Kiniksa Presents Rilonacept Final Phase 2 Clinical Data at the American Heart Association Scientific Sessions 2019

- Rapid resolution of both reported pain and inflammation as well as improvement in quality of life scores -

- Tapering and discontinuation of corticosteroids without pericarditis recurrence -

- Markedly lower annualized incidence of pericarditis episodes while on treatment -

- Pivotal Phase 3 trial of rilonacept in recurrent pericarditis (RHAPSODY) enrolling; data expected in 2H 2020 -

HAMILTON, BERMUDA – November 16, 2019 – 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 final 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 Heart Association (AHA) Scientific Sessions 2019.

"The final Phase 2 data showed the potential for rilonacept to improve clinically meaningful outcomes associated with unmet need in recurrent pericarditis," said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. "Rilonacept treatment in the Phase 2 trial led to a rapid resolution of recurrent pericarditis episodes that was sustained throughout the 6-month study as well as an improvement in overall quality of life scores. Treatment with rilonacept also led to a decrease in the annualized incidence of pericarditis episodes and importantly supported the discontinuation of corticosteroids without pericarditis recurrence. These data further confirm our confidence in the design of RHAPSODY, our pivotal Phase 3 trial of rilonacept in recurrent pericarditis, for which we expect top-line data in the second half of 2020."

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

The Phase 2 trial evaluated the treatment response to rilonacept in a range of pericarditis populations and was divided into five parts across two cohorts.

Symptomatic recurrent pericarditis patients:

- Part 1: Symptomatic patients with recurrent pericarditis and C-reactive protein (CRP) > 1 mg/dL;
- Part 2: Symptomatic patients with recurrent pericarditis and CRP ≤ 1 mg/dL but pericardial inflammation confirmed by magnetic resonance imaging (MRI); and
- Part 4: Symptomatic patients with post-pericardiotomy syndrome (PPS) and CRP > 1 mg/dL.

Corticosteroid-dependent recurrent pericarditis patients:

- Part 3: Asymptomatic patients with recurrent pericarditis who were dependent upon or unable to wean off corticosteroids; and
- Part 5: Asymptomatic patients with PPS who were dependent upon or unable to wean off corticosteroids.

In this study, all patients received 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 was an optional 18-week extension treatment period, during which physicians were given the option to wean patients off concomitant NSAIDs, colchicine, and/or corticosteroids. The assessed efficacy outcomes measures included an 11-point pain Numerical Rating Scale (NRS), CRP, electrocardiogram, and size of pericardial effusion. The co-principal investigators were Dr. Allan Klein of Cleveland Clinic and Dr. David Lin of Minneapolis Heart Institute Foundation.

25 unique patients enrolled in the 6-week base treatment period across Parts 1 through 5 of the Phase 2 trial, and 23 patients continued into the optional 18-week extension treatment period and completed 24 weeks of treatment.

The Phase 2 data provided first evidence that rilonacept treatment improved clinically meaningful outcomes associated with the unmet medical need in recurrent pericarditis.

Resolution of Pericarditis Episodes

Symptomatic recurrent pericarditis patients with CRP > 1mg/dL (Parts 1 and 4; n = 13), who were failing standard of care management with NSAIDs, colchicine, and/or corticosteroids, are

expected to be most relevant to the enrollment population for the ongoing Phase 3 clinical trial (RHAPSODY). There was a rapid reduction in both reported pain and inflammation after the first dose as well as a persistent and clinically meaningful response throughout the study for these patients.

- Mean patient-reported pericardial pain on an 11-point NRS decreased from 4.5 at baseline to 0.5 at 24 weeks;
- mean CRP decreased from 4.6 mg/dL at baseline to 0.2 mg/dL at 24 weeks; mean time to CRP normalization was 9 days; and
- pericardial signs resolved or improved in all patients, including pericardial effusion (6/7 patients), PR depression (2/3 patients), widespread ST elevation (2/2 patients) and pericardial rub (2/2 patients).

Symptomatic recurrent pericarditis patients with CRP ≤ 1mg/dL (Part 2; n = 3), who were failing standard of care management with NSAIDs, colchicine, and/or corticosteroids, showed a reduction in both reported pain and inflammation after the first dose as well as a persistent and clinically meaningful response throughout the study.

- Mean patient-reported pericardial pain on an 11-point NRS decreased from 4.7 at baseline to 0.0 at 24 weeks; and
- mean CRP decreased from 0.46 mg/dL at baseline to 0.32 mg/dL at 24 weeks.

Tapering and Discontinuation of Corticosteroids without Pericarditis Recurrence
15 recurrent pericarditis patients on corticosteroids at baseline enrolled in the 6-week base treatment period, and 13 continued into the optional 18-week extension treatment period and completed 24 weeks of treatment. During the optional 18-week extension treatment period, investigators were given the option to wean patients from concomitant medications while continuing weekly rilonacept treatment. All patients on corticosteroids stopped or tapered corticosteroids during this part of the study without experiencing a recurrent pericarditis episode.

- 11 recurrent pericarditis patients discontinued corticosteroids completely: 4 symptomatic patients across Parts 1 and 2 as well as 7 corticosteroid-dependent patients across Parts 3 and 5.
- The corticosteroid dose was successfully tapered in the 2 remaining recurrent pericarditis patients by the end of the 24-week study period: 1 symptomatic patient in Part 2 and 1 corticosteroid-dependent patient in Part 3.

Reduction in Recurrences of Pericarditis Episodes

A comparison of the annualized incidence of pericarditis episodes during the study while receiving rilonacept versus patients' own natural history in the period prior to the study demonstrated a decrease in annualized incidence of pericarditis episodes across all parts from 3.9 episodes/year prior to the study to <0.18 episodes/year.

Improved Quality of Life Scores
Rilonacept treatment resulted in improvement of Patient Reported Outcomes Measurement Information System (PROMIS) Global Health scores for Physical and Mental Global Health (U.S. mean score = 50.0; standard deviation = 10).

- In symptomatic recurrent pericarditis patients (Parts 1, 2, and 4), the mean Physical and Mental Global Health baseline scores were 39.9 and 44.5, respectively, and improved to 51.3 and 50.5, respectively, at 24 weeks.
- In corticosteroid-dependent recurrent pericarditis patients (Parts 3 and 5), the mean Physical and Mental Global Health baseline scores were 43.3 and 46.5, respectively, and improved to 46.8 and 50.7, respectively, at 24 weeks.

Safety
Rilonacept was generally 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), 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.

"Recurrent pericarditis is a debilitating autoinflammatory disease with a clear unmet need," said Dr. Allan Klein, MD, of Cleveland Clinic. "The final Phase 2 data show a sustained clinical response over 6 months of treatment, supporting tapering of corticosteroids while on treatment and affirming the efficacy of the interim analysis reported at the American College of Cardiology's (ACC) Annual Scientific Sessions in March 2018. I look forward to further investigation of rilonacept in the ongoing Phase 3 trial, RHAPSODY."

Kiniksa is enrolling RHAPSODY, a global, randomized withdrawal (RW) design, pivotal Phase 3 clinical trial in the U.S., Australia, Israel, and Italy. 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. Top-line data are expected in the second half of 2020.

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 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 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 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 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. The co-principal investigators are Dr. Allan Klein of Cleveland Clinic and Dr. Massimo Imazio of the University of Torino, Italy.

About Rilonacept

Rilonacept is a weekly, subcutaneously-injected, recombinant fusion protein that blocks inflammatory cytokines interleukin-1α (IL-1α) and interleukin 1β (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 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 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 relevance of final data presented at AHA from our Phase 2 clinical trial in recurrent pericarditis, including its potential support for discontinuation of corticosteroids and our Phase 3 clinical trial design as well as its potential impact on unmet medical need; statements regarding the expected number of patients randomized in the RW period and objectives of the design of our Phase 3 clinical trial for rilonacept; and timing of potential 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: changes between final data from our Phase 2 clinical trial presented at AHA or otherwise and any 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; delays or difficulty in activating sites or enrolling patients in 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; 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 filed with the Securities and Exchange Commission ("SEC") on November 5, 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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