Kiniksa Pharmaceuticals Logo

Kiniksa Reports Third Quarter 2018 Financial Results and Pipeline Progress

November 1, 2018

– Rilonacept pivotal Phase 3 clinical trial initiating this year
– Mavrilimumab pre-IND meeting complete; plans for global Phase 2 clinical trial advancing
– KPL-716 advancing into multiple chronic pruritic diseases, starting with a planned Phase 2a/2b in prurigo nodularis in Q1 2019
– KPL-716 repeated-single-dose Phase 1b results expected in 2H 2019

HAMILTON, Bermuda, Nov. 01, 2018 (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 third quarter 2018 financial results and pipeline progress.

"Continued solid execution through the third quarter positions us to deliver multiple near-term milestones," said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. "We are advancing our pipeline as planned. In the fourth quarter of 2018, we expect to initiate our pivotal Phase 3 study for rilonacept in recurrent pericarditis and advance our plans for a global Phase 2 proof-of-concept trial for mavrilimumab in giant cell arteritis. Additionally, we are preparing KPL-716 to be evaluated in multiple chronic pruritic conditions, starting with a Phase 2a/2b study in prurigo nodularis planned for the first quarter of 2019."

Third Quarter 2018 and Recent Pipeline Progress

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

- Kiniksa plans to initiate a single, pivotal, placebo-controlled, randomized-withdrawal design, global Phase 3 clinical trial of rilonacept in approximately 60 subjects with symptomatic recurrent pericarditis later this year.

- Kiniksa continues to enroll subjects into the rilonacept open-label Phase 2 proof-of-concept clinical trial in order to gain further experience with rilonacept in different pericarditis populations. The response to treatment continues to be consistent with the first subjects who entered the trial in terms of rapid reduction in reported pain and inflammation. Kiniksa plans to report additional open-label Phase 2 proof-of-concept clinical trial data and close enrollment in the trial later this year.

Mavrilimumab (monoclonal antibody inhibitor targeting GM-CSFRα)

- Kiniksa completed its pre-investigational new drug application (IND) meeting with the U.S. Food and Drug Administration (FDA) and believes its Phase 2 clinical trial development plan for mavrilimumab in giant cell arteritis (GCA) is consistent with the feedback it received from the FDA.

  - The double-blind, randomized, placebo-controlled, global Phase 2 proof-of-concept clinical trial of mavrilimumab for the treatment of GCA will enroll approximately 60 subjects with new-onset and refractory disease. Subjects will be randomized 3:2 to mavrilimumab 150 mg or placebo injected subcutaneously once every 2 weeks co-administered with a corticosteroid taper. Treatment duration will be 26 weeks. The primary efficacy endpoint will be time to first flare.

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

- Kiniksa plans to advance KPL-716 into multiple chronic pruritic diseases, starting with an adaptive design Phase 2a/2b clinical trial in prurigo nodularis, in the first quarter of 2019.

- Kiniksa announced Phase 1a/1b clinical data for KPL-716. In this First-in-Human clinical trial, single intravenous (IV) and subcutaneous (SC) doses of KPL-716 were well-tolerated in both adult healthy volunteers and adult subjects with moderate-to-severe atopic dermatitis experiencing moderate-to-severe pruritus. Additionally, a single dose of KPL-716 7.5 mg/kg IV (n=10) versus placebo IV (n=10) provided evidence of target engagement and an early signal of efficacy in reducing pruritus as well as inflammation and disease severity in subjects with moderate-to-severe atopic dermatitis. Preliminary pharmacokinetic modeling supports testing once every 2 weeks and once-monthly SC dosing regimens in a Phase 2b.

- Kiniksa continues to enroll a repeated-single-dose cohort as an additional part of the Phase 1b clinical trial in the U.S. and Canada. In this double-blind, randomized, placebo-controlled study, approximately 50 subjects with moderate-to-severe atopic dermatitis experiencing moderate-to-severe pruritus receive KPL-716 360 mg or placebo via SC injection once weekly for 12 weeks. The study is designed to evaluate safety and chronic efficacy data on both pruritus and inflammatory disease response markers by providing similar and longer-term exposures compared to the single-dose IV cohort of the Phase 1b clinical trial. Results are expected in the second half of 2019.

KPL-045 (monoclonal antibody inhibitor of the CD30L co-stimulatory molecule)

- Kiniksa continues its preclinical activities with KPL-045 in inflammatory diseases driven by T-cell-dependent autoantibody

generation and dysregulated T_H effector memory responses and expects to file an IND application with the FDA in the second half of 2019.

KPL-404 (monoclonal antibody inhibitor of the CD40 co-stimulatory receptor)

- Kiniksa continues its preclinical activities with KPL-404 in T-cell-dependent, B-cell-mediated disorders and expects to file an IND application with the FDA in the second half of 2019.

Third Quarter 2018 Financial Results

- For the third quarter of 2018, Kiniksa reported a net loss of $24.4 million, compared to a net loss of $16.0 million for the third quarter of 2017.
- Total operating expenses for the third quarter of 2018 totaled $26.2 million compared to $16.2 million for the third quarter of 2017. Non-cash share-based compensation expense totaled $1.5 million for the third quarter of 2018, compared to $0.3 million for the third quarter of 2017.
- As of September 30, 2018, the company had cash, cash equivalents and short-term investments of $337.9 million and no outstanding debt.

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, currently 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 IL-1α and IL-1β signaling. Rilonacept was approved by the FDA for the treatment of cryopyrin-associated periodic syndrome (CAPS), which includes cold auto-inflammatory syndrome and Muckle-Wells syndrome, and has been commercially available in the U.S. from Regeneron Pharmaceuticals, Inc. for this indication since 2008. Kiniksa's lead indication for rilonacept is recurrent pericarditis, which is a recurring painful inflammation of the pericardium.

About Mavrilimumab
Mavrilimumab is an investigational fully-human monoclonal antibody that antagonizes 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 oncostatin M receptor beta (OSMRβ), which mediates signaling of 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-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.

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. Kiniksa obtained a license to conduct research and development on KPL-404 from Primatope Therapeutics, Inc. in 2017 and has an exclusive option to acquire all outstanding capital stock of Primatope, which, subject to extension and the payment of specified extension fees, is exercisable until January 2019.

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 and potential impact thereof; plans and timing for initiation of new clinical trials; the consistency of our Phase 2 clinical trial development plan for mavrilimumab in GCA with FDA feedback; potential designs of our new clinical trials; proposed indications for the investigation of our product candidates; plans and timing to report clinical trial 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 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 Quarterly Report on Form 10-Q for the period ended June 30, 2018 filed with the Securities and Exchange Commission (“SEC”) on August 6, 2018 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.

Every Second Counts!™

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

	As of September 30, 2018	December 31, 2017
Cash, cash equivalents, and short-term investments	$ 337,863	$ 45,555
Working capital	313,507	29,674
Total assets	347,101	47,492
Accumulated deficit	(151,645)	(90,998)
Total shareholders' equity (deficit)	318,913	(89,708)

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

	Three Months Ended September 30,		Nine Months Ended September 30,	
	2018	2017	2018	2017
Operating expenses:				
Research and development	$ 20,644	$ 14,008	$ 50,475	$ 26,426
General and administrative	5,515	2,241	13,550	6,263
Total operating expenses	26,159	16,249	64,025	32,689
Loss from operations	(26,159)	(16,249)	(64,025)	(32,689)
Interest income	1,622	169	2,992	396
Loss before provision for income taxes	(24,537)	(16,080)	(61,033)	(32,293)
Benefit (provision) for income taxes	131	51	386	121
Net loss and comprehensive loss	$ (24,406)	$ (16,029)	$ (60,647)	$ (32,172)
Net loss per share attributable to common shareholders —basic and diluted	$ (0.51)	$ (8.25)	$ (2.53)	$ (19.21)
Weighted average common shares outstanding—basic and diluted	48,183,424	1,942,106	23,983,410	1,675,133

Source: Kiniksa Pharmaceuticals, Ltd.