Kiniksa Pharmaceuticals Logo

Kiniksa Presents Preclinical Data on the GM-CSF Signaling Pathway in Temporal Arteries of GCA Patients at the Annual European Congress of Rheumatology

June 12, 2019

- Preclinical data support rationale for targeting GM-CSF in GCA –

- Mavrilimumab shows biological effect on genes relevant to GCA pathophysiology in ex vivo cultures of GCA arteries –

HAMILTON, Bermuda, June 12, 2019 (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 it presented preclinical data on the granulocyte macrophage colony stimulating factor (GM-CSF) signaling pathways in temporal arteries of patients with giant cell arteritis (GCA) at the European League Against Rheumatism's (EULAR) European Congress of Rheumatology in Madrid, Spain.

"The preclinical data presented at EULAR underscore the mechanistic rationale of targeting GM-CSFRα in patients with GCA," said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. "GM-CSF, GM-CSFRα, and their associated signaling molecules were shown to be significantly upregulated in temporal arteries of GCA patients. Furthermore, mavrilimumab was shown to suppress the increased expression of genes associated with this inflammatory pathway. We are continuing to advance our global Phase 2 clinical trial of mavrilimumab in GCA and look forward to presenting top-line data in the second half of 2020."

Dr. Maria C. Cid[1] is a lead author of the poster presentation entitled ***GM-CSF Pathway Signature Identified in Temporal Artery Biopsies of Patients with GCA***.

- Data from this preclinical study comparing two independent sources of temporal artery biopsies showed the GM-CSF signaling pathway molecular signature was confirmed to be upregulated in GCA biopsies versus control at both the mRNA and protein level. GM-CSF and T_H1 pathway signatures were demonstrated in GCA patient temporal arteries by independent analytical techniques. The data also demonstrated active GM-CSF signaling in diseased tissue is evidenced by increased expression of PU.1, a transcription factor downstream of GM-CSF signaling, in the vessel wall. Additionally, treatment of *ex vivo* cultures of GCA arteries with mavrilimumab suppressed expression of these genes, indicating the biological effect of mavrilimumab on genes relevant to GCA pathophysiology.

The materials are available through the Investors and Media section of Kiniksa's website (www.kiniksa.com).

Kiniksa is developing mavrilimumab, 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, for the potential treatment of GCA.

GCA is a chronic inflammatory disease of medium-large blood vessels that causes headaches, jaw and other muscle claudication as well as ischemic visual loss. Kiniksa estimates U.S. prevalence of approximately 75,000 to 150,000 patients with similar prevalence rates for other major markets.

Kiniksa is enrolling a randomized, double-blind, placebo-controlled, global Phase 2 proof-of-concept clinical trial of mavrilimumab in subjects with GCA. The trial is expected to enroll approximately 60 subjects with new-onset and refractory disease. Subjects will be randomized 3:2 to mavrilimumab 150 mg or placebo injected subcutaneously once every 2 weeks co-administered with a corticosteroid taper. The primary efficacy endpoint involves measuring GCA flares during the 26-week treatment period. Top-line data are expected in the second half of 2020.

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 giant cell arteritis, an inflammatory disease of blood vessels.

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.

[1] Hospital Clínic, University of Barcelona, Institut d'Investigacions Biomèdiques August Pi i Sunyer (IDIBAPS), Vasculitis Research Unit, Department of Autoimmune Diseases, Barcelona, Spain

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 development of mavrilimumab for the potential treatment of GCA; our conclusions from pre-clinical data for mavrilimumab; our global Phase 2 clinical trial of mavrilimumab in GCA; and our expected timing of topline data from our Phase 2 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 and any interim data we present; our potential inability to replicate in later clinical trials, including our Phase 2 clinical trial of mavrilimumab in GCA, the positive interim data from our pre-clinical and earlier clinical trials; potential impact of additional data from us or other companies; potential undesirable side effects caused by

mavrilimumab; our potential inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities; and our reliance on third parties to manufacture mavrilimumab and to conduct research, clinical trials and/or certain regulatory activities for mavrilimumab.

These and other important factors discussed under the caption “Risk Factors” in our Quarterly Report on Form 10-Q for the period ended March 31, 2019, filed with the Securities and Exchange Commission (“SEC”) on May 7, 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.

Every Second Counts!™

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