Kiniksa Announces New England Journal of Medicine Publication of Rilonacept Phase 3 Data in Recurrent Pericarditis and Late-Breaking Science Presentation at American Heart Association Scientific Sessions 2020

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HAMILTON, Bermuda, Nov. 17, 2020 (GLOBE NEWSWIRE) -- Kiniksa Pharmaceuticals, Ltd. (Nasdaq: KNSA) ("Kiniksa"), a biopharmaceutical company with a pipeline of assets designed to modulate immunological pathways across a spectrum of diseases, today announced that data from RHAPSODY, the pivotal Phase 3 trial of rilonacept in recurrent pericarditis, were published in *The New England Journal of Medicine.* Additionally, the RHAPSODY data were presented at the late-breaking science session during the American Heart Association (AHA) Scientific Sessions 2020. Kiniksa previously reported positive top-line RHAPSODY results in June 2020. Rilonacept is a weekly, subcutaneously-injected, recombinant fusion protein that blocks interleukin-1 alpha (IL-1α) and interleukin-1 beta (IL-1β) signaling.

The manuscript entitled *Phase 3 Trial of Interleukin-1 Trap Rilonacept in Recurrent Pericarditis*, is available on *The New England Journal of Medicine* website with open access for seven days.

The AHA presentation entitled *RHAPSODY: Rilonacept an IL-1α and IL-1β Trap Resolves Pericarditis Episodes and Reduces Risk of Recurrence in a Phase 3 Trial of Patients with Recurrent Pericarditis* was presented virtually by Allan Klein, MD, of Cleveland Clinic, co-principal investigator of the study and compensated member of a 2019 Kiniksa scientific advisory committee. Massimo Imazio, MD, of the University of Torino, Italy, is co-principal investigator.

"Recurrent pericarditis is a debilitating autoinflammatory disease with a clear unmet need," said Dr. Klein. "Data were reported which showed that patients treated with rilonacept experienced a 96% reduction in risk of recurrent pericarditis events. Furthermore, in acutely symptomatic patients who were failing standard management, rilonacept treatment in the study provided rapid and sustained reductions in pain and C-reactive protein as soon as after the first dose. These data suggest that targeted immunomodulation with rilonacept may signal a paradigm shift in the management of patients with recurrent pericarditis."

In RHAPSODY, the primary efficacy endpoint of time-to-first adjudicated pericarditis recurrence in the randomized withdrawal period was highly statistically significant (Hazard Ratio = 0.04, $p<0.0001$). Additionally, annualized incidence of pericarditis recurrence decreased from 4.42 episodes per year prior to the study to 0.15 episodes per year while on rilonacept treatment. All major secondary endpoints were also highly statistically significant.

Rilonacept was well-tolerated in the study, with adverse events consistent with the U.S. Food and Drug Administration (FDA)-approved label for the treatment of Cryopyrin-Associated Periodic Syndromes (CAPS). The most common adverse events were injection site reactions and upper respiratory tract infections. There were no drug-related serious adverse events.

About RHAPSODY
RHAPSODY is the global, randomized withdrawal design, pivotal Phase 3 clinical trial of rilonacept 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 nonsteroidal anti-inflammatory drugs (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 rilonacept 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 rilonacept were randomized 1:1 and received 160 mg SC weekly rilonacept or placebo; and a long-term extension treatment period with up to 24 months of open-label rilonacept 160 mg SC weekly. For more information, refer to ClinicalTrials.gov Identifier: NCT03737110.

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. 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. 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 interleukin1 receptor antagonist (DIRA) is currently under FDA review following the submission of a Supplemental Biologics License Application (sBLA) in June 2020. Rilonacept in recurrent pericarditis is an investigational drug. The FDA granted Breakthrough Therapy designation to rilonacept for the treatment of recurrent pericarditis in 2019 and Orphan Drug designation to rilonacept for the treatment of 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.

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 across a spectrum of diseases. For more information, please visit www.kiniksa.com.

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 forwardlooking 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: the belief that the Phase 3 data suggest that targeted immunomodulation with rilonacept may signal a paradigm shift in the management of patients with recurrent pericarditis; and the 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: impact of additional data from us or other companies; potential undesirable side effects caused by rilonacept; our potential inability to demonstrate safety and efficacy of rilonacept in recurrent pericarditis to the satisfaction of the FDA or other applicable regulatory authorities; the potential for applicable regulatory authorities to not accept sBLA filings or to delay or deny approval of any of our product candidates; our potential inability to demonstrate that the clinical data integrity and quality of the biologic manufacturing processes and facilities are sufficient for the FDA to approve rilonacept in recurrent pericarditis; our reliance on third parties to manufacture our product candidates, including our reliance on Regeneron to manufacture the clinical and commercial supply of rilonacept; ; the potential impact of the COVID-19 pandemic and measures taken in response to the pandemic; and changes in our operating plan and funding requirements.

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 November 5, 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 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.

Every Second Counts!™

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

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