# Kiniksa Announces Upcoming Rilonacept Analyst Day

September 21, 2020

**- Virtual event scheduled for Monday, September 28th, 2020 from 8:00 a.m. to 9:30 a.m. EDT-**

HAMILTON, Bermuda, Sept. 21, 2020 (GLOBE NEWSWIRE) -- Kiniksa Pharmaceuticals, Ltd. (Nasdaq: KNSA) ("Kiniksa"), a biopharmaceutical company with a pipeline of clinical-stage assets designed to modulate immunological pathways that are implicated across a spectrum of diseases, today announced that the company will host a virtual Rilonacept Analyst Day on Monday, September 28th, 2020 from 8:00 a.m. to 9:30 a.m. Eastern Daylight Time.

The event will feature presentations from the Kiniksa management team on the market opportunity for rilonacept in recurrent pericarditis as well as the company's continued commercial preparations and launch strategy. Additionally, guest speaker Paul Cremer, MD, Cardiovascular Medicine, Cleveland Clinic, will review the burden of recurrent pericarditis, the current treatment landscape, and the unmet need.

Rilonacept was discovered and developed by Regeneron Pharmaceuticals, Inc. (Regeneron) and is approved by the U.S. Food and Drug Administration (FDA) under the brand name ARCALYST® for the treatment of Cryopyrin-Associated Periodic Syndromes (CAPS). Kiniksa licensed rilonacept from Regeneron in 2017 for evaluation in diseases believed to be mediated by both interleukin-1 alpha (IL-1α) and interleukin-1 beta (IL-1β), including recurrent pericarditis. The FDA granted Breakthrough Therapy designation to rilonacept for recurrent pericarditis in 2019 and Orphan Drug designation to rilonacept for pericarditis in 2020. Based on highly statistically significant Phase 3 data in recurrent pericarditis, the Biologic License Application (BLA) for CAPS will transfer to Kiniksa, and the company plans to submit a supplemental Biologic License Application (sBLA) in recurrent pericarditis to the FDA this year. Upon receipt of FDA approval for rilonacept in recurrent pericarditis, Kiniksa will assume the sales and distribution of rilonacept for the approved indications in the United States and evenly split profits on sales with Regeneron.

**Webcast and Conference Call Information**

Kiniksa will host a webcast and conference call at 8:00 a.m. Eastern Daylight Time on Monday, September 28th, 2020. The presentation will be accessible through this link as well as through the Investors & Media section of the company's website. Individuals can also participate by dialing (866) 614-0636 (U.S. and Canada) or (409) 231-2053 (international) using conference ID number 3890078. The archived webcast will be available on Kiniksa's website for 14 days beginning approximately one hour after the conclusion of the event.

**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 the Rilonacept License Agreement with Regeneron**

In 2017, Regeneron granted Kiniksa an exclusive license to develop and commercialize rilonacept worldwide, aside from Israel, Egypt, Turkey and select countries in the Middle East and North Africa. In the United States and Japan, Kiniksa's license is initially for all indications other than those involving local administration to the eye or ear, oncology, deficiency of the interleukin1 receptor antagonist (DIRA) and CAPS. If Kiniksa is successful in receiving marketing approval for rilonacept in the United States for a new indication, the scope of the license granted to Kiniksa will automatically expand to include DIRA and CAPS in the United States and Japan, and Kiniksa will assume the sales and distribution of rilonacept in these additional indications. Outside the United States and Japan, Kiniksa's license is for all indications other than local application to the eye or ear, oncology, CAPS, DIRA and certain periodic fever syndromes. Kiniksa made an upfront payment of $5.0 million to Regeneron and is obligated to make regulatory milestone payments of up to $27.5 million in the aggregate. Thereafter, Kiniksa and Regeneron will evenly split profits on sales of rilonacept after deducting certain commercialization expenses subject to specified limits.

**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 DIRA is currently pending FDA approval following the submission of an sBLA in June 2020. Rilonacept in recurrent pericarditis is an investigational drug. The FDA granted Breakthrough Therapy designation to rilonacept for recurrent pericarditis in 2019 and Orphan Drug designation to rilonacept for 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 that are implicated 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: our expectation with respect to the BLA for CAPs transferring to Kiniksa; our timing for submitting an sBLA to the FDA; and the potential for all of our clinical stage product candidates to offer 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: the potential impact of the COVID-19 pandemic and measures taken in response to the pandemic; changes in our operating plan and funding requirements; existing or new competition; and our ability to attract and retain qualified personnel.

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 August 4, 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.



***Every Second Counts!™***

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