Kiniksa Presents Rilonacept Interim Phase 2 Clinical Data Poster at the American College of Cardiology's 68th Annual Scientific Session

March 18, 2019

- Interim clinical data show reductions in both inflammation and reported pain throughout the treatment period
- Enrollment is ongoing in RHAPSODY, Kiniksa's pivotal Phase 3 trial of rilonacept in recurrent pericarditis

HAMILTON, Bermuda, March 18, 2019 (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, reported additional interim data from an open-label Phase 2 clinical trial of rilonacept, a weekly, subcutaneously-injected, recombinant fusion protein that blocks IL-1α and IL-1β signaling. The data were included in a poster presentation at the American College of Cardiology's (ACC) 68 th Annual Scientific Session in New Orleans, LA.

Dr. Allan Klein, MD, of Cleveland Clinic and co-principal investigator for the trial, presented *Rilonacept in Recurrent Pericarditis: First Efficacy and Safety Data from an Ongoing Phase 2 Pilot Clinical Trial*. The materials are available through the Investors and Media section of Kiniksa's website (www.kiniksa.com).

Kiniksa closed enrollment in the ongoing, open-label Phase 2 clinical trial in December 2018, and expects to complete the trial in the second half of 2019.

The Phase 2 trial, which evaluates the treatment response to rilonacept in different pericarditis populations, is divided into five parts:

- Part 1: Symptomatic subjects with recurrent pericarditis and C-reactive protein (CRP) > 1 mg/dL;
- Part 2: Symptomatic subjects with recurrent pericarditis and CRP ≤ 1 mg/dL but pericardial inflammation confirmed by magnetic resonance imaging (MRI);
- Part 3: Asymptomatic subjects with recurrent pericarditis who are dependent upon or unable to wean off of corticosteroids;
- Part 4: Symptomatic subjects with post-pericardiotomy syndrome (PPS) and CRP > 1 mg/dL; and
- Part 5: Asymptomatic subjects with PPS who are dependent upon or unable to wean off of corticosteroids.

The clinical trial started with Part 1 and was later expanded to include Parts 2 through 5. All subjects receive a loading dose of rilonacept 320 mg subcutaneously (SC), followed by 160 mg SC weekly maintenance on top of any combination of co-administered nonsteroidal anti-inflammatory drugs (NSAIDs) and/or colchicine and/or corticosteroids during a 6-week base treatment period. There is an optional 18-week extension period, during which subjects are allowed to wean off of concomitant NSAIDs, colchicine, and/or corticosteroids. The assessed efficacy outcomes measures include an 11-point pain Numerical Rating Scale (NRS), CRP, electrocardiogram (ECG) and size of pericardial effusion. The co-principal investigators are Dr. Allan Klein of Cleveland Clinic and Dr. David Lin of Minneapolis Heart Institute Foundation.

As of the January 23rd interim data cutoff date for the presentation at ACC, 25 subjects enrolled in the 6-week base treatment period across Parts 1 through 5 of the Phase 2 trial, 23 subjects continued into the optional 18-week extension period and 11 completed 24 weeks of treatment. Part 1 and Part 4, which enrolled actively symptomatic patients, are expected to be most relevant to the enrollment population for the ongoing Phase 3 clinical trial (RHAPSODY).

In Part 1 (symptomatic recurrent pericarditis; CRP > 1mg/dL), 12 subjects enrolled in the 6-week base treatment period. Results showed a reduction in both inflammation and reported pain after the first dose and a persistent clinical response throughout the 6-week base treatment period:

- mean patient-reported pericardial pain on an 11-point NRS decreased from 4.6 at baseline to 0.8 at 6 weeks;
- mean CRP decreased from 4.9 mg/dL at baseline to 0.3 mg/dL at 6 weeks; median time to CRP normalization was 9 days; and
- pericardial signs resolved, including pericardial effusion (6/7 subjects), PR depression (2/3 subjects), widespread ST elevation (2/2 subjects) and pericardial rub (2/2 subjects).

As of January 23rd, all 12 of the subjects enrolled in Part 1 completed the 6-week base treatment period, and 7 completed 24 weeks of treatment. These 7 subjects exhibited a continued clinical response to rilonacept as follows:

- mean patient-reported pericardial pain on an 11-point NRS further decreased to 0.4, and mean CRP was 0.3 mg/dL at 24 weeks;
- the pericardial effusion and PR depression in the remaining subjects resolved during the extension period; and
- of the 4 subjects on corticosteroids at baseline, the 2 subjects who had completed 24 weeks of treatment successfully tapered off corticosteroids.

The first subject who enrolled in Part 1 completed 24 weeks of treatment symptom-free and experienced recurrence of pericarditis symptoms that required the addition of NSAIDs approximately 8 weeks after completing rilonacept treatment. The patient subsequently experienced a pericarditis flare with tamponade physiology. The subject re-enrolled in the study as the last subject before enrollment closed and experienced a reduction in both inflammation and reported pain similar to her first rilonacept treatment course.

As of January 23rd, the 3 subjects enrolled in Part 2 (symptomatic recurrent pericarditis; CRP ≤ 1 mg/dL) and the 1 subject enrolled in Part 4 (symptomatic PPS) completed the 6-week base treatment period and showed reductions in both inflammation and reported pain. Additionally, 2 of the subjects enrolled in Part 2 completed 24 weeks of treatment and showed continued clinical response to rilonacept.

As of January 23rd, inflammation and reported pain remained low for the 6 subjects enrolled in Part 3 (asymptomatic steroid-dependent recurrent pericarditis) and the 3 subjects enrolled in Part 5 (asymptomatic steroid-dependent PPS) who completed the 6-week base treatment period. Additionally, 2 of the subjects enrolled in Part 3 completed 24 weeks of treatment. For these subjects, inflammation and reported pain continued to remain low while corticosteroids were tapered and discontinued.

Rilonacept has been generally well-tolerated in the study, with adverse events (AEs) 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 AEs were injection site reactions. There was one treatment-related serious AE which resulted in discontinuation: a skin abscess which responded to medical treatment. Infections are reported in the rilonacept label for CAPS.

“These results are encouraging,” said Dr. John F. Paolini, MD, PhD, FACC, Chief Medical Officer at Kiniksa. “Subjects across all cohorts of this trial experienced normalized CRP and pain levels while on rilonacept treatment. We believe this data could help support a potential treatment solution for patients suffering from recurrent pericarditis across a range of etiologies and we look forward to the data from RHAPSODY.”

Kiniksa is enrolling RHAPSODY, a double-blind, placebo-controlled, randomized withdrawal (RW) design, global Phase 3 clinical trial. The study is intended to evaluate the efficacy and safety of rilonacept treatment in subjects with recurrent pericarditis. The company expects that up to 50 subjects will be randomized into the RW period. Eligible subjects 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 NRS and a CRP value ≥ 1 mg/dL within the 7-day period prior to first study drug administration. Subjects included in the study may be receiving concomitant NSAIDs and/or colchicine and/or oral corticosteroid treatment in any combination. The primary efficacy endpoint is time-to-first-pericarditis-recurrence in the RW period. The Clinical Endpoint Committee will adjudicate all suspected pericarditis recurrences for inclusion in the primary efficacy endpoint analysis. The co-principal investigators are Dr. Allan Klein of Cleveland Clinic and Dr. Massimo Imazio of the University of Torino, Italy.

“Recurrent pericarditis is a clinical syndrome with unmet medical need, as patients have significant morbidity from their frequent flares of debilitating chest pain due to limited therapeutic options,” said Dr. Allan Klein, MD, of Cleveland Clinic. “The data from this ongoing rilonacept open-label Phase 2 pilot study are encouraging, and I look forward to continuing to investigate this medication in the Phase 3 RHAPSODY trial.”

About RHAPSODY
RHAPSODY is the ongoing, pivotal Phase 3 clinical trial in recurrent pericarditis utilizing rilonacept. The study is comprised of 5 periods: a screening period; a single-blind run-in period during which subjects receive a loading dose of rilonacept 320 mg injected 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 subjects 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 subjects in the long-term extension period will be followed for 24 weeks for safety and pericarditis recurrences.

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 Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Autoinflammatory Syndrome and Muckle-Wells Syndrome. 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. Kiniksa exclusively licensed rilonacept from Regeneron for recurrent pericarditis and certain other indications. Rilonacept in recurrent pericarditis is an investigational drug.

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 has a pipeline of product candidates across various stages of development, focused on autoinflammatory and autoimmune conditions. 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 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 for data from our Phase 2 clinical trial having the potential to help support rilonacept as a potential treatment solution for patients suffering from recurrent pericarditis; expected completion of the Phase 2 clinical trial; our conclusions from the Phase 2 interim clinical trial data; the potential relevance of certain patient populations from our Phase 2 clinical trial to those participating in our Phase 3 clinical trial; and statements regarding the size and objectives of the design of our Phase 3 clinical trial for rilonacept and potential data from the 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: changes between final data and any interim data we announce; our potential inability to replicate in later clinical trials,

including our Phase 3 clinical trial, the positive interim data from our Phase 2 and earlier clinical trials; delays or difficulty in activating sites or enrolling subjects in our global Phase 3 clinical trial; potential complications in coordinating among requirements, regulations and guidelines of regulatory authorities across a number of jurisdictions for our global Phase 3 clinical trial; impact of additional data from us or other companies; 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 for the period ended December 31, 2018, filed with the Securities and Exchange Commission (“SEC”) on March 12, 2019, 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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