Kiniksa Announces Commercial Availability of ARCALYST ® (rilonacept) for Recurrent Pericarditis

April 1, 2021

- ARCALYST is the first and only FDA-approved therapy for recurrent pericarditis –
- ARCALYST now available on prescription basis in the U.S. –
- Kiniksa One Connect™ program provides ongoing patient access and support services –

HAMILTON, Bermuda, April 01, 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 that ARCALYST ® (rilonacept), a weekly, subcutaneously-injected, recombinant fusion protein that blocks interleukin-1 alpha (IL-1α) and interleukin-1 beta (IL-1β) signaling, is now commercially available for recurrent pericarditis. On March 18, 2021, the U.S. Food and Drug Administration (FDA) approved ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years and older.

“We are excited to announce that ARCALYST is now available as the first and only approved therapy for patients with recurrent pericarditis,” said Ross Moat, ARCALYST General Manager. “Our experienced commercial and medical affairs teams have already begun to engage with physicians, payers and patients to raise awareness of this innovative treatment option. Kiniksa is committed to patient access and has established Kiniksa One Connect™, our patient support program, which provides comprehensive access and support services for any patient prescribed ARCALYST.”

ARCALYST is available through a distribution network comprised of several specialty pharmacies, which provide extensive and timely access across the United States.

Kiniksa One Connect™ is available to assist patients on their treatment journey with ARCALYST. The program will help provide potential access, initiation, affordability solutions and ongoing patient support. For more information, call 1-833-KINIKSA (1-833-546-4572).

ARCALYST was discovered and developed by Regeneron Pharmaceuticals, Inc. (Regeneron). Upon the approval by the FDA for recurrent pericarditis, Kiniksa took responsibility for sales and distribution of ARCALYST for all the approved indications in the United States, including cryopyrin-associated periodic syndromes (CAPS) and deficiency of IL-1 receptor antagonist (DIRA), and will evenly split profits with Regeneron, as described in the ARCALYST License Agreement.

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

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. 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.

Kiniksa launched the RESONANCE Registry (REgiStry Of the NAtural history of recurreNt periCarditis in pEdiatric and adult patients; clinicaltrials.gov: NCT04687358), a voluntary patient registry database led by physician-researchers with experience in managing patients with recurrent pericarditis, in March 2021. This registry aims to capture retrospective and prospective, longitudinal, observational data in up to 500 patients across the U.S. The continued advancement in recurrent pericarditis research underscores Kiniksa’s commitment to partner with the healthcare and patient communities to improve the collective knowledge of this debilitating disease.

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 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 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. 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 with someone who has had tuberculosis, has or has had HIV, hepatitis B or hepatitis C, or takes 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 (e.g., 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 continued availability of ARCALYST; our targeted patient population; the potential impact of the Kiniksa One Connect program to help provide access, initiation and affordability solutions as well as 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: 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.

ARCALYST is a registered trademark of Regeneron Pharmaceuticals, Inc.

ENBREL, HUMIRA, KINERET, and REMICADE are trademarks of Immunex Corporation, AbbVie Biotechnology Ltd., Sobi, Inc., and Janssen Biotech, Inc., respectively.

Every Second Counts!™

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