Kiniksa Announces U.S. Orphan Drug Designation for Mavrilimumab for the Treatment of Giant Cell Arteritis

September 15, 2020

Management to present at the Morgan Stanley 18th Annual Global Healthcare Conference on Wednesday, September 16th at 2pm EDT

HAMILTON, Bermuda, Sept. 15, 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 the U.S. Food and Drug Administration (FDA) granted Orphan Drug designation to mavrilimumab for the treatment of giant cell arteritis (GCA). Mavrilimumab is a monoclonal antibody inhibitor targeting granulocyte macrophage colony stimulating factor receptor alpha (GM-CSFRα).

"We are pleased to announce that the FDA granted Orphan Drug designation to mavrilimumab for the treatment of GCA," said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. "Data from our Phase 2 study of mavrilimumab in GCA are expected in the fourth quarter of this year."

There will be a live webcast of Kiniksa's presentation at the Morgan Stanley 18th Annual Global Healthcare Conference on Wednesday, September 16th at 2:00 p.m. Eastern Daylight Time. The company will provide an update on key activities relating to rilonacept, mavrilimumab, vixarelimab and KPL-404. The presentation 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 Orphan Drug Designation
The FDA grants Orphan Drug designation status to products that treat rare diseases, providing incentives to sponsors developing drugs or biologics. The FDA defines rare diseases as those affecting fewer than 200,000 people in the U.S. at the time of designation. Orphan drug designation entitles a party to financial incentives such as opportunities for grant funding towards clinical trial costs, tax advantages and user-fee waivers. If a product that has orphan designation subsequently receives the first FDA approval for the disease or condition for which it has such designation, the product is entitled to orphan drug exclusivity, which means that the FDA may not approve any other applications to market the same drug for the same indication for seven years from the date of such approval, except in limited circumstances. Additionally, orphan drug designation waives the requirement to conduct pediatric studies for the product in the disease it is designated.

About Giant Cell Arteritis
Giant cell arteritis is a 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 steroid taper.

About Mavrilimumab
Mavrilimumab is an investigational fully-human monoclonal antibody that targets granulocyte macrophage colony stimulating factor receptor alpha (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 giant cell arteritis (GCA), an 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 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 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 belief that mavrilimumab targets the cytokine that may be responsible for the critical pathological steps of GCA and that the rare disease has significant unmet need; timing of data from our Phase 2 study of mavrilimumab in GCA; and our belief that our product candidates all offer the potential for 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; 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 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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