Kiniksa Reports First Quarter 2019 Financial Results and Highlights Recent Corporate and Pipeline Activity

May 2, 2019

–First patient dosed in Phase 2a trial of KPL-716 in prurigo nodularis
– Upcoming presentations at SID, ISPOR and EULAR scientific conferences on KPL-716, rilonacept and mavrilimumab, respectively
– Acquisition of KPL-404 (anti-CD40) complete; IND filing with FDA anticipated in 2H 2019

HAMILTON, Bermuda, May 02, 2019 (GLOBE NEWSWIRE) -- Kiniksa Pharmaceuticals, Ltd. (Nasdaq: KNSA) ("Kiniksa"), a biopharmaceutical company focused on discovering, acquiring, developing and commercializing therapeutic medicines for patients with significant unmet medical need, today reported first quarter 2019 financial results and highlighted recent corporate and pipeline activity.

"The first quarter of 2019 was marked by the advancement of our clinical-stage pipeline," said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. "Progress included the initiation of our Phase 2a trial for KPL-716 in prurigo nodularis as well as the continued enrollment of our pivotal Phase 3 trial for rilonacept in recurrent pericarditis and our global Phase 2 trial for mavrilimumab in giant cell arteritis. Going forward, we expect to build upon this momentum with focused investment behind our product candidates, including commercial readiness activities for rilonacept, and continued execution."

Clinical-Stage Pipeline Activity

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

- Kiniksa is advancing rilonacept for the potential treatment of recurrent pericarditis, a painful autoinflammatory cardiovascular disease with an estimated U.S. prevalent population of approximately 40,000 patients seeking and receiving medical treatment.
    - Kiniksa is enrolling RHAPSODY, a global, randomized-withdrawal (RW) design, pivotal Phase 3 clinical trial of rilonacept in subjects with recurrent pericarditis. The primary efficacy endpoint is time-to-first-adjudicated pericarditis-recurrence in the RW period. Top-line data are expected in the second half of 2020.
    - In March 2019, Kiniksa presented interim data from an open-label Phase 2 clinical trial of rilonacept in subjects with recurrent pericarditis at the American College of Cardiology's (ACC) 68 th Annual Scientific Session. As of the January 2019 data cutoff date, all symptomatic recurrent pericarditis and post-pericardiotomy syndrome (PPS) subjects showed reductions in both C-reactive protein (CRP) and reported pain as well as improvement in quality of life scores. Additionally, CRP and reported pain remained low for all asymptomatic recurrent pericarditis and PPS subjects. Rilonacept has been generally well-tolerated in the study, with adverse events (AEs) consistent with the U.S. Food and Drug Administration (FDA)-approved label of rilonacept for the treatment of Cryopyrin-Associated Periodic Syndromes (CAPS). The most common AEs were injection site reactions. There was one treatment-related serious AE which resulted in discontinuation: a skin abscess that responded to medical treatment. Enrollment has closed and Kiniksa expects to complete the trial in the second half of 2019.
- Kiniksa is advancing its commercial readiness activities for rilonacept, including the expansion of its commercial and medical affairs teams and raising awareness of unmet need through payer, physician and patient engagement.
- Kiniksa plans to present new data from a systematic literature review highlighting the economic and humanistic burden of recurrent pericarditis at the International Society for Pharmacoeconomics and Outcomes Research (ISPOR) in May 2019.

Mavrilimumab (monoclonal antibody inhibitor targeting GM-CSFRα)

- Kiniksa is advancing mavrilimumab for the potential treatment of giant cell arteritis (GCA), a chronic inflammatory disease of medium-large blood vessels with an estimated U.S. prevalence of approximately 75,000 to 150,000 patients.
    - Kiniksa is enrolling a global Phase 2 proof-of-concept clinical trial of mavrilimumab in subjects with GCA. The primary efficacy endpoint is time-to-first-flare. Top-line data are expected in the second half of 2020.
- In April 2019, Kiniksa presented the design of the global Phase 2 clinical trial of mavrilimumab in GCA at the 19thInternational Vasculitis Workshop.
- Kiniksa plans to present preclinical data on the granulocyte macrophage colony stimulating factor (GM-CSF) and Type 1 T helper (T_H1) signaling pathways in temporal arteries of GCA patients at the European League Against Rheumatism's (EULAR) European Congress of Rheumatology in June 2019.

KPL-716 (monoclonal antibody inhibitor of signaling through OSMRβ)

- Kiniksa is advancing KPL-716 for the potential treatment of a variety of pruritic diseases, including prurigo nodularis, a chronic inflammatory skin condition with an estimated U.S. prevalence of approximately 300,000 patients.
    - Kiniksa is enrolling a Phase 2a clinical trial of KPL-716 in subjects with prurigo nodularis. The primary efficacy endpoint is percent change from baseline in weekly average Worst-Itch Numeric Rating Scale (WI-NRS). Top-line data are expected in the first half of 2020.
    - Kiniksa plans to initiate an exploratory Phase 2 clinical trial in diseases characterized by chronic pruritus in the first half of 2019. The trial is designed to identify chronic pruritic conditions where interleukin-31 (IL-31) and/or oncostatin M (OSM) may be playing a role and to investigate the efficacy, safety and tolerability of KPL-716 in reducing the moderate-to-severe pruritus experienced by these subjects. Top-line data are expected in the second half of 2020.
    - Kiniksa completed enrollment in a repeated-single-dose Phase 1b clinical trial of KPL-716 in subjects with moderate-to-severe atopic dermatitis experiencing moderate-to-severe pruritus. The study is designed to provide safety and exploratory data on both pruritus and inflammation disease response markers. Top-line data are expected in the second half of 2019.
- In April 2019, Kiniksa presented encore KPL-716 Phase 1a/1b clinical data at Revolutionizing Atopic Dermatitis (RAD).
- Kiniksa plans to present clinical and preclinical data from KPL-716 at the Society for Investigational Dermatology (SID) in May 2019. The data provides further scientific rationale for targeting oncostatin M receptor beta (OSMRβ) and dual pathway inhibition of IL-31 and OSM via KPL-716 for the potential treatment of chronic pruritic diseases.

Preclinical Pipeline Activity

- Kiniksa is progressing its preclinical activities with KPL-404, a monoclonal antibody inhibitor of the CD40 co-stimulatory receptor, in T-cell-dependent, B-cell-mediated disorders. Kiniksa expects to file an investigational new drug (IND) application with the FDA in the second half of 2019 and initiate a Phase 1 clinical trial in the first half of 2020.
    - In April 2019, Kiniksa presented preclinical KPL-404 data at the Keystone Symposia, Antibodies as Drugs: New Horizons in the Therapeutic Use of Engineered Antibodies. The data showed favorable pharmacokinetic and pharmacodynamic profiles, including engagement of CD40 target and block of antigen-specific primary and secondary antibody responses in a T-cell dependent antibody response cynomolgus monkey model.
    - In March 2019, Kiniksa closed the acquisition of Primatope Therapeutics, Inc. (“Primatope”), the company that owned or controlled the intellectual property related to KPL-404.
- Kiniksa is evaluating the progression of KPL-045, a monoclonal antibody inhibitor of the CD30 ligand co-stimulatory molecule, pending preclinical data from the program in the context of the company’s portfolio.

Financial Results and Recent Corporate Activity

- In the first quarter of 2019, Kiniksa completed a public offering of 2,816,110 Class A common shares (inclusive of the underwriters’ overallotment option exercise) at a public offering price of $18.26 per share and a concurrent private placement of 2,000,000 Class A1 common shares, at the same offering price per share. Aggregate net proceeds to Kiniksa were approximately $83.0 million.
- For the first quarter of 2019, Kiniksa reported a net loss of $65.8 million, compared to a net loss of $16.0 million for the first quarter of 2018.
- Total operating expenses for the first quarter of 2019 totaled $67.6 million compared to $16.3 million for the first quarter of 2018. Non-cash share-based compensation expense totaled $2.9 million for the first quarter of 2019, compared to $0.6 million for the first quarter of 2018.
    - Total operating expenses for the first quarter of 2019 included $18.0 million related to the acquisition of Primatope and $10.0 million related to the first patient dosed in the Phase 2a clinical trial of KPL-716 in subjects with prurigo nodularis.
- As of March 31, 2019, the company had cash, cash equivalents and short-term investments of $326.5 million and no outstanding debt.

Financial Guidance

- Kiniksa expects that its cash, cash equivalents and short-investments will fund its current operating plan into 2021.

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 has a pipeline of product candidates across various stages of development, focused on autoinflammatory and autoimmune conditions. For more information, please visit www.kiniksa.com.

About Rilonacept
Rilonacept is a weekly, subcutaneously-injected, recombinant fusion protein that blocks interleukin-1α (IL-1α) and interleukin 1β (IL-1β) signaling. Rilonacept was discovered and developed by Regeneron Pharmaceuticals, Inc. and is approved by the FDA under the brand name ARCALYST® for

the treatment of CAPS, which includes Familial Cold Autoinflammatory Syndrome and Muckle-Wells Syndrome. 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. Kiniksa exclusively licensed rilonacept from Regeneron for recurrent pericarditis and certain other indications. Rilonacept in recurrent pericarditis is an investigational drug.

About Mavrilimumab
Mavrilimumab is an investigational fully-human monoclonal antibody that is designed to antagonize GM-CSF signaling by binding to the alpha subunit of the GM-CSF receptor. Kiniksa's lead indication for mavrilimumab is giant cell arteritis, an inflammatory disease of blood vessels.

About KPL-716
KPL-716 is an investigational fully-human monoclonal antibody that targets OSMRβ, which mediates signaling of IL-31 and OSM, two key cytokines implicated in pruritus, inflammation and fibrosis. Kiniksa believes KPL-716 to be the only monoclonal antibody in development that targets both pathways simultaneously.

About KPL-404
KPL-404 is an investigational humanized monoclonal antibody that is designed to inhibit the CD40-CD40-ligand interaction, a key T-cell co-stimulatory signal critical for B-cell maturation and immunoglobulin class switching. Dysregulation of the CD40-CD40L pathway has been implicated in multiple autoimmune disease pathologies such as Systemic Lupus Erythematosus, Rheumatoid Arthritis, Sjogren's Syndrome and Grave's Disease.

About KPL-045
KPL-045 is an investigational fully-human monoclonal antibody that is designed to inhibit the CD30-CD30 ligand interaction, a co-stimulatory signal involved in activating and sustaining memory T-cells.

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 execution, pre-commercial activities and investment focus, and potential results therefrom; expected timeframe for funding our operating plan with current cash, cash equivalents and short-term investments; plans and timing for completion of clinical trials; proposed indications for the investigation of our product candidates; estimated disease prevalence; plans and timing to report or present top-line clinical trial, pre-clinical and other data; plans and timing for the submission of investigational new drug and other applications and submissions to regulatory authorities; and plans and timing to advance additional pipeline programs into clinical trials.

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: potential delays or difficulty in enrollment of patients in, and activation of sites for, our clinical trials; potential complications in coordinating among requirements, regulations and guidelines of regulatory authorities across a number of jurisdictions for our global clinical trials; potential delays or difficulty in completing our clinical trials; potential undesirable side effects caused by our product candidates; our potential inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities; potential for changes between final data and any interim "top-line" and preliminary data we announce; impact of additional data from us or other companies; our potential inability to replicate in later clinical trials positive results from our earlier clinical trials; our ability to manufacture drug substance for Phase 1 clinical trials using our facilities; drug substance and/or drug product shortages caused by issues at our third-party manufacturers' facilities; our reliance on certain third parties as the sole source of supply of the drug substance and drug products used in our product candidates; our reliance on third parties to conduct our research, pre-clinical studies, clinical trials, and other trials for our product candidates; we face substantial competition; and our ability to attract and retain qualified personnel.

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 March 12, 2019 and our other reports 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.

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

	As of

	March 31, 2019	December 31, 2018
Cash, cash equivalents, and short-term investments	$ 326,533	$ 307,304
Working capital [1]	298,064	271,196
Total assets	346,159	321,965
Accumulated deficit	(260,046)	(194,225)
Total shareholders' equity	306,519	279,267

[1] We define working capital as current assets less current liabilities.

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

	Three Months Ended March 31,	
	2019	2018
Operating expenses:		
Research and development	$ 59,253	$ 12,630
General and administrative	8,394	3,710
Total operating expenses	67,647	16,340
Loss from operations	(67,647)	(16,340)
Interest income	1,809	305
Loss before benefit for income taxes	(65,838)	(16,035)
Benefit for income taxes	17	53
Net loss	$ (65,821)	$ (15,982)
Net loss per share attributable to common shareholders —basic and diluted	$ (1.27)	$ (6.45)
Weighted average common shares outstanding—basic and diluted	51,758,353	2,478,903

Source: Kiniksa Pharmaceuticals, Ltd.