Kiniksa Announces American College of Rheumatology Convergence 2020 Late-Breaking Abstracts Presentation of Mavrilimumab Phase 2 Giant Cell Arteritis Data

November 9, 2020

Primary and secondary efficacy endpoints achieved statistical significance; mavrilimumab reduced risk of flare and increased sustained remission

HAMILTON, Bermuda, Nov. 09, 2020 (GLOBE NEWSWIRE) -- Kiniksa Pharmaceuticals, Ltd. (Nasdaq: KNSA) ("Kiniksa"), a biopharmaceutical company with a pipeline of assets designed to modulate immunological pathways across a spectrum of diseases, today announced that data from the global Phase 2 clinical trial of mavrilimumab in giant cell arteritis (GCA) were presented at the late-breaking abstracts session during the American College of Rheumatology (ACR) Convergence 2020. Mavrilimumab is an investigational fully-human monoclonal antibody that targets granulocyte macrophage colony stimulating factor receptor alpha (GM-CSFRα). Both the primary and secondary efficacy endpoints achieved statistical significance, and there was a consistent trend of efficacy across the new onset and relapsing/refractory cohorts.

"There is a real need for novel therapies for patients with giant cell arteritis," said Dr. Maria Cid, Hospital Clínic, University of Barcelona, IDIBAPS. "My fellow co-principal investigator, Dr. Sebastian Unizony, and I presented data which show that mavrilimumab significantly reduced risk of flare and increased sustained remission compared to placebo. This is an encouraging potential therapeutic advancement for these patients, particularly for those with relapsed/refractory disease, as many are not able to achieve sustained remission on current standard of care. I look forward to further investigation of mavrilimumab in subsequent clinical trials."

Dr. Maria Cid[1], a co-principal investigator for the global Phase 2 trial, delivered a virtual presentation entitled *Mavrilimumab (Anti GM-CSF Receptor α Monoclonal Antibody) Reduces Risk of Flare and Increases Sustained Remission in a Phase 2 Trial of Patients with Giant Cell Arteritis* at the late-breaking abstracts session (L06 - L11) on Monday, November 9, 2020 at 11:30 a.m. Eastern Time. Dr. Sebastian Unizony[2] is a co-principal investigator.

The Phase 2 trial randomized 70 patients 3:2 to mavrilimumab 150 mg (n=42) or placebo (n=28) biweekly injected subcutaneously, co-administered with a protocol-defined 26-week oral corticosteroid taper. Patients were stratified by new onset (n=35) or relapsing/refractory (n=35) disease.

Both the primary efficacy endpoint of time-to-first adjudicated GCA flare by Week 26 in all treated patients (Hazard Ratio = 0.38, p=0.0263) and the secondary efficacy endpoint of sustained remission at Week 26 in all treated patients (33.3 percentage point increase; p=0.0038) were statistically significant. Additionally, while the trial was not powered for individual disease cohorts, there was a consistent trend of efficacy across the new onset and relapsing/refractory cohorts.

Mavrilimumab was well-tolerated; there were no drug-related serious adverse events, and the rates of drug-related treatment-emergent adverse events between mavrilimumab recipients and placebo recipients were similar.

The 12-week washout safety follow-up period is ongoing, and additional analyses of this Phase 2 trial are planned. Next steps for the development program in GCA will be further informed by anticipated discussions with the U.S. Food and Drug Administration (FDA).

The FDA recently granted Orphan Drug designation to mavrilimumab for the treatment of GCA.

Kiniksa is also evaluating mavrilimumab in severe COVID-19 pneumonia and hyperinflammation and is enrolling the Phase 2 portion of a global, randomized, double-blind, placebo-controlled adaptive design Phase 2/3 clinical trial. Additionally, data are expected from a randomized, double-blind, placebo-controlled investigator-initiated study in the U.S. in the fourth quarter of 2020.

[1] Hospital Clínic, University of Barcelona, Institut d'Investigacions Biomèdiques August Pi i Sunyer (IDIBAPS); [2] Massachusetts General Hospital, Harvard University

About the Global Phase 2 Clinical Trial of Mavrilimumab in GCA
The randomized, double-blind, placebo-controlled, global Phase 2 clinical trial of mavrilimumab in GCA consists of a 6-week screening period, a 26-week double-blind placebo-controlled treatment period, and a 12-week washout safety follow-up period. Patients age 50 to 85 years with active GCA, confirmed by temporal artery biopsy and/or imaging, with erythrocyte sedimentation rate (ESR) ≥ 30 mm/hour or C-reactive protein (CRP) ≥ 1 mg/dL, and symptoms of GCA within 6 weeks from randomization, were included. All patients were required to have achieved corticosteroid-induced remission (resolution of symptoms, ESR < 20 mm/hour, CRP < 1 mg/dL) prior to randomization.

About Giant Cell Arteritis
Giant cell arteritis is a rare chronic inflammatory disease of medium-to-large arteries. Cranial giant cell arteritis typically presents with headache and jaw claudication as well as constitutional symptoms of fever and fatigue. Acute events can include permanent vision loss from diminished blood flow to the eye. The large vessel form of giant cell arteritis affects the branches of the aorta supplying the trunk and limbs. There is currently one FDA-approved treatment for giant cell arteritis as an adjunct to a corticosteroid taper.

About Mavrilimumab
Mavrilimumab is an investigational fully-human monoclonal antibody that targets GM-CSFRα. 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. Kiniksa's lead indication for mavrilimumab is GCA, a rare inflammatory disease of medium-to-large arteries. Kiniksa is also evaluating mavrilimumab in COVID-19 pneumonia and hyperinflammation. The FDA granted Orphan Drug designation to mavrilimumab for the treatment of GCA in 2020.

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 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 across a spectrum of diseases. For more information, please visit www.kiniksa.com.

Forward-Looking Statements
The information contained in 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 need for novel therapies for patients with giant cell arteritis ("GCA"); mavrilimumab's potential to offer a treatment option for patients with GCA, particularly patients with relapsing/refractory disease; the unmet need for patients with GCA; continued analyses of the on-going washout period Phase 2 trial data; next steps for the development program in GCA being further informed by anticipated discussions with the FDA; the timing of data from our clinical trials or investigator initiated studies; and the potential for 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 impact of additional data from us, investigator-initiated studies or other companies; the potential for undesirable side effects to be caused by mavrilimumab; the potential inability to replicate in later studies or clinical trials positive results from earlier studies or clinical trials; the impact of discussions with the FDA on our development program in GCA; our reliance on third parties to manufacture our product candidates and conduct our clinical trials and/or perform certain regulatory activities for our product candidates; drug substance and/or drug product shortages, including in connection with our engagement of a manufacturing organization to produce mavrilimumab beyond our current inventory; 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 November 5, 2020 and our other reports subsequently filed with or furnished to 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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