Kiniksa Highlights Corporate Priorities and Expected 2021 Milestones

January 11, 2021

- PDUFA goal date of March 21, 2021 for rilonacept in recurrent pericarditis -
- Data from Phase 2 portion of mavrilimumab Phase 2/3 trial in severe COVID-19 pneumonia and hyperinflammation expected in 1H 2021 -
- Final Phase 1 KPL-404 data expected in 1H 2021 -

HAMILTON, Bermuda, Jan. 11, 2021 (GLOBE NEWSWIRE) -- 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 highlighted its corporate priorities and expected 2021 milestones. Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa will provide further detail in a corporate presentation at the Virtual 39th Annual J.P. Morgan Healthcare Conference today, Monday, January 11, 2021 at 4:30 p.m. Eastern Time.

"2020 was marked by significant progress across our entire pipeline, setting the stage to build long-term value across our portfolio," said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. "Moving forward, 2021 has the potential to be a transformational year for Kiniksa with multiple catalysts expected across our pipeline, notably with the potential commercial launch of rilonacept in recurrent pericarditis in the first half of the year. As we focus on our launch readiness preparations, our commitment to bringing novel therapies to patients with unmet need remains at the core of our goals."

Expected 2021 Milestones

Rilonacept (IL-1α and IL-1β cytokine trap)

- Kiniksa's Prescription Drug User Fee Act (PDUFA) goal date for rilonacept in recurrent pericarditis is March 21, 2021, as assigned by the U.S. Food and Drug Administration (FDA) upon the acceptance of the supplemental Biologics License Application (sBLA), with priority review. If approved by the FDA, Kiniksa expects the potential commercial launch of rilonacept in recurrent pericarditis in the first half of 2021 and consequently would evenly split profits on sales of all approved indications in the United States, including cryopyrin-associated periodic syndromes (CAPS) and deficiency of IL-1 receptor antagonist (DIRA), with Regeneron Pharmaceuticals, Inc. (Regeneron).

Mavrilimumab (monoclonal antibody inhibitor targeting GM-CSFRα)

- Kiniksa plans to provide next steps for the development of mavrilimumab, including for giant cell arteritis (GCA), in the first half of 2021.
- Kiniksa is conducting a Phase 2/3 clinical trial in severe COVID-19 pneumonia and hyperinflammation. The company expects to provide data from the Phase 2 portion of the trial in the first half of 2021.

Vixarelimab (monoclonal antibody inhibitor of signaling through OSMRβ)

- Kiniksa is conducting a Phase 2b dose-ranging trial of vixarelimab in prurigo nodularis. The Phase 2b trial is expected to enroll approximately 180 patients experiencing severe pruritus. Patients will be randomized to receive vixarelimab or placebo subcutaneously (SC) once-monthly.

KPL-404 (monoclonal antibody inhibitor of signaling between CD40 and CD40L)

- Full receptor occupancy through Day 29 shown in preliminary Phase 1 data from the single-ascending dose clinical trial of KPL-404 in healthy volunteers at the 3 mg/kg intravenous (IV) dose. This corresponded with complete suppression of the T-cell Dependent Antibody Response (TDAR) to keyhole limpet hemocyanin (KLH) through Day 29.
- The data to-date support further evaluation in patients, including potential monthly IV or SC administration. Kiniksa expects final data and safety follow-up from all cohorts in the first half of 2021.

Financial Guidance
Kiniksa ended 2020 with approximately $323 million in cash, cash equivalents and short-term investments (unaudited). The company expects that these cash reserves will fund its current operating plan into 2023.

Presentation at the Virtual 39th Annual J.P. Morgan Healthcare Conference
Kiniksa will webcast its corporate presentation at the Virtual 39th Annual J.P. Morgan Healthcare Conference today, Monday, January 11, 2021 at 4:30 p.m. Eastern Time. A live webcast of Kiniksa's presentation will be accessible through the Investors & Media section of the company's website (www.kiniksa.com). A replay of the webcast will be available on Kiniksa's website for 14 days following the conference.

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.

About Rilonacept
Rilonacept is a weekly, subcutaneously-injected, recombinant dimeric fusion protein that blocks interleukin-1 alpha (IL-1α) and interleukin-1 beta (IL-1β) signaling. Rilonacept was discovered and developed by Regeneron and is approved by the FDA under the brand name ARCALYST® for the treatment of CAPS, specifically Familial Cold Autoinflammatory Syndrome and Muckle-Wells Syndrome, and DIRA. Rilonacept in recurrent pericarditis is an investigational drug. The FDA granted Breakthrough Therapy designation to rilonacept for the treatment of recurrent pericarditis in 2019 and Orphan Drug designation to rilonacept for the treatment of pericarditis in 2020.

Important information about ARCALYST® (rilonacept) Injection
IL-1 blockade may interfere with immune response to infections. Serious, life-threatening infections have been reported in patients taking ARCALYST. ARCALYST should be discontinued if a patient develops a serious infection. Taking ARCALYST with TNF inhibitors is not recommended because this may increase the risk of serious infections.

Patients should not receive a live vaccine while taking ARCALYST. It is recommended that prior to initiation of therapy with ARCALYST patients receive all recommended vaccinations, as appropriate, including pneumococcal vaccine and inactivated influenza vaccine. In the initial development program for ARCALYST, six serious adverse reactions were reported by four patients: Mycobacterium intracellular infection, gastrointestinal bleeding and colitis, sinusitis and bronchitis and Streptococcus pneumoniae meningitis. The most commonly reported adverse reactions associated with ARCALYST were injection site reaction and upper respiratory tract infection. Patients should be monitored for changes in their lipid profiles and provided with medical treatment if warranted. Treatment with immunosuppressants, including ARCALYST, may result in an increase in risk of malignancies. Hypersensitivity reactions associated with ARCALYST administration in clinical studies have been rare. If a hypersensitivity reaction occurs, administration of ARCALYST should be discontinued and appropriate therapy initiated.

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 GCA, a rare 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 the treatment of GCA in 2020.

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-CD40 ligand (CD40L) 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-CD40L interaction is an attractive approach for multiple autoimmune disease pathologies such as rheumatoid arthritis, Sjogren's syndrome, Graves' disease, systemic lupus erythematosus and solid organ transplant. 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 that progress made across our pipeline in 2020, has set the stage to build long-term value across our portfolio; our belief that 2021 has the potential to be a transformational year for Kiniksa; expected multiple catalysts across our pipeline in 2021; the potential commercial launch of rilonacept in recurrent pericarditis in the first half of the year, if approved by the FDA; expected timing of data from clinical trials, including expected data from the Phase 2 portion of the adaptive design Phase 2/3 clinical trial of mavrilimumab in severe COVID-19 pneumonia and hyperinflammation in the first half of 2021, next steps for the development of mavrilimumab, including for giant cell arteritis (GCA), in the first half of 2021, and final data from the single-ascending-dose Phase 1 clinical trial of KPL-404 in healthy volunteers in the first half of 2021; our belief that KPL-404 has the potential to address a broad range of autoimmune diseases; our beliefs about the mechanisms of action of our product candidates and potential impact of their approach, including our beliefs that vixarelimab is the only monoclonal antibody in development that targets both interleukin-31 (IL-31) and oncostatin M (OSM) pathways simultaneously; that KPL-404's disruption of the CD40-CD40L interaction is an attractive approach for multiple autoimmune disease pathologies; our belief that all of our product candidates offer the potential for differentiation; and expectation about our cash reserves funding our current operating plan into 2023.

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: delays or difficulty in enrollment of patients in, and activation or continuation of sites for, our clinical trials; amendments to our clinical trial protocols initiated by us or required by regulatory authorities; delays or difficulty in completing our clinical trials, including as a result of the COVID-19 pandemic; potential for changes between final data and any preliminary, interim, top-line or other data from clinical trials conducted by us or third parties; 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; impact of

additional data from us or other companies; potential undesirable side effects caused by our product candidates; our inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities or otherwise producing negative, inconclusive or commercially uncompetitive results; potential for applicable regulatory authorities to not accept our BLA or sBLA filings or to delay or deny approval of any of our product candidates or to require additional 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 product candidates and to manufacture our product candidates; drug substance and/or drug product shortages; our reliance on third parties to conduct research, clinical trials, and/or certain regulatory activities for our product candidates; complications in coordinating requirements, regulations and guidelines of regulatory authorities across jurisdictions for our clinical trials; 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 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 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.

ARCALYST® is a registered trademark of Regeneron Pharmaceuticals, Inc.

Every Second Counts!™

Kiniksa Investor and Media Contact
Mark Ragosa
(781) 430-8289
mragosa@kiniksa.com

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