Kiniksa Pharmaceuticals Reports Second Quarter 2022 Financial Results and Provides Corporate Update

August 3, 2022

– ARCALYST® (rilonacept) net revenue of $27.0 million in Q2 2022 –

– ARCALYST full-year 2022 net revenue expected to be $115 - $130 million –

– Upfront and near-term proceeds of $100 million expected from vixarelimab global license agreement –

– Cash reserves after the close of the vixarelimab global license agreement expected to fund operations into at least 2025 –

– Conference call and webcast scheduled for 8:30 am ET today –

HAMILTON, Bermuda, Aug. 03, 2022 (GLOBE NEWSWIRE) -- Kiniksa Pharmaceuticals, Ltd. (Nasdaq: KNSA) (Kiniksa), a biopharmaceutical company with a portfolio of assets designed to modulate immunological pathways across a spectrum of diseases, today reported second quarter 2022 financial results and provided a corporate update.

“The continued momentum of ARCALYST in recurrent pericarditis in the second quarter of 2022 provides conviction in our full-year expectation for net revenue of between $115 to 130 million. Additionally, we believe the strong performance of ARCALYST since launch supports incremental investment to broaden our reach and help even more patients suffering from recurrent pericarditis,” said Sanj K. Patel, Chairman and Chief Executive Officer of Kiniksa. “We are also focused on expanding our portfolio by leveraging our cross-functional cardiovascular expertise. These efforts will be enabled in part by the non-dilutive proceeds from our global license agreement with Genentech.”

Corporate Update:

- Today, Kiniksa announced a global license agreement with Roche and Genentech, a member of the Roche Group (Genentech), for the rights to develop and commercialize vixarelimab.
  - Kiniksa will receive upfront and near-term proceeds of $100 million. In addition, the company is eligible to receive up to approximately $600 million in certain development, regulatory, and sales-based milestones, before fulfilling upstream financial obligations, as well as royalties on annual net sales.
  - Kiniksa completed screening patients for the Phase 2b clinical trial of vixarelimab in prurigo nodularis and plans to complete the trial. The company will not disclose data in the second half of 2022.
  - Kiniksa plans to use the non-dilutive proceeds received from the transaction to advance synergistic cardiovascular opportunities.

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

- ARCALYST net revenue was $27.0 million for the second quarter of 2022.
  - More than 550 prescribers have written ARCALYST prescriptions for recurrent pericarditis since launch, with a growing number of repeat prescribers.
  - More than 90% payer approval rate of completed patient cases for recurrent pericarditis in the second quarter of 2022.
  - ARCALYST use in recurrent pericarditis to date indicates continuous treatment durations of approximately 12 months.
- Kiniksa plans to evolve its sales operation with approximately 20 additional field sales representatives in the fourth quarter of 2022.

KPL-404 (monoclonal antibody inhibitor of CD40-CD154 interaction)

- Kiniksa is conducting a Phase 2 clinical trial of KPL-404 in rheumatoid arthritis which is designed to evaluate the efficacy, dose response, pharmacokinetics, and safety of chronic subcutaneous dosing over 12 weeks.

Mavrilimumab (monoclonal antibody inhibitor targeting GM-CSFRα)

- Kiniksa is evaluating the development of mavrilimumab in rare cardiovascular diseases where the granulocyte macrophage

colony stimulating factor (GM-CSF) mechanism has been implicated and that have synergies with the company's existing commercial infrastructure.

Financial Results

- Total net revenue for ARCALYST product sales in the second quarter of 2022 was $27.0 million, compared to $7.7 million for the second quarter of 2021.
- Total operating expenses for the second quarter of 2022 were $46.3 million, compared to $48.3 million for the second quarter of 2021.
    - Collaboration expense in the second quarter of 2022 was $3.7 million. Kiniksa did not report a collaboration expense in the second quarter of 2021.
    - Non-cash, share-based compensation expense for the second quarter of 2022 was $6.7 million, compared to $5.7 million for the second quarter of 2021.
- Net loss for the second quarter of 2022 was $20.0 million, compared to a net loss of $41.6 million for the second quarter of 2021.
- As of June 30, 2022, the company had $138.2 million of cash, cash equivalents, and short-term investments, and no debt.

Financial Guidance

- Kiniksa continues to expect ARCALYST net revenue for the full-year 2022 to be between $115 million and $130 million.
- Kiniksa expects that its cash and cash equivalents will fund its current operating plan into at least 2025 following the close of the vixarelimab global license agreement with Genentech.

Conference Call Information

- Kiniksa will host a conference call and webcast at 8:30 a.m. Eastern Time on Wednesday, August 3, 2022, to discuss second quarter 2022 financial results and to provide a corporate update.
- Individuals interested in participating in the call should dial (800) 715-9871 (U.S. and Canada) or (646) 307-1963 (international) using conference ID number 1606846. To access the webcast, please visit the Investors and Media section of Kiniksa's website. A replay of the webcast will also be available on Kiniksa's website within approximately 48 hours after the event.

About Kiniksa
Kiniksa is a biopharmaceutical company focused on discovering, acquiring, developing, and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. Kiniksa's portfolio assets, ARCALYST, KPL-404, and mavrilimumab, are based on strong biologic rationale or validated mechanisms, target underserved conditions, and offer the potential for differentiation. These assets are designed to modulate immunological pathways across a spectrum of diseases. For more information, please visit www.kiniksa.com.

About 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 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 designation to ARCALYST for the treatment of pericarditis in 2020. The European Commission granted Orphan Drug Designation to ARCALYST for the treatment of idiopathic pericarditis in 2020.

ARCALYST is indicated for:

- Treatment of Recurrent Pericarditis (RP) and reduction in risk of recurrence in adults and pediatric patients 12 years and older.
- Treatment of Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Auto-inflammatory Syndrome (FCAS), and Muckle-Wells Syndrome (MWS) in adults and children 12 years and older.
- Maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA) in adults and pediatric patients weighing 10 kg or more.

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 KPL-404
KPL-404 is an investigational humanized monoclonal antibody that is designed to inhibit 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 interaction is an attractive approach to address 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 GCA achieved their primary and secondary endpoints with statistical significance. Kiniksa is evaluating the development of mavrilimumab in rare cardiovascular diseases where the GM-CSF mechanism has been implicated and that have synergies with the company's existing commercial infrastructure.

Forward-Looking Statements
This press release contains forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995. In some cases, you can identify forward looking statements by terms such as "may," "will," "should," "expect," "plan," "anticipate," "could," "intend," "target," "project," "contemplate," "believe," "estimate," "predict," "potential" or "continue" or the negative of these terms or other similar expressions, although not all forward-looking statements contain these identifying words. All statements contained in this press release that do not relate to matters of historical fact should be considered forward-looking statements, including without limitation, statements regarding: the global license agreement between Kiniksa and Genentech, including (i) anticipated upfront, near-term, milestone and royalty payments under such agreement, (ii) statements regarding the use of proceeds from the agreement and (iii) Kiniksa's plan to complete its Phase 2b clinical trial of vixarelimab in prurigo nodularis; our expectation that ARCALYST net revenue for full-year 2022 will be between $115 million and $130 million; our plans to evolve our sales operation with approximately 20 additional field sales representatives in the fourth quarter of 2022; our expectation about our cash reserves funding our current operating plan into 2025 following the close of the vixarelimab global license agreement with Genentech; our expectations regarding our next steps for mavrilimumab; our beliefs about the mechanisms of action of our product candidates and potential impact of their approach, including that using KPL-404 to disrupt the CD40-CD154 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: ours and Genentech's ability to obtain antitrust clearance and close our global license agreement in a timely manner; our ability to realize anticipated near-term payments, milestones and royalty payments under such agreement; our ability to successfully utilize the proceeds we will receive from such agreement; 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; inability to successfully execute on our commercial strategy for ARCALYST; our reliance on third parties as the sole source of supply of the drug substance and drug product used in our products and product candidates; our reliance on Regeneron as the sole manufacturer of ARCALYST; raw materials, important ancillary products 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; the impact of the COVID-19 pandemic and any subsequent pandemic and measures taken in response to such pandemics on our business and operations as well as the business and operations of our manufacturers, CROs upon whom we rely to conduct our clinical trials, and other third parties with whom we conduct business or otherwise engage, including the FDA and other regulatory authorities; changes in our operating plan and 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.

ARCALYST® is a registered trademark of Regeneron Pharmaceuticals, Inc. All other trademarks are the property of their respective owners.

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 June 30,		Six Months Ended June 30,	
	2022	**2021**	**2022**	**2021**
Revenue:				
Product revenue, net	$ 26,972	$ 7,704	$ 49,161	$ 7,704
Collaboration revenue	—	—	10,000	—
Total revenue	26,972	7,704	59,161	7,704
Costs and operating expenses:				
Cost of goods sold	5,029	2,466	9,248	2,466
Collaboration expenses	3,672	—	11,926	—
Research and development	13,798	23,945	34,615	52,628
Selling, general and administrative	23,841	21,848	46,059	42,448
Total operating expenses	46,340	48,259	101,848	97,542
Loss from operations	(19,368)	(40,555)	(42,687)	(89,838)
Interest income	103	6	137	15
Loss before provision for income taxes	(19,265)	(40,549)	(42,550)	(89,823)
Provision for income taxes	(716)	(1,014)	(2,641)	(1,224)
Net loss	$ (19,981)	$ (41,563)	$ (45,191)	$ (91,047)
Net loss per share attributable to common shareholders—basic and diluted	$ (0.29)	$ (0.61)	$ (0.65)	$ (1.33)
Weighted average common shares outstanding—basic and diluted	69,289,972	68,395,703	69,213,860	68,332,943

KINIKSA PHARMACEUTICALS, LTD.
SELECTED CONSOLIDATED BALANCE SHEET DATA
(In thousands)
(Unaudited)

	As of	
	June 30, 2022	**December 31, 2021**
Cash, cash equivalents, and short-term investments	$ 138,208	$ 182,201
Working capital	136,394	151,622
Total assets	210,576	232,800
Accumulated deficit	(720,588)	(675,397)
Total shareholders' equity	153,494	185,037