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

August 3, 2021

- Q2 2021 ARCALYST® (rilonacept) net revenue of $7.7 million with greater than 100 prescribing physicians -

- Phase 3 data from mavrilimumab clinical trial in COVID-19-related ARDS on-track for Q1 2022; new Phase 2 data demonstrate persistent clinical effect through Day 90 -

- Initiation of KPL-404 Phase 2 proof-of-concept clinical trial expected in Q4 2021 -

- Conference call and webcast scheduled for 8:30 a.m. ET today -

HAMILTON, Bermuda, Aug. 03, 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 second quarter 2021 financial results and recent corporate and portfolio activity.

"Our commercial team is doing a superb job with the launch of ARCALYST in recurrent pericarditis," said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. "Feedback received from physicians and patients highlights the transformational potential of ARCALYST for patients suffering from recurrent pericarditis. We have seen strong uptake and look forward to using this momentum to accelerate broader adoption across this underserved population."

"We are executing on the development of our clinical-stage pipeline," said John F. Paolini, MD, PhD, Chief Medical Officer of Kiniksa. "The potential broad utility of mavrilimumab across multiple indications is increasingly promising, and we expect data from our Phase 3 clinical trial in COVID-19-related ARDS in the first quarter of 2022. We are enrolling a Phase 2b clinical trial of vixarelimab in prurigo nodularis and believe vixarelimab has the potential to make a meaningful impact on these patients' lives by addressing both the pruritus and the skin nodules associated with this devastating disease. KPL-404, our anti-CD40 program, has potential across a range of autoimmune diseases, and we plan to initiate a Phase 2 proof-of-concept clinical trial in rheumatoid arthritis in the fourth quarter of this year."

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

- ARCALYST net revenue in the second quarter of 2021 was $7.7 million.
- The second quarter of 2021 represented Kiniksa's first quarter of ARCALYST sales. ARCALYST became commercially available through Kiniksa on April 1st, 2021.
- Greater than 100 physicians who did not participate in Phase 3 RHAPSODY prescribed ARCALYST to at least one recurrent pericarditis patient.
- Greater than 65% of all recurrent pericarditis enrollments came from physicians practicing outside of the 12 Phase 3 RHAPSODY clinical trial sites in the U.S.
- Greater than 90% of completed new patient enrollment cases were approved for coverage under the payer medical exceptions process.
- Kiniksa continues to expect that the majority of payers will establish coverage policies within six months and that almost all payers will update their coverage policies within a year from launch.
- Kiniksa OneConnect™ provided access, initiation and ongoing support for existing ARCALYST cryopyrin-associated periodic syndromes (CAPS) and deficiency of IL-1 receptor antagonist (DIRA) patients transitioning to Kiniksa product as well as new ARCALYST patients for all approved indications.

Mavrilimumab (monoclonal antibody inhibitor targeting GM-CSFRα)

- Kiniksa's interactions with the U.S. Food and Drug Administration (FDA) resulted in defined paths for Phase 3 development of mavrilimumab in COVID-19-related acute respiratory distress syndrome (ARDS) and in giant cell arteritis (GCA).
- Kiniksa today announced additional data from the Phase 2 portion of the Phase 2/3 clinical trial of mavrilimumab in patients with severe COVID-19-related ARDS.
    - Follow-up overall survival data from the cohort of non-mechanically ventilated patients through Day 90 demonstrated persistent clinical effect, confirming and extending the previously-reported Day 29 data.
    - Day 29 data from the cohort of mechanically-ventilated patients did not show a reduction in death.

- Kiniksa continues to enroll non-mechanically ventilated patients in the Phase 3 clinical trial of mavrilimumab in COVID-19-related ARDS.
    - Kiniksa discontinued enrolling mechanically-ventilated patients in the Phase 3 trial.
    - Kiniksa expects data from the Phase 3 trial in non-mechanically-ventilated patients in the first quarter of 2022.

Vixarelimab (monoclonal antibody inhibitor of signaling through OSMRβ)

- Kiniksa continues to enroll patients in 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 the CD40 and CD154 interaction)

- Kiniksa plans to initiate a placebo-controlled Phase 2 proof-of-concept clinical trial of KPL-404 in rheumatoid arthritis in the fourth quarter of 2021.
    - The planned trial is designed to provide safety data, pharmacokinetic characterization, and efficacy of chronic SC dosing over 12 weeks.
    - Rheumatoid arthritis was selected for demonstration of KPL-404 proof of concept as it is a well-characterized autoimmune disease with decades of published clinical data across diverse mechanistic classes, allowing for objective evaluation in established endpoints.
    - The pharmacokinetic lead-in of the planned trial supports characterization of chronic administration of KPL-404 in a patient population and provides optionality to evaluate the therapeutic potential of KPL-404 across a range of other autoimmune diseases with pathologies believed to be mediated by the CD40-CD154 pathway.

Upcoming Scientific Conference Presentations

- Kiniksa plans to present additional data from RHAPSODY, the pivotal Phase 3 trial of rilonacept, as well as the study design of the RESONANCE Registry, at the European Society of Cardiology virtual congress, which will be held August 27, 2021 through August 30, 2021. Details of the poster presentations are as follows:
    - Antonio Brucato, MD, Department of Biomedical and Clinical Science, University of Milan, Fatebenefratelli Hospital, Milan, will present a poster entitled, *Health-Related Quality of Life in Patients with Recurrent Pericarditis: Results from RHAPSODY, a Phase 3 Study of Rilonacept*.
    - Alison Reid, PhD, Kiniksa Pharmaceuticals Corp., will present a poster entitled, *RESONANCE Registry: Rationale and Design of the Retrospective and Prospective Longitudinal, Observational Registry in Pediatric and Adult Patients with Recurrent Pericarditis.*

Financial Results

- Total revenue in the second quarter of 2021 was $7.7 million. Second quarter 2021 revenue is related to product sales for ARCALYST, which became commercially available through Kiniksa on April 1, 2021. Kiniksa did not generate product revenue in the second quarter of 2020.
- Net loss for the second quarter of 2021 was $41.6 million, compared to a net loss of $37.5 million for the second quarter of 2020.
- Total operating expenses for the second quarter of 2021 were $48.3 million, compared to $31.9 million for the second quarter of 2020.
    - Non-cash, share-based compensation expense for the second quarter of 2021 was $5.7 million, compared to $4.9 million for the second quarter of 2020.
- As of June 30, 2021, the company had cash, cash equivalents and short-term investments of $225.9 million and no debt.

Financial Guidance

- Kiniksa expects ARCALYST net revenue for the third quarter of 2021 to be between $9.0 million and $10.0 million.
- Kiniksa expects that its cash, cash equivalents and short-term investments will fund its current operating plan into 2023.

Conference Call Information

Kiniksa will host a conference call and webcast at 8:30 a.m. Eastern Time on Tuesday, August 3, 2021 to discuss second quarter 2021 financial results and recent corporate and portfolio activity. Individuals interested in participating in the call should dial (866) 614-0636 (U.S. and Canada) or (409) 231-2053 (international) using conference ID number 1485623. To access the webcast, please visit the Investors and Media section of Kiniksa's website at www.kiniksa.com. The archived webcast will be available on Kiniksa's website for 14 days beginning approximately one hour after the call has completed.

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 ARCALYST
ARCALYST is a weekly, subcutaneously-injected recombinant dimeric fusion protein that blocks interleukin-1 alpha and interleukin-1 beta signaling. ARCALYST was discovered by Regeneron and is approved by the FDA for recurrent pericarditis, CAPS, including Familial Cold Autoinflammatory Syndrome and Muckle-Wells Syndrome, and 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. The European Commission granted Orphan Drug designation to ARCALYST for the treatment of idiopathic 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. ARCALYST should not be started if you have an infection that keeps coming back.
- 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, and the Phase 2 clinical trial achieved both the primary and secondary efficacy endpoints with statistical significance. Kiniksa continues to evaluate mavrilimumab in COVID-19-related ARDS. 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 expectations regarding payers establishing coverage policies for ARCALYST; our beliefs about the future commercial demand for ARCALYST; our beliefs about the timing and results of the final data from the Phase 3 portion of our clinical trial for mavrilimumab in non-mechanically-ventilated patients in the first quarter of 2022; expected timing and design of clinical trials, including (i) the enrollment of patients in and the timing of results from our phase 3 clinical trial of mavrilimumab in COVID-19-related ARDS, (ii) the initiation our Phase 2 proof-of-concept trial of KPL-404 in rheumatoid arthritis by the fourth quarter of 2021, (iii) the enrollment of patients the Phase 2b portion of our clinical trial of vixarelimab in prurigo nodularis and (iv) 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; our belief that all of our product candidates offer the potential for differentiation; our ability to execute on our clinical stage pipeline; our expectations regarding ARCALYST net revenue for the third quarter of 2021; our plans to present at any future medical conferences; and our expectations 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 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, 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 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; our reliance on third parties 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 on 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.

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 June 30,		Six Months Ended June 30,	
	2021	**2020**	**2021**	**2020**
Revenue:				
Product revenue, net	$ 7,704	$ —	$ 7,704	$ —
Costs and operating expenses:				
Cost of goods sold	2,466	—	2,466	—
Research and development	23,945	22,324	52,628	43,225
Selling, general and administrative	21,848	9,536	42,448	18,022
Total operating expenses	48,259	31,860	97,542	61,247
Loss from operations	(40,555)	(31,860)	(89,838)	(61,247)
Interest income	6	266	15	1,055
Loss before provision for income taxes	(40,549)	(31,594)	(89,823)	(60,192)
Provision for income taxes	(1,014)	(5,875)	(1,224)	(3,696)

Net loss	$ (41,563)	$ (37,469)	$ (91,047)	$ (63,888)
Net loss per share attributable to common shareholders —basic and diluted	$ (0.61)	$ (0.65)	$ (1.33)	$ (1.13)
Weighted average common shares outstanding—basic and diluted	68,395,703	57,914,105	68,332,943	56,618,397

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

	As of	
	June 30, 2021	**December 31, 2020**
Cash, cash equivalents, and short-term investments	$ 225,866	$ 323,482
Working capital	206,028	301,403
Total assets	279,208	349,464
Accumulated deficit	608,520	517,473
Total shareholders' equity	235,785	311,935

Source: Kiniksa Pharmaceuticals, Ltd.