Kiniksa Announces FDA Approval of ARCALYST ® (rilonacept) for Recurrent Pericarditis

March 18, 2021

- ARCALYST is the first and only FDA-approved therapy for recurrent pericarditis -
- Commercial launch expected in April 2021 -
- Kiniksa launches Kiniksa One Connect™ patient support program -
- Conference call and webcast scheduled for 5:30 p.m. EDT today -

HAMILTON, Bermuda, March 18, 2021 (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 announced the U.S. Food and Drug Administration (FDA) approved ARCALYST ® (rilonacept), a weekly, subcutaneously-injected, recombinant fusion protein that blocks interleukin-1 alpha (IL-1α) and interleukin-1 beta (IL-1β) signaling, for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years and older. The commercial launch is expected in April 2021.

“The approval of ARCALYST in recurrent pericarditis offers patients the first and only FDA-approved therapy for this devastating disease and also represents a transformational event for Kiniksa,” said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. “I would like to thank the recurrent pericarditis community and specifically acknowledge the exceedingly dedicated patients, nurses, physicians and caregivers who participated in the clinical trials as well as the Kiniksa team whose absolute focus on patients made this possible. We look forward to launching ARCALYST for recurrent pericarditis with the support of our experienced commercial and medical affairs teams and, importantly, providing this breakthrough therapy to patients suffering with this debilitating disease as quickly as possible.”

Recurrent pericarditis is a painful autoinflammatory cardiovascular disease that typically presents with chest pain and is often associated with changes in electrical conduction and sometimes buildup of fluid around the heart, called pericardial effusion. Patients who have additional pericarditis episodes following a symptom-free period of 4-6 weeks or longer are identified as having recurrent pericarditis. Recurrent pericarditis symptoms have an impact on quality of life, limit physical activities, and lead to frequent emergency department visits and hospitalizations. Data show that approximately 40,000 patients in the U.S. seek and receive treatment for recurrent pericarditis each year. Of that group, approximately 14,000 patients experience a second or subsequent event (recurrence) due to persistent underlying disease or inadequate response to conventional therapies, such as nonsteroidal anti-inflammatory drugs (NSAIDs), colchicine and corticosteroids.

The FDA approval of ARCALYST in recurrent pericarditis follows positive data from RHAPSODY, a pivotal Phase 3 trial of ARCALYST in recurrent pericarditis. RHAPSODY met its prespecified primary endpoint of time-to-first adjudicated pericarditis recurrence in the randomized withdrawal period and all major secondary efficacy endpoints with statistical significance. The data showed that ARCALYST treatment in the trial improved clinically meaningful outcomes associated with the significant unmet medical need in recurrent pericarditis. There were rapid and sustained reductions in both reported pain and inflammation as early as after the first dose. Median time to treatment response was 5 days, with a 97% treatment response rate. Patients randomized to ARCALYST experienced a 96% reduction in the risk for a recurrent pericarditis event (Hazard Ratio = 0.04, $p<0.0001$), with 92% of trial days being pain free or at most experiencing minimal pain, compared to 40% of trial days on placebo ($p<0.0001$). The most common adverse events were injection site reactions and upper respiratory tract infections. RHAPSODY data were published in *The New England Journal of Medicine* simultaneously with a late-breaking scientific presentation at the American Heart Association’s Scientific Sessions 2020.

“Recurrent pericarditis is a debilitating disease that disrupts the lives of afflicted patients. Data have shown that recurrent pericarditis stems from an underlying autoinflammatory pathophysiology mediated by IL-1α and IL-1β,” said Allan Klein, MD, of Cleveland Clinic, co-principal investigator of RHAPSODY and compensated member of a 2019 Kiniksa scientific advisory committee. “This approval supports the concept of a targeted immunomodulation therapeutic strategy as a paradigm shift in the management of patients with this devastating disease.”

Kiniksa is providing Kiniksa One Connect™, a program that will assist patients on their treatment journey. The program will help provide access, initiation, affordability solutions and ongoing support for any patient receiving ARCALYST therapy. For more information, call 1-833-KINIKSA (1-833-546-4572).

“The approval of ARCALYST in recurrent pericarditis is an extremely important achievement for those suffering from this disease as there can be significant impact on a patient’s physical and emotional health as well as quality of life,” said Dona Winnowski, President of the Pericarditis Alliance. “The development and commercialization of new medicines for underserved and severe diseases are essential, and this first FDA-approved therapy in recurrent pericarditis brings hope to patients and their families.”

ARCALYST was discovered by Regeneron Pharmaceuticals, Inc. (Regeneron) and received initial FDA approval in February 2008 for the treatment of cryopyrin-associated periodic syndromes (CAPS), including Familial Cold Autoinflammatory Syndrome and Muckle-Wells Syndrome, and subsequent approval for the maintenance of remission of deficiency of IL-1 receptor antagonist (DIRA) in December 2020.

Kiniksa licensed ARCALYST from Regeneron in 2017 for evaluation in diseases believed to be mediated by both IL-1α and IL-1β, including recurrent pericarditis. Upon this approval by the FDA for recurrent pericarditis, Kiniksa takes responsibility for sales and distribution of ARCALYST for all the approved indications in the United States, including CAPS and DIRA, and will evenly split profits with Regeneron, as described in the ARCALYST License Agreement.

For more information on ARCALYST, visit www.arcalyst.com.

Conference Call Information
Kiniksa will host a conference call and webcast at 5:30 p.m. Eastern Time on Thursday, March 18, 2021 to discuss the FDA approval of ARCALYST for recurrent pericarditis. Individuals interested in participating in the call should dial (866) 614-0636 (U.S. and Canada) or (409) 231-2053 (international) using conference ID number 2458467. To access the webcast, please visit the Investors and Media section of Kiniksa's website at www.kiniksa.com. The archived webcast will be available on Kiniksa's website for 14 days beginning approximately one hour after the call has completed.

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 assets, ARCALYST, mavrilimumab, vixarelimab and KPL-404, 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 Recurrent Pericarditis
Recurrent pericarditis is a painful and debilitating autoinflammatory cardiovascular disease that typically presents with chest pain and is often associated with changes in electrical conduction and sometimes buildup of fluid around the heart, called pericardial effusion. Patients who have additional pericarditis episodes following a symptom-free period of 4-6 weeks are identified as having recurrent pericarditis. Recurrent pericarditis symptoms have an impact on quality of life, limit physical activities, and lead to frequent emergency department visits and hospitalizations.

About ARCALYST
ARCALYST is a weekly, subcutaneously-injected, recombinant dimeric fusion protein that blocks IL-1α and IL-1β signaling. ARCALYST was discovered by Regeneron and is approved by the FDA for recurrent pericarditis, CAPS, including Familial Cold Autoinflammatory Syndrome and Muckle-Wells Syndrome, and 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.

About RHAPSODY
RHAPSODY is the global, randomized withdrawal design, pivotal Phase 3 clinical trial of ARCALYST in recurrent pericarditis. Eligible patients presented at screening with at least a third pericarditis episode, defined as at least 1 day with pericarditis pain of ≥ 4 on the 11-point Numerical Rating Scale (NRS) and a C-reactive protein (CRP) value ≥ 1 mg/dL within the 7-day period prior to first study drug administration. Patients could be receiving concomitant NSAIDs and/or colchicine and/or oral corticosteroid treatment in any combination. The study was comprised of 4 periods: a screening period; a single-blind run-in period during which patients received a loading dose of ARCALYST 320 mg injected subcutaneously (SC) followed by 160 mg SC weekly while background pericarditis medications were tapered and discontinued; a double-blind, placebo-controlled randomized withdrawal period during which clinical responders to ARCALYST were randomized 1:1 and received 160 mg SC weekly ARCALYST or placebo; and a long-term extension treatment period with up to 24 months of open-label ARCALYST 160 mg SC weekly. The primary efficacy endpoint was time-to-first pericarditis-recurrence in the randomized withdrawal period. The Clinical Endpoint Committee adjudicated all suspected pericarditis recurrences for inclusion in the primary efficacy endpoint analysis. Kiniksa will continue to follow patients in the long-term extension treatment period for up to 24 months. The co-principal investigators are Dr. Allan Klein of Cleveland Clinic and Dr. Massimo Imazio of the University of Torino, Italy. For more information, refer to ClinicalTrials.gov Identifier: NCT03737110.

About the ARCALYST License Agreement with Regeneron
In 2017, Regeneron granted Kiniksa an exclusive license to develop and commercialize ARCALYST worldwide, excluding Israel, Egypt, Turkey and select countries in the Middle East and North Africa. In the United States and Japan, Kiniksa's license is for all indications other than those involving oncology and local administration to the eye or ear. Upon the approval of the supplemental Biologics License Application (sBLA) for ARCALYST in recurrent pericarditis, the scope of the license granted to Kiniksa automatically expanded to include DIRA and CAPS in the United States and Japan, and Kiniksa assumed the responsibility for sales and distribution of ARCALYST in these additional indications in the United States. Outside the United States and Japan, Kiniksa's license is for all indications other than CAPS and certain periodic fever syndromes, DIRA, oncology, and local application to the eye or ear. Upon approval of ARCALYST in recurrent pericarditis, Kiniksa is obligated to make a $20 million milestone payment to Regeneron. Kiniksa and Regeneron will evenly split profits on sales of ARCALYST after deducting certain commercialization expenses, subject to specified limits.

Important information about ARCALYST® Injection

- ARCALYST can 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. You should not begin ARCALYST if you have an infection, or have infections that keep coming back. After starting ARCALYST, if you get an infection or show any sign of an infection, including a fever, cough, flu-like symptoms, or have any open sores on your body, call your doctor right away. Treatment with ARCALYST should be stopped if you get a serious 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.
- Before starting ARCALYST, tell your doctor if you think you have an infection, are being treated for an infection, have signs of an infection, have any open sores, have a history of infections that keep coming back, have asthma, have diabetes or an immune system problem, have tuberculosis or have been in contact without someone who has had tuberculosis, have or have had HIV, hepatitis B or hepatitis C, or take other medicines that affect your immune system.
- Before you begin treatment with ARCALYST, talk with your healthcare provider about your vaccine history. Ask your healthcare provider whether you should receive any vaccines, including the pneumonia vaccine and flu vaccine, before you begin treatment with ARCALYST.
- ARCALYST can cause serious side effects:
    - 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 (eg, rash, swollen face, trouble breathing).
    - Your doctor will do blood tests to check for changes in your blood cholesterol and triglycerides.
- Common side effects of ARCALYST include injection-site reactions, upper respiratory tract infections, joint and muscle aches, rash, ear infection, sore throat, and runny nose.
- Tell your doctor if you are scheduled to receive any vaccines, if you are pregnant or plan to become pregnant, and if you are breastfeeding or plan to breastfeed.
- Tell your doctor if you take other medicines that affect the immune system such as interleukin-1 blockers, tumor necrosis factor blockers, or corticosteroids.

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

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 belief about the impact of FDA approval of ARCALYST in recurrent pericarditis on patients with this disease and on our company; the belief that regulatory approval supports the concept of a targeted immunomodulation therapeutic strategy as a paradigm shift in the management of patients with this disease; our bringing ARCALYST to patients with recurrent pericarditis as quickly as possible; the timing of our commercial launch of ARCALYST in recurrent pericarditis; our targeted patient population; the potential impact of the Kiniksa One Connect program to help provide access, initiation, affordability solutions and ongoing support for any patient receiving ARCALYST therapy; and our beliefs about the mechanisms of action of our product candidates and potential impact of their approach.

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: our inexperience as a company commercializing therapeutic products; our limited experience as a company establishing sales, marketing, distribution and general infrastructure either directly and/or through agreements with third parties; our potential inability to execute on our commercial strategy effectively or at all; the potential inability of our Kiniksa One Connect program to effectively help provide access, initiation, affordability solutions and ongoing support for any patient receiving ARCALYST therapy; the potential for ARCALYST to not gain market acceptance by physicians, patients, or third-party payers for the treatment of recurrent pericarditis; the potential delay or failure of ARCALYST to obtain or maintain coverage and adequate reimbursement for the treatment of recurrent pericarditis; the incidence and prevalence of our target patient population for ARCALYST in recurrent pericarditis may be smaller than we estimate; potential undesirable side effects caused by ARCALYST; our reliance on Regeneron as the sole source of supply of the drug substance and drug products used in ARCALYST and to manufacture our clinical and commercial supply of ARCALYST; drug substance and/or drug product shortages; the impact of the COVID-19 pandemic and measures taken in response to the pandemic on our business and operations as well as the business and operations of our manufacturers, including Regeneron, and other third parties with whom we conduct business or otherwise engage, including regulatory authorities; changes in our operating plan and funding requirements; and existing or new competition.

These and other important factors discussed under the caption “Risk Factors” in our Annual Report on Form 10-K filed with the Securities and Exchange Commission (“SEC”) on February 25, 2021 and our other reports subsequently filed with the SEC 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. While we may elect to update such forward-looking statements at some point in the future, we disclaim any obligation to do so, even if subsequent events cause our views to change. 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!™

Kiniksa Investor and Media Contact
Mark Ragosa
(781) 430-8289
mragosa@kiniksa.com

Source: Kiniksa Pharmaceuticals, Ltd.