Kiniksa Announces 28-Day Clinical Outcomes Data from Mavrilimumab Treatment Protocol in Severe COVID-19 Pneumonia and Active U.S. IND for Phase 2/3 Clinical Trial

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HAMILTON, Bermuda, June 08, 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 the presentation of 28-day clinical outcomes data from the open-label treatment protocol with mavrilimumab, an investigational fully-human monoclonal antibody that targets granulocyte macrophage colony stimulating factor receptor alpha (GM-CSFRα), in severe coronavirus 2019 (COVID-19) pneumonia and hyperinflammation at the European E-Congress of Rheumatology (EULAR) 2020. The company also announced an active investigational new drug application (IND) with the U.S. Food and Drug Administration (FDA) for its global placebo-controlled Phase 2/3 clinical trial of mavrilimumab in severe COVID-19 pneumonia and hyperinflammation. Additionally, an investigator-initiated placebo-controlled study in the U.S. is enrolling patients.

On Saturday, June 6, 2020, at EULAR 2020, Professor Lorenzo Dagna, MD, FACP, Head, Unit of Immunology, Rheumatology, Allergy and Rare Diseases, IRCCS San Raffaele Scientific Institute and Vita-Salute San Raffaele University in Milan, Italy, delivered 28-day clinical outcomes data from the open-label treatment protocol with mavrilimumab in non-mechanically ventilated patients with severe COVID-19 pneumonia and hyperinflammation. The presentation, entitled *Mavrilimumab Improves Outcomes in Severe COVID-19 Pneumonia and Systemic Hyper-Inflammation*, is available through the Science section of Kiniksa's website (www.kiniksa.com).

In the treatment protocol, 13 non-mechanically ventilated patients with severe COVID-19 pneumonia and hyperinflammation were treated with a single intravenous dose of mavrilimumab 6 mg/kg upon admission to the hospital. Twenty-six contemporaneous non-mechanically ventilated patients with severe COVID-19 pneumonia and hyperinflammation and with similar baseline characteristics upon admission to the hospital, including comorbidities, baseline inflammatory markers and respiratory dysfunction, were evaluated as a control-group. All patients received similar standard of care therapy, including antivirals and antibiotics.

Over the course of the 28-day follow-up period, mavrilimumab-treated patients experienced earlier and improved clinical outcomes than control-group patients, including earlier weaning from supplemental oxygen, shorter hospitalizations, and no deaths.

- Death occurred in 0% (n=0/13) of mavrilimumab-treated patients by Day 28, compared to 27% (n=7/26) of control-group patients (p=0.086).
- 8% (n=1/13) of mavrilimumab-treated patients progressed to mechanical ventilation by Day 28, compared to 35% (n=9/26) of control-group patients who progressed to mechanical ventilation or died (p=0.077).
- 100% (n=13/13) of mavrilimumab-treated patients and 65% (n=17/26) of control-group patients attained the clinical improvement endpoint (defined as improvement of ≥ 2 categories on a 7-point scale for assessment of clinical status) by Day 28 (p=0.0001).
- Fever resolved in 91% (n=10/11 febrile patients) of mavrilimumab-treated patients by Day 14, compared to 61% (n=11/18 febrile patients) of control-group patients (p=0.0093).
- Representative mavrilimumab-treated patients showed significant improvement in lung opacification on computerized tomography (CT) scans, consistent with the overall improvement in their clinical status.

P-values above are unadjusted for multiplicity. Mavrilimumab was well-tolerated in all patients, without infusion reactions.

"The 28-day clinical outcomes data from the treatment protocol with mavrilimumab in COVID-19 pneumonia and hyperinflammation are consistent with the recently-reported 14-day dataset," said John F. Paolini, MD, PhD, Chief Medical Officer of Kiniksa. "Mavrilimumab-treated patients showed earlier and improved clinical outcomes compared to matched contemporaneous control-group patients throughout the protocol. These data are encouraging, and we look forward to evaluating mavrilimumab further in this patient population through placebo-controlled studies. To this end, we are pleased to announce the active U.S. IND for our global placebo-controlled Phase 2/3 clinical trial of mavrilimumab in severe COVID-19 pneumonia and hyperinflammation as well as the ongoing enrollment of a placebo-controlled investigator-initiated study in the U.S."

Kiniksa's Phase 2/3 clinical trial protocol is a global, randomized, double-blind, placebo-controlled study designed to evaluate the efficacy and safety of mavrilimumab relative to placebo in addition to standard of care therapy in the treatment of patients with severe COVID-19 pneumonia and hyperinflammation.

About Kiniksa's Phase 2/3 Clinical Trial Protocol of Mavrilimumab in Severe COVID-19 Pneumonia and Hyperinflammation
Kiniksa's Phase 2/3 clinical trial protocol is a global, randomized, double-blind, placebo-controlled study encompassing 2 phases of development (Phase 2 and Phase 3). The Phase 2 portion of the trial is expected to enroll approximately 160 patients and is intended to evaluate the efficacy and safety of 2 dose levels of mavrilimumab relative to placebo in patients who have tested positive for COVID-19 and have x-ray/CT evidence of bilateral pneumonia, active or recent fever, and clinical laboratory results indicative of hyperinflammation. The Phase 3 portion is expected to enroll approximately 420 patients and is intended to confirm Phase 2 efficacy and safety findings. In both Phase 2 and Phase 3, patients are expected to be

enrolled into 2 cohorts: Cohort 1 will include non-intubated, hospitalized patients who require supplemental oxygen to maintain SpO2 ≥ 92%, (i.e., non-mechanically ventilated patients); and Cohort 2 will include hospitalized patients for whom mechanical ventilation was recently initiated within 48 hours prior to randomization (i.e., ventilated patients). Following screening, enrolled patients in each cohort will be randomized 1:1:1 to receive a single IV infusion of mavrilimumab 6mg/kg or 10 mg/kg or placebo (Day 1). The primary efficacy endpoint for the Phase 2 portion of the trial for Cohort 1 is the proportion of patients alive and without respiratory failure (defined as the need for high flow oxygen, non-invasive ventilation, invasive mechanical ventilation, or extracorporeal membrane oxygenation) at Day 15 and for Cohort 2 is the mortality rate by Day 15. There will be a seamless transition in enrollment of patients in both cohorts between the Phase 2 and Phase 3 portions of the trial. For each cohort, once the last patient in Phase 2 is enrolled, all subsequent patients will be considered Phase 3 patients. Once the last patient in Phase 2 completes Day 15, primary efficacy and safety analyses of the Phase 2 data will be conducted. Following demonstration of efficacy and safety in Phase 2, the Phase 3 portion of the trial will be continued/completed.

About the Investigator-Initiated Placebo-Controlled Study of Mavrilimumab in Severe COVID-19 Pneumonia and Hyperinflammation in the U.S.

The investigator-initiated Phase 2 trial is a randomized, double-blind, placebo-controlled study in the U.S. designed to evaluate the efficacy and safety of mavrilimumab versus placebo on top of standard of care therapy in the treatment of severe COVID-19 pneumonia and hyperinflammation. Standard of care therapy may include, but is not limited to, anti-viral treatment and/or supportive care. The clinical trial is expected to enroll up to approximately 60 patients with diagnosed COVID-19 pneumonia and hyperinflammation, initially at Cleveland Clinic. The clinical trial may expand to other centers depending on case rates in different geographies. Patients will be randomized 1:1 to mavrilimumab 6 mg/kg or placebo. The primary endpoint is the proportion of patients alive and off of supplemental oxygen at Day 14. For more information, refer to ClinicalTrials.gov Identifier: NCT04399980.

About the Treatment Protocol of Mavrilimumab in Severe COVID-19 Pneumonia and Hyperinflammation in Italy

The mavrilimumab open-label treatment protocol was a prospective, interventional, single-active-arm, single-center pilot experience in Italy conducted by Professor Lorenzo Dagna. within a COVID-19 Program directed by Professor Alberto Zangrillo, Head of Department of Anesthesia and Intensive Care of the Scientific Institute San Raffaele Hospital and Università Vita-Salute San Raffaele in Milan, Italy. Patients suffering from severe pulmonary involvement of COVID-19, acute respiratory distress, fever, and clinical and biological markers of systemic hyperinflammation status were treated with a single intravenous dose of mavrilimumab 6mg/kg. The objective of the treatment protocol was to determine whether mavrilimumab in addition to standard of care therapy could improve clinical outcomes in patients with COVID-19 pneumonia and hyperinflammation. A control-group was assembled consisting of contemporaneous patients receiving standard of care therapy and matched for age, sex, comorbidities, baseline inflammatory markers and respiratory dysfunction. Per standard of care therapy, all patients received on admission medical treatment with hydroxychloroquine, azithromycin, and lopinavir/ritonavir as well as respiratory support with supplemental oxygen and/or non-invasive ventilation with continuous positive airway pressure.

About Mavrilimumab

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

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, have 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

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 our Phase 2/3 clinical trial protocol and investigator initiated studies for mavrilimumab in severe COVID-19 pneumonia and hyperinflammation; our clinical collaboration with Kite in CAR T; our assessment of the 14-Day and 24-Day outcomes data from the treatment protocol with mavrilimumab in severe COVID-19 pneumonia and hyperinflammation in Italy; our plans to evaluate mavrilimumab further in that patient population through placebo-controlled studies; our planned clinical trial designs; and the potential for all of 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 inability to replicate in later clinical trials positive results from earlier studies, clinical trials or investigator initiated treatment protocols for mavrilimumab in severe COVID-19 pneumonia and hyperinflammation; the potential for undesirable side effects to be caused by mavrilimumab; changes to our clinical trial protocol; case rates of severe COVID-19 pneumonia and hyperinflammation in various geographies; our reliance on third parties to manufacture our product candidates and conduct our clinical trials; 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 4, 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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