Kiniksa Pharmaceuticals Provides Corporate and Portfolio Update

January 9, 2023

– ARCALYST® (rilonacept) Q4 2022 and full-year 2022 net product revenue of $39.9 million and $122.5 million, respectively (unaudited) –

– 2 of 3 cohorts enrolled in KPL-404 Phase 2 trial in rheumatoid arthritis; data expected in 1H 2024 –

– Cash reserves expected to fund operations into at least 2025 –

HAMILTON, Bermuda, Jan. 09, 2023 (GLOBE NEWSWIRE) -- Kiniksa Pharmaceuticals, Ltd. (Nasdaq: KNSA) (Kiniksa), a biopharmaceutical company with a pipeline of immune-modulating assets designed to target a spectrum of cardiovascular and autoimmune diseases, today provided a corporate and portfolio update.

"Kiniksa continues to optimize its portfolio and is well-positioned for both near- and long-term growth. We plan to advance our cardiovascular franchise through continued ARCALYST commercial execution in recurrent pericarditis and the pursuit of collaborative study agreements with mavrilimumab. Additionally, we are building an autoimmune franchise initially through the clinical trial of KPL-404, our CD40 antagonist, in rheumatoid arthritis," said Sanj K. Patel, Chairman and Chief Executive Officer of Kiniksa. "These efforts are supported by our profitable ARCALYST collaboration, non-dilutive capital from recent out-licensing transactions, and continued financial discipline. This combination provides cash runway into at least 2025 and allows for continued capital allocation to our existing programs as well as synergistic opportunities."

Corporate Update

- Kiniksa's year-end 2022 cash, cash equivalents, and short-term investments of $190.4 million (unaudited) are expected to fund its current operating plan into at least 2025.
- Kiniksa's resources are currently focused on growing its cardiovascular franchise through ARCALYST sales and mavrilimumab collaborative study agreements, as well as building an autoimmune franchise, initially with the clinical trial of KPL-404 in rheumatoid arthritis.

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

- ARCALYST net product revenue was $39.9 million and $122.5 million for the fourth quarter and full year 2022, respectively (unaudited).
- Since launch, greater than 800 prescribers have written ARCALYST prescriptions for recurrent pericarditis, with 22% having written prescriptions for 2 or more patients.
- As of the end of the fourth quarter of 2022, there was a greater than 90% payer approval rate of completed patient cases for recurrent pericarditis.
- As of the end of the fourth quarter of 2022, the average duration of initial therapy in the commercial setting was approximately 12 months, and approximately 45% of recurrent pericarditis patients who had discontinued ARCALYST therapy restarted treatment.
- As of the end of the fourth quarter of 2022, approximately 5% of the target 14,000 multiple-recurrent pericarditis patients were actively on ARCALYST treatment.
- Kiniksa expects to provide full-year 2023 ARCALYST net product revenue guidance with its fourth quarter and full-year 2022 financial results.

Portfolio Update
KPL-404 (monoclonal antibody inhibitor of CD40-CD154 signaling)

- Kiniksa has completed enrollment of the second and final cohort of the multiple ascending dose portion of the Phase 2 clinical trial of KPL-404 in rheumatoid arthritis. Following completion of this portion of the trial, the proof-of-concept portion will begin. The company expects data from the trial in the first half of 2024.

Mavrilimumab (monoclonal antibody inhibitor targeting GM-CSFRα)

- Kiniksa is pursuing collaborative study agreements to evaluate the potential of mavrilimumab in rare cardiovascular diseases where the granulocyte macrophage colony stimulating factor (GM-CSF) mechanism has been implicated.

41st Annual J.P. Morgan Healthcare Conference

Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa will provide a corporate presentation at the 41st Annual J.P. Morgan Healthcare Conference today, Monday, January 9, 2023, at 1:30 p.m. Pacific Time (4:30 p.m. Eastern Time). A live webcast of Kiniksa's presentation will be accessible through the Investors & Media section of the company's website at www.kiniksa.com. 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 of immune-modulating assets, ARCALYST, KPL-404, and mavrilimumab, are based on strong biologic rationale or validated mechanisms, target a spectrum of underserved cardiovascular and autoimmune conditions, and offer the potential for differentiation. 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.

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 giant cell arteritis 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.

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: our plan to (i) advance our cardiovascular franchise through continued ARCALYST commercial execution in recurrent pericarditis and the pursuit of collaborative study agreements with mavrilimumab and (ii) build an autoimmune franchise, initially with the clinical trial of KPL-404 in rheumatoid arthritis; our expectation that (i) the proof-of-concept portion of the Phase 2 clinical trial of KPL-404 in rheumatoid arthritis will begin after completion of the multiple ascending dose portion of such trial, and (ii) we will report data from such Phase 2 clinical trial in the first half of 2024; our pursuit of collaborative study agreements to evaluate the potential of mavrilimumab in rare cardiovascular diseases where the GM-CSF mechanism has been implicated; our expectation to provide full-year 2023 ARCALYST net product revenue guidance with our fourth quarter and full-year 2022 financial results; our expectation about our cash reserves funding our current operating plan into at least 2025; 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: 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 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; 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.

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