Kiniksa Provides Timing of Second Half 2020 Clinical Data Readouts

June 10, 2020

- Rilonacept pivotal Phase 3 data in recurrent pericarditis expected in Q3 2020 -

- Mavrilimumab Phase 2 data in GCA and KPL-404 Phase 1 data expected in Q4 2020 -

- Management to provide a corporate update at the 41st Annual Goldman Sachs Global Healthcare Conference on Thursday, June 11th at 3pm EDT -

HAMILTON, Bermuda, June 10, 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, today announced that data from RHAPSODY, the pivotal Phase 3 trial of rilonacept (IL-1α and IL-1β trap) in recurrent pericarditis, are expected in the third quarter of 2020. Additionally, the company expects mavrilimumab (anti-GM-CSFRα) Phase 2 data in giant cell arteritis (GCA) and KPL-404 (anti-CD40) single-ascending-dose Phase 1 data in the fourth quarter of 2020.

"Focused corporate and clinical execution year-to-date has led to significant advancements across our pipeline," said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. "We plan to build upon this progress throughout the second half of the year. Following the receipt of Breakthrough Therapy designation and a Type B meeting with the U.S. FDA, we were able to pull forward the timeline for pivotal Phase 3 rilonacept data into the third quarter. We also anticipate Phase 2 data for mavrilimumab and Phase 1 data for KPL-404 in the fourth quarter. Additionally, we plan to initiate a Phase 2/3 clinical trial for mavrilimumab in severe COVID-19 pneumonia and hyperinflammation and a Phase 2b dose-ranging trial for vixarelimab in prurigo nodularis."

"We believe there is urgent need to bring an approved therapy to patients suffering from recurrent pericarditis," said Qasim Rizvi, Chief Commercial Officer and SVP of Operations at Kiniksa. "We are focused on the key clinical outcomes measures in recurrent pericarditis that matter the most to patients and physicians: resolution of acute episodes, prevention of future recurrences while on treatment, tapering and discontinuation of corticosteroids, and improvement in quality of life. RHAPSODY is a pivotal Phase 3 clinical trial that has the potential to generate meaningful data to address these unmet clinical needs."

A live webcast of Kiniksa's presentation at the 41 st Annual Goldman Sachs Global Healthcare Conference on Thursday, June 11th will be accessible through the Investors & Media section of the company's website (www.investors.kiniksa.com). A replay of the webcast will be available on Kiniksa's website for 14 days following the conference.

About RHAPSODY
RHAPSODY is the ongoing global (U.S., Australia, Israel, and Italy), randomized withdrawal (RW) design, pivotal Phase 3 clinical trial of rilonacept in recurrent pericarditis. 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 third quarter of 2020. The co-principal investigators are Dr. Allan Klein of Cleveland Clinic and Dr. Massimo Imazio of the University of Torino, Italy. For more information, refer to ClinicalTrials.gov Identifier: NCT03737110.

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 Cryopyrin-Associated Periodic Syndromes (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 Mavrilimumab
Mavrilimumab is an investigational fully-human monoclonal antibody that targets granulocyte macrophage colony stimulating factor receptor alpha (GM-CSFRα). Kiniksa's lead indication for mavrilimumab is giant cell arteritis (GCA), an inflammatory disease of medium-to-large arteries. Mavrilimumab was dosed in over 550 patients with rheumatoid arthritis through Phase 2b clinical studies in Europe and achieved prospectively-defined primary endpoints of efficacy and safety. Additionally, Kiniksa and Kite, a Gilead company, have a clinical collaboration to evaluate mavrilimumab in combination with Yescarta® (axicabtagene ciloleucel) in patients with relapsed or refractory large B-cell lymphoma. Kiniksa also has an active investigational new drug (IND) application with the U.S. Food and Drug Administration (FDA) for a Phase 2/3 clinical trial evaluating mavrilimumab in severe COVID-19 pneumonia and hyperinflammation.

About Vixarelimab
Vixarelimab is an investigational fully-human monoclonal antibody that targets oncostatin M receptor beta (OSMRβ), which mediates signaling of interleukin-31 (IL-31) and oncostatin M (OSM), two key cytokines implicated in pruritus, inflammation and fibrosis. Kiniksa believes vixarelimab to be the only monoclonal antibody in development that targets both pathways simultaneously.

About KPL-404
KPL-404 is an investigational humanized monoclonal antibody that is designed to inhibit CD40-CD40 ligand (CD40L) interaction, a key T-cell co-stimulatory signal critical for B-cell maturation and immunoglobulin class switching. Kiniksa believes disrupting CD40-CD40L interaction is an attractive approach for blocking T-cell mediated, B-cell driven responses, drivers of multiple autoimmune disease pathologies such as rheumatoid arthritis, Sjogren's syndrome, Graves' disease, systemic lupus erythematosus and solid organ transplant.

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 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: our expectation of being able to build on the progress made in advancing our pipeline in the first half of 2020 during the second half of 2020; the timing of data from our clinical trials in 2020; the potential relevance of the data from our pivotal Phase 3 clinical trial of rilonacept in recurrent pericarditis, Phase 2 clinical trial of mavrilimumab in giant cell arteritis, and our Phase 1 clinical trial of KPL-404 in healthy volunteers; the timing of initiating our planned or potential clinical trials; the urgency in bringing an approved therapy to patients suffering from recurrent pericarditis; our clinical collaboration with Kite in CAR T; 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: our potential inability to replicate in later clinical trials the positive final data from our earlier clinical trials or investigator-initiated protocols or studies; impact of additional data from us or other companies; potential undesirable side effects caused by our product candidates; our potential inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities; our reliance on third parties to manufacture our product candidates; drug substance and/or drug product shortages; and our reliance on third parties to conduct research, clinical trials, and/or certain regulatory activities for our product candidates; delays or difficulty in activating sites or enrolling patients in our planned clinical trials; potential complications in coordinating among requirements, regulations and guidelines of regulatory authorities across a number of jurisdictions for our planned global clinical trials; 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 May 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.

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