Kiniksa Announces Results from Phase 3 Trial of Mavrilimumab in COVID-19-Related ARDS

December 28, 2021

HAMILTON, Bermuda, Dec. 28, 2021 (GLOBE NEWSWIRE) -- Kiniksa Pharmaceuticals, Ltd. (Nasdaq: KNSA) ("Kiniksa"), a biopharmaceutical company with a portfolio of assets designed to modulate immunological pathways across a spectrum of diseases, today announced that the Phase 3 portion of the Phase 2/3 trial of mavrilimumab in COVID-19-related acute respiratory syndrome (ARDS) did not meet the primary efficacy endpoint. Mavrilimumab is an investigational fully human monoclonal antibody that targets granulocyte macrophage colony stimulating factor receptor alpha (GM-CSFRα).

"The Phase 3 study of mavrilimumab in COVID-19-related ARDS did not provide the expected outcome, however we are proud of our efforts to help patients in need during this unprecedented time. We greatly appreciate the participation of the patients, their families, the investigators, and the Kiniksa employees who made this study possible," said Sanj K. Patel, Chairman and Chief Executive Officer of Kiniksa. "We continue to believe in the potential broad utility of mavrilimumab and are evaluating next steps for the molecule. Our current strategy focuses our resources on the ARCALYST franchise, including the commercial execution in recurrent pericarditis, as well as the development of vixarelimab and our anti-CD40 program, KPL-404."

The Phase 2/3 trial is a global, double-blind, placebo-controlled study designed to evaluate the safety and efficacy of mavrilimumab for the treatment of hospitalized, non-mechanically-ventilated adult patients with hypoxia and severe COVID-19 pneumonia/hyperinflammation. A total of 582 patients in the Phase 3 portion of the trial were randomized in a 1:1:1 ratio to receive a single intravenous dose of mavrilimumab 10 mg/kg, 6 mg/kg, or placebo. The Phase 3 portion of the trial did not meet its primary efficacy endpoint of proportion of patients alive and free of mechanical ventilation at Day 29.

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 portfolio of assets, ARCALYST, mavrilimumab, vixarelimab and KPL-404, are based on strong biologic rationale or validated mechanisms, target underserved conditions, and offer the potential for differentiation. These assets are designed to modulate immunological pathways across a spectrum of diseases. For more information, please visit www.kiniksa.com.

About Mavrilimumab
Mavrilimumab is an investigational fully human monoclonal antibody that blocks activity of granulocyte macrophage colony stimulating factor (GM-CSF) by specifically binding to the alpha subunit of the GM-CSF receptor. Mavrilimumab was previously evaluated in rheumatoid arthritis through Phase 2b clinical studies in Europe and achieved prospectively defined primary endpoints of efficacy and safety. Kiniksa is evaluating next steps for mavrilimumab in giant cell arteritis (GCA). The Phase 2 clinical trial of mavrilimumab in GCA achieved both the primary and secondary efficacy endpoints with statistical significance. The U.S. Food and Drug Administration (FDA) granted Orphan Drug designation to mavrilimumab for the treatment of GCA in 2020.

About ARCALYST®
ARCALYST is a weekly, subcutaneously injected recombinant dimeric fusion protein that blocks IL-1 alpha and IL-1 beta signaling. ARCALYST was discovered by Regeneron and is approved by the FDA for recurrent pericarditis, cryopyrin-associated periodic syndromes (CAPS), including Familial Cold Autoinflammatory Syndrome and Muckle-Wells Syndrome, and deficiency of IL-1 receptor antagonist (DIRA). The FDA granted Breakthrough Therapy designation to ARCALYST for the treatment of recurrent pericarditis in 2019 and Orphan Drug designation to ARCALYST for the treatment of pericarditis in 2020.

Important Information About ARCALYST Injection

- ARCALYST can affect your immune system and can lower the ability of your immune system to fight infections. Serious infections, including life-threatening infections and death have happened in patients taking ARCALYST. You should not begin ARCALYST if you have an infection or have infections that keep coming back. After starting ARCALYST, if you get an infection or show any sign of an infection, including a fever, cough, flu-like symptoms, or have any open sores on your body, call your doctor right away. Treatment with ARCALYST should be stopped if you get a serious infection.
- While taking ARCALYST, do not take other medicines that block interleukin-1, such as Kineret® (anakinra), or medicines that block tumor necrosis factor, such as Enbrel® (etanercept), Humira® (adalimumab), or Remicade® (infliximab), as this may increase your risk of getting a serious infection.
- Before starting ARCALYST, tell your doctor if you think you have an infection, are being treated for an infection, have signs of an infection, have any open sores, have a history of infections that keep coming back, have asthma, have diabetes or an immune system problem, have tuberculosis, or have been in contact with someone who has had tuberculosis, has or has had HIV, hepatitis B or hepatitis C, or takes other medicines that affect your immune system.
- Before you begin treatment with ARCALYST, talk with your healthcare provider about your vaccine history. Ask your healthcare provider whether you should receive any vaccines, including the pneumonia vaccine and flu vaccine, before you

begin treatment with ARCALYST.

- ARCALYST can cause serious side effects:
- Medicines that affect the immune system may increase the risk of getting cancer.
- Stop taking ARCALYST and call your doctor or get emergency care right away if you have any symptoms of an allergic reaction (e.g., rash, swollen face, trouble breathing).
- Your doctor will do blood tests to check for changes in your blood cholesterol and triglycerides.
- Common side effects of ARCALYST include injection-site reactions, upper respiratory tract infections, joint and muscle aches, rash, ear infection, sore throat, and runny nose.
- Tell your doctor if you are scheduled to receive any vaccines, if you are pregnant or plan to become pregnant, and if you are breastfeeding or plan to breastfeed.
- Tell your doctor if you take other medicines that affect the immune system such as interleukin-1 blockers, tumor necrosis factor blockers, or corticosteroids.
- For more information about ARCALYST, talk to your doctor and see the Product Information.

About Vixarelimab
Vixarelimab is an investigational fully human monoclonal antibody that targets oncostatin M receptor beta (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 KPL-404
KPL-404 is an investigational humanized monoclonal antibody that is designed to inhibit CD40-CD154 (CD40 ligand) interaction, a key T-cell co-stimulatory signal critical for B-cell maturation and immunoglobulin class switching and Type 1 immune responses. Kiniksa believes disrupting the CD40-CD154 interaction is an attractive approach to address multiple autoimmune disease pathologies. Kiniksa owns or controls the intellectual property related to KPL-404.

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 in the potential broad utility of mavrilimumab and our expectations regarding our next steps for mavrilimumab, including in giant cell arteritis, our strategy regarding the focus of our resources, our beliefs about the mechanisms of action of our product candidates and potential impact of their approach, including that vixarelimab is the only monoclonal antibody in development that targets both interleukin-31 (IL-31) and oncostatin M (OSM) pathways simultaneously and that disrupting the CD40-CD154 interaction is an attractive approach to address multiple autoimmune disease pathologies; our belief that all of our product candidates offer the potential for differentiation; and our ability to execute on our clinical stage pipeline.

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 inability to replicate in later clinical trials the positive final data from our earlier clinical trials or studies; impact of additional data from us or other companies, including the potential for our data to produce negative, inconclusive or commercially uncompetitive results; potential undesirable side effects caused by our products and product candidates; our inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities; potential for applicable regulatory authorities to not accept our filings or to delay or deny approval of any of our product candidates or to require additional data or trials to support any such approval; our reliance on third parties as the sole source of supply of the drug substance and drug products used in our products and product candidates; the 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 existing or new competition.

These and other important factors discussed in our filings with the U.S. Securities and Exchange Commission (the "SEC"), including under the caption "Risk Factors" contained therein, 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. Except as required by law, we disclaim any intention or obligation to update or revise any forward-looking statements. 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!®

Kiniksa Investor and Media Contact
Rachel Frank
(339) 970-9437
rfrank@kiniksa.com