May 9, 2017

Akebia Announces First Quarter 2017 Financial Results

- Recent Collaboration Agreements Position Vadadustat Global Phase 3 Development Program for Success, Offer Strong Financial and Commercial Support, and Drive Significant Value Creation -

CAMBRIDGE, Mass.--(BUSINESS WIRE)-- Akebia Therapeutics, Inc. (NASDAQ:AKBA), a biopharmaceutical company focused on delivering innovative therapies to patients with kidney disease through the biology of hypoxia-inducible factor (HIF), today announced financial results for the first quarter ended March 31, 2017.

"Our team at Akebia continues to execute on our goals in 2017, as we advance the Phase 3 program for vadadustat and expand our pipeline," said John P. Butler, President and Chief Executive Officer of Akebia. "We established two major collaborations with Otsuka Pharmaceutical in the last four months that support our global development program for vadadustat and drive long-term value for Akebia. The Otsuka agreements, coupled with an earlier collaboration with Mitsubishi Tanabe, provide for $573 million or more in committed capital and a total potential deal value of $2.2 billion plus royalties ranging from mid-single digit to tiered double digit. We believe that the strength of our collaborations is a testament to the potential of vadadustat to change the standard of care for patients with anemia associated with chronic kidney disease. With cash plus committed development funding of over $600 million, Akebia is in a very strong financial position."

First Quarter 2017 and Recent Corporate Highlights

- Expanded relationship with Otsuka for vadadustat from a profit share agreement in the U.S. to include a collaboration and license agreement for Europe, China and other territories, excluding Latin America. This agreement provides for committed capital and potential milestone payments from Otsuka of up to $865 million, including $208 million or more in upfront and development funding and up to $657 million in milestone payments;
- Signed an exclusive agreement with Johnson & Johnson Innovation to in-license HIF product candidates and access an extensive library of HIF compounds, including AKB-5169, a differentiated, oral, non-absorbed preclinical compound for the treatment of inflammatory bowel disease, which is poised for IND submission in the second half of 2018;
- Published positive Phase 2a study results in the *American Journal of Nephrology*, demonstrating that vadadustat increased hemoglobin levels in a dose-dependent manner and improved iron mobilization in non-dialysis chronic kidney disease (CKD) patients; and
- The Independent Data Monitoring Committee for Akebia's global Phase 3 PRO2TECT and INNO$_2$VATE programs met and recommended continuing the studies without modification.

Financial Results

The company reported a net loss of ($44.5) million, or ($1.15) per share, for the first quarter of 2017 as compared to a net loss for the first quarter of 2016 of ($25.8) million or ($0.70) per share.

Collaboration revenue was $20.9 million for the first quarter of 2017, which related to the Company's agreement with Otsuka. Collaboration revenue in connection with our agreement with Mitsubishi Tanabe Pharma Corporation is expected to commence in the second half of 2017.

Research and development expenses were $60.0 million for the first quarter of 2017 compared to $20.2 million for the first quarter of 2016. The increase is primarily attributable to external costs related to the global PRO$_2$TECT and INNO$_2$VATE Phase 3 programs. Research and development expenses were further increased by headcount and compensation-related costs.

General and administrative expenses were $5.8 million for both the first quarters of 2017 and 2016 due to offsetting increases and decreases in associated costs.

The company ended the first quarter of 2017 with cash, cash equivalents and marketable securities of $251.8 million. The company is also entitled to receive $373.0 million or more in committed capital from collaborators, which is expected to be

received over the course of the global development program for vadadustat, of which $73.0 million was received in April 2017 in connection with the expanded collaboration with Otsuka. Based on the timing of payments from collaborators, Akebia expects existing and committed cash resources to fund the company's current operating plan into the first quarter of 2019. However, the remaining committed research and development funding will continue to be received from Otsuka on a prepaid, quarterly basis up to an estimated aggregate of $60.0 million.

About Akebia Therapeutics

Akebia Therapeutics, Inc. is a biopharmaceutical company headquartered in Cambridge, Massachusetts, focused on delivering innovative therapies to patients with kidney disease through hypoxia-inducible factor biology. Akebia's lead product candidate, vadadustat, is an oral investigational therapy in development for the treatment of anemia related to chronic kidney disease in both non-dialysis and dialysis patients. Akebia's global Phase 3 program for vadadustat, which includes the PRO$_2$TECT studies for non-dialysis patients with anemia associated with chronic kidney disease and the INNO$_2$VATE studies for dialysis-dependent patients, is currently ongoing. For more information, please visit our website at www.akebia.com.

Forward-Looking Statements

This press release includes forward-looking statements. Such forward-looking statements include those about Akebia's strategy, future plans and prospects, including statements regarding the potential commercialization of vadadustat if approved by regulatory authorities, the potential indications and benefits of vadadustat and Akebia's other product candidates, anticipated financial contributions from Otsuka Pharmaceutical and Mitsubishi Tanabe under Akebia's collaboration agreements, and the timing of the potential filing of an IND for AKB-5169. The words "anticipate," "appear," "believe," "estimate," "expect," "intend," "may," "plan," "predict," "project," "target," "potential," "will," "would," "could," "should," "continue," and similar expressions are intended to identify forward-looking statements, although not all forward-looking statements contain these identifying words. Each forward-looking statement is subject to risks and uncertainties that could cause actual results to differ materially from those expressed or implied in such statement, including the risk that existing preclinical and clinical data may not be predictive of the results of ongoing or later clinical trials; the funding required to develop Akebia's product candidates and operate the company, and the actual expenses associated therewith; the actual costs incurred in the Phase 3 studies of vadadustat and the availability of financing to cover such costs; early termination of Akebia's collaboration or license agreements; Akebia's ability to satisfy its obligations under its collaboration and license agreements; the timing and content of decisions made by the FDA and other regulatory authorities; the actual time it takes to initiate and complete research and development; the success of competitors in developing product candidates for diseases for which Akebia is currently developing its product candidates; and Akebia's ability to obtain, maintain and enforce patent and other intellectual property protection for vadadustat and its other product candidates. Other risks and uncertainties include those identified under the heading "Risk Factors" in Akebia's Annual Report on Form 10-Q for quarter ended March 31, 2017, and other filings that Akebia may make with the Securities and Exchange Commission in the future. Akebia does not undertake, and specifically disclaims, any obligation to update any forward-looking statements contained in this press release.

AKEBIA THERAPEUTICS, INC
Consolidated Statements of Operations
(in thousands except share and per share data)
(unaudited)

	Three Months Ended	
	March 31, 2017	**March 31, 2016**
Collaboration revenue	$ 20,865	$ —
Operating expenses:		
Research and development	60,049	20,235
General and administrative	5,788	5,811
Total operating expenses	65,837	26,046
Operating loss	(44,972)	(26,046)
Other income, net	429	248
Net loss	$ (44,543)	$ (25,798)
Net loss per share - basic and diluted	$ (1.15)	$ (0.70)
Weighted-average number of common shares - basic and diluted	38,759,221	36,873,594

AKEBIA THERAPEUTICS, INC.
Selected Balance Sheet Data
(in thousands)
(unaudited)

	March 31, 2017	December 31, 2016
Cash, cash equivalents and available for sale securities	$ 251,805	$ 260,343
Working capital	121,590	182,053
Total assets	259,256	300,216
Total stockholders' equity	30,617	68,120

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