Kiniksa Pharmaceuticals Logo

Kiniksa Announces Rilonacept Interim Phase 2 Clinical Data and Initiates Pivotal Phase 3 Clinical Trial

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- Phase 2 enrollment completed; interim clinical data show reductions in both inflammation and reported pain after first dose and throughout treatment period

- Pivotal Phase 3 clinical trial, RHAPSODY, now actively recruiting and screening subjects

HAMILTON, Bermuda, Dec. 11, 2018 (GLOBE NEWSWIRE) -- Kiniksa Pharmaceuticals, Ltd. (Nasdaq: KNSA) ("Kiniksa"), a biopharmaceutical company with a pipeline of five product candidates across various stages of development, today announced 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, in subjects with symptomatic recurrent pericarditis. The data show reductions in both inflammation and reported pain after the first dose which persisted throughout the treatment period. The company also reported that it has initiated RHAPSODY, a pivotal Phase 3 clinical trial in recurrent pericarditis, and is actively recruiting and screening subjects.

"We are focused on rapidly developing rilonacept in recurrent pericarditis as a potential treatment for this unmet medical need," said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. "Pericarditis is a debilitating disease where recurrence leads to higher risk of relapse and impacts quality of life. Rilonacept has the potential to be the first approved therapy for patients suffering from recurrent pericarditis."

Kiniksa recently completed enrollment in an open-label Phase 2 pilot study, which is evaluating the treatment response to rilonacept in subjects with both symptomatic recurrent pericarditis as well as other patient subsets within pericarditis, including asymptomatic steroid-dependent subjects with recurrent pericarditis and subjects with post-pericardiotomy syndrome. In this study, 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. An optional 18-week extension period follows during which weaning off of concomitant NSAIDs, colchicine and corticosteroids is allowed. The assessed efficacy outcomes measures include an 11-point pain Numerical Rating Scale (NRS), C-reactive protein (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 November 1st, 12 subjects, each with at least 3 episodes of pericarditis and elevated CRP (>1mg/dL), enrolled in a 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 0.9 at 6 weeks;
- mean CRP decreased from 4.9 mg/dL at baseline to 0.37 mg/dL at 6 weeks; median time to CRP normalization was 9 days; and
- pericardial signs resolved, including pericardial effusion (5/6 subjects), PR depression (3/4 subjects), widespread ST elevation (2/2 subjects), and pericardial rub (3/3 subjects).

As of November 1st, 10 of the 12 enrolled subjects received at least 6 weeks of treatment with rilonacept, 6 continued into the optional 18-week extension period, and 4 completed 24 weeks of treatment. These 10 subjects exhibited a continued clinical response to rilonacept as described below:

- mean patient-reported pericardial pain on an 11-point NRS further decreased to 0.3, and mean CRP was 0.44 mg/dL at 24 weeks;
- the pericardial effusion in the 1 remaining subject resolved during the extension period; and
- of the 4 subjects on corticosteroids at baseline, the 1 subject who had completed 24 weeks of treatment successfully tapered off corticosteroids.

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 gastrointestinal disorders and 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.

"We are pleased to announce the commencement of RHAPSODY, a pivotal Phase 3 clinical trial of rilonacept in recurrent pericarditis," said John F. Paolini, MD, PhD, FACC, Chief Medical Officer of Kiniksa. "RHAPSODY was supported by the interim data from our Phase 2 pilot study, which showed that rilonacept provided a reduction in both inflammation and reported pain as well as a durability of response in subjects with a symptomatic recurrent pericarditis episode. We believe we are now closer to providing a potential treatment option for patients suffering from recurrent pericarditis, a disease with significant unmet medical need."

Kiniksa has initiated RHAPSODY, a double-blind, placebo-controlled, randomized withdrawal (RW) study with an open-label extension period, and is actively recruiting and screening subjects. 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.

Conference Call Information
Kiniksa will host a conference call and webcast at 9:00 a.m. Eastern Time on Tuesday, December 11, 2018 to review the company's interim Phase 2 clinical data and its recently initiated pivotal Phase 3 clinical trial of rilonacept in subjects with recurrent pericarditis. Individuals interested in participating in the call should dial (866) 614-0636 (U.S. and Canada) or (409) 231-2053 (international) using conference ID number 9898327. To access the webcast and presentation slides, please visit the Investors and Media section of Kiniksa's website at www.kiniksa.com. The call will be available for replay via telephone starting at 12:00 p.m. Eastern Time on December 11, 2018 running through 12:00 p.m. Eastern Time on December 18, 2018. To listen to the replay, dial (855) 859-2056 (U.S. and Canada) or (404) 537-3406 (international) using conference ID number 9898327. The archived webcast will be available on Kiniksa's website for 14 days beginning approximately one hour after the call has completed.

About RHAPSODY
RHAPSODY is a 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), which includes 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 five 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 rilonacept to be the first approved therapy for patients suffering from recurrent pericarditis and our stage in the process for providing such an approved therapy; our conclusions from the Phase 2 interim clinical trial data; and statements regarding objectives of the design of our Phase 3 clinical trial for rilonacept.

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 the positive interim data from our Phase 2 and earlier clinical trials; delays or difficulty in 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 Quarterly Report on Form 10-Q for the period ended September 30, 2018 filed with the Securities and Exchange Commission ("SEC") on November 6, 2018 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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