Kiniksa Highlights Phase 2 Study Data Showing the Corticosteroid-Sparing Effect of Rilonacept in Patients with Recurrent Pericarditis at the American College of Cardiology's 69th Annual Scientific Session

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- Phase 2 study data show the potential for rilonacept treatment to eliminate or reduce the risk of corticosteroid-associated morbidity in recurrent pericarditis -

- Enrollment complete for the pivotal Phase 3 trial of rilonacept in recurrent pericarditis (RHAPSODY); data expected in 2H 2020 -

HAMILTON, Bermuda, March 30, 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, recently highlighted a supplemental analysis from its Phase 2 clinical trial of rilonacept, an IL-1α and IL-1β cytokine trap, in a range of recurrent pericarditis populations. The analysis, which showed the corticosteroid-sparing effect of rilonacept treatment in the Phase 2 trial, was included in a virtual poster at the American College of Cardiology's (ACC) 69 th Annual Scientific Session.

"Novel therapies which could eliminate or reduce the risk of significant corticosteroid-associated morbidity in recurrent pericarditis are needed," said John F. Paolini, MD, PhD, FACC, Chief Medical Officer of Kiniksa. "The supplemental analysis presented virtually at ACC highlights the corticosteroid-sparing effect of rilonacept treatment in the study by enabling a reduction in corticosteroid dose or by obviating the need for corticosteroid use while on treatment."

Dr. Allan Klein, MD, of Cleveland Clinic and co-principal investigator for the trial, is the lead author of the virtual poster *Corticosteroid Tapering and Discontinuation in a Phase 2 Study of Rilonacept in Recurrent Pericarditis*. The materials are available through the Science section of Kiniksa's website (www.kiniksa.com).

Final data from the Phase 2 trial were presented at the American Heart Association (AHA) Scientific Sessions in November 2019. The data showed that rilonacept treatment in the study improved clinically meaningful outcomes associated with the unmet medical need in recurrent pericarditis, including resolution of pericarditis episodes, tapering and discontinuation of corticosteroids without pericarditis recurrence, reduction in recurrences of pericarditis episodes while on treatment and improved quality of life scores.

In the supplemental analysis highlighted at ACC, patients from the Phase 2 clinical trial were divided into groups based on their use of concomitant therapies at baseline:

- Corticosteroid-failure patients with active pericarditis experienced rapid, sustained and clinically meaningful reductions in pericarditis pain and C-reactive protein (CRP), a biomarker of inflammation, and tapered or discontinued corticosteroids without recurrence of disease while on rilonacept treatment.
- Corticosteroid-dependent patients tapered or discontinued corticosteroids without pericarditis recurrence while on rilonacept treatment.
- Colchicine-failure patients with active pericarditis experienced rapid, sustained and clinically meaningful reductions in pain and CRP while on rilonacept treatment.

Rilonacept was generally well-tolerated in the trial, with adverse events consistent with the U.S. Food and Drug Administration (FDA)-approved label for the treatment of Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Autoinflammatory Syndrome and Muckle-Wells Syndrome. The most common adverse events were mild, transient injection site reactions that did not cause discontinuation. There was one treatment-related serious adverse event which resulted in discontinuation: a skin abscess which responded to medical treatment. Infections are reported in the rilonacept label for CAPS.

Kiniksa has completed enrollment in RHAPSODY, a randomized withdrawal (RW) design, pivotal Phase 3 clinical trial. The primary efficacy endpoint is time-to-first pericarditis-recurrence in the RW period. Top-line data are expected in the second half of 2020.

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, KPL-716 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 signaling pathways that are implicated across a spectrum of diseases. For more information, please visit www.kiniksa.com.

About Rilonacept
Rilonacept is a weekly, subcutaneously-injected, recombinant fusion protein that blocks interleukin-1 alpha (IL-1α) and interleukin-1 beta (IL-1β) signaling. Rilonacept was discovered and developed by Regeneron Pharmaceuticals, Inc. (Regeneron) and is approved by the FDA under the brand name ARCALYST® for the treatment of CAPS, which includes Familial Cold Autoinflammatory Syndrome and Muckle-Wells Syndrome. Interleukin-1

(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 tumor necrosis factor (TNF) inhibitors is not recommended because this may increase the risk of serious infections. Kiniksa exclusively licensed rilonacept from Regeneron for recurrent pericarditis and certain other indications. Rilonacept in recurrent pericarditis is an investigational drug. The FDA has granted Breakthrough Therapy designation to rilonacept for recurrent pericarditis.

About RHAPSODY

RHAPSODY is the ongoing, pivotal Phase 3 clinical trial in recurrent pericarditis utilizing rilonacept. The company expects that at least 50 patients will be randomized into the RW period. Eligible patients must present 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 CRP value ≥ 1 mg/dL within the 7-day period prior to first study drug administration. Patients included in the study may be receiving concomitant nonsteroidal anti-inflammatory drugs (NSAIDs) and/or colchicine and/or oral corticosteroid treatment in any combination. The study is comprised of 5 periods: a screening period; a single-blind run-in period during which patients receive a loading dose of rilonacept 320 mg subcutaneous (SC) followed by 160 mg SC weekly while background pericarditis medications are tapered and discontinued; a double-blind, placebo-controlled 24-week RW period during which clinical responders to rilonacept are randomized 1:1 and receive 160 mg SC weekly rilonacept or placebo for at least 24 weeks; a long-term extension treatment period after trial completion during which all patients completing the RW period have the option to receive up to 24 weeks of open-label rilonacept 160 mg SC weekly; and a long-term extension follow-up period during which all patients in the long-term extension period will be followed for 24 weeks for safety and pericarditis recurrences.

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: the potential relevance of data presented at ACA from our Phase 2 clinical trial in recurrent pericarditis, including its potential support for discontinuation of corticosteroids and its potential impact on unmet medical need; statements regarding the objectives of the design of our Phase 3 clinical trial for rilonacept; and timing of data from the Phase 3 clinical trial.

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 or research disclosed by us or others with respect to recurrent pericarditis; our potential inability to replicate in later clinical trials, including our Phase 3 clinical trial, the positive final data from our Phase 2 and earlier clinical trials and other studies; potential coronavirus impact on study visits, data, protocol implementation, samples, and shipments of drug substance and product with respect to our global Phase 3 clinical trial; patients failing to complete the clinical trial; patients failing to experience pre-specified events during the clinical trial within an expected time-frame, if at all; potential complications in coordinating among requirements, regulations and guidelines of regulatory authorities across a number of jurisdictions for our global Phase 3 clinical trial; potential undesirable side effects caused by rilonacept; our potential inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities; our reliance on Regeneron to manufacture rilonacept; drug substance and/or drug product shortages; and our reliance on third parties to conduct research, clinical trials, and/or certain regulatory activities for rilonacept.

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 March 5, 2020 and our other reports 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.

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