Kiniksa Reports First Quarter 2021 Financial Results and Recent Corporate and Portfolio Activity

May 4, 2021

- ARCALYST® (rilonacept) launched as the first and only FDA-approved therapy for recurrent pericarditis -
- Mavrilimumab Phase 2 severe COVID-19 data demonstrated a reduction in mechanical ventilation and death at Day 29; Phase 3 enrollment ongoing -
- Final KPL-404 Phase 1 data support further development in patients; Phase 2 proof-of-concept trial initiation planned for 2H 2021 -
- Cash reserves of approximately $264 million -

HAMILTON, Bermuda, May 04, 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 reported first quarter 2021 financial results and recent corporate and portfolio activity.

"The first quarter was transformational for Kiniksa with the approval of ARCALYST as the first and only FDA-approved therapy for patients with recurrent pericarditis. We are focused on the launch of ARCALYST and are confident in our commercialization strategy," said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. "Additionally, we are executing across our broader portfolio of immune-modulating assets. We recently reported positive data for mavrilimumab in severe COVID-19 pneumonia and hyperinflammation and remain engaged with the FDA and other government agencies to identify pathways for accelerated availability of mavrilimumab as a potential therapeutic option for this patient population. We also reported positive final Phase 1 data for our potentially best-in-class anti-CD40 program, KPL-404, and plan to initiate a Phase 2 proof-of-concept trial in the second half of 2021."

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

- Kiniksa received approval from the U.S. Food and Drug Administration (FDA) on March 18, 2021, for ARCALYST for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and children 12 years and older. The commercial launch of ARCALYST in recurrent pericarditis commenced in April 2021.
    - Kiniksa is responsible for sales and distribution of ARCALYST for all the approved indications in the United States, including cryopyrin-associated periodic syndromes (CAPS) and deficiency of IL-1 receptor antagonist (DIRA), and will evenly split profits with Regeneron Pharmaceuticals, Inc. (Regeneron).
- Kiniksa is executing on its commercial strategy, including engagement with priority accounts and payers to enable rapid and broad access to ARCALYST for patients.
- Kiniksa is enrolling pediatric and adult patients with recurrent pericarditis in the RESONANCE registry.

Mavrilimumab (monoclonal antibody inhibitor targeting GM-CSFRα)

- Kiniksa expects to provide next steps for mavrilimumab, including for giant cell arteritis (GCA), in the second quarter of 2021.
- Kiniksa announced data from the Phase 2 portion of the Phase 2/3 clinical trial of mavrilimumab in non-mechanically ventilated patients with severe COVID-19 pneumonia and hyperinflammation receiving local standard of care. Non-mechanically ventilated patients treated with mavrilimumab demonstrated a reduction in mechanical ventilation and death at Day 29 pooled across dose cohorts.
    - Kiniksa continues to advance its engagement activities with the FDA and other government agencies to identify pathways to potentially accelerate availability of mavrilimumab as a therapeutic option for severe COVID-19 patients. Enrollment in the Phase 3 portion of the trial is ongoing.

Vixarelimab (monoclonal antibody inhibitor of signaling through OSMRβ)

- Kiniksa is conducting a placebo-controlled Phase 2b clinical trial of vixarelimab in prurigo nodularis, evaluating a range of once-monthly dose regimens via subcutaneous (SC) injection.
    - The primary efficacy endpoint is the percent change from baseline in the weekly-average Worst-Itch Numeric Rating Scale at Week 16.

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

- Kiniksa announced final data today from the KPL-404 Phase 1 clinical trial in healthy volunteers. KPL-404 was well

tolerated and showed dose-dependent increases in concentration across cohorts. The data support further development in patients with optionality for intravenous and/or SC administration.
    - Kiniksa plans to initiate a Phase 2 proof-of-concept clinical trial of KPL-404 in rheumatoid arthritis in the second half of 2021. The planned trial will provide safety and characterization of chronic dosing with SC administration over 12 weeks as well as the potential to evaluate KPL-404 across a range of other autoimmune diseases.

Scientific Conference Presentations

- Kiniksa plans to present additional data from the RHAPSODY, the pivotal Phase 3 trial of rilonacept, at the American College of Cardiology virtual scientific conference, which will be available starting on May 15, 2021 at 8:00 a.m. Eastern Time. Details of the presentations are as follows:
    - Antonio Brucato, MD, Department of Biomedical and Clinical Science, University of Milan, Fatebenefratelli Hospital, Milan, will present a poster entitled, *Tapering and discontinuation of background therapies during the transition to rilonacept monotherapy in RHAPSODY, a phase 3 clinical trial of rilonacept in patients with recurrent pericarditis.*
    - Paul Cremer, MD, Department of Cardiovascular Medicine, Cleveland Clinic, Cleveland, will present a moderated poster entitled, *Cardiac magnetic resonance imaging for guiding decision-making on treatment duration: data from RHAPSODY, a phase 3 clinical trial of rilonacept in recurrent pericarditis.*

Financial Results

- Net loss for the first quarter of 2021 was $49.5 million, compared to a net loss of $26.4 million for the first quarter of 2020.
- Total operating expenses for the first quarter of 2021 were $49.3 million, compared to $29.4 million for the first quarter of 2020.
    - Non-cash, share-based compensation expense for the first quarter of 2021 was $7.1 million, compared to $4.2 million for the first quarter of 2020.
- Kiniksa made a $20.0 million milestone payment to Regeneron in the first quarter of 2021 upon the FDA approval of ARCALYST in recurrent pericarditis. The milestone payment was capitalized as an intangible asset and will be amortized through cost of goods sold on a straight-line basis over the 20-year life of the asset starting in the second quarter of 2021.
- As of March 31, 2021, the company had cash, cash equivalents and short-term investments of $264.0 million and no debt.

Financial Guidance

- Kiniksa expects that its cash, cash equivalents and short-term investments will fund its current operating plan into 2023.

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.

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 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 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 is evaluating mavrilimumab in GCA, a rare inflammatory disease of medium-to-large arteries. The company's Phase 2 trial in GCA achieved both the primary and secondary efficacy endpoints with statistical significance. Kiniksa is also evaluating mavrilimumab in severe 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-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 for 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 engaging with the FDA and other government agencies to identify pathways for the potential accelerated availability of mavrilimumab as a therapeutic option for severe COVID-19 patients; expected timing of next steps for mavrilimumab, including for giant cell arteritis (GCA), in the second quarter of 2021; our beliefs about our commercial strategy for ARCALYST; our beliefs about the final data from our Phase 1 clinical trial of KPL-404 in healthy volunteers ; expected timing and design of clinical trials, including initiating our Phase 2 proof-of-concept trial of KPL-404 in rheumatoid arthritis in the second half of 2021 and the potential to evaluate KPL-404 across a range of other autoimmune diseases; 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 that vixarelimab is the only monoclonal antibody in development that targets both interleukin-31 (IL-31) and oncostatin M (OSM) pathways simultaneously and that KPL-404 has the potential to be a best-in-class monoclonal antibody inhibitor; 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; 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, or emergency use authorization for, any of our product candidates or to require additional data or trials to support any such approval or authorization; delays, difficulty or inability 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 and to manufacture our products and 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 Annual Report on Form 10-K filed with the Securities and Exchange Commission ("SEC") on February 25, 2021 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. All other trademarks are the property of their respective owners.

Every Second Counts!™

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KINIKSA PHARMACEUTICALS, LTD.
CONSOLIDATED STATEMENTS OF OPERATIONS
(In thousands, except share and per share amounts)
(Unaudited)

	Three Months Ended March 31,	
	2021	**2020**
Operating expenses:		
Research and development	$ 28,683	$ 20,901
Selling, general and administrative	20,600	8,486
Total operating expenses	49,283	29,387
Loss from operations	(49,283)	(29,387)
Interest income	9	789
Loss before (provision) benefit for income taxes	(49,274)	(28,598)
(Provision) benefit for income taxes	(210)	2,179
Net loss	$ (49,484)	$ (26,419)
Net loss per share attributable to common shareholders—basic and diluted	$ (0.72)	$ (0.48)
Weighted average common shares outstanding—basic and diluted	68,269,486	55,322,690

KINIKSA PHARMACEUTICALS, LTD.
SELECTED CONSOLIDATED BALANCE SHEET DATA
(In thousands)
(Unaudited)

	As of	
	March 31, 2021	**December 31, 2020**
Cash, cash equivalents, and short-term investments	$ 264,025	$ 323,482
Working capital	240,111	301,403
Total assets	311,158	349,464
Accumulated deficit	(566,957)	(517,473)
Total shareholders' equity	270,696	311,935

Source: Kiniksa Pharmaceuticals, Ltd.