Kiniksa Pharmaceuticals Reports First Quarter 2024 Financial Results and Recent Portfolio Execution

April 23, 2024

– ARCALYST® (rilonacept) Q1 2024 net product revenue of $78.9 million, representing 85% year-over-year growth –
– ARCALYST 2024 expected net product revenue increased to $370 - $390 million –
– Abiprubart Phase 2b trial in Sjögren's Disease planned to initiate in 2H 2024 –
– Current operating plan expected to remain cash flow positive on an annual basis –
– Conference call and webcast scheduled for 8:30 am ET today –

HAMILTON, Bermuda, April 23, 2024 (GLOBE NEWSWIRE) -- Kiniksa Pharmaceuticals, Ltd. (Nasdaq: KNSA) (Kiniksa), a commercial-stage biopharmaceutical company with a pipeline of immune-modulating assets designed to target a spectrum of cardiovascular and autoimmune diseases, today reported first quarter 2024 financial results and recent portfolio execution.

"One of the drivers of strong ARCALYST growth is an expanding utilization of ARCALYST as a steroid-sparing therapy for patients suffering from recurrent pericarditis. In addition, with continued frequent prescriber engagement and high physician and patient satisfaction, we now expect 2024 ARCALYST net sales to increase from our previous guidance of between $360 and $380 million to $370 and $390 million," said Sanj K. Patel, Chairman and Chief Executive Officer of Kiniksa. "Additionally, we believe abiprubart has the potential to provide meaningful benefit through convenient subcutaneous dosing, and we plan to initiate a Phase 2b trial in Sjögren's Disease in the second half of 2024. This continued commercial execution and pipeline advancement is included in our current operating plan, which we expect to remain cash flow positive on an annual basis."

Portfolio Execution
ARCALYST (IL-1α and IL-1β cytokine trap)

- ARCALYST net product revenue was $78.9 million for the first quarter of 2024.
- Since launch in April 2021, approximately 2,000 prescribers have written ARCALYST prescriptions for recurrent pericarditis.
- As of the end of the first quarter of 2024, average total duration of ARCALYST therapy in recurrent pericarditis was approximately 23 months.
- Data from the RESONANCE patient registry were presented at the American College of Cardiology Scientific Session (ACC.24). The data highlight a shift in recurrent pericarditis management by pericarditis-focused cardiologists at 21 academic sites across the US and demonstrate increasing proportional ARCALYST use compared to and in advance of corticosteroids since ARCALYST commercial availability for recurrent pericarditis in 2021.

Abiprubart (anti-CD40 monoclonal antibody inhibitor of CD40-CD154 interaction)

- Kiniksa expects to initiate a Phase 2b trial designed to evaluate the efficacy of biweekly and monthly abiprubart administered subcutaneously in patients with Sjögren's Disease in the second half of 2024.

Mavrilimumab (monoclonal antibody inhibitor targeting GM-CSFRα)

- Kiniksa is evaluating potential partnership opportunities to advance development of mavrilimumab, which has generated positive data in mid-stage clinical trials across multiple indications.

Financial Results

- Total revenue for the first quarter of 2024 was $79.9 million, compared to $48.3 million for the first quarter of 2023.
  - Total revenue for the first quarter of 2024 included $1.0 million in license and collaboration revenue, compared to $5.7 million for the first quarter of 2023.
- Total operating expenses for the first quarter of 2024 were $96.4 million, compared to $59.5 million for the first quarter of 2023.
  - Total operating expenses for the first quarter of 2024 included $20.8 million in collaboration expenses, which are driven by ARCALYST collaboration profitability, compared to $8.3 million for the first quarter of 2023.
  - Total operating expenses for the first quarter of 2024, included $7.2 million in non-cash, share-based compensation expense, compared to $6.1 million for the first quarter of 2023.
- Net loss for the first quarter of 2024 was $17.7 million, compared to a net loss of $12.3 million for the first quarter of 2023.
- As of March 31, 2024, Kiniksa had $213.6 million of cash, cash equivalents, and short-term investments and no debt.

Financial Guidance

- Kiniksa expects 2024 ARCALYST net product revenue of between $370 million and $390 million, compared to prior guidance of between $360 million and $380 million.
- Kiniksa expects its current operating plan to remain cash flow positive on an annual basis.

Conference Call Information

- Kiniksa will host a conference call and webcast at 8:30 a.m. Eastern Time on Tuesday, April 23, 2024, to discuss first quarter 2024 financial results and recent portfolio execution.
- Individuals interested in participating in the call via telephone may register here. Upon registration, all telephone participants will receive a confirmation email detailing how to join the conference call, including the dial-in number along with a unique passcode and registrant ID that can be used to access the call. To access the webcast, please visit the Investors and Media section of Kiniksa's website. A replay of the event will also be available on Kiniksa's website within approximately 48 hours after the event.

About Kiniksa
Kiniksa is a commercial-stage biopharmaceutical company focused on discovering, acquiring, developing, and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. Kiniksa's immune-modulating assets, ARCALYST, abiprubart, and mavrilimumab, are based on strong biologic rationale or validated mechanisms, target a spectrum of underserved cardiovascular and autoimmune conditions, and offer the potential for differentiation. For more information, please visit www.kiniksa.com.

About ARCALYST
ARCALYST is a weekly, subcutaneously injected recombinant dimeric fusion protein that blocks interleukin-1 alpha (IL-1α) and interleukin-1 beta (IL-1β) signaling. ARCALYST was discovered by Regeneron Pharmaceuticals, Inc. (Regeneron) and is approved by the U.S. Food and Drug Administration (FDA) for recurrent pericarditis, cryopyrin-associated periodic syndromes (CAPS), including Familial Cold Autoinflammatory Syndrome and Muckle-Wells Syndrome, and deficiency of IL-1 receptor antagonist (DIRA). The FDA granted Breakthrough Therapy designation to ARCALYST for the treatment of recurrent pericarditis in 2019 and Orphan Drug exclusivity to ARCALYST in 2021 for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and pediatric patients 12 years and older. The European Commission granted Orphan Drug Designation to ARCALYST for the treatment of idiopathic pericarditis in 2021.

IMPORTANT SAFETY INFORMATION ABOUT ARCALYST

- ARCALYST may affect your immune system and can lower the ability of your immune system to fight infections. Serious infections, including life-threatening infections and death, have happened in patients taking ARCALYST. If you have any signs of an infection, call your doctor right away. Treatment with ARCALYST should be stopped if you get a serious infection. You should not begin treatment with ARCALYST if you have an infection or have infections that keep coming back (chronic infection).
- While taking ARCALYST, do not take other medicines that block interleukin-1, such as Kineret® (anakinra), or medicines that block tumor necrosis factor, such as Enbrel® (etanercept), Humira® (adalimumab), or Remicade® (infliximab), as this may increase your risk of getting a serious infection.
- Talk with your doctor about your vaccine history. Ask your doctor whether you should receive any vaccines before you begin treatment with ARCALYST.
- Medicines that affect the immune system may increase the risk of getting cancer.
- Stop taking ARCALYST and call your doctor or get emergency care right away if you have any symptoms of an allergic reaction.
- Your doctor will do blood tests to check for changes in your blood cholesterol and triglycerides.
- Common side effects include injection-site reactions (which may include pain, redness, swelling, itching, bruising, lumps, inflammation, skin rash, blisters, warmth, and bleeding at the injection site), upper respiratory tract infections, joint and muscle aches, rash, ear infection, sore throat, and runny nose.

For more information about ARCALYST, talk to your doctor and see the Product Information.

About Abiprubart
Abiprubart is an investigational humanized monoclonal antibody that binds to CD40 and is designed to inhibit the CD40-CD154 (CD40 ligand) interaction, a key T-cell co-stimulatory signal critical for B-cell maturation and immunoglobulin class switching and Type 1 immune responses. Kiniksa believes disrupting the CD40-CD154 co-stimulatory interaction is an attractive approach to addressing multiple autoimmune disease pathologies.

About Mavrilimumab
Mavrilimumab is an investigational fully human monoclonal antibody that blocks activity of GM-CSF by specifically binding to the alpha subunit of the GM-CSF receptor (GM-CSFRα). Phase 2 clinical trials of mavrilimumab in rheumatoid arthritis and giant cell arteritis achieved their primary and secondary endpoints with statistical significance. Kiniksa is evaluating potential partnership opportunities for mavrilimumab.

Forward-Looking Statements
This press release contains forward-looking statements. In some cases, you can identify forward looking statements by terms such as "may," "will," "should," "expect," "plan," "anticipate," "could," "intend," "target," "project," "contemplate," "believe," "estimate," "predict," "potential" or "continue" or the negative of these terms or other similar expressions, although not all forward-looking statements contain these identifying words. All statements contained in this press release that do not relate to matters of historical fact should be considered forward-looking statements, including without

limitation, statements regarding: our expectation that ARCALYST 2024 net product revenue will be between $370 million and $390 million; our plan to initiate a Phase 2b clinical trial of abiprubart in Sjögren's Disease in the second half of 2024; our expectation to remain cash flow positive on an annual basis within our current operating plan; future clinical trial design, including the design of our planned Phase 2b trial of abiprubart in Sjögren's Disease; our beliefs about the mechanisms of our product candidates and potential impact of their approach, including that using abiprubart to disrupt the CD40-CD154 co-stimulatory interaction is an attractive approach to address multiple autoimmune disease pathologies; and our belief that all of our product candidates offer the potential for differentiation.

These forward-looking statements are based on management's current expectations. These statements are neither promises nor guarantees, but involve known and unknown risks, uncertainties and other important factors that may cause 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, including without limitation, the following: risks arising from the planned redomiciliation of our principal holding company from Bermuda to the United Kingdom; delays or difficulty in enrollment of patients in, and activation or continuation of sites for, our clinical trials; delays or difficulty in completing our clinical trials as originally designed; potential for changes between final data and any preliminary, interim, top-line or other data from clinical trials; our inability to replicate results from our earlier clinical trials or studies; impact of additional data from us or other companies, including the potential for our data to produce negative, inconclusive or commercially uncompetitive results; potential undesirable side effects caused by our products and product candidates; our inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities; potential for applicable regulatory authorities to not accept our filings, delay or deny approval of any of our product candidates or require additional data or trials to support approval; our reliance on third parties as the sole source of supply of the drug substance and drug product used in our product candidates; raw material, important ancillary product and drug substance and/or drug product shortages; our reliance on third parties to conduct research, clinical trials, and/or certain regulatory activities for our product candidates; complications in coordinating requirements, regulations and guidelines of regulatory authorities across jurisdictions for our clinical trials; changes in our operating plan, business development strategy or funding requirements; and existing or new competition.

These and other important factors discussed in our filings with the U.S. Securities and Exchange Commission, including under the caption "Risk Factors" contained therein, could cause actual results to differ materially from those indicated by the forward-looking statements made in this press release. Any such forward-looking statements represent management's estimates as of the date of this press release. Except as required by law, we disclaim any intention or obligation to update or revise any forward-looking statements. These forward-looking statements should not be relied upon as representing our views as of any date subsequent to the date of this press release.

Every Second Counts! ®

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

	Three Months Ended March 31,	
	2024	**2023**
Revenue:		
Product revenue, net	$ 78,885	$ 42,659
License and collaboration revenue	973	5,686
Total revenue	79,858	48,345
Operating expenses:		
Cost of goods sold	10,583	7,036
Collaboration expenses	20,801	8,288
Research and development	26,334	15,172
Selling, general and administrative	38,682	29,045
Total operating expenses	96,400	59,541
Loss from operations	(16,542)	(11,196)
Other income	2,266	1,832
Loss before income taxes	(14,276)	(9,364)
Provision for income taxes	(3,428)	(2,906)
Net loss	$ (17,704)	$ (12,270)
Net loss per share attributable to common shareholders —basic and diluted	$ (0.25)	$ (0.18)
Weighted average common shares outstanding—basic and diluted	70,633,023	69,751,697

KINIKSA PHARMACEUTICALS, LTD.

SELECTED CONSOLIDATED BALANCE SHEET DATA
(In thousands)
(Unaudited)

	As of	
	March 31, 2024	**December 31, 2023**
Cash, cash equivalents, and short-term investments	$ 213,552	$ 206,371
Working capital	206,379	212,631
Total assets	519,673	526,322
Accumulated deficit	(495,654)	(477,950)
Total shareholders' equity	431,895	438,839