Kiniksa Pharmaceuticals Announces Development of KPL-387 in Recurrent Pericarditis and Updates Corporate Strategy

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– Kiniksa plans to initiate a Phase 2/3 clinical trial of KPL-387 in recurrent pericarditis in mid-2025 –

– KPL-387 Phase 1 single ascending dose data support potential monthly dosing –

– Kiniksa continues to focus development on diseases with unmet need, prioritizing cardiovascular indications –

– Kiniksa to discontinue abiprubart development in Sjögren's Disease –

LONDON, Feb. 25, 2025 (GLOBE NEWSWIRE) -- Kiniksa Pharmaceuticals International, plc (Nasdaq: KNSA) (Kiniksa), a biopharmaceutical company developing and commercializing novel therapies for diseases with unmet need, with a focus on cardiovascular indications, today announced the development program for KPL-387 in recurrent pericarditis and provided an update on its corporate strategy. KPL-387 is an independently developed monoclonal antibody that binds human interleukin-1 receptor 1 (IL-1R1), inhibiting the signaling activity of the cytokines interleukin-1α (IL-1α) and interleukin-1β (IL-1β).

"Through the successful development and commercialization of ARCALYST, Kiniksa has helped thousands of patients suffering from recurrent pericarditis. Since the launch in 2021, we have generated over $800 million in product revenue and have become cash flow positive on an annual basis. We plan to further increase our penetration into the recurrent pericarditis population with ARCALYST," said Sanj K. Patel, Chairman and Chief Executive Officer of Kiniksa. "We also continue to focus development on novel therapies for diseases with unmet need, prioritizing cardiovascular indications. Today, we are excited to extend our leadership in the recurrent pericarditis market through the development of KPL-387. We believe KPL-387 could expand the treatment options for recurrent pericarditis patients by providing a single monthly subcutaneous injection in a liquid formulation."

"KPL-387 could provide a meaningful addition to the therapeutic options available to patients suffering from recurrent pericarditis. Data generated from the single ascending dose portion of the ongoing Phase 1 study support our belief that KPL-387 could enable dosing with a single monthly subcutaneous injection in a liquid formulation," said John F. Paolini, M.D., Ph.D., FACC, Chief Medical Officer of Kiniksa. "We have interacted with the FDA, and we plan to initiate a Phase 2/3 clinical trial of KPL-387 in recurrent pericarditis in mid-2025, with Phase 2 data expected in the second half of 2026."

Corporate Update

- Kiniksa continues to focus development on diseases with unmet need, prioritizing cardiovascular indications.
- Kiniksa is developing KPL-387, a fully human immunoglobulin G2 (IgG2) monoclonal antibody that binds IL-1R1, inhibiting the signaling of the cytokines IL-1α and IL-1β, in recurrent pericarditis with a target profile of monthly subcutaneous (SC) dosing.
    - Kiniksa is conducting a single ascending dose (SAD) and multiple ascending dose Phase 1 clinical trial of KPL-387 in healthy volunteers.
        - Topline data from the SAD portion of the Phase 1 trial of KPL-387 support potential monthly SC dosing in recurrent pericarditis.
    - Kiniksa has interacted with the U.S. Food and Drug Administration (FDA) and expects to initiate a Phase 2/3 clinical trial of KPL-387 in recurrent pericarditis in mid-2025, with Phase 2 data expected in the second half of 2026.
- Kiniksa is advancing KPL-1161, an Fc-modified IgG2 monoclonal antibody that binds IL-1R1, inhibiting the signaling of the cytokines IL-1α and IL-1β, towards clinical development with a target profile of quarterly SC dosing.
- Kiniksa plans to discontinue the development of abiprubart in Sjögren's Disease. The company will explore strategic alternatives for the asset.
- Kiniksa has exercised its right to terminate its exclusive license agreement for mavrilimumab with MedImmune.

About Kiniksa
Kiniksa is a biopharmaceutical company dedicated to improving the lives of patients suffering from debilitating diseases by discovering, acquiring, developing, and commercializing novel therapies for diseases with unmet need, with a focus on cardiovascular indications. Kiniksa's portfolio of assets is based on strong biologic rationale or validated mechanisms and offers the potential for differentiation. For more information, please visit www.kiniksa.com.

About ARCALYST
ARCALYST® (rilonacept) 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 the treatment of recurrent pericarditis (RP) and reduction in risk of recurrence in adults and children 12 years and older. ARCALYST is also approved by the FDA for the treatment of Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Autoinflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS) in adults and children 12 years and older, and the maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA) in adults and pediatric patients weighing 10 kg or more. The FDA granted Orphan Drug Exclusivity to ARCALYST upon its approval for recurrent pericarditis in 2021. 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 KPL-387
KPL-387 is an independently developed, investigational, fully human IgG2 monoclonal antibody that binds IL-1R1, inhibiting the signaling of the cytokines IL-1α and IL-1β. Kiniksa believes KPL-387 could expand the treatment options for recurrent pericarditis patients by enabling dosing with a single monthly SC injection in a liquid formulation.

About KPL-1161
KPL-1161 is an independently developed, investigational, Fc-modified IgG2 monoclonal antibody that binds IL-1R1, inhibiting the signaling of the cytokines IL-1α and IL-1β, with a target profile of quarterly SC dosing. Kiniksa is currently engaging in IND-enabling development activities for KPL-1161.

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.

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 plan to initiate a Phase 2/3 clinical trial of KPL-387 in recurrent pericarditis in mid-2025, with Phase 2 data expected in the second half of 2026; our plan to discontinue the development of abiprubart in Sjögren’s Disease and explore strategic alternatives for the asset; our plan to prioritize development in cardiovascular indications; our plan to further increase our penetration into the recurrent pericarditis market with ARCALYST; our belief that KPL-387 could expand the treatment options for recurrent pericarditis patients by providing a single monthly SC injection in a liquid formulation; our target profile of monthly and quarterly SC dosing for KPL-387 and KPL-1161, respectively; our beliefs about the mechanisms of our assets and potential impact of their approach; and our belief that our portfolio of assets offers 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; 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; 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; business development activities and their impact on our financial performance and strategy; 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.

ARCALYST® is a registered trademark of Regeneron Pharmaceuticals, Inc.

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