Kiniksa Presents Data on the Burden of Disease in Patients with Recurrent Pericarditis at the European Society of Cardiology Congress 2020

August 31, 2020

HAMILTON, Bermuda, Aug. 31, 2020 (GLOBE NEWSWIRE) -- Kiniksa Pharmaceuticals, Ltd. (Nasdaq: KNSA) ("Kiniksa"), a biopharmaceutical company focused on discovering, acquiring, developing and commercializing therapeutic medicines for patients with significant unmet medical need, presented findings from a real-world patient survey assessing the substantial impact of recurrent pericarditis on health-related quality of life at ESC Congress 2020, the annual meeting of the European Society of Cardiology.

"The results of the patient survey highlight the severe and complex impact of recurrent pericarditis on both physical and mental quality of life. Furthermore, this study highlights the unmet need for a therapy that can rapidly resolve pericarditis episodes with the potential to prevent future recurrences," said Qasim Rizvi, Chief Commercial Officer and SVP of Operations at Kiniksa. "The pivotal Phase 3 trial of rilonacept in recurrent pericarditis demonstrated a high degree of statistical significance on the primary and all major secondary endpoints, therefore we plan to submit an sBLA to the FDA in recurrent pericarditis this year. In preparation for commercialization we are continuing to generate additional evidence on the burden of recurrent pericarditis and also increasing disease awareness among payers, physicians and advocacy groups. We look forward to bringing this potential treatment to patients as soon as possible."

Martin LeWinter, MD, Larner College of Medicine, University of Vermont, was the lead author of the virtual poster *Clinical Characteristics and Health-Related Quality of Life of Patients with Recurrent Pericarditis in the United States: Findings from a Patient Survey.* The materials are available through the Science section of Kiniksa's website (www.Kiniksa.com).

"This study represents the first real-world analysis of patient-reported health-related quality of life in recurrent pericarditis," said Martin LeWinter, MD, Larner College of Medicine, University of Vermont. "Quantifying the impact of recurrent pericarditis on important health-related quality of life measures, including physical and emotional well-being, has enabled a more complete appreciation of the overall burden of recurrent pericarditis. For example, we found that pericarditis pain significantly impacts the ability to work, to be with family members, and to sleep for many patients at a level greater than that reported for many other debilitating diseases. These insights can help clinicians and patients evaluate treatment options and ultimately may contribute to improving outcomes."

Kiniksa reported positive data from RHAPSODY, a global, randomized withdrawal design, pivotal Phase 3 clinical trial of rilonacept in recurrent pericarditis. RHAPSODY showed that rilonacept improved clinically meaningful outcomes associated with the unmet medical need in recurrent pericarditis for patients in the study. The primary efficacy endpoint of time-to-first adjudicated pericarditis recurrence in the randomized withdrawal period was highly statistically significant: rilonacept treatment resulted in a 96% reduction in risk of recurrent pericarditis events (Hazard Ratio = 0.04, $p<0.0001$). All major secondary endpoints were also highly statistically significant.

Rilonacept was discovered and developed by Regeneron Pharmaceuticals, Inc. (Regeneron) and is approved by the U.S. Food and Drug Administration (FDA) under the brand name ARCALYST® for the treatment of for Cryopyrin-Associated Periodic Syndromes (CAPS). Kiniksa licensed rilonacept from Regeneron in 2017 for evaluation in diseases believed to be mediated by both interleukin-1 alpha (IL-1α) and interleukin-1 beta (IL-1β), including recurrent pericarditis. The FDA granted Breakthrough Therapy designation to rilonacept for recurrent pericarditis in 2019 and Orphan Drug designation to rilonacept for pericarditis in 2020. Based on the Phase 3 RHAPSODY data, the Biologic License Application (BLA) for CAPS will transfer to Kiniksa, and the company plans to submit a supplemental Biologic License Application (sBLA) in recurrent pericarditis to the FDA this year. Upon receipt of FDA approval for rilonacept in recurrent pericarditis, Kiniksa will assume the sales and distribution of rilonacept for the approved indications in the United States and evenly split profits on sales with Regeneron.

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 clinical-stage product candidates, rilonacept, mavrilimumab, vixarelimab and KPL-404, are based on strong biologic rationale or validated mechanisms, target underserved conditions and offer the potential for differentiation. These pipeline assets are designed to modulate immunological pathways that are implicated 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 with pericarditis are deemed recurrent if they have an additional episode after a symptom-free period of 4-6 weeks and chronic if symptoms from any one episode last longer than three months. Recurrent pericarditis symptoms impair qualify of life, limit physical activities, and lead to frequent emergency department visits and hospitalizations. Claims analysis, cross validated with published research estimates, supports a prevalent population of approximately 40,000 patients in the U.S. seeking and receiving medical treatment. There are currently no FDA-approved treatments for recurrent pericarditis.

About Rilonacept
Rilonacept is a weekly, subcutaneously-injected, recombinant fusion protein that blocks IL-1α and IL-1β signaling. Rilonacept was discovered and developed by Regeneron and is approved by the FDA under the brand name ARCALYST® for the treatment of CAPS, specifically Familial Cold

Autoinflammatory Syndrome and Muckle-Wells Syndrome. Rilonacept for the treatment of deficiency of the interleukin-1 receptor antagonist (DIRA) is currently pending FDA approval following the submission of an sBLA in June 2020. Rilonacept in recurrent pericarditis is an investigational drug. The FDA granted Breakthrough Therapy designation to rilonacept for recurrent pericarditis in 2019 and Orphan Drug designation to rilonacept for pericarditis in 2020.

Important information about ARCALYST® (rilonacept) Injection

IL-1 blockade may interfere with immune response to infections. Serious, life-threatening infections have been reported in patients taking ARCALYST. ARCALYST should be discontinued if a patient develops a serious infection. Taking ARCALYST with TNF inhibitors is not recommended because this may increase the risk of serious infections.

Patients should not receive a live vaccine while taking ARCALYST. It is recommended that prior to initiation of therapy with ARCALYST patients receive all recommended vaccinations, as appropriate, including pneumococcal vaccine and inactivated influenza vaccine. In the initial development program for ARCALYST, six serious adverse reactions were reported by four patients: Mycobacterium intracellular infection, gastrointestinal bleeding and colitis, sinusitis and bronchitis and Streptococcus pneumoniae meningitis. The most commonly reported adverse reactions associated with ARCALYST were injection site reaction and upper respiratory tract infection. Patients should be monitored for changes in their lipid profiles and provided with medical treatment if warranted. Treatment with immunosuppressants, including ARCALYST, may result in an increase in risk of malignancies. Hypersensitivity reactions associated with ARCALYST administration in clinical studies have been rare. If a hypersensitivity reaction occurs, administration of ARCALYST should be discontinued and appropriate therapy initiated.

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: plans and timing of submitting a supplemental BLA to the FDA in recurrent pericarditis; our beliefs about the potential to bring rilonacept as a potential treatment option for patients with recurrent pericarditis; 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 plans, estimates or 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 potential inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities or otherwise producing negative, inconclusive or commercially uncompetitive results; potential for changes between final data and any preliminary, interim, top-line or other data we announce; impact of additional data from us or other companies; our reliance on third parties as the sole source of supply of the drug substance and drug products used in our product candidates; substantial existing or new competition; potential 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, 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 our ability to attract and retain qualified personnel.

These and other important factors discussed under the caption “Risk Factors” in our Quarterly Report on Form 10-Q filed with the Securities and Exchange Commission (SEC) on August 4, 2020 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 plans, estimates, or expectations 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. and Yescarta® is a registered trademark of Gilead Sciences, Inc., or its related companies.

Every Second Counts!™

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

Source: Kiniksa Pharmaceuticals, Ltd.