

Kiniksa Reports Second Quarter 2018 Financial Results and Pipeline Progress

August 6, 2018

– Rilonacept advancing toward pivotal Phase 3 clinical trial in 2H 2018 following recent End-of-Phase 2 communication with FDA–

–KPL-716 single-dose Phase 1a/1b Last Subject Last Visit achieved; clinical trial data expected in Q3 2018 –

– Mavrilimumab advancing toward Phase 2 clinical trial in 2H 2018; ex-U.S. IMPD/CTA filings ongoing; pre-IND meeting scheduled with FDA –

– Completed initial public offering raising \$155.5 million in net proceeds –

HAMILTON, Bermuda, Aug. 06, 2018 (GLOBE NEWSWIRE) -- [Kiniksa Pharmaceuticals, Ltd.](#) (Nasdaq: KNSA) (“Kiniksa”), a clinical-stage biopharmaceutical company focused on discovering, acquiring, developing and commercializing therapeutic medicines for patients with significant unmet medical need, today reported second quarter 2018 financial results and pipeline progress.

“We have made significant progress in driving our portfolio forward,” said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. “We plan to continue the positive momentum in the second half of the year with Phase 1a/1b data from KPL-716, the initiation of a pivotal Phase 3 trial for rilonacept in recurrent pericarditis and the initiation of a Phase 2 trial for mavrilimumab in giant cell arteritis.”

Second Quarter 2018 and Recent Pipeline Progress

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

- Following recent End-of-Phase 2 communication with the U.S. Food and Drug Administration (FDA) on the clinical development plan for rilonacept in recurrent pericarditis, Kiniksa plans to advance to a placebo-controlled, randomized-withdrawal design pivotal Phase 3 clinical trial in approximately 60 subjects with recurrent pericarditis in the second half of 2018.
- 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 both cadence and magnitude of effect. Kiniksa expects to report additional data later in 2018.

KPL-716 (monoclonal antibody inhibitor of signaling through OSMR β)

- Kiniksa achieved Last Subject Last Visit in the single-dose portion of its Phase 1a/1b clinical trial of KPL-716 and expects to report results in the third quarter of 2018. The clinical trial utilized a randomized, double-blind, placebo-controlled, single-ascending-dose, sequential-group design to evaluate the safety, tolerability and pharmacokinetics of KPL-716 in healthy volunteers and subjects with moderate-to-severe atopic dermatitis experiencing moderate-to-severe pruritus following intravenous or subcutaneous administration. Atopic dermatitis served as a proxy for a range of pruritic diseases in order to provide exploratory endpoint evidence of target engagement and an early signal of efficacy for KPL-716 in reducing pruritus.
- Kiniksa completed enrollment of LOTUS-PN, an observational study of patients with prurigo nodularis, in the second quarter of 2018.
- Pending data from the single-dose portion of the Phase 1a/1b clinical trial of KPL-716 and the LOTUS-PN study, Kiniksa plans to initiate a Phase 2 clinical trial of KPL-716 in prurigo nodularis.
- Kiniksa is enrolling a repeat single-dose cohort as an additional part of the Phase 1b clinical trial in the U.S. and Canada. In this study, which is designed to provide longer-term exposures to evaluate safety and additional exploratory data on both pruritus and inflammatory disease response markers, subjects with moderate-to-severe atopic dermatitis experiencing moderate-to-severe pruritus receive repeat single doses of KPL-716 or placebo administered subcutaneously.

Mavrilimumab (monoclonal antibody inhibitor targeting GM-CSFR α)

- Kiniksa plans to advance to a Phase 2 clinical trial of mavrilimumab for the treatment of giant cell arteritis in the second half of 2018. Submissions of investigational medicinal product dossiers and clinical trial applications with ex-U.S. regulatory authorities are ongoing. The company has also scheduled a pre-investigational new drug (IND) application meeting with the FDA.

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

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

Second Quarter 2018 Financial Results

- Kiniksa completed an initial public offering, including the exercise of the underwriters' overallotment option, in the second quarter of 2018. The company raised net proceeds of \$155.5 million after deducting underwriting discounts, commissions and offering expenses through the sale of 9,484,202 Class A common shares at a public offering price of \$18.00 per share.
- For the second quarter of 2018, Kiniksa reported a net loss of \$20.3 million, compared to a net loss of \$11.2 million for the second quarter of 2017.
- Total operating expenses for the second quarter of 2018 totaled \$21.5 million compared to \$11.4 million for the second quarter of 2017. Non-cash share-based compensation expense totaled \$1.1 million for the second quarter of 2018, compared to \$0.1 million for the second quarter of 2017.
- As of June 30, 2018, the company had cash, cash equivalents and short-term investments of \$359.2 million and no outstanding debt.

About Kiniksa

Kiniksa is a clinical-stage 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-1 α and IL-1 β 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 Mavrimumab

Mavrimumab 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 mavrimumab 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 (OSMR β), 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 strategy and momentum, plans and timing for initiation of new clinical trials, potential designs of our new clinical trials, proposed indications for the investigation of our product candidates, plans and timing to report clinical trial data, timing for the initiation of clinical trial sites, 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: our potential inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities; potential delays or difficulty in enrollment of patients, which could delay or prevent the clinical development of our product candidates; potential undesirable side effects caused by our product candidates; our dependence on pre-clinical and clinical development conducted by third parties prior to our acquisition of our product candidates; our inability to replicate in later clinical trials positive results from pre-clinical studies or generate any human data demonstrating efficacy; potential for changes between final data and any interim "top-line" and preliminary data we announce; our reliance on third parties to manufacture our product candidates; 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 active pharmaceutical ingredient, drug product, and drug substance 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 final prospectus filed with the Securities and Exchange

Commission ("SEC") on May 24, 2018 relating to our Registration Statement on Form S-1, 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 June 30, 2018	December 31, 2017
Cash, cash equivalents, and short-term investments	\$ 359,190	\$ 45,555
Working capital	340,725	29,674
Total assets	365,129	47,492
Accumulated deficit	(127,239)	(90,998)
Total shareholders' equity (deficit)	341,799	(89,708)

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

	Three Months Ended June 30,		Six Months Ended June 30,	
	2018	2017	2018	2017
Operating expenses:				
Research and development	\$ 17,200	\$ 9,272	\$ 29,831	\$ 12,417
General and administrative	4,327	2,120	8,036	4,022
Total operating expenses	21,527	11,392	37,867	16,439
Loss from operations	(21,527)	(11,392)	(37,867)	(16,439)
Interest income	1,066	153	1,371	226
Loss before provision for income taxes	(20,461)	(11,239)	(36,496)	(16,213)
Benefit (provision) for income taxes	202	36	255	70
Net loss and comprehensive loss	\$ (20,259)	\$ (11,203)	\$ (36,241)	\$ (16,143)
Net loss per share attributable to common shareholders—basic and diluted	\$ (0.97)	\$ (6.69)	\$ (3.09)	\$ (10.47)
Weighted average common shares outstanding—basic and diluted	20,787,288	1,673,500	11,735,578	1,541,691

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