Kiniksa Provides Corporate Update and Outlines Anticipated 2022 Milestones

January 10, 2022

- ARCALYST® full-year 2022 guidance to be provided with Q4 2021 financial results -
- Vixarelimab Phase 2b data in prurigo nodularis expected in 2H 2022 –
- KPL-404 Phase 2 trial in rheumatoid arthritis now enrolling and dosing patients -
- Year-end 2021 cash reserves expected to fund current operating plan into 2024 -

HAMILTON, Bermuda, Jan. 10, 2022 (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 provided a corporate update and announced anticipated milestones for 2022.

“2021 was a transformational year for Kiniksa. We progressed to a commercial-stage company with the successful launch of ARCALYST for the treatment of recurrent pericarditis. We are pleased with the strong physician and patient adoption and expect to provide annual guidance along with fourth quarter and full-year 2021 financial results,” said Sanj K. Patel, Chairman and Chief Executive Officer of Kiniksa. “This year, we look forward to continued commercial excellence with ARCALYST, as well as reporting data from the Phase 2b trial of vixarelimab in prurigo nodularis, and conducting the Phase 2 trial of KPL-404 in rheumatoid arthritis.”

Corporate Update

- Kiniksa’s resources are currently focused on growing the ARCALYST franchise and progressing the clinical trials of vixarelimab in prurigo nodularis and KPL-404 in rheumatoid arthritis.
- Kiniksa’s year-end 2021 cash, cash equivalents and short-term investments of approximately $182 million (unaudited) are expected to fund its current operating plan into 2024.

Portfolio Update

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

- Kiniksa expects to provide full-year 2022 ARCALYST net revenue guidance with its fourth quarter and full-year 2021 financial results.

Mavrilimumab (monoclonal antibody inhibitor targeting GM-CSFRα)

- As previously reported, the Phase 3 portion of the Phase 2/3 trial of mavrilimumab in COVID-19-related acute respiratory distress syndrome (ARDS) did not meet its primary efficacy endpoint of the proportion of patients alive and free of mechanical ventilation at Day 29.
- Kiniksa continues to believe in the potential broad utility of mavrilimumab and is evaluating next steps for the molecule.

Vixarelimab (monoclonal antibody inhibitor of signaling through OSMRβ)

- Kiniksa expects data from the Phase 2b dose-ranging clinical trial of once-monthly subcutaneous vixarelimab in prurigo nodularis in the second half of 2022.

KPL-404 (monoclonal antibody inhibitor of CD40-CD154 signaling)

- Kiniksa is enrolling and dosing patients in a Phase 2 clinical trial of KPL-404 in rheumatoid arthritis which is designed to enable potential development in a spectrum of autoimmune diseases believed to be mediated by the CD40-CD154 pathway.

40th Annual J.P. Morgan Healthcare Conference

Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa will provide a corporate presentation at the 40th Annual J.P. Morgan Healthcare Conference today, Monday, January 10, 2022, at 1:30 p.m. Eastern Time. A live webcast of Kiniksa’s presentation will be accessible through the Investors & Media section of the company’s website at www.kiniksa.com. A replay of the webcast will also be available on Kiniksa’s website within approximately 48 hours after the event.

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 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 ARCALYST
ARCALYST is a weekly, subcutaneously injected recombinant dimeric fusion protein that blocks interleukin-1 alpha (IL-1α) and interleukin-1 beta (IL-1β) signaling. ARCALYST was discovered by Regeneron and is approved by the U.S. Food and Drug Administration (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 SAFETY INFORMATION ABOUT ARCALYST

- ARCALYST may 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. If you have any signs of an infection, call your doctor right away. Treatment with ARCALYST should be stopped if you get a serious infection. You should not begin treatment with ARCALYST if you have an infection or have infections that keep coming back (chronic 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.
- Talk with your doctor about your vaccine history. Ask your doctor whether you should receive any vaccines before you begin treatment with ARCALYST.
- 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.
- Your doctor will do blood tests to check for changes in your blood cholesterol and triglycerides.
- Common side effects include injection-site reactions (which may include pain, redness, swelling, itching, bruising, lumps, inflammation, skin rash, blisters, warmth, and bleeding at the injection site), upper respiratory tract infections, joint and muscle aches, rash, ear infection, sore throat, and runny nose.

For more information about ARCALYST, talk to your doctor and see the Product Information.

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 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-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 beliefs about the future commercial demand for ARCALYST; our expectation about our year-end cash reserves and those cash reserves funding our current operating plan into 2024; expected timing of data from the dose-ranging Phase 2b clinical trial of vixarelimab in prurigo nodularis in the second half of 2022; our expectation about continued enrollment and dosing of patients in a Phase 2 trial of KPL-404 in rheumatoid arthritis; our belief in the potential broad utility of mavrilimumab and our expectations regarding our next steps for mavrilimumab, including in giant cell arteritis; our belief that KPL-404 has broad utility across multiple indications; 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 plans to present at any future conferences.

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 ongoing 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; 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; delays, difficulty or inability to successfully execute on our commercial strategy for ARCALYST; 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; our reliance on Regeneron as the sole manufacturer of ARCALYST; raw materials, 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 in our filings with the U.S. Securities and Exchange Commission, 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.

ARCALYST® is a registered trademark of Regeneron Pharmaceuticals, Inc. All other trademarks are the property of their respective owners.

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