Kiniksa Pharmaceuticals Provides Corporate Update

January 4, 2024

– ARCALYST® (rilonacept) 2023 net product revenue grew ~90% year-over-year to $233.1 million (unaudited) –

– ARCALYST 2024 net product revenue expected to be $360 - $380 million –

– Abiprubart (KPL-404) Phase 2 rheumatoid arthritis trial met the primary efficacy endpoint in Cohort 3 at the weekly dose level –

– Abiprubart Phase 2 rheumatoid arthritis data from Cohort 4 expected in Q2 2024 –

– Cash reserves of $206.3 million (unaudited) expected to fund operations into at least 2027–

HAMILTON, Bermuda, Jan. 04, 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 provided a corporate update.

"Strong execution to date has laid the foundation for the continued advancement of Kiniksa's portfolio in 2024. ARCALYST 2023 net product revenue grew ~90% year-over-year to $233.1 million, underscoring our robust commercial performance. We believe there is substantial opportunity with ARCALYST in recurrent pericarditis and expect to drive continued revenue growth and collaboration profitability by reaching an increasing number of patients. In fact, at the end of 2023 Kiniksa penetrated approximately 9% into the multiple-recurrence population, compared to approximately 5% at the end of 2022," said Sanj K. Patel, Chairman and Chief Executive Officer of Kiniksa. "Additionally, abiprubart showed clinical effect in the first three cohorts of the Phase 2 trial in rheumatoid arthritis. Despite a high placebo response rate, the 5 mg/kg weekly dose level in Cohort 3 achieved statistical significance, but the 5 mg/kg biweekly dose level did not. We look forward to evaluating results from Cohort 4, and we will use the totality of the data to determine next steps for the program. Importantly, our strong financial position provides optionality to continue to invest across our business, including ARCALYST commercialization as well as both pipeline and business development."

Portfolio Execution

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

- ARCALYST net product revenue was $71.2 million and $233.1 million for the fourth quarter and full year 2023, respectively (unaudited).
- Since launch in April 2021, more than 1,700 prescribers have written ARCALYST prescriptions for recurrent pericarditis.
- As of the end of the fourth quarter of 2023, average total duration of ARCALYST therapy in recurrent pericarditis increased to approximately 23 months.
- As of the end of the fourth quarter of 2023, approximately 9% of the target 14,000 multiple-recurrence patients were actively on ARCALYST treatment.
- Kiniksa increased the size of its salesforce to approximately 85 representatives by the end of 2023 to help drive further physician adoption and patient enrollments in 2024.
- Kiniksa expects 2024 ARCALYST net product revenue of between $360 million and $380 million.

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

- Kiniksa today announced that the Phase 2 clinical trial of abiprubart in rheumatoid arthritis met its primary efficacy endpoint, change from baseline in Disease Activity Score of 28 Joints Using C-reactive Protein (DAS28-CRP) versus placebo.
    - In Cohorts 1 and 2 (pharmacokinetic (PK)-lead in), multiple doses of abiprubart were well-tolerated and enabled the proof-of-concept portion of the study. Although these cohorts were not powered for DAS28-CRP (Secondary Efficacy Endpoint), the following results were observed:
        - In Cohort 1, in the abiprubart 2 mg/kg subcutaneous (SC) biweekly dosing group (n=6), the mean change from baseline in DAS28-CRP at Week 12 was -3.16 points compared to -1.09 points in pooled placebo recipients (n=4), (Mean Difference = -2.07, p=0.0312).
        - In Cohort 2, in the abiprubart 5 mg/kg SC biweekly dosing group (n=6), the mean change from baseline in DAS28-CRP at Week 12 was -3.44 points compared to pooled placebo recipients (n=4), (Mean Difference = -2.35, p=0.0338).
    - In Cohort 3, in the abiprubart 5 mg/kg SC weekly dosing group (n=27), the Least Squares (LS) mean change [95% confidence interval (CI)] from baseline in DAS28-CRP at Week 12 was -2.21 [-2.62, -1.80] points compared to -1.65

[-2.07, -1.23] points in placebo recipients (n=26), (LS Mean Difference = -0.56, p=0.0487).
    - In Cohort 3, in the abiprubart 5 mg/kg SC biweekly dosing group (n=25), the LS mean change [95% CI] from baseline in DAS28-CRP at Week 12 was -2.00 [-2.43, -1.58] points compared to -1.65 [-2.07, -1.23] points in placebo recipients (n=26), (LS Mean Difference = -0.35, p=0.2140).
    - Abiprubart significantly reduced Rheumatoid Factor (a clinical marker of disease activity and autoantibody pharmacodynamic marker of CD40 target engagement) by over 40% in both Cohort 3 dose levels.
    - Abiprubart was well-tolerated, with no dose-related adverse experiences observed.
- Kiniksa has now completed enrollment in a fourth cohort (Cohort 4) of the Phase 2 clinical trial of abiprubart in rheumatoid arthritis. Cohort 4 will evaluate a fixed dose level administered as a single subcutaneous injection once monthly. The company expects data from Cohort 4 in the second quarter of 2024.

Mavrilimumab (monoclonal antibody inhibitor targeting GM-CSFRα)

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

Corporate Update

- Kiniksa's year-end 2023 cash, cash equivalents, and short-term investments of $206.3 million (unaudited) are expected to fund its current operating plan into at least 2027.

42nd 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 42nd Annual J.P. Morgan Healthcare Conference on January 8, 2024, 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 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 (KPL-404)
Abiprubart (KPL-404) 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 the Phase 2 Clinical Trial of Abiprubart in Rheumatoid Arthritis
The ongoing Phase 2 rheumatoid arthritis trial is a randomized, double-blind, placebo-controlled trial designed to evaluate pharmacokinetics, safety, and efficacy of chronic subcutaneous administration of abiprubart and to provide optionality to evaluate abiprubart across a range of autoimmune diseases. This trial enrolled patients with active rheumatoid arthritis who had an inadequate response or were intolerant to a Janus kinase inhibitor (JAKi) or at least one biologic disease-modifying anti-rheumatic drug (bDMARD).

The multiple ascending-dose PK lead-in portion randomized 8 patients each in a 3:1 ratio to receive abiprubart 2 mg/kg or placebo (Cohort 1) or 5 mg/kg or placebo (Cohort 2), administered subcutaneously biweekly over a period of 12 weeks. The primary objective of this part of the trial was to evaluate pharmacokinetics, safety, and tolerability over 12 weeks. The secondary efficacy endpoint was change from baseline in DAS28-CRP versus placebo.

The first part of the proof-of-concept portion of the trial (Cohort 3) randomized 78 patients in a 1:1:1 ratio to receive abiprubart 5 mg/kg SC weekly, abiprubart 5 mg/kg SC biweekly, or placebo over a period of 12 weeks. The final part of the proof-of-concept portion of the trial (Cohort 4) randomized 51 patients in a 3:2 ratio to receive a fixed 600 mg loading dose on Day 1 followed by 400 mg SC every four weeks or placebo over a period of 12 weeks. The primary efficacy endpoint of the proof-of-concept portion of the trial is change from baseline in DAS28-CRP versus placebo.

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 now evaluating potential partnership opportunities for mavrilimumab.

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 expectation that ARCALYST 2024 net product revenue will be between $360 million and $380 million; our plan to report data from Cohort 4 of our Phase 2 clinical trial of abiprubart in rheumatoid arthritis in the second quarter of 2024; our expectation about our cash reserves funding our current operating plan into at least 2027; our expectation that we will drive continued ARCALYST revenue growth and collaboration profitability by reaching an increasing number of patients; 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: 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 current sole manufacturer of ARCALYST; risks arising from our ongoing technology transfer of ARCALYST drug substance manufacturing; 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.

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