Kiniksa Pharmaceuticals Reports Third Quarter 2021 Financial Results and Recent Portfolio Execution

November 1, 2021

- Q3 2021 ARCALYST® (rilonacept) net revenue of $12.1 million -
- ARCALYST prescribed by more than 200 physicians for recurrent pericarditis since approval -
- Data from Phase 3 trial of mavrilimumab in COVID-19-related ARDS expected in Q1 2022 -
- Data from Phase 2b trial of vixarelimab in prurigo nodularis expected in 2H 2022 -
- Conference call and webcast scheduled for 8:30 am ET today -

HAMILTON, Bermuda, Nov. 01, 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 third quarter 2021 financial results and recent portfolio execution.

"We are delighted with the ARCALYST launch performance in recurrent pericarditis. The continued momentum underscores the dedication of our first-rate team as well as the unmet need for patients with this debilitating disease," said Sanj K. Patel, Chairman and Chief Executive Officer of Kiniksa. "In addition to our ongoing commercial efforts, we are executing on our clinical-stage pipeline. Mavrilimumab demonstrated potentially best-in-class mortality reduction in a Phase 2 study in non-mechanically ventilated patients with COVID-19-related ARDS, and Phase 3 data are on track for the first quarter of 2022. We are also conducting a dose-ranging Phase 2b study of vixarelimab in prurigo nodularis with data expected in the second half of 2022. Furthermore, we expect to initiate a Phase 2 proof-of-concept trial of KPL-404 in rheumatoid arthritis by the end of 2021."

Portfolio Execution

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

- ARCALYST net revenue for the third quarter of 2021 was $12.1 million.
- More than 200 physicians have prescribed ARCALYST for recurrent pericarditis since approval.
- More than 90% of completed patient enrollment cases for recurrent pericarditis were approved for coverage.
- Kiniksa has achieved broad access for its recurrent pericarditis target population in the United States, with greater than 190 million lives receiving favorable coverage.
- Two thirds of ARCALYST prescriptions for recurrent pericarditis have been written for 12 months of therapy.
- Recent data from the ongoing Phase 3 clinical trial of ARCALYST in recurrent pericarditis (RHAPSODY) showed median duration of continuous ARCALYST therapy at the 1-year anniversary of the Long-Term Extension portion of the trial of approximately 20 months.

Mavrilimumab (monoclonal antibody inhibitor targeting GM-CSFRα)

- Mavrilimumab has demonstrated broad utility across multiple indications including COVID-19-related acute respiratory distress syndrome (ARDS) and giant cell arteritis (GCA).
- Kiniksa expects data from the Phase 3 clinical trial of mavrilimumab in hospitalized non-mechanically ventilated patients with COVID-19-related ARDS in the first quarter of 2022.
    - The primary efficacy endpoint for the Phase 3 trial is the proportion of patients alive and without mechanical ventilation at Day 29. A key secondary efficacy endpoint is mortality at Day 29.
    - Phase 2 clinical data in hospitalized non-mechanically ventilated patients with COVID-19-related ARDS showed mavrilimumab reduced the risk of mechanical ventilation or death by 65% versus placebo at Day 29.

Vixarelimab (monoclonal antibody inhibitor of signaling through OSMRβ)

- Kiniksa expects data from the dose-ranging Phase 2b clinical trial of vixarelimab in prurigo nodularis in the second half of 2022.
    - The primary efficacy endpoint for the Phase 2b trial is the percent change from baseline in the weekly-average Worst-Itch Numeric Rating Scale (WI-NRS) at Week 16.

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

- Kiniksa plans to initiate a Phase 2 clinical trial of KPL-404 in rheumatoid arthritis in the fourth quarter of 2021.
    - The Phase 2 trial is designed to evaluate the efficacy, dose response, pharmacokinetics, and safety of chronic

subcutaneous dosing in rheumatoid arthritis over 12 weeks.
    - The pharmacokinetic lead-in of the Phase 2 trial is designed to provide 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.

Financial Results

- Total net revenue for ARCALYST product sales in the third quarter of 2021 was $12.1 million.
- Net loss for the third quarter of 2021 was $30.5 million, compared to a net loss of $43.8 million for the third quarter of 2020.
- Total operating expenses for the third quarter of 2021 were $42.8 million, compared to $43.2 million for the third quarter of 2020.
    - Non-cash, share-based compensation expense for the third quarter of 2021 was $6.2 million, compared to $5.6 million for the third quarter of 2020.
- As of September 30, 2021, the company had cash, cash equivalents, and short-term investments of $200.2 million and no debt.

Financial Guidance

- Kiniksa expects ARCALYST net revenue for the fourth quarter of 2021 to be between $16.0 million and $17.0 million.
- Kiniksa continues to expect that its cash, cash equivalents and short-term investments will fund its current operating plan into 2023.

Upcoming Scientific Conference Presentations

- Presentations of new data from several of Kiniksa's pipeline programs are planned at the upcoming American College of Rheumatology (ACR) Convergence 2021, being held virtually from November 3, 2021 to November 9, 2021. In addition, encore presentations of RHAPSODY data are planned. Details of the pipeline presentations are as follows:
    - Marc Corbera-Bellalta, Ph.D., University of Barcelona, will give an oral presentation titled, *Transcriptomic Changes Induced by Mavrilimumab versus Tocilizumab in ex-vivo Cultured Arteries from Patients with Giant-cell Arteritis.*
    - Sebastian Unizony, MD, Massachusetts General Hospital, will present a poster entitled, *Utility of CRP and ESR in the Assessment of Giant Cell Arteritis Relapse in a Phase 2 Trial of Mavrilimumab.*
    - Manoj Samant, Ph.D., Kiniksa Pharmaceuticals, will present a poster entitled, *Safety, Tolerability, Pharmacokinetics, Receptor Occupancy, and Suppression of T-cell-Dependent Antibody Response in a Phase 1 Study with KPL-404, an anti-CD40 Monoclonal Antibody.*
- Kiniksa plans to present details on the RESONANCE registry in pediatric and adult patients with recurrent pericarditis at the American Heart Association (AHA) Scientific Sessions 2021, being held virtually from November 13, 2021 to November 15, 2021. Details of the presentation are as follows:
    - Sushil Allen Luis, MD, Mayo Clinic, will present a poster entitled, *Including the Patient's Perspective in the Planning of Data Collection in the Recurrent Pericarditis Registry, RESONANCE.*

Conference Call Information
Kiniksa will host a conference call and webcast at 8:30 am ET on Monday, November 1, 2021 to discuss third quarter 2021 financial results and to provide a corporate update.

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 1699563. To access the webcast, please visit the Investors and Media section of Kiniksa'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 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, 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 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 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; expected timing and design of our clinical trials, including (i) the timing of data from the Phase 3 portion of our clinical trial for mavrilimumab in COVID-19-related ARDS in the first quarter of 2022, (ii) the initiation of our Phase 2 proof-of-concept trial of KPL-404 in rheumatoid arthritis by the end of 2021 and (iii) the timing of data from the dose-ranging Phase 2b clinical trial of vixarelimab in prurigo nodularis in the second half of 2022; 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; 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 fourth 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; 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 (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 September 30,		Nine Months Ended September 30,	
	2021	2020	2021	2020
Revenue:				
Product revenue, net	$ 12,095	$ —	$ 19,799	$ —
Costs and operating expenses:				
Cost of goods sold	2,767	—	5,233	—
Research and development	19,236	31,419	71,864	74,644
Selling, general and administrative	20,759	11,799	63,207	29,821
Total operating expenses	42,762	43,218	140,304	104,465
Loss from operations	(30,667)	(43,218)	(120,505)	(104,465)
Interest income	5	49	20	1,104
Loss before benfit (provision) for income taxes	(30,662)	(43,169)	(120,485)	(103,361)
Benefit (provision) for income taxes	118	(667)	(1,106)	(4,363)
Net loss	$ (30,544)	$ (43,836)	$ (121,591)	$ (107,724)
Net loss per share attributable to common shareholders—basic and diluted	$ (0.44)	$ (0.66)	$ (1.78)	$ (1.80)
Weighted average common shares outstanding—basic and diluted	68,662,673	65,958,513	68,444,061	59,754,495

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

	As of	
	September 30, 2021	December 31, 2020

Cash, cash equivalents, and short-term investments	$	200,183	$	323,482
Working capital		183,085		301,403
Total assets		252,859		349,464
Accumulated deficit		639,064		517,473
Total shareholders' equity		214,048		311,935