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

August 12, 2019

– Rilonacept pivotal Phase 3 study dosing patients in the U.S., Australia, Israel and Italy; expect to present final Phase 2 data this year –
– Mavrilimumab global Phase 2 study dosing patients in fifteen countries –
– KPL-716 repeated-single-dose Phase 1b interim data showed rapid and sustained anti-pruritic effect throughout the 12-week treatment period; no meaningful difference from placebo on other efficacy endpoints specific to atopic dermatitis –
– Enrollment in KPL-716 Phase 2a in prurigo nodularis progressing; first patients dosed in exploratory Phase 2 study in diseases characterized by chronic pruritus –

HAMILTON, Bermuda, Aug. 12, 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 second quarter 2019 financial results and highlighted recent corporate and pipeline activity.

"Kiniksa is advancing a pipeline of autoimmune and autoinflammatory product candidates based on validated mechanisms or strong biologic rationale and focused on rare diseases," said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. "Enrollment in our clinical trials continues to progress across our portfolio, and interim KPL-716 repeated-single-dose data support our ongoing Phase 2 development in prurigo nodularis and diseases characterized by chronic pruritus. Additionally, we look forward to clinical readouts from multiple programs over the next twelve months, starting later this year with final data from our rilonacept Phase 2 study."

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 and debilitating autoinflammatory cardiovascular disease.
    - Kiniksa is enrolling RHAPSODY, a randomized-withdrawal (RW) design, pivotal Phase 3 clinical trial of rilonacept in subjects with recurrent pericarditis in the U.S., Australia, Israel and Italy. 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.
    - Kiniksa expects to present final data from an open-label Phase 2 clinical trial of rilonacept in different pericarditis populations later this year.
    - Kiniksa continues to advance its launch readiness activities for rilonacept through expansion of its commercial and medical affairs teams, generation of evidence on unmet need and disease burden, and research and education with payers, physicians and advocacy groups.

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.
    - Kiniksa is enrolling a global Phase 2 proof-of-concept clinical trial of mavrilimumab in subjects with GCA in fifteen countries. The primary efficacy endpoint is time-to-first-flare. Top-line data are expected in the second half of 2020.

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

- Kiniksa announced interim data today from a repeated-single-dose Phase 1b clinical trial of KPL-716 in subjects with moderate-to-severe atopic dermatitis. The data showed a rapid and sustained reduction in pruritus throughout the 12-week treatment period. There was no meaningful benefit of repeated-single-doses of KPL-716 on other efficacy endpoints specific to atopic dermatitis, including Eczema Area and Severity Index (EASI) and Scoring Atopic Dermatitis (SCORAD). There were no serious adverse events. However, there were more atopic dermatitis flares in the KPL-716-treated population versus placebo. All flares were successfully managed with topical corticosteroids. KPL-716 was otherwise well-tolerated by all subjects. Kiniksa has no current plan to invest in atopic dermatitis development.

- Kiniksa is advancing KPL-716 for the potential treatment of a variety of pruritic diseases, including prurigo nodularis, a chronic inflammatory skin condition.
    - 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 is enrolling an exploratory Phase 2 clinical trial in diseases characterized by chronic pruritus. The trial is designed to identify chronic pruritic conditions where signaling through oncostatin M receptor beta (OSMRβ) 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. Kiniksa expects to provide interim data from this study on a cohort-by-cohort basis throughout 2020.

Preclinical Pipeline Activity

- Kiniksa is progressing its preclinical activities with KPL-404, a monoclonal antibody inhibitor of the CD40 co-stimulatory receptor in diseases characterized by T-cell-dependent, B-cell-mediated pathology. Kiniksa expects to file an investigational new drug (IND) application with the U.S. Food and Drug Administration (FDA) later this year.
- Kiniksa is evaluating the progression of KPL-045, a monoclonal antibody inhibitor of the CD30 ligand co-stimulatory molecule, based on preclinical data from the program in the context of the company's portfolio.

Third Quarter 2019 Scientific Conferences

- Kiniksa plans to present at the following scientific conferences in the third quarter of 2019:
    - European Society of Cardiology (ESC) in September 2019; rilonacept Phase 3 methods.
    - American College of Epidemiology (ACE) in September 2019; retrospective claims analysis of recurrent pericarditis epidemiology in the U.S.
    - European Society for Dermatological Research (ESDR) in September 2019; preclinical data analyzing expression of OSMRβ in chronic pruritic diseases.

Financial Results

- For the second quarter of 2019, Kiniksa reported a net loss of $37.2 million, compared to a net loss of $20.3 million for the second quarter of 2018.
- Total operating expenses for the second quarter of 2019 totaled $39.3 million compared to $21.5 million for the second quarter of 2018. Non-cash share-based compensation expense totaled $3.5 million for the second quarter of 2019, compared to $1.1 million for the second quarter of 2018.
- As of June 30, 2019, the company had cash, cash equivalents and short-term investments of $287.4 million and no outstanding debt.

Financial Guidance

- Kiniksa expects that its cash, cash equivalents and short-term 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. (Regeneron) and is approved by the FDA under the brand name ARCALYST® for the treatment of Cryopyrin-Associated Periodic Syndromes (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 granulocyte macrophage colony stimulating factor (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 interleukin-31 (IL-31) and oncostatin M (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: plans and timing to advance our product candidates; proposed indications for the investigation of our product candidates; plans and timing to report or present interim, final and top-line clinical trial, pre-clinical and other data; our conclusions from interim pre-clinical and clinical trial data for KPL-716; plans and timing for the submission of investigational new drug and other applications and submissions to regulatory authorities; and expected timeframe for funding our operating plan with current cash, cash equivalents and short-term investments.

These forward-looking statements are based on management's current plans, estimates or 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 amendments to our clinical trial protocols initiated by us or required by regulatory authorities; 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 or otherwise producing negative, inconclusive or commercially uncompetitive results; potential for changes between final data and any preliminary, interim and "top-line" 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; 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 Quarterly Report on Form 10-Q filed with the Securities and Exchange Commission ("SEC") on May 7, 2019 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 plans, estimates, or expectations 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.

Every Second Counts!™

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

	As of	
	June 30,	**December 31,**

	2019	2018
Cash, cash equivalents, and short-term investments	$ 287,447	$ 307,304
Working capital [1]	266,280	271,196
Total assets	308,137	321,965
Accumulated deficit	(297,237)	(194,225)
Total shareholders' equity	275,197	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 June 30,		Six Months Ended June 30,	
	2019	2018	2019	2018
Operating expenses:				
Research and development	$ 30,848	$ 17,200	$ 90,101	$ 29,831
General and administrative	8,441	4,327	16,835	8,036
Total operating expenses	39,289	21,527	106,936	37,867
Loss from operations	(39,289)	(21,527)	(106,936)	(37,867)
Interest income	1,724	1,066	3,533	1,371
Loss before benefit for income taxes	(37,565)	(20,461)	(103,403)	(36,496)
Benefit for income taxes	374	202	391	255
Net loss	$ (37,191)	$ (20,259)	$ (103,012)	$ (36,241)
Net loss per share attributable to common shareholders —basic and diluted	$ (0.68)	$ (1.11)	$ (1.94)	$ (3.45)
Weighted average common shares outstanding—basic and diluted	54,475,476	18,328,402	53,225,710	10,492,474

Source: Kiniksa Pharmaceuticals, Ltd.