



## **Kiniksa Reports Fourth Quarter and Full-Year 2018 Financial Results and Highlights Recent Corporate and Pipeline Activity**

March 7, 2019

- Enrollment and dosing of patients commenced in rilonacept pivotal Phase 3 trial
- Enrollment and dosing of patients commenced in mavrilimumab global Phase 2 trial
- KPL-716 advancing into two Phase 2 trials in 1H 2019; repeated-single-dose Phase 1b data expected in 2H 2019
- Additional interim rilonacept Phase 2 data to be presented at the American College of Cardiology's 68<sup>th</sup> Annual Scientific Session
- Completed public offering and concurrent private placement raising approximately \$83 million in net proceeds

HAMILTON, Bermuda, March 07, 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 fourth quarter and full-year 2018 financial results and highlighted recent corporate and pipeline activity.

"Execution in the fourth quarter and full-year 2018 created a step-change in status for our clinical-stage pipeline targeting underserved autoimmune and autoinflammatory indications," said Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa. "Rilonacept advanced into a pivotal Phase 3 for recurrent pericarditis, and mavrilimumab progressed into a global Phase 2 for giant cell arteritis. Additionally, we plan to initiate Phase 2 trials for KPL-716 in chronic pruritic diseases, starting with prurigo nodularis, in the first half of 2019. We expect that our focused investment in these clinical-stage programs will generate clinical data for nine indications, which we believe will drive value for both patients and shareholders."

### **Clinical-Stage Pipeline Activity**

#### **Rilonacept (IL-1 $\alpha$ and IL-1 $\beta$ 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 double-blind, placebo-controlled, randomized-withdrawal (RW) design, global Phase 3 clinical trial of rilonacept in subjects with recurrent pericarditis. The primary efficacy endpoint is time-to-first-pericarditis-recurrence in the RW period. Top-line data are expected in the second half of 2020.
  - In December 2018, Kiniksa completed enrollment in an open-label Phase 2 clinical trial of rilonacept in subjects with recurrent pericarditis and announced interim data. As of the November 2018 data cutoff date, 12 symptomatic subjects participating in one portion of the Phase 2 trial showed a reduction in both C-reactive protein (CRP) and reported pain. Ten of the subjects had completed the 6-week base treatment period and entered into the optional 18-week extension period. Four of the 10 subjects had completed the optional 18-week extension period. All subjects showed a persistent clinical response as measured by CRP and pain levels at each measurement point during the study.
  - Kiniksa plans to present additional interim data from the open-label Phase 2 clinical trial of rilonacept in subjects with recurrent pericarditis at the American College of Cardiology's 68<sup>th</sup> Annual Scientific Session in March 2019.

#### **Mavrilimumab (monoclonal antibody inhibitor targeting GM-CSFR $\alpha$ )**

- Kiniksa announced an active investigational new drug (IND) application and 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 randomized, double-blind, placebo-controlled, 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.

#### **KPL-716 (monoclonal antibody inhibitor of signaling through OSMR $\beta$ )**

- 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 plans to initiate a randomized, double-blind, placebo-controlled, adaptive design Phase 2a/2b clinical trial of

KPL-716 in subjects with prurigo nodularis in the first half of 2019. The primary efficacy endpoint of the Phase 2a cohort is percent change from baseline in weekly average Worst-Itch Numeric Rating Scale (WI-NRS). Phase 2a top-line data are expected in the first half of 2020.

° Kiniksa plans to initiate an exploratory, multi-indication, randomized, double-blind, placebo-controlled, Phase 2 pilot clinical trial in a number of diseases characterized by chronic pruritus in the first half of 2019. The exploratory study 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. Kiniksa expects the study cohorts to be chronic idiopathic urticaria, chronic idiopathic pruritus, plaque psoriasis, lichen planus and lichen simplex chronicus. Each cohort will be an independent sub-study. Top-line data are expected in the second half of 2020.

° Kiniksa is enrolling a randomized, double-blind, placebo-controlled, repeated-single-dose Phase 1b clinical trial of KPL-716 in subjects with moderate-to-severe atopic dermatitis experiencing moderate-to-severe pruritus. Top-line data are expected in the second half of 2019.

- Kiniksa plans to present four abstracts at the Society for Investigational Dermatology in May 2019.

#### **Preclinical Pipeline Activity**

- Kiniksa continues 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 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 January 2019, Kiniksa exercised its exclusive option to acquire all of the outstanding capital stock of Primatope Therapeutics, Inc., the company that owns or controls the intellectual property related to KPL-404. Kiniksa expects to close this transaction in March 2019.
- 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. Kiniksa no longer expects to file an IND application with the FDA in the second half of 2019.

#### **Financial Results and Recent Corporate Activity**

- In the first quarter of 2019, Kiniksa completed a public offering, including the exercise of the underwriters' over-allotment option, of 2,816,110 Class A common shares at a public offering price of \$18.26 per share. Concurrent with the public offering, Kiniksa sold 2,000,000 Class A1 common shares to certain existing shareholders affiliated with certain of Kiniksa's directors at a sale price equal to the price of the public offering. The net proceeds to Kiniksa from these offerings after deducting underwriting discounts, commissions and offering expenses were approximately \$83.0 million.
- For the fourth quarter of 2018, Kiniksa reported a net loss of \$42.6 million, compared to a net loss of \$32.7 million for the fourth quarter of 2017.
- For the full-year 2018, Kiniksa reported a net loss of \$103.2 million, compared to a net loss of \$64.9 million for the full-year 2017.
- Total operating expenses for the fourth quarter of 2018 totaled \$44.1 million compared to \$32.7 million for the fourth quarter of 2017. Non-cash share-based compensation expense totaled \$2.6 million for the fourth quarter of 2018, compared to \$0.3 million for the fourth quarter of 2017.
- Total operating expenses for the full-year 2018 totaled \$108.2 million compared to \$65.4 million for the full-year 2017. Non-cash share-based compensation expense totaled \$5.7 million for the full-year 2018, compared to \$0.9 million for the full-year 2017.
- As of December 31, 2018, the company had cash, cash equivalents and short-term investments of \$307.3 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, focused on autoinflammatory and autoimmune conditions. For more information, please visit [www.kiniksa.com](http://www.kiniksa.com).

#### **About Rilonacept**

Rilonacept is a weekly, subcutaneously-injected, recombinant fusion protein that blocks IL-1 $\alpha$  and IL-1 $\beta$  signaling. Rilonacept was discovered and developed by Regeneron and is approved by the FDA under the brand name ARCALYST<sup>®</sup> 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 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 (OSMR $\beta$ ), 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.

**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 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 initiation of new clinical trials; potential designs of our new clinical trials; proposed indications for the investigation of our product candidates; estimated disease prevalence; plans and timing to report or present clinical trial data; plans and timing for the submission of investigational new drug and other applications and submissions to regulatory authorities; plans and timing to advance additional pipeline programs into clinical trials; and plans and timing to close our acquisition of Primatope.

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 final prospectus filed with the Securities and Exchange Commission (“SEC”) on February 1, 2019 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!<sup>TM</sup>****Kiniksa Investor and Media Contact**

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

	As of	
	December 31, 2018	December 31, 2017
Cash, cash equivalents, and short-term investments	\$ 307,304	45,555
Working capital	271,196	29,674
Total assets	321,965	47,492
Accumulated deficit	(194,225)	(90,998)
Total shareholders' equity (deficit)	279,267	(89,708)

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

	Three Months Ended		Years Ended	
	December 31,		December 31,	
	2018	2017	2018	2017
Operating expenses:				
Research and development	\$ 36,122	\$ 29,931	\$ 86,597	\$ 56,357
General and administrative	8,013	2,780	21,563	9,043
Total operating expenses	44,135	32,711	108,160	65,400
Loss from operations	(44,135)	(32,711)	(108,160)	(65,400)
Interest income	1,727	133	4,719	529
Loss before benefit (provision) for income taxes	(42,408)	(32,578)	(103,441)	(64,871)
Benefit (provision) for income taxes	(172)	(123)	214	(2)
Net loss and comprehensive loss	\$ (42,580)	\$ (32,701)	\$ (103,227)	\$ (64,873)
Net loss per share attributable to common shareholders —basic and diluted	\$ (0.88)	\$ (14.79)	\$ (3.49)	\$ (35.85)
Weighted average common shares outstanding—basic and diluted	48,458,892	2,210,713	29,547,427	1,809,751



Source: Kiniksa Pharmaceuticals, Ltd.