Kiniksa Announces Upcoming Presentation on Mavrilimumab in COVID-19 Pneumonia and Hyperinflammation at the European E-Congress of Rheumatology 2020

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HAMILTON, Bermuda, May 29, 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 an upcoming presentation, entitled *Mavrilimumab Improves Outcomes in Severe COVID-19 Pneumonia and Systemic Hyper-Inflammation,* showing 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 patients with severe coronavirus 2019 (COVID-19) pneumonia and hyperinflammation will be delivered at the European E-Congress of Rheumatology (EULAR) 2020. Additionally, preclinical data analyzing the role of the granulocyte macrophage colony stimulating factor (GM-CSF) pathway in giant cell arteritis (GCA) pathophysiology will be included in a poster presentation.

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 will deliver an oral presentation of outcomes data from the mavrilimumab treatment protocol in COVID-19 pneumonia and hyperinflammation in Italy.

Oral Presentation Details:

- **Abstract #CO0001:** *Mavrilimumab Improves Outcomes in Severe COVID-19 Pneumonia and Systemic Hyper-Inflammation*
- **Session:** EULAR COVID-19 Recommendations
- **Date and Time:** Saturday, June 6, 2020 from 10:15am to 10:25am Central European Time*****
- **Lead Authors:** Lorenzo Dagna, Giacomo De Luca[1,2], Corrado Campochiaro[1,2]

There will also be a presentation of preclinical data analyzing the role of the GM-CSF pathway in GCA pathophysiology delivered by Dr. Maria C. Cid, MD, Hospital Clínic, University of Barcelona, Institut d'Investigacions Biomèdiques August Pi i Sunyer (IDIBAPS), Vasculitis Research Unit, Department of Autoimmune Diseases, Barcelona, Spain.

Poster Presentation Details:

- **Abstract #FRI0010:** *GM-CSFR Pathway is Implicated in Pathogenic Inflammatory Mechanisms in Giant Cell Arteritis*
- **Session:** A Journey into the Innate Immune Mechanisms Driving Rheumatic Diseases and Novel Therapeutic Strategies
- **Date and Time:** Friday, June 5, 2020 from 11:50am to 1:30pm Central European Time
- **Lead Authors:** Maria C. Cid, Sujatha Muralidharan[3], Marc Corbera-Bellalta[4]

Kiniksa intends to make the presentations available through the Science section of Kiniksa's website (www.kiniksa.com) after the EULAR embargo lifts, which is expected to be at the time of each presentation.

***Information updated as of 6/3/2020**

[1]IRCCS San Raffaele Scientific Institute, Milan, Italy; [2]Vita-Salute San Raffaele University, Milano, Italy; [3]Kiniksa Pharmaceuticals, Lexington, United States of America; [4]Vasculitis Research Unit, Hospital Clinic, University of Barcelona, IDIBAPS.

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 have a clinical collaboration to evaluate mavrilimumab in combination with Yescarta® (axicabtagene ciloleucel) in patients with relapsed or refractory large B-cell lymphoma.

About the Mavrilimumab Treatment Protocol in COVID-19 Pneumonia & 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, 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 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. The objective of the treatment protocol was to determine whether mavrilimumab in addition to standard management could improve clinical outcomes in patients with COVID-19 pneumonia and

hyperinflammation. A control-group was assembled consisting of contemporaneous patients receiving local standard of care and matched for age, sex, comorbidities, baseline inflammatory markers and respiratory dysfunction. Per standard of care of the hospital, 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 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 signaling pathways that are implicated across a spectrum of diseases. For more information, please visit www.kiniksa.com.

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