Kiniksa Commences Dosing of Vixarelimab Phase 2b Clinical Trial in Prurigo Nodularis

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– Evaluating vixarelimab across a range of once-monthly dose regimens injected subcutaneously –

HAMILTON, BERMUDA – December 15, 2020 – 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 the commencement of dosing in the Phase 2b clinical trial of vixarelimab in prurigo nodularis, a chronic inflammatory skin condition characterized by severely pruritic skin nodules. Vixarelimab is a fully-human monoclonal antibody that targets oncostatin M receptor beta (OSMRβ). The U.S. Food and Drug Administration (FDA) granted Breakthrough Therapy designation to vixarelimab for the treatment of pruritus associated with prurigo nodularis in 2020.

"We are pleased to announce the commencement of dosing in the Phase 2b trial of vixarelimab in patients with prurigo nodularis. The study builds upon learnings from our prior preclinical and clinical work in the disease and will evaluate vixarelimab across a range of once-monthly dose regimens," said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. "By targeting the OSMRβ receptor, we believe vixarelimab has the potential to make a meaningful impact on the lives of patients suffering with prurigo nodularis by addressing both the pruritus and the skin nodules associated with this devastating disease."

The Phase 2b trial is a randomized, double-blind, placebo-controlled study designed to investigate the efficacy, safety, and pharmacokinetics of vixarelimab in patients with prurigo nodularis. The trial is expected to enroll approximately 180 patients experiencing severe pruritus. Patients will be randomized 1:1:1:1 to receive vixarelimab 540 mg, 360 mg, 120 mg, or placebo as a once-monthly subcutaneous injection. The primary efficacy endpoint is the percent change from baseline in the weekly-average Worst-Itch Numeric Rating Scale (WI-NRS) at Week 16. Key secondary endpoints include the proportion of patients achieving greater-than-or-equal-to 4-point weekly-average WI-NRS reduction at Week 16 and the proportion of patients achieving a 0/1 score (clear/almost clear) on the prurigo nodularis-investigator's global assessment (PN-IGA) at Week 16.

Kiniksa's Breakthrough Therapy application was based on data from the Phase 2a clinical trial of vixarelimab in prurigo nodularis. The Phase 2a trial met its primary efficacy endpoint, as there was a statistically significant reduction in the weekly-average WI-NRS from baseline at Week 8 in vixarelimab recipients compared to placebo recipients. Additionally, the majority of vixarelimab recipients showed a clinically-meaningful greater-than-or-equal-to 4-point weekly-average WI-NRS reduction at Week 8, and a statistically significant percentage of vixarelimab recipients achieved a PN-IGA score of 0/1 at Week 8 compared to placebo recipients. Data from the Phase 2a trial were recently presented at the European Academy of Dermatology and Venereology Virtual Congress.

About Vixarelimab Phase 2b Trial in Prurigo Nodularis

The Phase 2b trial is a randomized, double-blind, placebo-controlled study designed to investigate the efficacy, safety, and pharmacokinetics of vixarelimab in reducing pruritus in subjects with prurigo nodularis. The trial is enrolling patients with moderate-to-severe prurigo nodularis experiencing severe pruritus (WI-NRS ≥ 7 at the screening visit and a mean weekly WI-NRS of ≥ 7 for the week (7 consecutive days) immediately prior to randomization). Patients are required to stop antihistamines and topical treatments, including corticosteroids, for at least two weeks prior to dosing. Prurigo nodularis treatments, other than study drug, are not allowed except for rescue.

About Vixarelimab

Vixarelimab is an investigational fully-human monoclonal antibody that targets 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. Kiniksa's lead indication for vixarelimab is prurigo nodularis, a chronic inflammatory skin condition characterized by severely pruritic skin nodules. The FDA granted Breakthrough Therapy designation to vixarelimab for the treatment of pruritus associated with prurigo nodularis 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

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 beliefs about the potential to impact the lives of patients with prurigo nodularis by addressing both the pruritus and inflammatory skin nodules associated with the debilitating disease; expected

number of patients to be enrolled in the dose-ranging Phase 2b clinical trial of vixarelimab in prurigo nodularis; our beliefs about the mechanisms of action of our product candidates and potential impact of their approach; and our beliefs about vixarelimab being the only monoclonal antibody in development that targets both pathways simultaneously.

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: delays or difficulty in enrollment of patients in, and activation or continuation of sites for, our dose-ranging Phase 2b clinical trial of vixarelimab in prurigo nodularis; potential amendments to our clinical trial protocols initiated by us or required by regulatory authorities; delays or difficulty in completing our dose-ranging Phase 2b clinical trial of vixarelimab in prurigo nodularis, including as a result of the COVID-19 pandemic; potential undesirable side effects caused by vixarelimab; our potential inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities or otherwise producing negative, inconclusive or commercially uncompetitive results; impact of additional data from us or other companies; potential inability to replicate in later clinical trials positive results from earlier pre-clinical and clinical trials or studies of vixarelimab in subsequent clinical trials conducted by us or third parties; drug substance and/or drug product shortages; our reliance on 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; substantial existing or new competition; impact of the COVID-19 pandemic, and measures taken in response to the pandemic, on our business and operations as well as the business and operations of our manufacturers, CROs upon whom we rely to conduct our clinical trials, and other third parties with whom we conduct business or otherwise engage, including the FDA and other regulatory authorities; changes in our operating plan and funding requirements; 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 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.

Every Second Counts!™

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