
| Weighted-average shares outstanding | 77,942,082 | 71,004,640 | 76,984,393 | 70,818,831 |
| | | | | |
| Basic net income (loss) per share | $ 0.24 | $ (0.06) | $ 0.36 | $ (0.31) |
| Diluted net income (loss) per share | $ 0.23 | $ (0.06) | $ 0.34 | $ (0.31) |

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

For the three and six months ended June 30, 2025 diluted earnings per share includes the assumed exercise of dilutive options, assumed exercise of dilutive PSOs for which the performance condition has been met, and the assumed issuance of ordinary shares for unvested RSUs and PSUs for which the market or 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 share options and the average unrecognized compensation expense for unvested share-based compensation awards, would be used to purchase the Company's ordinary shares at the average market price during the period.

For the three and six months ended June 30, 2024 the Company's potentially dilutive securities, which include options, unvested RSUs and unvested PSUs, have been excluded from the computation of diluted net loss per share attributable to ordinary shareholders as the effect would be to reduce the earnings per share ("EPS") attributable to ordinary shareholders. Therefore, the weighted average number of ordinary shares outstanding used to calculate both basic and diluted EPS attributable to ordinary shareholders is the same. The Company excluded the following potential ordinary shares, presented based on amounts outstanding at each period end, from the computation of diluted EPS attributable to ordinary shareholders for the periods indicated because including them would have had an anti-dilutive effect:

| | Three Months Ended June 30, | | Six Months Ended June 30, | |
|---|---|---|---|---|
| | 2025 | 2024 | 2025 | 2024 |
| Share options to purchase ordinary shares | 3,026,168 | 11,780,526 | 3,804,284 | 11,780,526 |
| Performance share options to purchase ordinary shares | — | — | — | — |
| Unvested RSUs | 62,701 | 2,328,169 | 512,753 | 2,328,169 |
| Unvested PSUs | — | 28,824 | — | 28,824 |
| Total anti-dilutive shares | 3,088,869 | 14,137,519 | 4,317,037 | 14,137,519 |

PSUs and PSOs that are outstanding and contain performance-based or market-based vesting criteria for which the performance or market conditions have not been met are excluded from the presentation of common stock equivalents outstanding in the table above.

## 12. Income Taxes

Prior to the Redomiciliation, the Company was incorporated and principally subject to taxation in Bermuda. Following the Redomiciliation, the Company is incorporated and principally subject to taxation in the UK. Under the laws of Bermuda, tax on a company's income is assessed at a zero percent tax rate. As a result, the Company has not recorded any income tax benefits from its losses incurred in Bermuda during the reporting periods in which the Company was incorporated there, and no net operating loss carryforwards will be available to the Company for those losses. Following the Redomiciliation, the Company's income is subject to the enacted UK statutory corporate tax rate and net operating losses incurred have an indefinite carryforward. The Company's wholly owned United States subsidiaries, Kiniksa US and Primatope, are subject to federal and state income taxes in the United States. The Company's wholly owned subsidiary Kiniksa Bermuda remains subject to taxation, if any, in Bermuda. The Company's wholly owned subsidiary Kiniksa UK, and Kiniksa UK's wholly owned subsidiaries, Kiniksa Germany, Kiniksa France, and Kiniksa Switzerland, are subject to taxation in their respective countries. Certain of the Company's subsidiaries operate under cost plus arrangements.

The Company's income tax rate for the three and six months ended June 30, 2025 is attributable to Kiniksa Switzerland's, Kiniksa UK's Swiss branch office's, Kiniksa UK's and Kiniksa US's income or loss subject to taxation in each of their respective countries. Income tax provision for the three and six months ended June 30, 2025 was $5,045 and $12,071, respectively. The provision for income taxes is primarily driven by income earned in Switzerland, UK, United States and uncertain tax positions offset in part by tax benefits from Foreign Derived Intangible Income ("FDII") deduction and United States federal and state research and development credits ("R&D Credits").

Although Kiniksa Bermuda had no corporate income tax, the Company's income tax rate for the three and six months ended June 30, 2024 was due to Kiniksa Switzerland's, Kiniksa UK's Swiss branch office's, Kiniksa UK's and Kiniksa US's income or loss subject to taxation in each of their respective countries. Income tax provision for the three and six months ended June 30, 2024 was $6,212 and $9,640, respectively. The provision for income taxes is primarily driven by income earned in the UK, Switzerland and United States offset in part by tax benefits from FDII deduction and R&D 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.

Since 2021, the Company has engaged in a series of intra-entity asset transfers and allocations to contribute assets to its wholly owned Switzerland subsidiary, UK subsidiary and its UK Swiss branch office.

In January 2021, in connection with its launch readiness activities, Kiniksa Bermuda contributed all of its rights, title and interest in, among other things, certain contracts (including the Regeneron Agreement), intellectual

property rights, product filings and approvals and other information, plans and materials owned or controlled by Kiniksa Bermuda insofar as they related exclusively or primarily to ARCALYST to Kiniksa UK.

In February 2022, Kiniksa Bermuda contributed its exclusive rights to develop and commercialize mavrilimumab in the Huadong Territory to Kiniksa UK. Subsequent to the contribution there was a triggering event in 2024 resulting in decreases to the tax asset balance related to mavrilimumab intangible assets.

In July 2022, Kiniksa Bermuda contributed all of its rights, title and interest in, among other things, certain contracts (including the Biogen Agreement), intellectual property rights, product filings and approvals and other information, plans and materials owned or controlled by Kiniksa Bermuda insofar as they related exclusively or primarily to vixarelimab to Kiniksa UK.

The consolidated Company did not incur tax liabilities on any of these intra-entity transfers since the transferor, Kiniksa Bermuda, is exempt from income tax in Bermuda, its jurisdiction of incorporation. Kiniksa UK accounted for the 2021 and 2022 intra-entity transfers as transfers of assets between related parties and received stepped up tax bases in the contributed intellectual property assets, equal to the fair value of the assets at the time of transfer. The Company recorded UK deferred tax assets as a result of these contributions, which represent the difference between the stepped-up tax bases and the book bases for financial statement purposes. At the time of the 2021 and 2022 transfers of the relevant assets, the Company recorded a valuation allowance on the full amount of the recognized deferred tax assets.

The fair value of the January 2021 transfer of ARCALYST intellectual property assets was determined utilizing forecasted cash flows attributable to commercial operations and estimated probabilities of success of such cash flows, discounted to present value utilizing the discounted cash flow method. The fair values of the transferred mavrilimumab and vixarelimab intellectual property assets were determined utilizing future cash flows related to agreements with third parties for the use of the applicable intellectual property and estimated probabilities of success of such cash flows, discounted to present value utilizing the discounted cash flow method.

In December 2023, Kiniksa UK allocated all of its rights, title and interest in, among other things, certain contracts (including the Regeneron Agreement), intellectual property rights, product filings and approvals and other information, plans and inventory owned or controlled by the Company insofar as they related exclusively or primarily to ARCALYST to Kiniksa UK's Swiss branch office.

The December 2023 allocation of the assets to the Swiss branch did not result in a taxable disposal for Kiniksa UK as the allocation was to a branch within the entity. The future results of Kiniksa UK's Swiss branch office are subject to income taxes in Switzerland. Kiniksa UK's Swiss branch office received a step up in basis resulting in a Swiss deferred tax asset. The fair value of the allocated ARCALYST intellectual property assets was determined utilizing forecasted cash flows attributable to commercial operations and estimated probabilities of success of such cash flows, discounted to present value utilizing the discounted cash flow method. The fair value of the ARCALYST inventory was determined utilizing the average net selling price less estimated costs to sell.

In January 2024, Kiniksa Bermuda transferred to Kiniksa Switzerland all rights, title and interest in, among other things, certain contracts, intellectual property rights, product filings and approvals and other information, plans and materials owned insofar as they related exclusively or primarily to abiprubart, mavrilimumab, KPL-387, KPL-1161 and certain preclinical assets. In June 2024, Kiniksa UK terminated its exclusive rights to develop and commercialize mavrilimumab in the Huadong Territory, with such rights reverting to Kiniksa Switzerland. Thereafter Kiniksa Switzerland held worldwide rights to develop and commercialize mavrilimumab until the MedImmune Agreement was terminated in May 2025. In October 2024, Kiniksa UK contributed all of its rights, title and interest in, among other things, certain contracts (including the Biogen Agreement), intellectual property rights, product filings and approvals and other information, plans and materials owned or controlled by Kiniksa UK insofar as they related exclusively or primarily to vixarelimab to Kiniksa Switzerland.

The consolidated Company did not incur tax liabilities on any of the January 2024 intra-entity transfers since the transferor, Kiniksa Bermuda, is exempt from income tax in Bermuda. Kiniksa Switzerland accounted for the intra-entity transfers as transfers of assets between related parties and received stepped up tax bases in the contributed intellectual property assets, equal to the fair value of the assets at the time of transfer. In relation to the June 2024 transaction, Kiniksa UK received consideration in exchange for the termination of exclusive rights. Neither Kiniksa UK

nor Kiniksa Switzerland incurred any tax liabilities as a result of the transaction. The consolidated Company did not incur tax liabilities on any of the October 2024 intra-entity transfers since the transferor, Kiniksa UK, is the sole direct shareholder of Kiniksa Switzerland. Kiniksa Switzerland accounted for the intra-entity transfers as transfers of assets between related parties and received stepped up tax bases in the contributed intellectual property assets, equal to the fair value of the assets at the time of transfer.

The fair values of the transferred assets were determined utilizing future cash flows of projected operations and estimated probabilities of success of such cash flows, discounted to present value utilizing the discounted cash flow method. The Company recorded deferred tax assets as a result of these contributions, which represent the difference between the stepped-up tax bases and the book bases for financial statement purposes. The Company maintains a valuation allowance on the full amount of the Kiniksa Switzerland deferred tax assets. There are no material deferred tax assets in the jurisdictions outside the United States, UK and Switzerland.

## 13. Commitments and Contingencies

***License Agreements***

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

***Manufacturing Commitments***

The Company has a supply agreement with Regeneron pursuant to which the Company may order both clinical and commercial product (see Note 10). In June 2024, the Company entered into a Master Services Agreement and a Product Specific Agreement with Samsung as part of its technology transfer of the manufacturing process for ARCALYST drug substance. The Company has additionally entered into agreements with several contract development and manufacturing organizations to provide the Company with preclinical and clinical trial materials for its non-ARCALYST assets. As of June 30, 2025, the Company had committed to minimum purchase commitments under all of these agreements totaling $161,555, of which $25,621 is due within one year.

The Company issued termination notices to CDMOs in February 2025 to terminate the clinical supply agreements for the production of abiprubart. During the six months ended June 30, 2025, the Company recorded and paid $2,500 in research and development expenses because of these terminations. During the three months ended June 30, 2025, the Company did not record any expenses in connection with these terminations. The Company does not expect to incur any additional expenses because of these terminations.

***Performance Cash Awards***

Beginning in the second quarter of 2025, the Company began granting cash awards (“Performance Cash Awards”) to certain eligible employees pursuant to the 2018 Plan, which were eligible to be received upon the achievement of certain specified development and regulatory milestones and that are subject to earnout percentages based upon the date of applicable milestone achievement. As of June 30, 2025, the Company estimates the future cash payments under such Performance Cash Awards to be $22,309 if the milestones are achieved. The Performance Cash Awards will be recognized when the applicable milestones are deemed probable of achievement with a cumulative catch-up and recognized over the remaining term.

***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, 2025 or December 31, 2024.

***Legal Proceedings***

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

**14. Segment Information and Geographic Data**

The Company manages its operations as a single operating segment for the purposes of assessing performance and making operating decisions. The Company's singular focus is on developing and commercializing novel therapies that target cardiovascular diseases with significant unmet medical need. The Company's Chief Operating Decision Maker ("CODM") is the Chief Executive Officer. The Company's CODM reviews consolidated operating results and decides how to allocate resources based on net income that also is reported on the income statement as consolidated net income. The measure of segment assets is reported on the balance sheet as total consolidated assets. The CODM utilizes net income to make key decisions about how to allocate resources across the Company's commercial product and development programs.

The following table presents selected financial information with respect to the Company's single operating segment for the three and six months ended June 30, 2025 and 2024:

| | Three Months Ended June 30, | | Six Months Ended June 30, | |
|---|---|---|---|---|
| | 2025 | 2024 | 2025 | 2024 |
| Revenue: | | | | |
| Product revenue, net | $ 156,797 | $ 103,394 | $ 294,582 | $ 182,279 |
| License and collaboration revenue | - | 5,237 | - | 6,210 |
| Total revenue | 156,797 | 108,631 | 294,582 | 188,489 |
| Operating expenses: | | | | |
| Cost of goods sold | 18,603 | 12,322 | 36,471 | 22,905 |
| Collaboration expenses | 52,418 | 30,014 | 96,208 | 50,815 |
| Direct research and development expenses by program: | | | | |
| ARCALYST | 171 | 597 | 526 | 710 |
| KPL-387 | 8,485 | 1,631 | 13,663 | 2,718 |
| KPL-1161 | 370 | 100 | 470 | 100 |
| Abiprubart | 635 | 12,368 | 5,002 | 27,457 |
| Vixarelimab | - | 279 | - | 1,033 |
| Mavrilimumab | - | 155 | 120 | 378 |
| Unallocated research and development expenses | 9,092 | 8,887 | 18,297 | 17,955 |
| Selling, general and administrative | 46,863 | 42,395 | 90,393 | 81,077 |
| Total operating expenses | 136,637 | 108,748 | 261,150 | 205,148 |
| Other income (1) | 2,717 | 2,421 | 5,010 | 4,687 |
| Income before income taxes | 22,877 | 2,304 | 38,442 | (11,972) |
| Provision for income taxes | (5,045) | (6,212) | (12,071) | (9,640) |
| Net income (loss) | $ 17,832 | $ (3,908) | $ 26,371 | $ (21,612) |
| | | | | |
| Other significant noncash items: | | | | |
| Share-based compensation expense | $ 8,876 | $ 7,363 | $ 16,624 | $ 14,569 |
| Non-cash lease expense | 924 | 836 | 1,759 | 1,615 |
| Deferred income taxes | 2,644 | 10,569 | 5,637 | 14,934 |

(1) Includes interest income of $2,731 and $5,021 for the three and six months ended June 30, 2025, respectively. Includes interest income of $2,338 and $4,494 for the three and six months ended June 30, 2024, respectively.

The following table presents total revenue by geographic region of the customer for the three and six months ended June 30, 2025 and 2024:

| | Three Months Ended June 30, | | Six Months Ended June 30, | |
|---|---|---|---|---|
| | 2025 | 2024 | 2025 | 2024 |
| Revenue: | | | | |
| United States | $ 156,506 | $ 108,495 | $ 294,102 | $ 187,318 |
| United Kingdom | 291 | 59 | 480 | 296 |
| Rest of world | - | 77 | - | 875 |
| Total revenue | $ 156,797 | $ 108,631 | $ 294,582 | $ 188,489 |

## 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, 2024 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 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 developing and commercializing novel therapies for diseases with unmet need, with a focus on cardiovascular indications. Our portfolio of assets is based on strong biologic rationale or validated mechanisms and offers the potential for differentiation.

ARCALYST is an interleukin-1α ("IL-1α") and interleukin-1β ("IL-1β") cytokine trap. In 2017, we licensed ARCALYST from Regeneron, which 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 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 United States prevalent population of approximately 40,000 patients seeking and receiving medical treatment. ARCALYST is also approved in the United States for the treatment of Cryopyrin-Associated Periodic Syndromes ("CAPS"), including Familial Cold Autoinflammatory Syndrome ("FCAS") 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. ARCALYST is commercially available across the United States through a select network of specialty pharmacies. 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 2022, we granted Hangzhou Zhongmei Huadong Pharmaceutical Co., Ltd. ("Huadong") exclusive rights to develop and commercialize ARCALYST in the Huadong Territory (as defined below). In 2023, Regeneron initiated a technology transfer of the manufacturing process for ARCALYST drug substance, and we are working to qualify Samsung as our replacement CDMO.

KPL-387 is an investigational, fully human immunoglobulin G2 monoclonal antibody that binds human interleukin-1 receptor 1 ("IL-1R1"), inhibiting IL-1α- and IL-1β-mediated signaling. KPL-387 is an independently developed asset that we believe has the potential to further advance recurrent pericarditis treatment options for patients by providing the added convenience of monthly subcutaneous dosing with a liquid formulation. In June 2024, we initiated a Phase 1 clinical trial of KPL-387 in normal healthy volunteers. In July 2025, we announced that the Phase 2 dose-focusing portion of the pivotal Phase 2/3 clinical trial of KPL-387 in recurrent pericarditis had begun recruiting. We expect data from the Phase 2 portion of the trial in the second half of 2026.

KPL-1161 is an independently developed, pre-clinical, Fc-modified immunoglobulin G2 monoclonal antibody that binds IL-1R1, inhibiting IL-1α- and IL-1β-mediated signaling. KPL-1161 is a modified version of KPL-387 designed to have an increased drug half-life that we believe could support quarterly subcutaneous dosing.

Abiprubart is an investigational monoclonal antibody inhibitor of CD40-CD154 costimulatory interaction, which we believe to be an attractive approach to address multiple autoimmune disease pathologies. We hold an exclusive worldwide license to abiprubart from Beth Israel Deaconess Medical Center, Inc. ("BIDMC"). We previously announced a Phase 2b clinical trial of abiprubart in Sjögren's Disease. In February 2025, we announced our plans to discontinue development of abiprubart in the indication and explore strategic alternatives for the asset.

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, which has since been acquired by AstraZeneca PLC ("MedImmune"). In

February 2025, we announced our termination of our license agreement from MedImmune for convenience, effective in May 2025. In April 2025, we entered into a mutual termination agreement with Huadong with respect to the mavrilimumab Huadong Collaboration Agreement (as defined below), pursuant to which we agreed to terminate the agreement and release all claims related thereto.

We were initially incorporated under the laws of Bermuda in July 2015 and, in April 2024, subsequently announced the completion of the change of place of incorporation of our principal holding company from Bermuda to the United Kingdom (the "UK"), pursuant to a scheme of arrangement approved by both the Bermuda Supreme Court and our shareholders (the "Redomiciliation"), which caused the shareholders of our former parent company, Kiniksa Pharmaceuticals, Ltd. ("Kiniksa Bermuda"), to become the shareholders of our current parent company, Kiniksa Pharmaceuticals International, plc (the "Company" or "Kiniksa International"). As used herein, and unless the context otherwise requires, references to "we," "us," "our" and similar words or phrases prior to the Redomiciliation shall refer to Kiniksa Bermuda and from and after the Redomiciliation, to Kiniksa International. In addition, references to "ordinary shares" prior to the Redomiciliation are to Kiniksa Bermuda's common shares and from and after the Redomiciliation are to Kiniksa International's ordinary shares.

Our ability to generate product revenue sufficient to sustain our organization will depend heavily on a number of factors, including the continued commercialization of ARCALYST, the development and eventual commercialization of one or more of our current or future product candidates, if approved, and the management of our costs consistent with our current operating plan. For the six months ended June 30, 2025, our net income was $26.4 million. As of June 30, 2025, we had an accumulated deficit of $494.8 million.

As of June 30, 2025, we had cash, cash equivalents and short-term investments of $307.8 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*."

**Components of Our Results of Operations**

***Product revenue, net***

We have been generating product revenue from sales of ARCALYST since April 2021. ARCALYST is sold through a third party logistics provider that distributes primarily through a select network of specialty pharmacies (collectively, "customers"), which deliver the medication to patients by mail. ARCALYST is currently approved for sale only in the United States, and, therefore, we expect to derive substantially all of our product revenue from the United States for the foreseeable future.

Net revenue from product sales is recognized at the transaction price when the customer obtains 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 606 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, milestone payments and products sold under collaboration agreements.

In February 2022, we entered into two collaboration and license agreements (the "Huadong Collaboration Agreements"), with Huadong, pursuant to which we granted Huadong exclusive rights to develop and commercialize ARCALYST and mavrilimumab, in the Asia Pacific region excluding Japan (the "Huadong Territory"). In April 2025, we entered into a mutual termination agreement with Huadong pursuant to which we agreed to terminate the mavrilimumab Huadong Collaboration Agreement and release all claims related thereto.

Under the Huadong Collaboration Agreements, we received a total upfront cash payment of $22.0 million, which included $12.0 million for the Huadong Territory license of ARCALYST and $10.0 million for the Huadong Territory license of mavrilimumab. In the fourth quarter of 2024, following the achievement of a regulatory milestone under the ARCALYST Huadong Collaboration Agreement, Huadong became obligated to make an additional cash payment of $20.0 million, which was received in the first quarter of 2025. In addition, we will be eligible to receive additional contingent sales-based milestones payments related to ARCALYST. Huadong will also be obligated to pay us tiered percentage royalties ranging from the low-to-mid teens on annual net sales of ARCALYST in the Huadong Territory, subject to certain reductions tied to ARCALYST manufacturing costs and certain other customary reductions, with an aggregate minimum floor. Royalties will be payable on a country-by-country or region-by-region basis until the later of (i) 12 years after the first commercial sale of ARCALYST in such country or region in the Huadong Territory, (ii) the date of expiration of the last valid patent claim of our patent rights or any joint collaboration patent rights that covers ARCALYST in such country or region in the Huadong Territory, and (iii) the expiration of the last regulatory exclusivity for ARCALYST in such country or region in the Huadong Territory. We recognized the $10.0 million related to the mavrilimumab license during the year ended December 31, 2022 and do not expect to recognize any additional license and collaboration revenue following the termination of the mavrilimumab Huadong Collaboration Agreement. We have recognized $0.2 million of revenue of the $32.0 million transaction price under the ARCALYST license agreement as of June 30, 2025, and will recognize the remaining revenue as materials are shipped.

We are party to a license agreement (the "Genentech License Agreement") with Genentech, effective September 2022, pursuant to which we granted Genentech exclusive worldwide rights to develop and commercialize vixarelimab and related antibodies (each, a "Genentech Licensed Product").

Under the Genentech License Agreement, we received an upfront payment of $80.0 million for the license. Additionally, in 2023, we received a total of $35.0 million in additional payments from Genentech related to delivery of certain drug material to Genentech and Genentech's achievement of a development milestone. In the fourth quarter of 2023, following the achievement of a development milestone related to a second indication under the Genentech License Agreement, Genentech became obligated to make an additional cash payment of $10.0 million, which was received in the first quarter of 2024. In the second quarter of 2024, we received $5.0 million following the achievement of a development milestone related to a third indication under the Genentech License Agreement. We will be eligible to receive up to a total of approximately $600.0 million in contingent payments, including specified development, regulatory and sales-based milestones, of which approximately $570.0 million remains as of June 30, 2025. We 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 our 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. In 2024, we completed our remaining obligations under the Genentech License Agreement and we have recognized as revenue all of the considerations received.

***Operating Expenses***

*Cost of Goods Sold*

Cost of goods sold includes production and distribution costs of ARCALYST, 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 supply chain, quality, and regulatory activities, and the technology transfer of the manufacturing process for the ARCALYST drug substance.

Following the technology transfer of drug substance manufacturing from Regeneron's US-based manufacturing facilities to Samsung's South Korea-based facilities, the importation of such drug substance into the United States may be subject to tariffs in the future. In such an event, the cost of drug substance that we import may increase. However, Kiniksa UK's Swiss branch office, which manufactures and sells ARCALYST, owns all of Kiniksa's ARCALYST intellectual property and, therefore, is not obligated to pay royalties to any other entity, which royalty payments would be subject to tariffs under the currently proposed US tariff policy. As a result, we do not expect that tariffs, if implemented as proposed, will have a material effect on our overall business, financial condition or results of operations.

*Collaboration expenses*

Collaboration expenses consist of Regeneron's share of the profit related to ARCALYST sales under the Regeneron Agreement and the cost of products sold under collaboration agreements. 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 ARCALYST drug substance manufacturing from Regeneron to Samsung, 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.

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 revenue from the sale of our current or future product candidates. 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, including 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 expect that our selling, general and administrative expenses will continue to increase in the future as we continue to expand our infrastructure related to the commercialization of ARCALYST and our other product candidates, if approved.

***Other Income***

Other income consists of interest income recognized from investments in money market funds, United States Treasury notes and other miscellaneous income offset by expenses related to investments.

***Income Taxes***

Prior to the Redomiciliation, our principal holding company was incorporated and principally subject to taxation in Bermuda. Following the Redomiciliation, our principal holding company is incorporated and principally subject to taxation in the UK. Under the 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 the reporting periods in which we were incorporated there, and no net operating loss carryforwards are currently available to us for those losses. Following the Redomiciliation, our income is subject to the enacted UK statutory corporate tax rate and net operating losses incurred have an indefinite carryforward. Our wholly owned United States subsidiaries, Kiniksa Pharmaceuticals, Inc. ("Kiniksa US"), and Primatope Therapeutics, Inc. are subject to federal and state income taxes in the United States. Our wholly owned subsidiary Kiniksa Pharmaceuticals (UK), Ltd. ("Kiniksa UK"), its Swiss branch office, and Kiniksa UK's wholly owned subsidiaries, Kiniksa Pharmaceuticals (Germany) GmbH, Kiniksa Pharmaceuticals (France) SARL, and Kiniksa Pharmaceuticals, GmbH ("Kiniksa Switzerland") are subject to taxation in their respective countries.

On July 4, 2025, new U.S tax legislation was signed into law (known as the "One Big Beautiful Bill Act" or "OBBBA") which makes permanent many of the tax provisions enacted in 2017 as part of the Tax Cuts and Jobs Act that were set to expire at the end of 2025. In addition, the OBBBA makes changes to certain U.S. corporate tax provisions, but many are generally not effective until 2026. We are currently evaluating the impact of the new legislation on our operations.

In the first quarter of 2024, Kiniksa Bermuda transferred to Kiniksa Switzerland all rights, title and interest in, among other things, certain contracts, intellectual property rights, product filings and approvals and other information, plans and materials owned insofar as they related exclusively or primarily to abiprubart, mavrilimumab, KPL-387, KPL-1161 and other preclinical assets. In connection with the foregoing transfer, we recognized a step-up in basis and did not incur any material tax liabilities.

In the second quarter of 2024, Kiniksa UK terminated its exclusive rights to develop and commercialize mavrilimumab in the Huadong Territory, with such rights reverting to Kiniksa Switzerland. Kiniksa Switzerland held worldwide rights to develop and commercialize mavrilimumab until the cancellation of the license agreement from MedImmune, effective in May 2025. In the fourth quarter of 2024, Kiniksa UK contributed all of its rights, title and interest in, among other things, certain contracts, intellectual property rights, product filings and approvals and other information, plans and materials owned or controlled by Kiniksa UK insofar as they related exclusively or primarily to vixarelimab to Kiniksa Switzerland. In connection with the termination of Kiniksa UK rights and the contribution, we revalued the assets at fair market value and did not incur any material tax liabilities.

**Results of Operations**

***Comparison of the Three Months Ended June 30, 2025 and 2024***

The following table summarizes our results of operations for the three months ended June 30, 2025 and 2024:

| | Three Months Ended June 30, | | |
|---|---|---|---|
| | 2025 | 2024 | Change |
| | | (in thousands) | |
| Revenue: | | | |
| Product revenue, net | $ 156,797 | $ 103,394 | $ 53,403 |
| License and collaboration revenue | — | 5,237 | (5,237) |
| Total revenue | 156,797 | 108,631 | 48,166 |
| Costs and Operating expenses: | | | |
| Cost of goods sold | 18,603 | 12,322 | 6,281 |
| Collaboration expenses | 52,418 | 30,014 | 22,404 |
| Research and development | 18,753 | 24,017 | (5,264) |
| Selling, general and administrative | 46,863 | 42,395 | 4,468 |
| Total operating expenses | 136,637 | 108,748 | 27,889 |
| Income (loss) from operations | 20,160 | (117) | 20,277 |
| Other income | 2,717 | 2,421 | 296 |
| Income before income taxes | 22,877 | 2,304 | 20,573 |
| Provision for income taxes | (5,045) | (6,212) | 1,167 |
| Net income (loss) | $ 17,832 | $ (3,908) | $ 21,740 |

*Product Revenue, Net*

We recognized net revenue from the sale of ARCALYST of $156.8 million for the three months ended June 30, 2025, compared to $103.4 million for the three months ended June 30, 2024, an increase of $53.4 million. The increase in product revenue was primarily driven by an increase in patient enrollment.

*License and Collaboration Revenue*

We reported no license and collaboration revenue for the three months ended June 30, 2025. We reported $5.2 million of license and collaboration revenue for the three months ended June 30, 2024, driven by the achievement of a $5.0 million development milestone related to a third indication under the Genentech License Agreement.

*Cost of Goods Sold*

We recognized cost of goods sold of $18.6 million for the three months ended June 30, 2025, compared to $12.3 million for the three months ended June 30, 2024, an increase of $6.3 million. The increase in cost of goods sold relates primarily to the increase in sales of ARCALYST and unfavorable production variances. Cost of goods sold related to the technology transfer of the manufacturing process were $1.7 million for the three months ended June 30, 2025, compared to $2.8 million for the three months ended June 30, 2024, a decrease of $1.1 million.

*Collaboration Expenses*

Collaboration expenses were $52.4 million for the three months ended June 30, 2025, compared to $30.0 million for the three months ended June 30, 2024, an increase of $22.4 million. Collaboration expenses increased due to increased revenue from sales of ARCALYST.

*Research and Development Expenses*

| | Three Months Ended June 30, | | |
|---|---|---|---|
| | 2025 | 2024 | Change |
| | (in thousands) | | |
| ARCALYST | $ 171 | $ 597 | $ (426) |
| KPL-387 | 8,485 | 1,631 | 6,854 |
| KPL-1161 | 370 | 100 | 270 |
| Abiprubart | 635 | 12,368 | (11,733) |
| Vixarelimab | — | 279 | (279) |
| Mavrilimumab | — | 155 | (155) |
| Unallocated research and development expenses: | | | |
| Personnel related (including share-based compensation) | 5,723 | 5,765 | (42) |
| Other | 3,369 | 3,122 | 247 |
| Total research and development expenses | $ 18,753 | $ 24,017 | $ (5,264) |

Research and development expenses were $18.8 million for the three months ended June 30, 2025, compared to $24.0 million for the three months ended June 30, 2024, a decrease of $5.3 million.

Direct costs for our KPL-387 program were $8.5 million during the three months ended June 30, 2025, compared to $1.6 million during the three months ended June 30, 2024. The increase in expenses incurred primarily related to our Phase 1 clinical trial in normal healthy volunteers and the start-up of our Phase 2/3 clinical trial of KPL-387 in recurrent pericarditis during the three months ended June 30, 2025, as compared to the manufacturing of clinical supply during the three months ended June 30, 2024.

Direct costs for our KPL-1161 program were $0.4 million for the three months ended June 30, 2025, compared to $0.1 million during the three months ended June 30, 2024. For the three months ended June 30, 2025 and 2024, expenses incurred primarily related to pre-clinical development.

The direct costs for our abiprubart program were $0.6 million during the three months ended June 30, 2025, compared to $12.4 million during the three months ended June 30, 2024, a decrease of $11.7 million. The decrease in expenses incurred primarily related to the close-out of our Phase 2b clinical trial in Sjögren's Disease during the three months ended June 30, 2025, as compared to the manufacturing of clinical material, continuation of cohort four of our Phase 2 clinical trial in rheumatoid arthritis ("RA") and start-up costs of our Phase 2b clinical trial in Sjögren's Disease during the three months ended June 30, 2024.

Unallocated research and development expenses were $9.1 million and $8.9 million for the three months ended June 30, 2025 and June 30, 2024, respectively. Personnel-related costs for the three months ended June 30, 2025 and 2024 included share-based compensation of $1.7 million and $1.5 million, respectively.

*Selling, General and Administrative Expenses*

Selling, general and administrative expenses were $46.9 million for the three months ended June 30, 2025, compared to $42.4 million for the three months ended June 30, 2024. The increase of $4.5 million was primarily due to an increase of $4.4 million in personnel-related costs largely attributable to an increase in headcount. Personnel-related costs for the three months ended June 30, 2025 and 2024 included share-based compensation of $6.6 million and $5.5 million, respectively.

*Provision for Income Taxes*

For the three months ended June 30, 2025, we recorded an income tax provision of $5.0 million relating primarily to income earned in the UK, Switzerland and the United States, net of the Foreign Derived Intangible Income

("FDII") deduction, benefits related to share-based compensation and United States federal and state research and development credits ("R&D Credits"). For the three months ended June 30, 2024, we recorded an income tax provision of $6.2 million relating primarily to income earned in the UK, Switzerland and the United States, net of the FDII deduction and R&D Credits.

***Comparison of the Six Months Ended June 30, 2025 and 2024***

The following table summarizes our results of operations for the six months ended June 30, 2025 and 2024:

| | Six Months Ended June 30, | | |
|---|---|---|---|
| | 2025 | 2024 | Change |
| | | (in thousands) | |
| Revenue: | | | |
| Product revenue, net | $ 294,582 | $ 182,279 | $ 112,303 |
| License and collaboration revenue | — | 6,210 | (6,210) |
| Total revenue | 294,582 | 188,489 | 106,093 |
| Operating expenses: | | | |
| Cost of goods sold | 36,471 | 22,905 | 13,566 |
| Collaboration expenses | 96,208 | 50,815 | 45,393 |
| Research and development | 38,078 | 50,351 | (12,273) |
| Selling, general and administrative | 90,393 | 81,077 | 9,316 |
| Total operating expenses | 261,150 | 205,148 | 56,002 |
| Income (loss) from operations | 33,432 | (16,659) | 50,091 |
| Other income | 5,010 | 4,687 | 323 |
| Income (loss) before income taxes | 38,442 | (11,972) | 50,414 |
| Provision for income taxes | (12,071) | (9,640) | (2,431) |
| Net income (loss) | $ 26,371 | $ (21,612) | $ 47,983 |

*Product Revenue, Net*

We recognized net revenue from the sale of ARCALYST of $294.6 million for the six months ended June 30, 2025, compared to $182.3 million for the six months ended June 30, 2024, an increase of $112.3 million. The increase in product revenue was primarily driven by an increase in patient enrollment.

*License and Collaboration Revenue*

We reported no license and collaboration revenue for the six months ended June 30, 2025. We reported $6.2 million of license and collaboration revenue for the six months ended June 30, 2024, primarily driven by the achievement of a $5.0 million development milestone related to a third indication under the Genentech License Agreement and $0.7 million of products sold under the ARCALYST Huadong Collaboration Agreements

*Cost of Goods Sold*

We recognized cost of goods sold of $36.5 million for the six months ended June 30, 2025, compared to $22.9 million for the six months ended June 30, 2024, an increase of $13.6 million. The increase in cost of goods sold relates primarily to the increase in sales of ARCALYST and unfavorable production variances. Cost of goods sold related to the technology transfer of the manufacturing process were $3.9 million for the three months ended June 30, 2025, compared to $4.9 million for the three months ended June 30, 2024, a decrease of $1.0 million.

*Collaboration Expenses*

Collaboration expenses were $96.2 million for the six months ended June 30, 2025, compared to $50.8 million for the six months ended June 30, 2024, an increase of $45.4 million. Collaboration expenses increased due to increased revenue from sales of ARCALYST.

*Research and Development Expenses*

| | Six Months Ended June 30, | | |
|---|---|---|---|
| | 2025 | 2024 | Change |
| | (in thousands) | | |
| ARCALYST | $ 526 | $ 710 | $ (184) |
| KPL-387 | 13,663 | 2,718 | 10,945 |
| KPL-1161 | 470 | 100 | 370 |
| Abiprubart | 5,002 | 27,457 | (22,455) |
| Vixarelimab | — | 1,033 | (1,033) |
| Mavrilimumab | 120 | 378 | (258) |
| Unallocated research and development expenses: | | | |
| Personnel related (including share-based compensation) | 11,646 | 11,889 | (243) |
| Other | 6,651 | 6,066 | 585 |
| Total research and development expenses | $ 38,078 | $ 50,351 | $ (12,273) |

Research and development expenses were $38.1 million for the six months ended June 30, 2025, compared to $50.4 million for the six months ended June 30, 2024, a decrease of $12.3 million.

Direct costs for our KPL-387 program were $13.7 million during the six months ended June 30, 2025, compared to $2.7 million during the six months ended June 30, 2024. The increase in expenses incurred primarily related to our Phase 1 clinical trial in normal healthy volunteers and the start-up of our Phase 2/3 clinical trial of KPL-387 in recurrent pericarditis during the six months ended June 30, 2025, as compared to the manufacturing of clinical supply and our Phase 1 study in normal healthy volunteers during the six months ended June 30, 2024.

Direct costs for our KPL-1161 program were $0.5 million for the six months ended June 30, 2025, compared to $0.1 million during the six months ended June 30, 2024. For the six months ended June 30, 2025 and 2024, expenses incurred primarily related to pre-clinical development.

The direct costs for our abiprubart program were $5.0 million during the six months ended June 30, 2025, compared to $27.5 million during the six months ended June 30, 2024, a decrease of $22.5 million. The decrease in expenses incurred primarily related to the close-out of our Phase 2b clinical trial in Sjögren's Disease during the six months ended June 30, 2025, as compared to the manufacturing of clinical material, continuation of cohort four of our Phase 2 clinical trial in RA and start-up costs of our Phase 2b clinical trial in Sjögren's Disease during the six months ended June 30, 2024.

Unallocated research and development expenses were $18.3 million for the six months ended June 30, 2025, compared to $18.0 million for the six months ended June 30, 2024. Personnel-related costs for the six months ended June 30, 2025 and 2024 included share-based compensation of $3.2 million and $3.0 million, respectively.

*Selling, General and Administrative Expenses*

Selling, general and administrative expenses were $90.4 million for the six months ended June 30, 2025, compared to $81.1 million for the six months ended June 30, 2024. The increase of $9.3 million was primarily due to an increase of $6.8 million in personnel-related costs largely attributable to an increase in headcount and an increase in

sales and marketing costs of $3.4 million largely attributable to promotional activities. Personnel-related costs for the six months ended June 30, 2025 and 2024 included share-based compensation of $12.4 million and $10.9 million, respectively.

*Provision for Income Taxes*

For the six months ended June 30, 2025, we recorded an income tax provision of $12.1 million relating primarily to income earned in the UK, Switzerland and the U.S., net of the FDII deduction, benefits related to share-based compensation and R&D Credits utilized. For the six months ended June 30, 2024, we recorded an income tax provision of $9.6 million relating primarily to income earned in the UK, Switzerland and the United States, net of the FDII deduction and R&D Credits.

**Liquidity and Capital Resources**

As of June 30, 2025, our principal source of liquidity was cash, cash equivalents and short-term investments, which totaled $307.8 million. Net income (loss) was $26.4 million and ($21.6) million for the six months ended June 30, 2025 and 2024, respectively. We expect our cash balance and our expected cash inflows from operations to allow us to meet our current operating plan.

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 $14.2 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 $11.9 million, of which $4.1 million are due within one year. In connection with our ongoing technology transfer of ARCALYST drug substance manufacturing, we have entered into a Master Services Agreement and a Product Specific Agreement with Samsung. Our commitments under such agreements, which include the purchase of raw materials and related service fees, will obligate us to minimum payments of $136.7 million, $0.7 million of which are due within one year. As of June 30, 2025, we have capitalized $18.4 million of production cost into inventory as semi-finished goods related to drug substance manufactured at Samsung. We have additionally entered into agreements with several CDMOs to provide us with preclinical and clinical trial materials for our non-ARCALYST assets, which obligate us to minimum payments of $10.7 million all of which are due within one year. We have long-term incentive plans for our employees that may result in cash award payments of $22.3 million, based upon the achievement of certain regulatory milestones, none of which are expected to be achieved in the next year.

Under various agreements with third parties, we are entitled to receive upfront payments, milestone payments, and royalties, each based upon specified milestones. In 2024, we received $15.0 million in development milestone payments from Genentech related to a second and third indication under the Genentech License Agreement. In 2025, we received a $20.0 million milestone payment related to Huadong’s achievement of a regulatory milestone under the ARCALYST Huadong Collaboration Agreement.

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

***Cash Flows***

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

| | Six Months Ended June 30, | |
|---|---|---|
| | 2025 | 2024 |
| | (in thousands) | |
| Net cash provided by operating activities | $ 50,414 | $ 9,155 |
| Net cash provided by (used in) investing activities | (55,424) | (21,273) |
| Net cash provided by financing activities | 13,466 | 3,435 |
| Net increase (decrease) in cash and cash equivalents | $ 8,456 | $ (8,683) |

*Operating Activities*

Net cash provided by operations was $50.4 million and $9.2 million for the six months ended June 30, 2025 and 2024, respectively. The increase in cash provided by operating activities is primarily due to an increase in net contribution from higher ARCALYST sales and an increase in cash received from licensing agreements of $5.0 million.

*Investing Activities*

Net cash used in investing activities was $55.4 million and $21.3 million for the six months ended June 30, 2025 and 2024, respectively. The increase in net cash used in investing activities was driven by managing our cash and short-term investment portfolio mix.

*Financing Activities*

During the six months ended June 30, 2025 and 2024, net cash provided by financing activities was $13.5 million and $3.4 million, respectively, consisting of proceeds from the exercise of share options offset by payments in connection with ordinary shares tendered for employee tax obligations.

***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. We may also incur expenses in connection with collaboration, licensing or other strategic transactions. Further, we may incur 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. We expect to incur expenses as we:

- support our sales, marketing and distribution capabilities, infrastructure and organization to commercialize ARCALYST and any product candidates for which we may obtain marketing approval;
- conduct new and ongoing research and pre-clinical and clinical development of our product candidates, including our Phase 2/3 clinical trial of KPL-387 in recurrent pericarditis, our ongoing Phase 1 clinical trial of KPL-387 in normal healthy volunteers and our pre-clinical investigations of KPL-1161;
- manufacture our products and product candidates for clinical or commercial use, increase our manufacturing capabilities, add additional manufacturers or suppliers and perform activities related to our technology transfer of the process for manufacturing ARCALYST drug substance;
- seek regulatory and marketing approvals for our product candidates that successfully complete clinical trials, if any;

- identify, assess and study new or expanded indications for our products and product candidates and/or new or alternative dosing levels, dosing frequencies or administrations of our products and product candidates;
- make milestone or other payments under any current or future license, acquisition, collaboration or other strategic transaction agreements;
- seek 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;
- seek to identify, assess, acquire or develop additional product candidates;
- address any 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;
- enter into licensing, acquisition, collaboration or other strategic transaction agreements;
- seek to maintain, protect and expand our intellectual property portfolio;
- seek to attract and retain skilled personnel;
- create additional infrastructure to support our product development and commercialization efforts; and
- experience delays or encounter 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 disruptions to the national or global economy.

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 collaboration, licensing or other strategic transactions. We expect to continue to incur significant expenses related to product manufacturing, including technology transfer costs, 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.

**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 risks in the ordinary course of our business. These risks primarily include interest rate sensitivities related to our short-term investments. There were no material changes to our quantitative and qualitative disclosures about market risk related to our investment activities during the three months ended June 30, 2025 as disclosed in "Item 7A. Quantitative and Qualitative Disclosures About Market Risks" of the Annual Report.

**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 (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, 2025. Based on that evaluation, our Chief Executive Officer and Chief Financial Officer concluded that our disclosure controls and procedures were effective at the reasonable assurance level as of June 30, 2025.

***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, 2025 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 ordinary 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 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.***

Since our commercial launch of ARCALYST, 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 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, could be materially adversely impacted by a number of foreseen and unforeseen factors, including:

- any delays in our ability to produce sufficient quantities of ARCALYST, or any of our future products, at an acceptable cost or quality, including such delays arising out of quality assurance concerns or changes in regulatory guidance, or those caused by our reliance on our third party manufacturers;
- our inability 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;
- an inadequate number of prescribers or accounts prescribing our current and 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 provide acceptable evidence of safety and efficacy;
- our inability to enable our products to be viewed as the product of choice within any indications for which they are approved;

- the prevalence and severity of side effects associated with any future product;
- our inability to compete with current or future competitor products and/or biosimilars;
- the convenience and ease of administration of our products relative to alternative therapies, if any;
- 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 address product labeling or product insert requirements, including any changes mandated by regulatory authorities after initial approval;
- our inability to equip customer-facing personnel with effective materials, including medical and sales literature to help them educate physicians and other healthcare professionals regarding applicable diseases relevant to ARCALYST or any of our future products;
- any delays in the ongoing technology transfer of the process for manufacturing ARCALYST drug substance;
- 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;
- publications of scientific literature, consensus papers and treatment guidelines unfavorable to the administration of our products and product candidates and/or the positioning of the class of drugs to which each of our products and product candidates belongs;
- our inability to develop or obtain and sustain sufficient operational functions and infrastructure to support our commercial activities;
- our inability to establish and maintain patent and trade secret protection or regulatory exclusivity for our products;
- our inability to enforce and defend our intellectual property rights and claims; 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.

***We rely on a select network of third party specialty pharmacies to market and sell ARCALYST that may not meet our or our patients' needs.***

We rely on a select network of third party specialty pharmacies to distribute ARCALYST in the United States, which is the only country where it is currently approved for sale. We expect to use a similar strategy for our current and future product candidates, if approved. We rely on such specialty pharmacies to effectively distribute products in a timely manner, provide certain patient support services, manage prescription intake, collect accurate patient and inventory data and collect payments from payors. While we have entered into agreements with each of these specialty

pharmacies, they may not perform as agreed, our strategic priorities may change or they may terminate their agreements with us. Further, an inability of our specialty pharmacies to meet our patients' needs may lead to reputational harm or patient loss. In the event that such network fails to properly meet our or our patients' needs, we may need to partner with other specialty pharmacies to replace or supplement our current network and there is no guarantee that we will be able to do so on commercially reasonable terms or at all. In addition, there is a risk that patients may discontinue or suspend their ARCALYST treatment in the process of transitioning between specialty pharmacies, and it may take time to re-integrate such patients into our network, if at all. In such an event, our business, results of operations, financial condition and prospects may be materially affected.

***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.***

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, the adequacy of reimbursement (including affordability of patient cost-sharing obligations) 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; implementing utilization management controls such as requiring prior authorization; limiting the amount of reimbursement for a particular product; restricting the prices that manufacturers may charge for their products and increasing the proportion of the cost for which the patient is responsible. Future government action to control costs is likely. See "*Risk Factors – General Risk Factors – Current and future healthcare legislation or executive or administrative action may have a material adverse effect on our business and results of operations.*" 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 including health technology assessment bodies in the European Union (the "EU") and UK, 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 at which we can sell, 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. In addition, obtaining and maintaining favorable coverage and adequate reimbursement may require us to offer pricing concessions to third party payors.

On January 12, 2025, the EU Regulation on Health Technology Assessment became applicable. The new Regulation introduces a single EU level submission for joint clinical assessments to evaluate the relative clinical effectiveness of new medicines and medical devices. The joint health technology assessment process seeks to assist national authorities in making more timely and informed decisions on the pricing and reimbursement of health technologies and streamline the procedure for health technology developers. From January 12, 2025, all new cancer medicines and advanced therapy medicinal products are subject to the joint clinical assessment. The rules will be extended to orphan medicines in January 2028 and from 2030, all new medicines will be subject to the new rules.

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 biosimilars and lower cost brands with or without the same approved indication. The result of such a change would be a more challenging value/benefit assessment and the potential for a worse relative outcome, including such payors refusing to provide coverage and reimbursement entirely, or finding the evidence not sufficiently compelling to support our desired pricing and reimbursement. Similarly, payors may implement coverage criteria that further restrict 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 before the payer will cover the use of ARCALYST or any of our product candidates, if approved.

We may be unable to sustain any favorable coverage and reimbursement on an ongoing basis. Third party payors may also revisit their previously established coverage policies from time to time including their assessment of the relative value/benefit provided by a drug product compared to clinical alternatives, such as any competitive products with the same or similar indications and biosimilars. 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 often introduce more challenging price negotiation methodologies when competitors exist or enter into the market. 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 biosimilar products enter the market, there are mandatory price reductions for the innovator product. 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 "*Risk Factors—Risks Related to Commercialization – It may be difficult for us to realize the benefit of increasing the price of certain of our commercialized products.*"

***It may be difficult for us to realize the benefit of increasing the price of certain of our commercialized products.***

We have and may continue to periodically increase the price of ARCALYST and may implement similar pricing practices for future products, if approved, and 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. 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, patients may have difficulty obtaining access to, or affording, such products and we may see materially negative impacts on our business operations.

In addition, President Trump, through an Executive Order, has directed government agencies and officials to take action to implement "most-favored nation" pricing for American purchasers. See "*Risk Factors – General Risk Factors – Current and future healthcare legislation or executive or administrative action may have a material adverse effect on our business and results of operations."* The scope and nature of such actions remain unclear but any future required compliance with such order could impede our ability to establish price increases with certain payors and purchasers.

Any price concessions will reduce our overall revenue generation and may impair the benefit of any price increases we may take. Price concessions that reduce our product revenue may require us to rely on potentially dilutive capital-raising efforts to fund our operations, which may impact the price of our ordinary 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 medical 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 impose onerous reimbursement policies that limit patient access. We cannot assure you that current payor coverage and reimbursement policies for ARCALYST will continue. 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 medical need, the loss of access to our products may materially harm our targeted patient populations who cannot source adequate alternative therapies.

We are also required to provide discounts or rebates under government healthcare programs or to certain government and private purchases in order to obtain coverage under federal healthcare programs. In addition, price increases that outpace inflation may also trigger additional rebate obligations, including under the Medicaid Drug Rebate Program.

***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 our targeted patient population, our revenue and ability to sustain 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 ARCALYST and any other of our current or future product candidates, if approved, 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, the country where substantially all ARCALYST sales occur, 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 expenditures 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 – Current and future healthcare legislation and executive or administrative action 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.

***Any future growth outside of the United States would be subject to additional regulatory burdens and other risks and uncertainties.***

We are currently authorized to market ARCALYST, our sole product, only in the United States, where we derive substantially all of our revenue. Our future growth 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, 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 United States Foreign Corrupt Practices Act (the "FCPA"), the UK Economic Crime and Corporate Transparency Act 2023 or similar foreign regulations;
- different medical practices and customs in such countries affecting acceptance in the marketplace;
- import or export licensing requirements;
- tariffs, taxes and other restrictions on international trade;
- 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; and
- foreign currency exchange rate fluctuations.

In some countries, particularly 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 or in population groups not previously observed. 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 expenditures 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 regulatory 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. In addition, approvals may come with potentially burdensome conditions. 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.

The pharmaceutical manufacturing process is highly regulated. 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 ("cGMP") or similar foreign regulations. Before approving a BLA, the FDA will typically conduct a pre-approval inspection of the manufacturing facilities for the product to determine whether the manufacturing processes and facilities comply with cGMPs. The FDA will not approve a product unless it determines that the manufacturing processes and facilities comply with cGMP requirements and are adequate to ensure consistent production of the product within required specifications. After approval, the FDA conducts periodic inspections of manufacturing facilities. Accordingly, we and our CDMOs and others with which we work must continue to expend time, money and effort in all areas of regulatory compliance, including manufacturing, production and quality control. In addition, to the extent that regulatory and policy changes implemented by the current presidential administration cause reductions in the FDA's workforce or budget, or other disruptions that affect the FDA's ability to exercise its regulatory authority, the ability of the FDA to conduct its pre- or post-approval inspections of the manufacturing facilities on which we rely may be impacted, which could delay or limit the ability of the FDA to provide the approvals necessary for the manufacture and/or sale of our current and future products and product candidates.

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. The FDA and other regulatory authorities may place other conditions on approvals including a Risk Evaluation Mitigation Strategy

("REMS") or similar risk management measures, to assure the safe use of the product. If the FDA or other regulatory authority concludes a REMS or similar risk management measures are needed, the applicant of the BLA or Marketing Authorization Application ("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 and quality 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 the patient 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 ordinary shares to decrease.

Additionally, we may in the future seek an accelerated approval for one or more of our product candidates. Under the accelerated approval program, the FDA may grant accelerated approval to a product candidate designed to treat a serious or life-threatening condition that provides meaningful therapeutic benefit over available therapies upon a determination that the product candidate has an effect on a surrogate endpoint or intermediate clinical endpoint that is reasonably likely to predict clinical benefit. As a condition of approval, the FDA requires that a sponsor of a product receiving accelerated approval perform one or more post-marketing confirmatory clinical trials. An unsuccessful confirmatory trial or failure to complete such a trial could result in the withdrawal of marketing approval. The FDA also requires as a condition of accelerated approval the pre-submission of promotional materials for FDA review.

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 we fail to comply with such requirements; 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; if problems arise with the facility where a product is manufactured; or if a regulatory agency disagrees with the promotion, marketing or labeling of a product, such regulatory agency may impose restrictions on that product or us, including requiring suspension of sales and 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; fail to comply with regulatory requirements; or a regulatory agency or enforcement authority disagrees with the promotion, marketing or labeling of our products, such 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;
- require us to withdraw or correct our marketing materials; or
- seize or detain products or require a product recall.

Any government investigation of alleged violations of law and regulations could require us to expend significant time, cost and resources in response, and could generate negative publicity or reputational harm. 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, impose restrictions on such product or its manufacture or require us to recall or remove such 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 an affected 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 are subject to extensive healthcare regulation and enforcement by various government entities, and our failure to strictly adhere to these regulatory requirements could have a detrimental impact on our business.***

The development and marketing of pharmaceutical products and related arrangements with healthcare professionals, third party payors, patients, and other third parties in the healthcare industry are subject to a wide range of 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.

Given the broad scope and evolving government interpretation and enforcement of these laws, our business activities could be subject to challenge under one or more of such laws. 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.

In addition, the development of our marketing and sales capabilities has required, and will continue to require, significant financial and management resources. Our direct sales and marketing efforts may not be successful or may be limited by future government policies or initiatives. For example, the current Secretary of the Department of Health & Human Services has expressed interest in banning direct-to-consumer advertising for prescription drugs. Several bills have been introduced in the current Congress that are aimed at reducing or eliminating direct-to-consumer prescription drug advertising across all media platforms. Although a ban on direct-to-consumer advertising would require legislative action, the FDA may implement regulatory or policy changes that materially limit our ability and that of our third party contractors to promote our products to consumers, which could materially impact our business. The current FDA Commissioner has indicated that the agency is closely reviewing direct-to-consumer prescription drug advertisements to ensure that they comply with the FDA's requirements.

Ensuring that our business arrangements with third parties comply with applicable healthcare laws and regulations may involve substantial costs. Any action against us for violation of these laws, even if we successfully defend against it, could cause us to incur significant legal expenses and divert our management's attention from the operation of our business. 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

***If we are unable to advance our product candidates in clinical development and obtain regulatory approval, or experience significant delays in doing so, our business may be significantly harmed.***

Our product candidates are in various stages of clinical development. We base our projections about the future development and potential approval of our product candidates on indirect data from other companies and the results of our preclinical and clinical trials, but ultimate success is uncertain and involves significant risk.

We cannot be certain that any of our product candidates will be successful in their clinical trials. We also cannot be certain that they will receive regulatory approval, even after completing a successful pivotal clinical trial. We may also choose to cease development of a product candidate prior to conducting a pivotal trial for any reason, including capital conservation purposes. We may also choose not to commercialize a product candidate that has completed a pivotal trial or received regulatory approval, for a number of reasons, including commercial viability. Such decisions may be for a particular indication or be for the product candidate entirely. In the event that a product candidate is unsuccessful in its clinical trials, fails to receive regulatory approval or is unviable for another reason, our business may be materially harmed by limiting our ability to recoup our development expenses through a successful commercial launch.

Each of our product candidates requires substantial preclinical or clinical development and manufacturing support as part of our product development strategy. The clinical success of our current and future product candidates depends upon several factors, including, but not limited to, the following:

- submission to and authorization to proceed with clinical trials by the FDA under investigational new drug applications ("INDs"), EU Clinical Trials Regulations (the "CTR") 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 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;
- 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 before and 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 may harm our ability to continue our operations by limiting our potential commercial prospects. In such an instance, we may need to seek capital elsewhere. See "*Risk Factors – General Risk Factors – We have a history of operating losses and may require substantial additional financing in the future."*

***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 the 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, leading us to abandon our pursuit of a particular indication or the development of a product candidate entirely. 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 – If we are unable to advance our product* candidates *in clinical development and obtain regulatory approval, or experience significant delays in doing so, our business may 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 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; 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. Any of the foregoing 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"), an ethics committee and/or other applicable committees for each clinical trial site before a trial may be initiated, which approval could be delayed, rejected or suspended. 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 national competent authorities of the EU member states, or other applicable regulatory authorities in other countries for each product candidate and, consequently, is a prerequisite to us 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, require redesign, have timely site activation and participant enrollment or 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, including due to competition from other companies' clinical trials for a particular indication, 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 global conflict;
- 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, including the cost to manufacture product candidates or acquire important ancillary products, 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 applicable 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 have occurred and may occur in the future. 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 national competent authorities, the EMA, the UK Medicines and Healthcare products Regulatory Agency (the "MHRA") 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, the EU, the UK 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, the EU and the UK may subject us to risks associated with the engagement of non-United States, non-EU and non-UK CROs who are unknown to the FDA, the EMA or the EU national component authorities or the MHRA and may have different standards of diagnosis, screening and medical care. Such trial sites may also incur 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, the EU and the UK.

In addition, the FDA's and other regulatory authorities' policies with respect to clinical trials may change and additional government regulations may be enacted. Such changes may require us to dedicate time, resources and capital to comply and, if we are unable to do so effectively or on a timely basis, our development plans may be impacted and our business may suffer material harm.

***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.***

The timely completion of clinical trials in accordance with their protocols depends, among other things, on our ability to enroll a sufficient number of patients who remain in the trial until its conclusion, particularly given that many of the conditions for which we are evaluating our current product candidates or may evaluate in the future are in small disease populations. Accordingly, when we encounter 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 fact that our product candidates modulate the immune system and carry unique risks associated with immunosuppression, including the risk of serious infections, potential interference with vaccines and other potential serious health risks;
- the availability and nature of competing clinical trials;
- the availability of standard of care and/or new or existing drugs approved for the indication the clinical trial is investigating;
- failure to obtain, maintain and/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 participants 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.

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 being developed or commercialized by third parties show the same or similar side effects as those we observed in our trials but to a greater degree or report new previously-unreported side effects, it could have an impact on the entire class of molecules, and the applicable regulatory authority may modify, suspend, or terminate our clinical trials; not authorize further clinical trials; require post-marketing clinical trial commitments or safety monitoring (e.g., REMS); or even suspend commercialization of any products or product candidates, as applicable, that contain a molecule within such 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 in the Huadong Territory. 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 United States 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 (e.g., our collaborative study agreement exploring ARCALYST as a treatment for cardiac sarcoidosis) or investigator initiated studies could increase the possibility of generating adverse safety results that impact our commercialization of such products or 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 after use by a significantly larger number of patients, including patients with different demographic characteristics than those that participated in our clinical trials. If we or others later identify undesirable side effects caused by our products or 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 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 professionals 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 candidates 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 ordinary 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 legislation, regulation or policy governing the development, approval and marketing of biological products may cause delays in our plans for submitting marketing applications and obtaining approvals for such applications, or we may be unable to obtain marketing approvals. For instance, comprehensive proposals have been made for the complete overhaul of the existing EU pharmaceutical legislation, 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 received a positive first reading of the European Parliament and in June 2025, the European Council adopted its position on the legislative proposal. Following such adoption, negotiations will take place between the European Parliament and the European Council for the final legislative text to be agreed through the so-called "co-decision" legislative process. It is unlikely that the new law will be adopted through the EU legislative process before 2026. When adopted, the new law 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, MAA or other submission or to obtain regulatory approval in the United States, Europe 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 believe that we have not 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 conclude that on-site inspections and data audits have not 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 policies, regulations and guidelines of regulatory authorities regarding the development, approval and marketing of biologic products may significantly change, including in the United States, as a result of the 2025 change in presidential administration, which may render our clinical data, biologic manufacturing process and other supporting information insufficient for approval or restrict us from marketing our product candidates in the manner in which we anticipate.

In addition, even if we were to obtain approval for one or more of our current or future product candidates, regulatory authorities may approve such 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 United States 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 biosimilar 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. 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. Even if we obtain Orphan Drug designation for certain product candidates for a particular orphan indication in the United States or the EU, we may not be the first to obtain marketing approval for such orphan indication due to the uncertainties associated with developing pharmaceutical products. In such case, subject to applicable laws in those jurisdictions, Orphan Drug exclusivity may no longer be available for our product candidates, if approved, unless we can show a significant benefit over the already approved orphan drug. Moreover, in the event our drug is deemed similar to the first approved orphan drug, we may be denied regulatory approval for our drug in such orphan indication for the duration of the Orphan Drug exclusivity period.

Regulatory authorities in some jurisdictions, including the United States and the EU, may designate drugs or biologics intended to treat small patient populations as Orphan Drug products, which are subject to a number of region-specific (e.g., tax credits, user fee exemptions and potential market exclusivity) rules and regulations.

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, which, if granted, offers the potential for a rolling review of a BLA if a number of conditions are met, which would allow data to be submitted and reviewed as they become available rather than waiting for the full data package to

become available to be submitted. Rolling review is often faster than the FDA's standard review process. 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 a PRIME designation from the EMA, a conditional MA or other designations, schemes or tools 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 a PRIME (Priority Medicines) designation from the EMA, a conditional MA or other designations, schemes or tools for one or more of our product candidates, each of which offer incentives similar to a United States Breakthrough Therapy designation. 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 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.

The 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.

***Due to the recent change in presidential administration, we face substantial uncertainty regarding potential regulatory developments in the United States that may adversely affect our business.***

We face substantial uncertainty regarding the potential for changes in the regulatory environment in the United States following the change in presidential administration in January 2025. While many of the current administration's policies have been focused on deregulation, the new administration and federal government could adopt legislation, regulation or policies that adversely affect our business, including by making it more difficult to continue to market ARCALYST or by creating a more challenging and costly environment to pursue the development and commercialization of our current or future product candidates. For example, the federal government, including the FDA, may implement legislative, regulatory or policy changes regarding the standards for approving biologic products that we may be unable to satisfy or changes regarding the marketing of approved biologics that may limit or prohibit the advertising and promotion of ARCALYST and, if approved, our current or future product candidates. The impending uncertainty could present new challenges or potential opportunities as we navigate the clinical development and approval process for our product candidates.

The current administration has also undertaken significant efforts to reduce the size and spending of the federal government, including at the FDA. A significant reduction in the FDA's workforce or the FDA's budget, or other disruptions at the FDA, could materially impact the FDA's ability to engage in a variety of activities that may affect our business, including routine regulatory and oversight activities. The current administration has substantially reduced the FDA's workforce and may make further reductions, which may lead to disruptions and delays in the FDA's review and oversight of our product candidates and impact the FDA's ability to provide timely feedback on our development

programs. Additionally, although it has been reported that there have not been reductions in workforce in the review or inspection divisions, any such reductions could extend review timelines, delay or prevent pre-approval inspections, and limit opportunities for FDA feedback on pending applications. Any of these actions may delay or limit our ability to obtain FDA approval and commercialize our product candidates, including our ability to obtain regulatory approval for the technology transfer of ARCALYST drug substance manufacturing to Samsung.

Furthermore, the current administration has discussed several changes to the reach and oversight of the FDA, which could affect its relationship with the pharmaceutical industry, transparency in decision making and ultimately the cost and availability of prescription drugs.

For additional discussion on risks related to the current administration's policy agenda, see "*Risk Factors – General Risk Factors – Current and future healthcare legislation or executive or administrative action may have a material adverse effect on our business and results of operations.*"

**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, which is highly regulated and complex, 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, package and ship our products and product candidates at sufficient quality and quantity to support our and our collaboration partners' commercialization and research and development efforts.

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. 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 products and 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.

Our reliance increases the risk that we will have insufficient quantities of ARCALYST and our product candidates or that ARCALYST and our product candidates may not be produced at an acceptable cost or quality; or that production may not be done in a timely manner due to, for example, deviations in operations or manufacturing facility control, or production interruptions caused by equipment failure and an inability to source adequate replacement parts and equipment; any of 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 materially impact our ability to source sufficient ARCALYST material to cover our needs. If, in the future, we are unable to source sufficient finished material, we may stock out or otherwise be unable to meet patient demand for ARCALYST, adversely affecting our business, results of operations and financial condition. In addition, 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. Further, reagents used for the analytical testing of ARCALYST have and may in the future become outdated, requiring qualification before new reagents may be used. These issues may be exacerbated by increased clinical or

commercial demand by us or our collaboration partners, or if we decide to develop ARCALYST in one or more additional indications or in additional territories.

We may be unable to adequately address current and future issues with the ARCALYST manufacturing process, which could prevent additional finished material from being distributed in a timely manner or within specifications. If we are unable to source additional commercial supply of ARCALYST, if needed, 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 is the sole manufacturer of ARCALYST drug substance and will remain so until we complete the technology transfer of the manufacturing process for ARCALYST drug substance to Samsung. 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 (including increased demand arising from our need to replace material lost to manufacturing issues). 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 we have with Regeneron. Further, we rely on a third party CDMO to package and label ARCALYST. Our reliance on Regeneron (including its respective CDMOs) and our other CDMO as our sole manufacturers and/or service providers means that we do not have control over ARCALYST 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. Many of these risks may still be present after successful completion of the technology transfer of ARCALYST drug substance manufacturing and there is no guarantee that such technology transfer will materially diminish our ARCALYST manufacturing risk profile.

We have qualified or engaged, as applicable, CDMOs to produce our clinical product candidates. 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 are party to a collaboration agreement with Huadong for ARCALYST. Until such time as Huadong is able to manufacture ARCALYST, either on its own or through a third party CDMO, we are its only source of drug supply. If our current supplier of drug substance and drug product for ARCALYST 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. Our replacement CDMO, Samsung, will utilize a modified manufacturing process from 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 Samsung 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, label and package ARCALYST and our current and future product candidates may be inspected by regulatory authorities in connection with the submission of our MAs to, and review by, regulatory authorities or based on their work for other clinical trial sponsors. While we provide oversight of

such 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 manufacturing, labeling, and packaging of current and future products and product candidates. If our CDMOs cannot successfully perform such functions in conformity with our specifications and the strict regulatory requirements of regulatory authorities, they will not be able to secure or maintain regulatory approval for their 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 manufacturing, labeling and packaging of our product candidates or if they withdraw any such approval in the future, we may need to find alternative facilities or CDMOs, 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 CDMOs, 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.

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; or an epidemic, 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 select 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, the costs for our products and product candidates, ancillary products and placebo may be affected by increased tariffs implemented by the current presidential administration. See "*Risk Factors — General Risk Factors – Changes in United States trade policy, including tariffs imposed by the United States, and any reciprocal tariffs imposed in response, could materially impact our business and results of operations.*"

Our products and product candidates may also compete with other product candidates and approved products for access to and capacity within manufacturing, packaging, and labeling facilities. There are a limited number of CDMOs that operate under cGMP regulations and that might be capable of performing such functions for us. Furthermore, given the limited capacity at such CDMOs and the long lead times needed to reserve capacity, CDMOs may require monetary commitments in connection with such reservations as well as fees for changes or cancellations or additional fees to accommodate expediting of manufacturing, packaging, and labeling. For our product candidates, we may wait to reserve capacity until we can be informed by data from the clinical trials of our product candidates, which may take several months. 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 capacity "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 Samsung, following the technology transfer of ARCALYST drug substance manufacturing, 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 CDMOs 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 them. Although we believe that there are several potential alternative CDMOs who could provide the services we currently contract for, 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, establishing replacement CDMOs 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 contractor 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 contractor, such replacement contractor 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 Samsung 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. Since then, we have worked to qualify Samsung, who will serve as the new manufacturer of ARCALYST drug substance, and new CTLs who will serve as the new testing labs of ARCALYST drug substance and drug product. We have also contracted with Samsung to document the technology transfer and enable the commercial manufacturing of ARCALYST drug substance should the technology transfer succeed.

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 Samsung as our replacement CDMO and have selected replacement CTLs, we are still in the technology transfer process and still must determine whether Samsung and such 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 Samsung will be manufacturing ARCALYST drug substance at a new manufacturing site and with a potentially different manufacturing process, and such CTLs will 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 replacement CDMO be able to manufacture drug substance at sufficient levels of comparability with the materials produced by the original manufacturer. We are still in the process of confirming comparability between the drug substance produced by Samsung and the drug substance produced by Regeneron. 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.

In addition, because the Samsung manufacturing facility is located in South Korea, unlike Regeneron's United States-based manufacturing facility, we may face new risks arising from tariffs, import/export restrictions, customs proceedings, product being lost or damaged during international shipping, differing regulations, supply chain interruptions and other risks inherent to international operations. See "*Risk Factors — General Risk Factors – Changes in United States trade policy, including tariffs imposed by the United States, and any reciprocal tariffs imposed in response, could materially impact our business and results of operations.*" These risks, should they occur, could increase our costs and affect our ability to meet clinical and commercial demand for ARCALYST, which could materially impact our business, financial condition and results of operations.

Regeneron is contractually obligated to continue manufacturing ARCALYST drug substance for at least a portion of the time that it will take us to qualify Samsung as 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 Samsung 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 technology transfer 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.

***The third parties upon whom we rely for the supply of 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 and KPL-387 are supplied to us from single-source suppliers and we obtain the drug substance and drug product used in abiprubart from a limited number of sources. For KPL-1161, we plan to manufacture drug substance in our in-house manufacturing facility and use a single supplier to manufacture drug product. While our in-house manufacturing capabilities have the limited capabilities to produce pre-clinical and early-stage clinical drug supply, we lack internal large-scale manufacturing capabilities necessary to support commercial requirements. Regeneron is currently our sole source manufacturer of ARCALYST drug substance and will remain so until we qualify Samsung as a replacement CDMO. We expect that Samsung will be our sole source manufacturer of ARCALYST drug substance following such qualification. 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 Samsung 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.*" Further, we have historically outsourced all ARCALYST packaging and labeling activities to a single CDMO and expect to do so for any future approved products. 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 and package and label such drugs, as applicable, in each case 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, we do not currently have arrangements in place for a redundant or second-source manufacturer of drug substance or drug product, or a redundant or second-source packager and labeler, in the event any of our current vendors cease or have a substantial delay in their operations or stop offering us sufficient quantities of these materials for any reason, as applicable. With respect KPL-387, KPL-1161 and abiprubart, while we anticipate having more than one source for drug substance now or in the future, as applicable, such sources are nonetheless limited and subject to similar risks as our other products and product candidates.

We are not certain that our suppliers will be able to meet our demand for our products and product candidates, 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 given their manufacturing capacity constraints. 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. 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 such drugs 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.

***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.

***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;
- 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 or unsuccessful. In addition, 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.

**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. ARCALYST currently faces competition in its CAPS and DIRA indications and is facing potential future competition in its recurrent pericarditis indication. KPL-387 is being developed for recurrent pericarditis and we believe, if commercialized, would likely face additional competition from drugs that may offer either more convenient dosing methods or frequencies than what is currently available. We have not yet announced an indication for KPL-1161, but expect that it will compete with a number of drugs that inhibit IL-1 or other mechanisms. Competition may come from major pharmaceutical companies, specialty pharmaceutical companies and biotechnology companies worldwide, each of whom may market and sell drugs or biologics or pursue 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.

**ARCALYST**

We are not aware of any other FDA-approved therapies for recurrent pericarditis, but we are aware of several competitors developing treatments for this indication. CardiolRx is an oral cannabidiol being developed by Cardiol Therapeutics in a Phase 3 clinical trial, targeting patients who are discontinuing treatment with an IL-1 blocker. R-Pharm International is developing goflikicept, which inhibits IL-1α/IL-1β-induced signaling. This drug is approved and marketed in Russia but with no currently active trial in the United States. Ventyx Biosciences is developing VTX2735, which is 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. In addition to their development program in CAPS, Ventyx Biosciences is conducting a Phase 2 trial of VTX2735 in recurrent pericarditis, which began in January 2025. Monte Rosa Therapeutics is developing MRT-8102 and has announced plans to conduct Phase 1 proof-of-concept studies in individuals with high levels of C-reactive protein and in individuals with cardio-immunology indications, such as pericarditis, among other prospective indications.

There are also drugs that inhibit IL-1β-induced signaling but do not inhibit IL-1α-induced signaling. 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), FMF, AOSD, Systemic Juvenile Idiopathic Arthritis (SJIA) and gout flares.

We are also aware of several other molecules that do not directly compete with our approved indications for ARCALYST but nonetheless target IL-1α and/or IL-1β directly or indirectly. Clinical stage development programs targeting IL-1α and/or IL-1β directly or indirectly via the NLRP3 inflammasome include: lutikizumab (by Abbvie for the treatment of hidradenitis suppurativa, ulcerative colitis, atopic dermatitis and Crohn's Disease); ZYIL-1 (by Zydus Lifesciences in amyotrophic lateral sclerosis); HT-6184 (by Halia in myelodysplastic syndromes, inflammatory pain); OLT1177 (by Olatec Therapeutics in osteoarthritis of the knee and acute gout flares); DFV-890 (by Novartis in FCAS); Selnoflast (by Roche, no indications announced); NT-00249 (by NodThera, no indications announced); NT-0796 (by NodThera in obesity); Somalix and Inzomelid (by Roche, no indications announced); VTX-3232 (by Ventyx Bioscience in Parkinson's); SSGJ-613 (by Sunshine Guojian Pharmaceuticals in acute gout); Natrunix (by XBIOTECH Inc in

rheumatoid arthritis, axial spondylarthritis); AVTX-009 (by Avalo Therapeutics in hidradenitis suppurativa); CAN-10 (by Cantargia in healthy subjects and plaque psoriasis) and VENT-02 (by Ventus Therapeutics, no indications announced). There are also therapies that modulate IL-1α-induced signaling in preclinical and clinical development for diseases other than recurrent pericarditis from Johnson & Johnson and XBIOTECH USA, INC.

**KPL-387**

Since we currently expect to develop KPL-387 for the treatment of recurrent pericarditis, we believe that it will compete with the same assets as those described above, which includes a number of drugs that target IL-1α and/or IL-1β. For recurrent pericarditis, there are a number of drugs in development that explore alternative dosing methods and frequencies to the once-weekly subcutaneous dosing method of ARCALYST, the only currently FDA-approved treatment. Both CardiolRx and VTX2735, in Phase 3 and 2 development by Cardiol Therapeutics and Ventyx Biosciences, respectively, are being investigated for oral administration. Goflikicept, currently marketed solely in Russia, though with prior clinical studies conducted in the United States, is dosed subcutaneously every two weeks following an initial loading dose. We expect that additional therapies offering even more convenient dosing, including those listed in the sections above, may enter the market, including therapies that offer quarterly dosing. For KPL-387 to succeed on a commercial basis, if approved, we expect that it will need to compete against such drugs by offering a more convenient dosing regimen and/or an improved risk-benefit profile than other available options.

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 and biotechnology 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. Further, our clinical trials may need to compete for participants and trial sites against other drugs in clinical development for the same indication. We believe the key competitive factors affecting the success of ARCALYST and any product candidates that we successfully develop and commercialize, are 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 biosimilar 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 portfolio. 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:

- we 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, partners or acquisition targets;
- 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; use of a substantial portion of our available cash for all or a portion of the consideration; or causing 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 our products and 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 products and 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 products and product candidates instead of developing or commercializing our products and product candidates ourselves. For

example, in February 2022, we granted Huadong exclusive rights to develop and commercialize ARCALYST 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. 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 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;

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

- 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 includes 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;

- 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;

- 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; 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.

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 recruitment and training of qualified personnel. This may result in weaknesses of our systems and infrastructure; 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. Our future financial performance and our ability to commercialize our product candidates, if approved, 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. 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 our business. We

also rely on trade secrets, proprietary know-how and continuing technological innovation to develop and maintain our proprietary or 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 rights include 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 and an exclusive license under our license agreement with BIDMC to patent applications and patents related to abiprubart.

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. As with 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 our current and future products and product candidates. A United States patent covering ARCALYST as a composition of matter expired in 2020, and relevant composition of matter patents issued outside of the United States expired in October 2023. A United States 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. We are unable to obtain composition of matter patents covering amino acid sequences, or corresponding nucleic acid sequences, of KPL-387. We own a pending patent application covering methods of using KPL-387 in the treatment of recurrent pericarditis. If issued, patents covering methods of using KPL-387 in treating recurrent pericarditis will have statutory expiration dates in 2046, not including any patent term extensions or adjustments. The issued composition of matter patents for abiprubart owned by us have statutory expiration dates in 2036, not including any extensions. The issued composition of matter patents licensed from BIDMC related to abiprubart have statutory expiration dates in 2032, not including any patent term extensions or adjustments.

In the United States, the natural (i.e., statutory) 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 that 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 and Europe covering ARCALYST as a composition of matter have expired. We are unable to obtain composition of matter patents covering the amino acid sequences, or corresponding nucleic acid sequences, of KPL-387. 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, such as data exclusivity or orphan exclusivity as applicable, 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 are party to agreements granting us the rights to develop and commercialize ARCALYST, abiprubart and vixarelimab. Each of these agreements requires us to use commercially reasonable efforts to develop and commercialize such drugs, make timely milestone and other payments, provide certain information regarding our activities with respect to such drugs 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. Further, disputes may arise between us and any of these counterparties regarding such obligations under, or the intellectual property subject to, such agreements, including:

- our diligence obligations to develop and commercialize the licensed technology, and what activities satisfy those diligence obligations;
- the scope of rights granted under the agreement and other interpretation-related issues;
- whether our use of the licensed technology is within the scope of the rights granted to us or otherwise consistent with the agreement;

- our obligations to make milestone, royalty or other payments under those agreements;
- other parties' performance being maintained under these 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 and vixarelimab, including the ARCALYST Huadong Collaboration Agreement 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. These collaborations may be subject to a number of risks, including those listed under "*—Risks*

*Related to Competition, Executing our Strategy and Managing Growth – 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 products and 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 products and product candidates"* above.

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.

***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 and antibody-related technologies. 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 abiprubart. If the claims of any of these patents are asserted against us, we do not believe our proposed activities related to abiprubart 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 United States patent in court, we would need to overcome a statutory presumption of validity that attaches to every United States 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 products and product candidates treat or are being developed to treat indications 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 products or 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 products or 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 our 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 ordinary 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 their 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. Current and proposed patent reform in the United States and other countries may contribute to those uncertainties and costs.

The Supreme Court of the United States 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, future actions by the United States Congress, the United States Courts, the USPTO and relevant law-making bodies in other countries 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. For example, with respect to patent term adjustment, the Federal Circuit's recent holding in *In re Cellect, LLC*, 81 F.4th 1216 (Fed. Cir. 2023), that obviousness-type double patent analysis for a patent that has received patent term adjustment must be based on the expiration date of the patent after the patent term adjustment has been added, may negatively impact the term of certain United States patents.

Finally, Europe's 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, by default automatically fall under the jurisdiction of the UPC. The UPC provides 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. 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.

Depending on future actions by governmental authorities, including legislative bodies, administrative authorities and court systems, the laws and regulations governing patents could change in unpredictable ways that would weaken our ability to obtain new patents, or may weaken the patent rights of existing patents in certain situations or to enforce our existing patents and patents that we might obtain in the future. If such an event were to occur, our business, financial condition, results of operations and future prospects may be adversely affected.

***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 be unable 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 be unable 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 be unable 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.

***If our trademarks and trade names are not adequately protected, then we may be unable 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 be unable 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.***

Although the trademark ARCALYST has been registered by Regeneron (and we have permission to use it pursuant to the Regeneron License Agreement), 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

***We have a history of operating losses and may require substantial additional financing in the future.***

Biopharmaceutical product development is a highly speculative undertaking and involves a substantial degree of risk. Our ability to generate product revenue sufficient to sustain our organization will depend heavily on a number of factors, including the continued commercialization of ARCALYST, the development and eventual commercialization of one or more of our current or future product candidates, if approved, and the management of our costs consistent with our current operating plan. Our future capital expenditures are expected to be substantial, and we may incur operating losses in the future if we encounter greater than expected expenses as we:

- support our sales, marketing and distribution capabilities, infrastructure and organization to commercialize ARCALYST and any product candidates for which we may obtain marketing approval;
- conduct new and ongoing research and pre-clinical and clinical development of our product candidates, including our Phase 2/3 clinical trial of KPL-387 in recurrent pericarditis, our ongoing Phase 1 clinical trial of KPL-387 in normal healthy volunteers and our pre-clinical investigations of KPL-1161;
- manufacture our products and product candidates for clinical or commercial use, increase our manufacturing capabilities, add additional manufacturers or suppliers and perform activities related to our technology transfer of the process for manufacturing ARCALYST drug substance;
- seek regulatory and marketing approvals for our product candidates that successfully complete clinical trials, if any;
- identify, assess and study new or expanded indications for our products and product candidates and/or new or alternative dosing levels, dosing frequencies or administrations of our products and product candidates;
- make milestone or other payments under any current or future license, acquisition, collaboration or other strategic transaction agreements;
- seek 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;
- seek to identify, assess, acquire or develop additional product candidates;
- address any 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;
- enter into licensing, acquisition, collaboration or other strategic transaction agreements;
- seek to maintain, protect and expand our intellectual property portfolio;
- seek to attract and retain skilled personnel;
- create additional infrastructure to support our product development and commercialization efforts; and
- experience delays or encounter 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 disruptions to the national or global economy.

Further, our financial results 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 may not be sustained in subsequent periods.

If we are unable to fund our operations through commercial ARCALYST 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 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.

Financing our activities also carries risk. The sale of additional equity or convertible securities would dilute all of our shareholders. Further, new investors could gain rights superior to our existing 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.

***If we fail to comply with reporting and payment obligations under the MDRP 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, including the Medicaid Drug Rebate Program (the "MDRP"), the Federal Supply Schedule (the "FSS") and the PHS 340B Drug Pricing Program. If we are found to have violated the requirements of such programs, 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. The Centers for Medicare & Medicaid Services 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.

***Current and future healthcare legislation or executive or administrative action may have a material adverse effect on our business and results of operations.***

In the United States, the UK, the EU and other jurisdictions, there have been and we expect there will continue to be a number of executive, legislative and regulatory initiatives and proposed changes to the healthcare system generally, and drug pricing and reimbursement policies particularly, that could affect our operations.

Under the current United States presidential administration, there have been significant and wide-ranging reforms to federal policy and the federal government. In particular, as in recent preceding presidential administrations, drug pricing and reimbursement reform have been a particular area of focus. For example, President Trump issued an Executive Order in April 2025 with multiple directives aimed at lowering drug prices, including refining the Medicare drug price negotiation program established by the Inflation Reduction Act of 2022; accelerating competition for high-cost prescription drugs by accelerating approval of generics and biosimilars and facilitating the process for re-classifying prescription drugs as over-the-counter drugs; and increasing drug importation. As another example, in May 2025, President Trump issued another Executive Order that directed government agencies and officials to identify most-favored nation pricing targets for prescription drugs (and looked to pharmaceutical manufacturers to make significant progress towards delivering target prices to patients); prevent foreign countries from disproportionately shifting the cost of global pharmaceutical research and development to the United States; and facilitate direct-to-consumer purchasing

programs for pharmaceutical manufacturers to sell their products to patients at the most-favored-nation price. Many of these reform initiatives will require additional legal and/or administrative action to implement.

Other healthcare reform efforts or other actions under the current presidential administration may adversely affect the development of new drug therapies, access to healthcare coverage or the funding of health care benefits, although the full impact of such efforts or actions cannot be predicted. For example, the Congressional Budget Office has estimated that recent restrictions in Medicaid eligibility and reductions in funding would increase the number of uninsured Americans by millions. In addition, Congress passed legislation expanding the orphan drug exclusion in the Medicare drug price negotiation program. As another example, reductions in the workforce at healthcare agencies may potentially limit or delay agency action, including action related to regulation on the development and marketing of drug products or the coverage and reimbursement of drug products.

The nature and extent of future healthcare reforms cannot be predicted. There is ongoing uncertainty regarding the nature or impact of any healthcare reform implemented by the current presidential administration through executive or administrative action or by Congress and the extent to which such actions may be subject to litigation or other challenges. As a result, there can be no assurance that the reform will not have a significant adverse impact on our business.

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

***Our information technology systems, or those of our third party CDMOs, CROs, specialty pharmacies, third party logistics providers and other contractors, consultants and service providers, may fail or suffer cyberattacks or security incidents, which could result in a material disruption of our or such third party's business or operations, impede our development programs for our product candidates or materially impact our ability to commercialize our products.***

Despite the implementation of security measures, our information technology systems and those of our third party CDMOs, CROs, specialty pharmacies, third party logistics providers and other contractors, consultants and service providers 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. Technologies such as artificial intelligence and machine learning are additionally being used to create more sophisticated attacks on targets, including targeted social engineering attempts. We may also face increased cybersecurity risks due to our reliance on internet technology and the number of our employees, such as our commercial field force, 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, because we have outsourced elements of our information technology infrastructure to vendors, such vendors may or could have access to our confidential information. A cyberattack 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 incidents may remain undetected for an extended period. Even if identified, we may be unable to adequately investigate or remediate incidents or cyberattacks 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 current or past significant system failure, accident or security incident that has materially affected or would be reasonably likely to materially affect us, including our business strategy, results of operations or financial condition 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, specialty pharmacies, third party logistics providers and other contractors, consultants and service providers, the costs associated with the investigation, remediation and potential notification of a cyberattack to counter-parties and data subjects could be material. A cyberattack could result in a material disruption of our or such third party's business or operations. 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 incident results in a loss of or damage to our data or applications or inappropriate disclosure or theft of confidential or proprietary information, the further development of our product candidates could be delayed. Further disruptions to our or our third party providers' infrastructure may inhibit our ability to commercialize ARCALYST through, among other things, interruptions in our logistics fulfillment, loss of patient and prescriber information, interruptions in our ability to communicate with the third party providers upon which we rely and impairments in our ability to service our patients and address their concerns. Any of these events could adversely impact our business and ability to generate product revenue. Although we maintain cybersecurity insurance coverage, it may not be adequate to cover all liabilities that we may incur from cyberattacks or security incidents 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 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; impede our ability 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.

In the United States, most healthcare professionals, including research institutions from which we obtain patient health information, are subject to privacy and security regulations promulgated under United States federal Health Insurance Portability and Accountability Act, as amended ("HIPAA"). We may obtain health information from third parties, such as research institutions with which we collaborate, that are subject to privacy and security requirements under HIPAA. Although we are not directly subject to HIPAA, other than potentially with respect to providing certain employee benefits, we could be subject to criminal penalties if we knowingly obtain or disclose individually identifiable health information maintained by a HIPAA covered entity in a manner that is not authorized or permitted by HIPAA. In the event that we are subject to or affected by HIPAA, any liability from failure to comply with the requirements of these laws could adversely affect our financial condition.

In addition, 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, as amended by the California Privacy Rights Act (together, the "CCPA") gives California residents expanded rights to access, correct, and delete their personal information, opt out of certain personal information sharing, receive detailed information about how their personal information is used and also imposes limitations on data uses, new audit requirements for higher risk data and opt outs for certain uses of sensitive data. 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 created a California data protection agency authorized to enforce the CCPA and issue substantive regulations, which 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. The Washington My Health My Data Act, which is applicable to companies doing business in Washington or targeting products or services to consumers in Washington, imposes disclosure and consent requirements, among other things, with respect to broadly defined consumer health data, and is enforceable through consumer class actions. Additional compliance investment and potential business process changes may also be required.

Furthermore, the United States 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 European Union General Data Protection Regulation 2016/679 ("GDPR"), and legislation of EU member states and EEA countries. 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 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 or 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.

The Swiss Federal Act on Data Protection (the "DPA") also applies to the collection and processing of personal data by companies located in Switzerland, or in certain circumstances, by companies located outside of Switzerland. The DPA may lead to an increase in our costs of compliance, risk of noncompliance and penalties for noncompliance as we potentially expand our footprint in Switzerland.

Failure or perceived failure to comply with the GDPR, the UK GDPR, the DPA 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. In addition, we make public statements about our use, collection, disclosure and other processing of personal data through our privacy policies and information provided on our website. Although we endeavor to comply with our public statements and documentation, we may at times fail to do so or be alleged to have failed to do so. If we or our third party CDMOs, CROs or other contractors, consultants or service providers fail to comply, or are perceived to have failed to comply, with applicable regulatory requirements, applicable policies or notices relating to privacy or data protection, contractual or other obligations to third parties, or any other legal obligations, laws, rules, regulations and standards relating to privacy or data protection, 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 investigation or enforcement action, litigation, claims or other proceedings could also generate adverse publicity, harm our reputation, result in significant liability and require that we devote substantial resources that could otherwise be used in other aspects of our business.

***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. An executive terminating their employment or taking an extended leave of absence without sufficient notice may leave a gap in the organization that we may be unable to fill on a timely basis, if at all. 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 corporate, 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 corporate, 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. In addition, laws and regulations may restrict our ability to attract, motivate and retain the required level of qualified personnel. For example, our business operations may rely on foreign personnel who require work permits. Any changes in immigration policies, work permit regulations, or visa requirements could adversely affect our ability to retain skilled employees. If work permits are denied, revoked, or not renewed, we may face disruptions in our operations, increased costs for hiring and training replacements, and potential delays in project execution. 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 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.

***Changes in United States trade policy, including tariffs imposed by the United States and any reciprocal tariffs imposed in response, could materially impact our business and results of operations.***

Changes to United States trade policy, including tariffs imposed by the United States on imported goods, as well as reciprocal tariffs that may be imposed by foreign governments, may adversely affect our business and the global

macroeconomic environment. In April 2025, the current presidential administration announced that the United States would impose baseline tariffs on all other countries and individualized reciprocal higher tariffs on countries with which the United States has purportedly high trade deficits, including South Korea. In addition, the administration has initiated or is considering imposing tariffs on certain types of foreign goods, and has announced plans to impose tariffs on pharmaceuticals, including pharmaceutical products and components manufactured outside of the United States. Such tariffs are currently paused; however, if enacted, they are expected to dramatically increase the price of goods imported into the United States, including pharmaceutical products and important products that are ancillary to the administration, processing or testing of pharmaceutical products. While we plan to conduct fill-finish and packaging for ARCALYST in the United States following the technology transfer of drug substance manufacturing to Samsung, we expect to import drug substance from South Korea into the United States following such date.

It remains unclear what the current administration or foreign governments will or will not do with respect to tariffs or other international trade agreements and policies. A trade war or other governmental action related to tariffs or international trade agreements or policies has the potential to disrupt our research activities, affect our suppliers, increase the cost of materials purchased to manufacture our products, and/or affect the United States or global economy or certain sectors thereof and, thus, could adversely impact our business. For example, any increase in prices of consumer goods may materially impact patient purchasing power, which could decrease patient compliance, duration of therapy or prevent patients from enrolling in ARCALYST therapy altogether. Such events may lead to a reduction in ARCALYST product revenue, which would materially impact our business and results of operations.

In response to the current presidential administration's announcement of its intention to impose tariffs, a number of countries and economic blocs have announced reciprocal tariffs on goods imported from the United States. While we currently do not sell ARCALYST outside of the United States, should we pursue commercialization in another region, such as the EU, we may be impacted by a number of factors, including increased costs for our product, more aggressive negotiations by private and public payers and lower consumer purchasing power.

There is substantial uncertainty with respect to what extent new (or modified) tariffs will be imposed in the long-term in the United States and other countries, or the ultimate impact such tariffs will have on us, our industry and the patients we seek to serve. We currently expect that tariffs, as proposed, will impact our ARCALYST gross margin by an immaterial amount because the price of ARCALYST drug substance that we expect to import from South Korea is a relatively small part of our overall cost of goods sold and because Kiniksa UK's Swiss branch office, which manufactures and sells ARCALYST, owns all of Kiniksa's ARCALYST intellectual property and, therefore, is not obligated to pay royalties to any other entity, which royalty payments would be subject to tariffs under the current presidential administration's proposed tariff scheme. However, the ultimate impact of tariffs, trade restrictions and resulting economic instability is uncertain and our business, financial condition and results of operations may be materially and adversely affected.

***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, terrorism, political unrest, outbreak of disease, labor disputes and boycotts;
- imposition of tariffs, 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 FCPA, 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.

***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. We may also receive pushback from other stakeholders regarding our initiatives related to ESG matters. If we do not successfully manage expectations across varied stakeholder interests, it could erode stakeholder trust or impact our reputation, and our financial results may suffer. In addition, even if we are effective at addressing such concerns, we may experience increased costs as a result of balancing competing interests related to ESG matters and executing upon our ESG goals, which costs may not be offset by any benefit to our reputation, and which could have an adverse impact on our business and financial condition.

Outside of the United States, continued global focus on ESG matters has resulted in the adoption of new laws and regulations, including reporting requirements imposed by the UK, which will impact the annual reports we are required to file in the UK as a result of the Redomiciliation. 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 ordinary 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, hurricanes, 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 Ordinary Shares

***The concentration of ownership of our Class B ordinary 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 ordinary shares, which shares are held primarily by entities affiliated with certain of our directors, and Class B1 ordinary 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 ordinary shares and may result in our Class A ordinary shares being undervalued.***

Each Class A ordinary share is entitled to one vote per Class A ordinary share and each Class B ordinary share is entitled to ten votes per Class B ordinary share. Our Class A1 ordinary shares and Class B1 ordinary 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 ordinary shares and Class B ordinary shares. As a result of the multi-class voting structure of our ordinary shares, our executive officers and certain other members of our senior management collectively control a substantial amount of the voting power of our ordinary shares and therefore are able to control the outcome of certain matters submitted to our shareholders for approval. As of June 30, 2025, the holders of Class A ordinary shares accounted for approximately 71% of our aggregate voting power and the holders of Class B ordinary shares accounted for approximately 29% of our aggregate voting power. Our executive officers and certain other members of our senior management hold Class A ordinary shares and Class B ordinary shares representing approximately 26% of our aggregate voting power as of June 30, 2025 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 ordinary shares, Class A1 ordinary shares or Class B1 ordinary shares as set forth in our articles of association. For example, as of June 30, 2025, entities affiliated with certain members of our directors could convert their Class A1 ordinary shares and Class B1 ordinary shares upon 61-days' prior written notice into Class A ordinary shares and Class B ordinary shares, respectively, which in the aggregate would result in such entities holding approximately 75% 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 ordinary shares and our Class B1 ordinary 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 ordinary 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 ordinary shares. Holders of our Class B ordinary shares, which have ten votes per share on most matters, may have significant control over 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 ordinary 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 ordinary shares may be volatile and fluctuate substantially, which could result in substantial losses for holders of our Class A ordinary 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 ordinary shares at or above the price they paid for their shares. The market price for our Class A ordinary 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, economic, political 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 war, 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 ordinary 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. In the past, following periods of volatility in the market, securities class action litigation has often been instituted against companies. Such litigation, if instituted against us, could result in substantial costs and diversion of management's attention and resources, which could materially and adversely affect our business and financial condition.

***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 ordinary 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 ordinary 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 ordinary shares could decrease, which in turn could cause the price of our Class A ordinary shares or its trading volume to decline.

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

A significant number of our Class A ordinary shares are issuable upon conversion of our Class B, Class A1, and Class B1 ordinary shares, subject to certain limitations on conversion. As of June 30, 2025, approximately 2.0 million Class A ordinary shares directly held by our executive officers and directors, inclusive of Class A ordinary shares issuable upon conversion of our Class B, Class A1, and Class B1 ordinary 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, 2025, there were approximately 13.9 million Class A ordinary shares subject to outstanding share options, PSOs, RSUs and PSUs 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 ordinary 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, 2025, on an as-converted to Class A ordinary shares basis, these shareholders collectively held approximately 33.9 million of our Class A ordinary shares. If any of these shareholders sell, convert or transfer, or indicate an intention to sell, convert or transfer, a

substantial amount of their ordinary shares (after certain restrictions on conversion or resale lapse), the market price of our Class A ordinary shares could decline.

Pursuant to our amended and restated investor rights agreement (our "Investors Rights Agreement"), certain shareholders are entitled to certain registration rights with respect our Class A ordinary shares, including Class A ordinary shares issuable upon conversions of our Class B, Class A1, and Class B1 ordinary shares and upon the exercise of certain rights to acquire Class A ordinary shares, or collectively registerable securities, under the Securities Act. As of June 30, 2025, on an as-converted to Class A ordinary shares basis, we have registered approximately 31.8 million Class A ordinary 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 ordinary shares basis, approximately 1.7 million Class A ordinary shares held by certain of our executive officers as of June 30, 2025. If any of these Class A ordinary shares are sold, or if it is perceived that they will be sold, in the public market, the market price of our Class A ordinary shares could decline.

***We have anti-takeover provisions in our articles of association that may discourage a change of control.***

Our articles of association 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 a limited number of reasons;
- limitations on the acquisition of more than 30% or more of our voting rights, except through certain defined permitted acquisitions;
- our multiclass ordinary share structure, which provides our holders of Class B ordinary 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 ordinary shares; and
- restrictions on the time period in which directors may be nominated.

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 ordinary 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 ordinary 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 ordinary shares will be the sole source of gain for our shareholders for the foreseeable future.

**Risks Related to Our Jurisdiction of Incorporation and Certain Tax Risks**

***As a result of increased shareholder voting requirements in the UK relative to Bermuda, we will have less flexibility with respect to our ability to issue new shares.***

Prior to the Redomiciliation, our principal holding company was incorporated in Bermuda. Under Bermuda law, a company's directors may issue, without shareholder approval, any authorized but unissued common shares. English law allows our shareholders to authorize the allotment of share capital which can be issued by our board of directors without further shareholder approval, but this authorization must be approved by our shareholders via an ordinary resolution from time to time (i.e., approval from shareholders holding more than 50% of the voting rights), with such authority capable of applying in respect of any period specified in such resolution up to a maximum of five years. At our annual meeting of shareholders in June 2025, our shareholders authorized us to issue new ordinary or preferred shares (up to a maximum of 35% of our outstanding shares on the record date for our 2025 annual meeting) for a period of five years from the date of our 2025 annual meeting. However, there is no guarantee that any subsequent authorizations will be approved. In the event that we do not receive such authorization, we would face significant impediments to our business and operations as we would be unable to use equity as consideration in business development deals or honor our outstanding employee equity awards without seeking shareholder approval in each case. Such a scenario would materially harm our business due to the administrative and financial costs related thereto.

Additionally, subject to specified exceptions, English law grants statutory preemptive rights to existing shareholders to subscribe for new issuances of shares for cash. English law requires that this opt-out must be renewed by the shareholders at least every five years, and we cannot guarantee that the opt-out of preemptive rights will always be approved. A waiver of pre-emption rights under English law requires approval of the shareholders holding at least 75% of the voting rights in an English company. At our annual meeting of shareholders in June 2025, our shareholders voted to approve such a waiver for a period of five years from the date of our 2025 annual meeting. If, in the future, we do not receive such a waiver, this would cause a material adverse effect due to the administrative and financial costs related thereto, as a waiver of preemption rights would need to be sought from shareholders in respect of each new issuance of shares, including each instance that an employee would seek to exercise their share options.

While both the general authority to allot and waiver of pre-emption rights could be approved on an annual (or multi-year) basis by shareholders at the annual general meeting, it cannot be guaranteed.

***The rights afforded to our shareholders are governed by English law. Not all rights available to shareholders under United States law will be available to holders of our ordinary shares.***

Our parent company is organized under the laws of England and Wales. The rights of holders of our ordinary shares are governed by English law and our articles of association, and these may not provide the same rights as shares offered by American companies.

In addition, English law may be subject to change in the future in ways that are disadvantageous to United States-based shareholders, which could adversely affect the rights of our investors. Rights afforded to shareholders under English law differ in certain respects from the rights of shareholders in companies incorporated in the United States. In particular, English law currently significantly limits the circumstances in which the shareholders of English companies may bring derivative actions (i.e., legal actions brought by a shareholder on behalf of a company against a third party). Under English law, in most cases, only Kiniksa International may be the proper plaintiff for the purposes of maintaining proceedings in respect of wrongful acts committed against it and, generally, neither an individual shareholder, nor any group of shareholders, has any right of action in such circumstances. In addition, English law does not afford appraisal rights to dissenting shareholders in the form typically available to shareholders in an American company.

It also may be difficult to enforce foreign civil liabilities against us because of our country of incorporation. See "*Risk Factors—Risks Related to our Jurisdiction of Incorporation and Certain Tax Risks—United States investors may find it difficult to enforce their civil liabilities against us.*"

***Investors in the United States may find it difficult to enforce their civil liabilities against us.***

It may be difficult for United States investors to bring and/or effectively enforce suits against us outside of the United States. We are a public limited company incorporated in England and Wales. If a judgment is obtained in the United States courts based on civil liability provisions of the United States federal securities laws against us or our directors or officers, it may, depending on the jurisdiction, be difficult to enforce the judgment in the non-United States courts against us. Accordingly, United States shareholders may be forced to bring legal proceedings against us under English law and in the English courts in order to enforce any claims that they may have against us or our directors and officers. The enforceability of a United States judgment in the UK will depend on the particular facts of the case as well as the laws and treaties in effect at the time. The United States and the UK do not currently have a treaty providing for reciprocal recognition and enforcement of judgments (other than arbitration awards) in civil and commercial matters. Nevertheless, it may be difficult for United States shareholders to bring an original action in the English courts to enforce liabilities based on the United States federal securities laws against us.

***We may become subject to unanticipated tax liabilities, including liabilities arising from the reallocation of our taxable income among our subsidiaries.***

Although we are incorporated under the laws of England and Wales, 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 tax liability, if greater than our overall effective tax rate, could materially adversely affect our results of operations.

For example, we are currently incorporated under the laws of England and Wales and have subsidiaries in the United States, the UK, Bermuda, 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 and uncertainties in the tax system in the countries in which we have operations, could materially adversely affect our financial condition and results of operations, and reduce net returns to our shareholders.***

We are unable to predict what tax reform may be proposed or enacted in the future by the United States, UK, Switzerland or the OECD or what effect such changes would have on our business and results of operations. Changes in tax rates, laws, practices, treaties, policies or regulations, or the change in interpretation thereof, could increase our effective tax rate or otherwise affect our financial position, results of operations and financial condition and/or increase the complexity, burden and cost of tax compliance.

***We may be treated as a passive foreign investment company ("PFIC") for United States federal income tax purposes. If we were to be classified a PFIC, this could result in adverse United States federal income tax consequences to United States Holders.***

We completed an analysis of the Company's and its subsidiaries sources of income and character of their assets for United States 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, 2024. Although we believe that we were not a PFIC for 2024 and do not expect to become a PFIC in 2025, there can be no guarantee that we, or our subsidiaries, will not be treated as a PFIC for any taxable period. In this regard, the determination of PFIC classification is not made until after the close of the year and it depends on the amount and character of our annual income and assets, which in turn can depend on the interpretation of regulations and authorities, the application of which can be unclear. A non-United States 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 beneficial owner of our Class A ordinary shares who is (a) an individual who is a citizen of the United States, (b) a corporation organized under the laws of the United States or any state, district or territory thereof, (c) an estate taxable with income subject to United States federal income tax or (d) certain trusts (each, a "United States Holder") owns our Class A ordinary shares, we will continue to be treated as a PFIC with respect to such United States Holder in all succeeding years during which the United States Holder owns the Class A ordinary shares, regardless of whether we continue to meet the PFIC test described above, unless we cease to be a PFIC and the United States Holder made a "qualified electing fund" election or "mark-to-market" election for (a) the first taxable year the United States 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 United States Holder made a "deemed sale" election or was qualified to and made a "deemed dividend" election.

If we, or our subsidiaries, are classified as a PFIC for any taxable year during which a United States Holder holds our Class A ordinary shares, certain adverse United States federal income tax consequences could apply to such United States 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 United States Holder is treated as owning at least 10% of our shares, by vote or by value, such holder may be subject to adverse United States 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, 2025. Even if we were not classified as a controlled foreign corporation, certain of our non-United States subsidiaries could be treated as controlled foreign corporations because our group includes one or more United States subsidiaries. If a United States Holder is treated as owning (directly, indirectly or constructively) at least 10% of the value or voting power of our shares, such United States 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 United States taxable income its pro rata share of "Subpart F income," "global intangible low-taxed income," and investments in United States 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 United States 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 United States 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-United States 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. United States Holders should consult their tax advisors regarding the potential application of these rules to any investment in our Class A ordinary 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.**

**Trading Arrangements**

The following table shows the "Rule 10b5-1 trading arrangements" or "non-Rule 10b5-1 trading arrangements" (as such terms are defined under Item 408 of Regulation S-K) adopted, amended or terminated by our directors and officers during the three months ended June 30, 2025:

| | | | | Trading Arrangement | | | |
|---|---|---|---|---|---|---|---|
| Name | Title | Action | Effective Date | Rule 10b5-1 | Non Rule 10b5-1 | Scheduled Expiration Date of Trading Plan[(1)] | Maximum Shares Subject to Trading Plan |
| Sanj K. Patel | CEO and Chairman of the Board | Adoption | May 7, 2025 | X | | October 19, 2026 | 355,590[(2)] |
| Michael R. Megna | SVP, Finance and Chief Accounting Officer | Adoption | May 24, 2025 | X | | September 15, 2026 | 36,145[(3)] |

(1) A trading arrangement may expire on an earlier date if all contemplated transactions are completed before such trading arrangement's expiration date, upon termination by broker or the holder of the trading arrangement or as otherwise provided in the trading arrangement.
(2) This figure includes a number of securities covered by a currently active plan and, therefore, may be less at the time that this plan goes into effect.
(3) Includes Class A ordinary shares to be received upon the vesting of outstanding RSUs. The final amount of shares available for sale will be lower due to our net vesting policy for employee RSUs.

**Item 6. Exhibits**

| Exhibit Number | Exhibit Description | Incorporated by Reference | | | | Filed/ Furnished Herewith |
|---|---|---|---|---|---|---|
| | | Form | File No. | Exhibit | Filing Date | |
| 10.1#† | KPL-387 Long-Term Incentive Plan for Executive Officers | 8-K | 001-38492 | 10.1 | 4/23/2025 | |
| 10.2# | Form of Milestone 1 Cash Award Grant Notice and Agreement under the KPL-387 Long-Term Incentive Plan | 8-K | 001-38492 | 10.2 | 4/23/2025 | |
| 10.3# | Form of Milestone 2 Cash Award Grant Notice and Agreement under the KPL-387 Long-Term Incentive Plan | 8-K | 001-38492 | 10.3 | 4/23/2025 | |
| 10.4# | Form of Milestone 1 PSU Award Grant Notice and Agreement under the KPL-387 Long-Term Incentive Plan | 8-K | 001-38492 | 10.4 | 4/23/2025 | |
| 10.5# | Form of Milestone 2 PSU Award Grant Notice and Agreement under the KPL-387 Long-Term Incentive Plan | 8-K | 001-38492 | 10.5 | 4/23/2025 | |
| 10.6# | Form of Milestone 1 Option Award Grant Notice and Agreement under the KPL-387 Long-Term Incentive Plan | 8-K | 001-38492 | 10.6 | 4/23/2025 | |
| 10.7# | Form of Milestone 2 Option Award Grant Notice and Agreement under the KPL-387 Long-Term Incentive Plan | 8-K | 001-38492 | 10.7 | 4/23/2025 | |
| 10.8# | Consulting Agreement by and between Kiniksa Pharmaceuticals International, plc and Dr. Richard Levy | | | | | * |
| 10.9# | 2018 Incentive Award Plan and forms of award agreements thereunder, as amended | | | | | * |
| 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 | | | | | ** |

| Exhibit Number | Exhibit Description | Incorporated by Reference | | | | Filed/ Furnished Herewith |
|---|---|---|---|---|---|---|
| | | Form | File No. | Exhibit | Filing Date | |
| 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 | | | | | *** |

# Indicates management contract or compensatory plan
† Portions of the exhibit have been redacted in compliance with Regulation S-K Item 601(b)(10)(iv)
* 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 INTERNATIONAL, PLC**

Date: July 29, 2025

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

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

# CONSULTING AGREEMENT

This Consulting Agreement (the "**Agreement**") is made as of July 1, 2025 (the "**Effective Date**") by and between Kiniksa Pharmaceuticals, GmbH a Swiss limited liability company with a business address at Grafenaustrasse 5, 6300 Zug, Switzerland ("**Kiniksa**"), and Dr. Richard S. Levy with an address at [***] ("**Consultant**").

**WHEREAS**, Kiniksa wants the benefit of Consultant's knowledge and expertise; and

**WHEREAS**, Consultant wants to provide Services (as defined below) to Kiniksa, its designees and affiliates, in connection with its global programs and operations, as provided in and subject to this Agreement;

**NOW THEREFORE**, in consideration of the premises and of the following mutual promises, covenants and conditions herein contained, and intending to be legally bound, Kiniksa and Consultant agree as follows:

**1. Services**. Kiniksa retains Consultant and Consultant agrees to provide consulting services (the "**Services**") to Kiniksa, its designees and affiliates as Kiniksa may from time to time reasonably request and as specified in Exhibit A attached hereto. Any changes to the Services (and any related compensation adjustments) must be agreed upon in writing between Consultant and Kiniksa prior to commencement of the changes.

**1.1 Performance**. Consultant agrees to render the Services to Kiniksa or to its designees and affiliates (a) under the general supervision of Kiniksa or its designees and affiliates, and (b) in accordance with prevailing industry standards and practices for the performance of similar services. Consultant will comply with all rules, procedures and standards promulgated from time to time by Kiniksa with regard to Consultant's access to and use of Kiniksa's property, information, equipment and facilities.

**1.2 Third Party Confidential Information**. Consultant agrees not to use any trade secrets or other confidential information of any other person, firm, corporation, institution or other entity in connection with any of the Services.

**1.3 No Conflicts**. Consultant is under no contractual or other obligation or restriction which is inconsistent with Consultant's execution of this Agreement or the performance of the Services. During the Term (as defined below), Consultant will not enter into any agreement, either written or oral, in conflict with Consultant's obligations under this Agreement. Consultant will arrange to provide the Services in such manner and at such times that the Services will not conflict with Consultant's responsibilities under any other agreement, arrangement or understanding or pursuant to any employment relationship Consultant has at any time with any third party.

**1.4 Compliance with Applicable Laws.** Consultant shall comply with all federal, state, and local applicable laws and regulations in Consultant's performance of the Services.

**1.5 Absence of Debarment**. Consultant represents that Consultant has not been (a) debarred, convicted, or is not subject to a pending debarment or conviction, pursuant to section 306 of the United States Food Drug and Cosmetic Act, 21 U.S.C. § 335a, (b) listed by any government or regulatory agencies as ineligible to participate in any government healthcare programs or government procurement or non-procurement programs (as that term is defined in 42 U.S.C. 1320a-7b(f)), or excluded, debarred, suspended or otherwise made ineligible to participate in any such program, or (c) convicted of a criminal offense related to the provision of healthcare items or services, or is not subject to any such pending action.

Consultant agrees to inform Kiniksa in writing promptly if Consultant is subject to the foregoing, or if any action, suit, claim, investigation, or proceeding relating to the foregoing is pending, or to the best of Consultant's knowledge, is threatened.

**1.6 Non-Referral.** The parties agree that Consultant is under no obligation to solicit, refer, or solicit referrals of patients for any Kiniksa business. Consultant will not receive any benefit of any kind for making any referrals nor suffer any detriment for not making such referrals. The parties further agree that no amount paid hereunder is intended to be, nor shall be construed as, an inducement or payment for referral of or recommending referral of patients for any Kiniksa business by Consultant to Kiniksa or by Kiniksa to Consultant. In addition, the fees charged hereunder do not include any discount, rebate, kickback, or other reduction in charge, and the fees charged hereunder are not intended to be, nor shall they be construed as, an inducement or payment for referral, or recommendation of referral, of business by Consultant to Kiniksa or by Kiniksa to Consultant. The sole purpose of the fee paid to Consultant hereunder is to pay fair market value for the Services provided by Consultant to Kiniksa hereunder.

**1.7 Disclosure Requirements**. The parties to this Agreement acknowledge that certain states, the United States government, and/or governments and industry groups outside of the United States and/or the federal government require pharmaceutical companies to disclose information on compensation, gifts or other remuneration provided to physicians and other health care professionals and health care organizations. Kiniksa may report information about remuneration provided under this Agreement, as required by law or industry group code. Once reported, such information may be publicly accessible.

**2. Compensation.**

In consideration for the Services rendered by Consultant to Kiniksa, Kiniksa agrees to pay Consultant the fees set forth in Exhibit A attached hereto. The parties represent and warrant that the fees were determined by the parties through good faith and arms' length bargaining, constitute fair market value for the Services, and have not been determined in a manner that takes into account the volume or value of any business between the parties. Consultant is not required to use or recommend Kiniksa products, and the parties represent and warrant that the fees are not intended to reward Consultant for the use or recommendation of Kiniksa products or to induce Consultant to use or recommend Kiniksa products.

**3. Materials; Deliverables.**

**3.1 Materials**. All documentation, information, and biological, chemical and other materials controlled by Kiniksa and furnished to Consultant by or on behalf of Kiniksa ("**Materials**") and all associated intellectual property rights will remain the exclusive property of Kiniksa. Consultant will use Materials provided by Kiniksa only as necessary to perform the Services and will treat them in accordance with the requirements of this Section 3.1. Consultant agrees that it will not use or evaluate those Materials or any portions thereof for any other purpose except as directed or permitted in writing by Kiniksa. Without Kiniksa's prior express written consent, Consultant agrees that it will not analyze the Materials, or transfer or make the Materials available to third parties.

**3.2 Deliverables**. Consultant shall assign, and hereby assigns, to Kiniksa all rights in and to inventions, discoveries, improvements, ideas, designs, processes, formulations, products, computer programs, works of authorship, databases, mask works, trade secrets, know-how, information, data, documentation, reports, research, creations and other products arising from or made in the performance of the Services (whether or not patentable or subject to copyright or trade secret protection) (collectively, "**Deliverables**"). For purposes of the copyright laws of the United States, Deliverables will constitute "works made for hire," except to the extent such Deliverables cannot by law be "works made for hire." Kiniksa will have the right to use Deliverables for any and all purposes. During and after the term of this Agreement, Consultant will cooperate fully in obtaining patent and other proprietary protection for any

patentable Deliverables, all in the name of Kiniksa and at Kiniksa's cost and expense. Such cooperation will include, without limitation, executing and delivering all requested applications, assignments and other documents, and taking such other measures as Kiniksa may reasonably request in order to perfect and enforce Kiniksa's rights in the Deliverables. Consultant appoints Kiniksa its attorney-in-fact to execute and deliver any such documents on behalf of Consultant if Consultant fails to do so. Consultant will, however, retain full ownership rights in and to all templates, programs and other materials developed or obtained or licensed from third parties by Consultant ("**Consultant Property**") prior to or independent of the Services, regardless of whether such Consultant Property is used in the performance of the Services. Consultant hereby grants to Kiniksa a perpetual, non-exclusive, fully paid-up worldwide license to use Consultant Property solely to the extent required for Kiniksa's use of the Deliverables.

**3.3 Third Party Intellectual Property.** Consultant will not use any third party intellectual property in performing the Services without Kiniksa's prior written consent.

**3.4 Records; Records Storage.** Consultant will maintain all materials and all other data and documentation obtained or generated by Consultant in the course of preparing for and providing the Services, including all computerized records and files (the "**Records**") in a secure area reasonably protected from fire, theft and destruction. These Records will be "works made for hire" and will remain the exclusive property of Kiniksa. Upon written instruction of Kiniksa, all Records will, at Kiniksa's option either be (a) delivered to Kiniksa or to its designee, or (b) disposed of, unless such Records are otherwise required to be stored or maintained by Consultant as a matter of law or regulation. In no event will Consultant dispose of any such Records without first giving Kiniksa sixty (60) days' prior written notice of Consultant's intent to do so. Consultant may, however, retain copies of any Records as are reasonably necessary for regulatory or insurance purposes, subject to Consultant's obligation of confidentiality**.**

**4. Confidential Information and Publicity.**

**4.1 Definition.** "**Confidential Information**" means all scientific, technical, financial or business information owned, possessed or used by Kiniksa or its affiliates, learned of by Consultant or developed by Consultant in connection with the Services, whether or not labeled "Confidential", including but not limited to (a) Deliverables, Materials, scientific data and sequence information, (b) marketing plans, business strategies, financial information, forecasts, personnel information and customer lists of Kiniksa and its affiliates, and (c) all information of third parties that Kiniksa has an obligation to keep confidential.

**4.2 Obligations of Confidentiality**. During the Term and for a period of five (5) years thereafter, Consultant will not directly or indirectly publish, disseminate or otherwise disclose, use for Consultant's own benefit or for the benefit of a third party, deliver or make available to any third party, any Confidential Information, other than in furtherance of the purposes of this Agreement, and only then with the prior written consent of Kiniksa. Consultant will exercise all reasonable precautions to physically protect the integrity and confidentiality of the Confidential Information.

**4.3 Exceptions.** Consultant will have no obligations of confidentiality and non-use with respect to any portion of the Confidential Information which:

**(a)** is or later becomes generally available to the public by use, publication or the like, through no fault of Consultant;

**(b)** is obtained from a third party who had the legal right to disclose it to Consultant; or

**(c)** Consultant already possesses, as evidenced by Consultant's written records that predate the receipt thereof.

In the event that Consultant is required by law or court order to disclose any Confidential Information, Consultant will give Kiniksa prompt notice thereof so that Kiniksa may seek an appropriate protective order. Consultant will reasonably cooperate with Kiniksa in its efforts to seek such a protective order.

**5. Term and Termination.**

**5.1 Term.** This Agreement will commence on the Effective Date and remain in effect until December 31, 2025 (the "**Term**"). This Agreement may be extended only by written agreement between the parties.

**5.2 Termination.** Either party may terminate this Agreement immediately at any time upon written notice to the other party.

**5.3 Effect of Expiration/Termination.** Upon expiration or termination of this Agreement, neither Consultant nor Kiniksa will have any further obligations under this Agreement, except that (a) Consultant will terminate all Services in progress in an orderly and non-disruptive manner as soon as practical and in accordance with a schedule agreed to by Kiniksa, unless Kiniksa specifies in the notice of termination that Services in progress should be completed, (b) Consultant will deliver to Kiniksa any Materials in Consultant's possession or control and all Deliverables made through expiration or termination, (c) Kiniksa will pay Consultant any monies due and owing Consultant, up to the time of the effective date of termination or expiration, for Services actually performed and all authorized expenses actually incurred, (d) Consultant will promptly refund to Kiniksa any monies paid by Kiniksa in advance for Services not rendered, (e) Consultant will immediately return to Kiniksa all Confidential Information and copies thereof provided to Consultant under this Agreement except for one (1) copy which Consultant may retain solely to monitor Consultant's surviving obligations of confidentiality, (f) Consultant will immediately return to Kiniksa any and all equipment and supplies provided to Consultant under this Agreement, and (g) the terms, conditions and obligations under Sections 1.5, 1.7, 3, 4, 5.3, and 6 will survive expiration or termination for any reason.

**6. Miscellaneous.**

**6.1 Independent Contractor**. All Services will be rendered by Consultant as an independent contractor and this Agreement does not create an employer-employee relationship between Kiniksa and Consultant. Consultant will have no rights to receive any employee benefits, such as bonuses, options, health and accident insurance, sick leave or vacation which are accorded to regular employees of Kiniksa or its affiliates. Consultant will not in any way represent itself to be an employee, partner, joint venturer, or agent of Kiniksa. Consultant shall have no authority to make any statements, representations or commitments of any kind, or to take any action, which shall be binding on Kiniksa. In performing the Services, the amount of time devoted by Consultant on any given day will be within Consultant's control, and Kiniksa will rely on Consultant to devote the amount of time necessary to fulfill the requirements of the Agreement in an efficient and timely manner. Consultant is responsible for providing all equipment and supplies required to perform the Services. In the event Kiniksa provides to Consultant any equipment or supplies in connection with the Services, such equipment and supplies shall remain the sole property of Kiniksa, be used solely for performing the Services and, upon Kiniksa's request, Consultant shall promptly return to Kiniksa all such equipment and supplies. Upon reasonable notice, Consultant shall meet with representatives of Kiniksa or one of its affiliates at a location to be designated by the Parties. Consultant shall not in any way represent itself to be an employee, partner joint venturer, or agent of Kiniksa. Consultant shall have no authority to make any statements, representations or commitments of any kind, or to take any action, which shall be binding on Kiniksa.

**6.2 Taxes.** Consultant will be solely and unconditionally responsible for any and all federal, state, or local taxes, social security withholding, and other self-employment tax obligations with

respect to payments made to Consultant under this Agreement. Consultant will provide Kiniksa with Consultant's taxpayer identification number or social security number, as applicable.

**6.3 Assignability and Binding Effect.** The Services to be rendered by Consultant are personal in nature. Consultant may not assign or transfer this Agreement or any of Consultant's rights or obligations hereunder except to a corporation of which Consultant is the sole stockholder. In no event will Consultant assign or delegate responsibility for actual performance of the Services to any other natural person. This Agreement will be binding upon and inure to the benefit of the parties and their respective legal representatives, heirs, successors and permitted assigns.

**6.4 Notices.** All notices required or permitted under this Agreement must be in writing and must be given by addressing the notice to the address for the recipient set forth in this Agreement or at such other address as the recipient may specify in writing under this procedure. Notices to Kiniksa must include a copy to Kiniksa Pharmaceuticals Corp., 100 Hayden Avenue, Lexington, MA 02421, USA, Attention: Legal Department. Notices will be deemed to have been given (a) three (3) business days after deposit in the mail with proper postage for first class registered or certified mail prepaid, or (b) one (1) business day after sending by nationally recognized overnight delivery service.

**6.5 No Modification.** This Agreement may be changed only by a writing signed by Consultant and an authorized representative of Kiniksa.

**6.6 Remedies.** It is understood and agreed that Kiniksa may be irreparably injured by a breach of this Agreement; that money damages would not be an adequate remedy for any such breach; and that Kiniksa will be entitled to seek equitable relief, including injunctive relief and specific performance, without having to post a bond, as a remedy for any such breach, and such remedy will not be Kiniksa's exclusive remedy for any breach of this Agreement.

**6.7 Severability.** Any of the provisions of this Agreement which are determined to be invalid or unenforceable in any jurisdiction will be ineffective to the extent of such invalidity or unenforceability in such jurisdiction, without rendering invalid or unenforceable the remaining provisions hereof and without affecting the validity or enforceability of any of the other terms of this Agreement in such jurisdiction, or the terms of this Agreement in any other jurisdiction. The parties will substitute for the invalid or unenforceable provision a valid and enforceable provision that conforms as nearly as possible with the original intent of the parties.

**6.8 Waivers.** No waiver of any term, provision or condition of this Agreement in any one or more instances will be deemed to be or construed as a further or continuing waiver of any other term, provision or condition of this Agreement. Any such waiver must be evidenced by an instrument in writing executed by Consultant or, in the case of Kiniksa, by an officer authorized to execute waivers.

**6.9 Entire Agreement.** This Agreement (including any exhibits or schedules attached hereto) constitutes the entire agreement of the parties with regard to the subject matter, and, with the exception of any written agreement between the parties relating to the disclosure or exchange of confidential information, supersedes all previous written or oral representations, agreements and understandings between the parties on the subject matter.

**6.10 Governing Law.** This Agreement and all acts and transactions pursuant hereto and the rights and obligations of the parties hereto shall be governed, construed and interpreted in accordance with the laws of the State of New York, without giving effect to the principles of conflicts of law.

**6.11 Counterparts.** This Agreement may be executed in any number of counterparts, each of which will be deemed an original and all of which together shall constitute one and the same

instrument. Signatures delivered via facsimile or electronic means shall be binding and treated as if they were original signatures.

**6.12 Headings.** The section headings are included solely for convenience of reference and will not control or affect the meaning or interpretation of any of the provisions of this Agreement.

**IN WITNESS WHEREOF**, duly authorized representatives of the parties have executed this Agreement as of the Effective Date.

**KINIKSA PHARMACEUTICALS, GMBH**

By: _____
Name:

Title:

**DR. RICHARD S. LEVY**

By: _____

**EXHIBIT A**

**1. Services:**

Consultant will provide advice on matters relating to (a) Kiniksa's pipeline development strategy and (b) the review and design of Kiniksa's clinical trial protocols.

During the Term, Consultant shall provide Services for an average of six (6) hours per week.

**2. Compensation:**

As compensation for the Services, Kiniksa, on its own behalf or through its affiliates, shall pay Consultant as follows:

(a) $30,000.00 USD within thirty (30) days of the Effective Date; and

(b) Restricted Stock Units ("**RSUs**"), representing the right to receive Class A ordinary shares of Kiniksa's parent company, Kiniksa Pharmaceuticals International, plc, with a combined value equal to $30,000.00 USD as of market closing on the Effective Date, rounded down to the nearest whole share. Such RSUs will vest immediately on the Effective Date.

Notwithstanding anything to the contrary in this Agreement, Kiniksa or its affiliates shall not pay Consultant compensation for Services provided pursuant to this Agreement in excess of $120,000 during any period of twelve consecutive months.

Consultant shall bear Consultant's own day-to-day expenses, such as expenses for telephone calls, faxes and mail, that Consultant incurs in providing Services. For all other out-of-pocket expenses, Kiniksa agrees to reimburse Consultant for those expenses that Kiniksa has authorized in advance**.**

**Exhibit 10.9**

As Amended June 11, 2025

**KINIKSA PHARMACEUTICALS INTERNATIONAL, PLC**
**2018 INCENTIVE AWARD PLAN**

**(FORMERLY, KINIKSA PHARMACEUTICALS, LTD.**
**2018 INCENTIVE AWARD PLAN)**

## ARTICLE I
## PURPOSE

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

## ARTICLE II
## ELIGIBILITY

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

## ARTICLE III
## ADMINISTRATION AND DELEGATION

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

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

## ARTICLE IV
## SHARES AVAILABLE FOR AWARDS

4.1 Number of Shares. Subject to adjustment under Article VIII and the terms of this Article IV, Awards may be made under the Plan covering up to the Overall Share Limit. As of the Plan's original effective date under Section 10.3, the Company's predecessor ceased granting awards under the Prior Plans; however, Prior Plan Awards remain subject to the terms of the

applicable Prior Plan. Shares issued under the Plan may consist of authorized but unissued Shares, Shares purchased on the open market or Shares held in treasury.

4.2 Share Recycling. If all or any part of an Award or Prior Plan Award expires, lapses or is terminated, exchanged for cash, surrendered, repurchased, canceled without having been fully exercised or forfeited, in any case, in a manner that results in the Company acquiring Shares covered by the Award or Prior Plan Award at a price not greater than the price (as adjusted to reflect any Equity Restructuring) paid by the Participant for such Shares or not issuing any Shares covered by the Award or Prior Plan Award, the unused Shares covered by the Award or Prior Plan Award will, as applicable, become or again be available for Award grants under the Plan. Further, Shares delivered (either by actual delivery or attestation) to the Company by a Participant to satisfy the applicable exercise or purchase price of an Award or Prior Plan Award and/or to satisfy any applicable tax withholding obligation, will, as applicable, become or again be available for Award grants under the Plan. The payment of Dividend Equivalents in cash in conjunction with any outstanding Awards shall not count against the Overall Share Limit.

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

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

## ARTICLE V
## SHARE OPTIONS AND SHARE APPRECIATION RIGHTS

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

5.2 Exercise Price. The Administrator will establish each Option's and Share Appreciation Right's exercise price and specify the exercise price in the Award Agreement. The exercise price will not be less than 100% of the Fair Market Value or nominal value per Share, whichever is greater, on the grant date of the Option or Share Appreciation Right.

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

suspended from the time of the delivery of such notice until the earlier of (i) such time as it is determined or otherwise agreed that the Participants service as a Service Provider will not be terminated for Cause as provided in such notice or (ii) the effective date of the Participants Termination of Service by the Company or any of its Subsidiaries for Cause (in which case the right of the Participant and the Participants transferees to exercise any Option or Share Appreciation Right issued to the Participant will terminate immediately upon the effective date of such termination of Service).

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

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

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

(b) if there is a public market for Shares at the time of exercise, unless the Company otherwise determines, (A) delivery (including telephonically to the extent permitted by the Company) of an irrevocable and unconditional undertaking by a broker acceptable to the Company to deliver promptly to the Company sufficient funds to pay the exercise price, or (B) the Participant's delivery to the Company of a copy of irrevocable and unconditional instructions to a broker acceptable to the Company to deliver promptly to the Company cash or a check sufficient to pay the exercise price; provided that such amount is paid to the Company at such time as may be required by the Administrator;

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

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

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

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

## ARTICLE VI
## RESTRICTED SHARES; RESTRICTED SHARE UNITS

6.1 General. The Administrator may grant Restricted Shares, or the right to purchase Restricted Shares, to any Service Provider, subject to the Company's right to repurchase all or part of such shares at their issue price or other stated or formula price from the Participant (or to require forfeiture of such shares) if conditions the Administrator specifies in the Award Agreement are not satisfied before the end of the applicable restriction period or periods that the Administrator establishes for such Award and subject to Applicable Laws. In addition, the Administrator may grant to Service Providers Restricted Share Units, which may be subject to vesting and forfeiture conditions during the applicable restriction period or periods, as set forth in an Award Agreement. The Administrator will determine and set forth in the Award Agreement the terms and conditions for each Restricted Share and Restricted Share Unit Award, subject to the conditions and limitations contained in the Plan.

6.2 Restricted Shares.

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

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

6.3 Restricted Share Units.

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

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

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

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

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

## ARTICLE VIII
## ADJUSTMENTS FOR CHANGES IN ORDINARY SHARES

## AND CERTAIN OTHER EVENTS

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

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

(a) To provide for the cancellation of any such Award in exchange for either an amount of cash or other property with a value equal to the amount that could have been obtained upon the exercise or settlement of the vested portion of such Award or realization of the Participants rights under the vested portion of such Award, as applicable; provided that, if the amount that could have been obtained upon the exercise or settlement of the vested portion of such Award or realization of the Participants rights, in any case, is equal to or less than zero, then the Award may be terminated without payment;

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

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

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

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

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

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

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

affect or restrict in any way the Company's right or power to make or authorize (i) any adjustment, recapitalization, reorganization or other change in the Company's capital structure or its business, (ii) any merger, amalgamation, consolidation dissolution or liquidation of the Company or sale of Company assets or (iii) any sale or issuance of securities, including securities with rights superior to those of the Shares or securities convertible into or exchangeable for Shares. The Administrator may treat Participants and Awards (or portions thereof) differently under this Article VIII.

## ARTICLE IX
## GENERAL PROVISIONS APPLICABLE TO AWARDS

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

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

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

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

9.5 Withholding. Each Participant must pay the Company, or make provision satisfactory to the Administrator for payment of, any taxes required by law to be withheld in connection with such Participant's Awards by the date of the event creating the tax liability. The Company may deduct a cash amount sufficient to satisfy such tax obligations based on the applicable statutory withholding rates (or such other rate as may be determined by the Company after considering any accounting consequences or costs) from any payment otherwise due to a Participant. Subject to Section 10.8 and any Company insider trading policy (including blackout periods), Participants may satisfy such tax obligations (i) in cash, by wire transfer of immediately available funds, by check made payable to the order of the Company, provided that the Company may limit the use of the foregoing payment forms if one or more of the payment forms below is permitted, (ii) to the extent permitted by the Administrator, in whole or in part by delivery of Shares valued at their Fair Market Value or elect to have the Company repurchase Shares otherwise issuable under an Award limited to the number of Ordinary Shares which have a Fair Market Value

on the date of repurchase necessary to pay the aggregate amount of tax liability, (iii) if there is a public market for Shares at the time the tax obligations are satisfied, unless the Company otherwise determines, (A) delivery (including telephonically to the extent permitted by the Company) of an irrevocable and unconditional undertaking by a broker acceptable to the Company to deliver promptly to the Company sufficient funds to satisfy the tax obligations, or (B) delivery by the Participant to the Company of a copy of irrevocable and unconditional instructions to a broker acceptable to the Company to deliver promptly to the Company cash or a check sufficient to satisfy the tax withholding; provided that such amount is paid to the Company at such time as may be required by the Administrator, or (iv) to the extent permitted by the Company, any combination of the foregoing payment forms approved by the Administrator. If any tax withholding obligation will be satisfied under clause (ii) of the immediately preceding sentence by the Company's retention of Shares from the Award creating the tax obligation and there is a public market for Shares at the time the tax obligation is satisfied, the Company may elect to instruct any brokerage firm determined acceptable to the Company for such purpose to sell on the applicable Participant's behalf some or all of the Shares retained and to remit the proceeds of the sale to the Company or its designee, and each Participant's acceptance of an Award under the Plan will constitute the Participant's authorization to the Company and instruction and authorization to such brokerage firm to complete the transactions described in this sentence.

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

9.7 Conditions on Delivery of Shares. The Company will not be obligated to deliver any Shares under the Plan or remove restrictions from Shares previously delivered under the Plan until (i) all Award conditions have been met or removed to the Company's satisfaction, (ii) as determined by the Company, all other legal matters regarding the issuance and delivery of such Shares have been satisfied, including any applicable securities laws and stock exchange or stock market rules and regulations, and (iii) the Participant has executed and delivered to the Company such representations or agreements as the Administrator deems necessary or appropriate to satisfy any Applicable Laws. The Company's inability to obtain authority from any regulatory body having jurisdiction, which the Administrator determines is necessary to the lawful issuance and sale of any securities, will relieve the Company of any liability for failing to issue or sell such Shares as to which such requisite authority has not been obtained. For additional clarity, if an Award is settled in Shares, the Company will have the right to withhold, in any manner permitted by Section 9.5, or require a Participant to remit to the Company, an amount sufficient to satisfy the Participant's obligation to pay the nominal value of the Shares issued to a Participant under such Award.

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

9.9 Additional Terms of Incentive Stock Options. The Administrator may grant Incentive Stock Options only to employees of the Company, any of its present or future parent or subsidiary corporations, as defined in Sections 424(e) or (f) of the Code, respectively, and any other entities the employees of which are eligible to receive Incentive Stock Options under the Code. If an Incentive Stock Option is granted to a Greater Than 10% Shareholder, the exercise price will not be less than 110% of the Fair Market Value on the Option's grant date, and the term of the Option will not exceed five years. All Incentive Stock Options will be subject to and construed consistently with Section 422 of the Code. By accepting an Incentive Stock Option, the Participant agrees to give prompt notice to the Company of dispositions or other transfers (other than in connection with a Change in Control) of Shares acquired under the Option made within (i) two years from the grant date of the Option or (ii) one year after the transfer of such Shares to the Participant, specifying the date of the disposition or other transfer and the amount the Participant realized, in cash, other property, assumption of indebtedness or other consideration, in such disposition or other transfer. Neither the Company nor the Administrator will be liable to a Participant, or any other party, if an Incentive Stock Option fails or ceases to qualify as an "incentive stock option" under Section 422 of the Code. Any Incentive Stock Option or portion thereof that fails to qualify as an "incentive stock option^^ under Section 422 of the Code for any reason, including becoming exercisable with respect to Shares having a fair market value exceeding the $100,000 limitation under Treasury Regulation Section L422-4, will be a Non-Qualified Stock Option.

## ARTICLE X
## MISCELLANEOUS

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

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

10.3 Effective Date and Tenn of Plan. The Plan originally became effective on May 14, 2018 and was amended and restated effective June 27, 2024. The Plan will remain in effect until

the tenth anniversary of the earlier of (i) the date the board of directors of Kiniksa Pharmaceuticals, Ltd. originally adopted the Plan or (ii) the date the shareholders of Kiniksa Pharmaceuticals, Ltd. originally approved the Plan, but Awards previously granted may extend beyond that date in accordance with the Plan.

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

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

10.6 Section 409A.

(a) General. The Company intends that all Awards be structured to comply with, or be exempt from, Section 409A, such that no adverse tax consequences, interest, or penalties under Section 409A apply. Notwithstanding anything in the Plan or any Award Agreement to the contrary, the Administrator may, without a Participants consent, amend this Plan or Awards, adopt policies and procedures, or take any other actions (including amendments, policies, procedures and retroactive actions) as are necessary or appropriate to preserve the intended tax treatment of Awards, including any such actions intended to (A) exempt this Plan or any Award from Section 409A, or (B) comply with Section 409A, including regulations, guidance, compliance programs and other interpretative authority that may be issued after an Award's grant date. The Company makes no representations or warranties as to an Award's tax treatment under Section 409A or otherwise. The Company will have no obligation under this Section 10.6 or otherwise to avoid the taxes, penalties or interest under Section 409A with respect to any Award and will have no liability to any Participant or any other person if any Award, compensation or other benefits under the Plan are determined to constitute noncompliant "nonqualified deferred compensation" subject to taxes, penalties or interest under Section 409A.

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

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

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

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

10.9 Data Privacy. As a condition for receiving any Award, each Participant explicitly and unambiguously consents to the collection, use and transfer, in electronic or other form, of personal data as described in this section by and among the Company and its Subsidiaries and affiliates exclusively for implementing, administering and managing the Participant's participation in the Plan. The Company and its Subsidiaries and affiliates may hold certain personal information about a Participant, including the Participant's name, address and telephone number; birthdate; social security, insurance number or other identification number; salary; nationality; job title(s); any Shares held in the Company or its Subsidiaries and affiliates; and Award details, to implement, manage and administer the Plan and Awards (the "Data"). The Company and its Subsidiaries and affiliates may transfer the Data amongst themselves as necessary to implement, administer and manage a Participant's participation in the Plan, and the Company and its Subsidiaries and affiliates may transfer the Data to third parties assisting the Company with Plan implementation, administration and management. These recipients may be located in the Participants country, or elsewhere, and the Participant's country may have different data privacy laws and protections than the recipients' country. By accepting an Award, each Participant authorizes such recipients to

receive, possess, use, retain and transfer the Data, in electronic or other form, to implement, administer and manage the Participant's participation in the Plan, including any required Data transfer to a broker or other third party with whom the Company or the Participant may elect to deposit any Shares. The Data related to a Participant will be held only as long as necessary to implement, administer, and manage the Participant's participation in the Plan. A Participant may, at any time, view the Data that the Company holds regarding such Participant, request additional information about the storage and processing of the Data regarding such Participant, recommend any necessary corrections to the Data regarding the Participant or refuse or withdraw the consents in this Section 10.9 in writing, without cost, by contacting the local human resources representative. The Company may cancel Participant's ability to participate in the Plan and, in the Administrator's discretion, the Participant may forfeit any outstanding Awards if the Participant refuses or withdraws the consents in this Section 10.9. For more information on the consequences of refusing or withdrawing consent, Participants may contact their local human resources representative.

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

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

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

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

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

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

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

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

## ARTICLE XI
## DEFINITIONS

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

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

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

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

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

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

11.6 "Cause" means, with respect to a Participant, (A) dishonesty with respect to the Company or any Subsidiary; (B) insubordination, substantial malfeasance or nonfeasance of duty;

(C) unauthorized disclosure of confidential information; (D) breach by a Participant of any provision of any employment, consulting, advisory, nondisclosure, non-competition or similar agreement between the Participant and the Company or any Subsidiary; and (E) conduct substantially prejudicial to the business of the Company or any Subsidiary; provided, however, that any provision in an agreement between a Participant and the Company or an Subsidiary, which contains a conflicting definition of Cause for termination and which is in effect at the time of such termination, shall supersede this definition with respect to that Participant. The determination of the Administrator as to the existence of Cause will be conclusive on the Participant and the Company.

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

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

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

whether a Change in Control is a "change in control event" as defined in Treasury Regulation Section 1.409A-3(i)(5) shall be consistent with such regulation.

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

11.9 "Committee" means one or more committees or subcommittees of the Board, which may include one or more Company directors or executive officers, to the extent Applicable Laws permit. To the extent required to comply with the provisions of Rule 16b-3, it is intended that each member of the Committee will be, at the time the Committee takes any action with respect to an Award that is subject to Rule 16b-3, a "non-employee director" within the meaning of Rule 16b-3; however, a Committee member's failure to qualify as a "non-employee director" within the meaning of Rule 16b-3 will not invalidate any Award granted by the Committee that is otherwise validly granted under the Plan.

11.10 "Company" means Kiniksa Pharmaceuticals International, plc, a public limited company organized under the laws of England and Wales, or any successor.

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

11.12 "Designated Beneficiary?, means the beneficiary or beneficiaries the Participant designates, in a manner the Administrator determines, to receive amounts due or exercise the Participant's rights if the Participant dies or becomes incapacitated. Without a Participant's effective designation, "**Designated Beneficiary**" will mean the Participants estate.

11.13 "Director" means a Board member.

11.14 "Disability" means a permanent and total disability under Section 22(e)(3) of the Code, as amended.

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

11.16 "Employee" means any employee of the Company or its Subsidiaries.

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

11.18 "Exchange Act" means the Securities Exchange Act of 1934, as amended.

11.19 "Fair Market Value" means, as of any date, the value of Ordinary Shares determined as follows: (i) if the Ordinary Shares are listed on any established stock exchange, a share's Fair Market Value will be the closing sales price for such Ordinary Shares as quoted on such exchange for such date, or if no sale occurred on such date, the last day preceding such date during which a sale occurred, as reported in The Wall Street Journal or another source the Administrator deems reliable; (ii) if the Ordinary Shares are not traded on a stock exchange but is quoted on a national market or other quotation system, the closing sales price on such date, or if no sales occurred on such date, then on the last date preceding such date during which a sale occurred, as reported in The Wall Street Journal or another source the Administrator deems reliable; or (iii) without an established market for the Ordinary Shares, the Administrator will determine the Fair Market Value in its discretion. Notwithstanding the foregoing, with respect to any Award granted on the pricing date of the Company's initial public offering, the Fair Market Value shall mean the initial public offering price of a Share as set forth in the Company's final prospectus relating to its initial public offering filed with the Securities and Exchange Commission.

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

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

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

11.23 "Ordinary Shares" means the Class A Ordinary Shares of the Company, nominal value of $0.000273235 per share.

11.24 "Option" means an option to purchase Shares.

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

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

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

11.28 "Performance Criteria" mean the criteria (and adjustments) that the Administrator may select for an Award to establish performance goals for a performance period, which may

include the following: net earnings or losses (either before or after one or more of interest, taxes, depreciation, amortization, and non-cash equity-based compensation expense); gross or net sales or revenue or sales or revenue growth; net income (either before or after taxes) or adjusted net income; profits (including but not limited to gross profits, net profits, profit growth, net operation profit or economic profit), profit return ratios or operating margin; budget or operating earnings (either before or after taxes or before or after allocation of corporate overhead and bonus); cash flow (including operating cash flow and free cash flow or cash flow return on capital); return on assets; return on capital or invested capital; cost of capital; return on shareholders' equity; total shareholder return; return on sales; costs, reductions in costs and cost control measures; expenses; working capital; earnings or loss per share; adjusted earnings or loss per share; price per share or dividends per share (or appreciation in or maintenance of such price or dividends); regulatory achievements or compliance; implementation, completion or attainment of objectives relating to research, development, regulatory, commercial, or strategic milestones or developments; market share; economic value or economic value added models; division, group or corporate financial goals; customer satisfaction/growth; customer service; employee satisfaction; recruitment and maintenance of personnel; human resources management; supervision of litigation and other legal matters; mergers, acquisitions, and other strategic partnerships, licenses, and other transactions; financial ratios (including those measuring liquidity, activity, profitability or leverage); debt levels or reductions; sales-related goals; financing and other capital raising transactions; cash on hand; acquisition activity; investment sourcing activity; and marketing initiatives, any of which may be measured in absolute terms or as compared to any incremental increase or decrease. Such performance goals also may be based solely by reference to the Company's performance or the performance of a Subsidiary, division, business segment or business unit of the Company or a Subsidiary, or based upon performance relative to performance of other companies or upon comparisons of any of the indicators of performance relative to performance of other companies. The Committee may provide for exclusion of the impact of an event or occurrence which the Committee determines should appropriately be excluded, including (a) restructurings, discontinued operations, extraordinary items, and other unusual, infrequently occurring or non-recurring charges or events, (b) asset write-downs, (c) litigation or claim judgments or settlements, (d) acquisitions or divestitures, (e) reorganization or change in the corporate structure or capital structure of the Company, (f) an event either not directly related to the operations of the Company, Subsidiary, division, business segment or business unit or not within the reasonable control of management, (g) foreign exchange gains and losses, (h) a change in the fiscal year of the Company, (i) the refinancing or repurchase of bank loans or debt securities, (j) unbudgeted capital expenditures, (k) the issuance or repurchase of equity securities and other changes in the number of issued and outstanding shares, (1) conversion of some or all of convertible securities to Ordinary Shares, (m) any business interruption event (n) the cumulative effects of tax or accounting changes in accordance with U.S. generally accepted accounting principles, or (o) the effect of changes in other laws or regulatory rules affecting reported results.

11.29 "Plan" means this 2018 Incentive Award Plan, as amended and restated.

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

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

11.32 “Restricted Share” means a Share awarded to a Participant under Article VI subject to certain vesting conditions and other restrictions.

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

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

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

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

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

11.38 “Shares” means Ordinary Shares.

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

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

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

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

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

**KINIKSA PHARMACEUTICALS INTERNATIONAL, PLC**
**2018 INCENTIVE AWARD PLAN**

## SHARE OPTION GRANT NOTICE

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

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

**Participant:**
**Grant Date:**
**Grant Number:**
**Exercise Price per Share:**
**Shares Subject to the Option:**
**Final Expiration Date:**
**Vesting Commencement Date:**
**Vesting Schedule:**
**Type of Option:**

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

| **KINIKSA PHARMACEUTICALS INTERNATIONAL, PLC** | **PARTICIPANT** |
|---|---|
| By: /s/ Sanj K. Patel | _____ |
| Name: Sanj K. Patel | |
| Title: CEO and Chairman of the Board | |

## SHARE OPTION AGREEMENT

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

### ARTICLE I
### GENERAL

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

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

### ARTICLE II
### PERIOD OF EXERCISABILITY

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

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

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

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

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

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

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

## ARTICLE III
## EXERCISE OF OPTION

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

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

3.3 Tax Withholding.

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

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

## ARTICLE IV
## OTHER PROVISIONS

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

4.2 Notices. Any notice to be given under the terms of this Agreement to the Company must be in writing and addressed to the Company in care of the Company's Secretary at the Company's principal office or the Secretary's then-current email address or facsimile number. Any notice to be given under the terms of this Agreement to Participant must be in writing and addressed to Participant (or, if Participant is then deceased, to the person entitled to exercise the

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

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

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

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

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

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

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

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

receive the Shares as a general unsecured creditor with respect to the Option, as and when exercised pursuant to the terms hereof.

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

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

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

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

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

**KINIKSA PHARMACEUTICALS INTERNATIONAL, PLC**
**2018 INCENTIVE AWARD PLAN**

# RESTRICTED SHARE UNIT GRANT NOTICE

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

The Company has granted to the participant listed below "**Participant**" the Restricted Share Units described in this Grant Notice (the subject to the terms and conditions of the Plan and the Restricted Share Unit Agreement attached as

**Exhibit A** (the "**Agreement**"), both of which are incorporated into this Grant Notice by reference.

Participant:
Grant Date:
Grant Number:
Number of RSUs:
Vesting Commencement Date:
Vesting Schedule:

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

| **KINIKSA PHARMACEUTICALS INTERNATIONAL, PLC** | **PARTICIPANT** |
|---|---|
| By: /s/ Sanj K. Patel | _____ |
| Name: Sanj K. Patel | |
| Title: CEO and Chairman of the Board | |

**Exhibit A**

## RESTRICTED SHARE UNIT AGREEMENT

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

### ARTICLE I
### GENERAL

1.1 Award of RSUs and Dividend Equivalents.

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

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

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

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

### ARTICLE II
### VESTING; FORFEITURE AND SETTLEMENT

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

2.2 Settlement.

(a) RSUs and Dividend Equivalents (including any Dividend Equivalent Account balance) will be paid in Shares or cash at the Company's option as soon as administratively practicable after the vesting of the applicable RSU, but in no event more than sixty (60) days after the RSU's vesting date. Notwithstanding the foregoing, the Company may delay any payment under this Agreement that the Company reasonably determines would violate Applicable Law until the earliest date the Company reasonably determines the making of the payment will not cause such a violation (in accordance with Treasury Regulation Section 1.409A-2(b)(7) (ii)), provided the Company reasonably believes the delay will not result in the imposition of excise taxes under Section 409A.

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

## ARTICLE III
## TAXATION AND TAX WITHHOLDING

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

3.2 Tax Withholding.

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

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

## ARTICLE IV
## OTHER PROVISIONS

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

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

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

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

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

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

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

4.8 Agreement Severable. In the event that any provision of the Grant Notice or this Agreement is held illegal or invalid, the provision will be severable and the illegality or invalidity

of the provision will not be construed to have any effect on, the remaining provisions of the Grant Notice or this Agreement.

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

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

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

**CERTIFICATIONS**

I, Sanj K. Patel, certify that:

1. I have reviewed this Quarterly Report on Form 10-Q of Kiniksa Pharmaceuticals International, plc;

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.

July 29, 2025

/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 International, plc;

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.

July 29, 2025

/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 International, plc (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, 2025 (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.

July 29, 2025

/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 International, plc (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, 2025 (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.

July 29, 2025

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