



Kiniksa Announces Pipeline Progress and Reiterates 2020 Clinical Data Readouts

January 13, 2020

- Enrollment target achieved for rilonacept Phase 3 trial in recurrent pericarditis; top-line data expected in 2H 2020 -
- Enrollment target achieved for mavrimumab Phase 2 trial in giant cell arteritis; top-line data expected in 2H 2020 -
 - Top-line data from KPL-716 Phase 2a trial in prurigo nodularis expected in 1H 2020 -
- Interim data from cohorts of KPL-716 Phase 2 trial in diseases characterized by chronic pruritus expected in 1H 2020 -
 - Top-line data from KPL-404 first-in-human study with antigen challenge TDAR expected in 2H 2020 -

HAMILTON, Bermuda, Jan. 13, 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 recent pipeline progress and upcoming 2020 clinical milestones. Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa will provide further detail in a corporate presentation at the 38th Annual J.P. Morgan Healthcare Conference today, Monday, January 13, 2020 at 2:00 p.m. Pacific Time / 5:00 p.m. Eastern Time at the Westin St. Francis Hotel in San Francisco, California.

"Kiniksa's clinical-stage assets, including rilonacept, mavrimumab, KPL-716 and KPL-404, are on track to generate clinical data this year," said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. "Each readout has the potential to be an important value-driving event and to help inform our portfolio strategy and capital allocation decisions. We have the capital to achieve multiple clinical data readouts and project that our 2019 year-end cash reserves of approximately \$233 million will fund our operating plan into the second half of 2021."

Recent Pipeline Progress and Expected 2020 Clinical Data Readouts

Rilonacept (IL-1 α and IL-1 β cytokine trap)

- Kiniksa has achieved its enrollment target for RHAPSODY, a global, randomized-withdrawal design, pivotal Phase 3 trial of rilonacept in patients with recurrent pericarditis. The company expects top-line data in the second half of 2020.
- Kiniksa recently announced the U.S. Food and Drug Administration (FDA) granted Breakthrough Therapy designation for rilonacept for the treatment of recurrent pericarditis. Kiniksa's Breakthrough Therapy application was based on final data from an open-label Phase 2 clinical trial of rilonacept in a range of recurrent pericarditis populations.

Mavrimumab (monoclonal antibody inhibitor targeting GM-CSFR α)

- Kiniksa has achieved its enrollment target for a global Phase 2 proof-of-concept trial of mavrimumab in patients with giant cell arteritis (GCA). The company expects top-line data in the second half of 2020.
- Kiniksa and Kite, a Gilead company (Kite), recently announced a clinical collaboration evaluating the investigational combination of Yescarta[®] (axicabtagene ciloleucel) and mavrimumab in relapsed or refractory large B-cell lymphoma. The objective of the study is to determine the effect of mavrimumab on the safety of Yescarta. Preclinical evidence shows the potential for interruption of granulocyte macrophage colony stimulating factor (GM-CSF) signaling to disrupt chimeric antigen receptor T (CAR T) cell-mediated inflammation without disrupting anti-tumor efficacy.

KPL-716 (monoclonal antibody inhibitor of signaling through OSMR β)

- Kiniksa expects top-line data from a Phase 2a clinical trial of KPL-716 in patients with prurigo nodularis in the first half of 2020.
- Kiniksa expects interim data from cohorts of a Phase 2 clinical trial of KPL-716 in diseases characterized by chronic pruritus in the first half of 2020.

KPL-404 (monoclonal antibody inhibitor of signaling between CD40 and CD40L)

- Kiniksa is enrolling and dosing subjects in a single-ascending-dose Phase 1 clinical trial of KPL-404 in healthy volunteers. The first-in-human trial will provide safety data and pharmacokinetics as well as receptor occupancy and T-cell Dependent Antibody Response (TDAR). Top-line data are expected in the second half of 2020.

KPL-045 (monoclonal antibody inhibitor of the CD30L co-stimulatory molecule)

- Based on preclinical data in the context of Kiniksa's portfolio, the company no longer plans to progress KPL-045 into clinical development.

Financial Guidance

Kiniksa ended 2019 with approximately \$233 million in cash, cash equivalents and short-term investments (unaudited). The company expects that these reserves will fund its operating plan into the second half of 2021.

Presentation at the 38th Annual J.P. Morgan Healthcare Conference

Kiniksa will webcast its corporate presentation at the 38th Annual J.P. Morgan Healthcare Conference today, Monday, January 13, 2020 at 2:00 p.m. Pacific Time / 5:00 p.m. Eastern Time. A live webcast of Kiniksa's presentation will be accessible through the Investors & Media section of the company's website (www.kiniksa.com). A replay of the webcast will be available on Kiniksa's website for 14 days following the conference.

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.

About Rilonacept

Rilonacept is a weekly, subcutaneously-injected, recombinant fusion protein that blocks interleukin-1 alpha (IL-1 α) and interleukin-1 beta (IL-1 β) 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. The FDA has granted Breakthrough Therapy designation to rilonacept for recurrent pericarditis.

About Mavrilimumab

Mavrilimumab is an investigational fully-human monoclonal antibody that is designed to antagonize GM-CSF signaling by binding to the alpha subunit of the GM-CSF receptor. Kiniksa's lead indication for mavrilimumab is GCA, an inflammatory disease of medium to large arteries. Additionally, Kiniksa and Kite have a clinical collaboration to evaluate mavrilimumab in combination with Yescarta[®] (axicabtagene ciloleucel) in patients with relapsed or refractory large B-cell lymphoma.

About KPL-716

KPL-716 is an investigational fully-human monoclonal antibody that targets oncostatin M receptor beta (OSMR β), which mediates signaling of interleukin-31 (IL-31) and oncostatin M (OSM), two key cytokines implicated in pruritus, inflammation and fibrosis. Kiniksa believes KPL-716 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 Sjogren's syndrome, systemic lupus erythematosus, rheumatoid arthritis, solid organ transplant and Graves' disease.

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 expectations for fiscal year 2020; plans and timing of enrollment of our clinical trials; proposed indications for the investigation of our product candidates; our beliefs about the approach of our product candidates and potential impact; our clinical collaboration with Kite evaluating the combination of Yescarta[®] and mavrilimumab; the potential for mavrilimumab with respect to CAR T cell mediated inflammation and otherwise; plans and timing to report or present preliminary, interim and final top-line clinical trial data and the potential impact of that data, including on our portfolio strategy and capital allocation; expected cash, cash equivalents and short-term investments at fiscal year-end 2019; projected timeframe for funding our operating plan with current cash, cash equivalents and short-term investments; and our expectations around not needing to raise additional capital prior to delivering our anticipated 2020 clinical trial data readouts.

These forward-looking statements are based on management's current plans, estimates or 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: potential delays or difficulty in enrollment of patients in, and activation of sites for, our clinical trials; potential complications in coordinating among requirements, regulations and guidelines of regulatory authorities across a number of jurisdictions for our global clinical trials; potential amendments to our clinical trial protocols initiated by us or required by regulatory authorities; potential delays or difficulty in completing our clinical trials, including as a result of our clinical trial design; potential for lower accrual of events in our clinical trials; potential undesirable side effects caused by our product candidates; our potential inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities or otherwise producing negative, inconclusive or commercially uncompetitive results; potential for changes between final data and any preliminary and interim "top-line" data we announce; impact of additional data from us or other companies; our potential inability to replicate in later clinical trials positive results from our earlier pre-clinical and clinical trials; drug substance and/or drug product shortages caused by issues at our third-party manufacturers' facilities; our reliance on certain third parties as the sole source of supply of the drug substance and drug products used in our product candidates; our reliance on third parties to conduct our research, pre-clinical studies, clinical trials, and other trials for our product candidates; changes in our operating plan and funding requirements; changes in the capital markets; market reaction to our anticipated 2020 clinical trial data readouts; substantial 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 November 5, 2019 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 plans, estimates, or expectations 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.

ARCALYST® is a registered trademark of Regeneron Pharmaceuticals, Inc. and Yescarta® is a registered trademark of Gilead Sciences, Inc., or its related companies.

Every Second Counts!™

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Source: Kiniksa Pharmaceuticals, Ltd.