December 20, 2016

Akebia and Otsuka Pharmaceutical Announce Collaboration to Develop and Commercialize Vadadustat in the U.S.

- Funds Vadadustat Global Phase 3 PRO$_2$TECT and INNO$_2$VATE Studies -

- Committed Capital and Potential Milestone Payments Could Exceed $1 Billion -

- Akebia and Otsuka Share Revenue and Commercialization Costs Equally -

- Akebia to Host Conference Call at 8:30 a.m. Eastern Time Today -

CAMBRIDGE, Mass. & TOKYO--(BUSINESS WIRE)-- Akebia Therapeutics, Inc. (NASDAQ: AKBA) and Otsuka Pharmaceutical Co., Ltd. today announced they have entered into a collaboration and license agreement in the U.S. for vadadustat, an oral hypoxia-inducible factor (HIF) stabilizer currently in development for the treatment of anemia associated with chronic kidney disease (CKD). Anemia related to CKD affects an estimated 1.8 million patients in the U.S. and arises from the kidney's failure to produce adequate amounts of erythropoietin, a key hormone stimulating the production of red blood cells.[1] Left untreated, anemia significantly accelerates patients' overall deterioration of health with increased morbidity and mortality. [2]

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The collaboration provides capital for the global development program for vadadustat, and commercial resources for a U.S. launch of vadadustat upon approval by the Food and Drug Administration. Under the terms of the agreement, Akebia will receive $265 million in committed funds plus development and commercial milestones, representing a total transaction value that could exceed $1 billion. The companies intend to contribute equally to commercialization efforts and share equally all costs and revenue in the U.S., where sales of erythropoiesis stimulating agents (ESAs), the current standard of care, are estimated to be $3.5 billion.[3] Akebia will continue to lead the ongoing global Phase 3 development program for vadadustat.

"Vadadustat has the potential to significantly change the current standard of care for patients with anemia associated with CKD and addresses a high unmet need for those suffering with this disease," said Mr. Tatsuo Higuchi, President and Representative Director of Otsuka. "With Akebia's renal expertise, this collaboration will enable Otsuka to expand our cardio-renal portfolio while demonstrating our commitment to delivering new treatment options to patients worldwide."

Under the terms of the agreement, Otsuka will pay $265 million or more in committed capital. This includes a payment of $125 million upon signing and a payment of approximately $35 million in the first quarter of 2017. The agreement also provides for Otsuka to pay $105 million or more of the costs of the global development program for vadadustat. Additionally, Otsuka will pay potential development and commercial milestones up to $765 million.

"This collaboration achieves our goal of funding our global PRO$_2$TECT and INNO$_2$VATE Phase 3 studies for vadadustat while retaining significant long-term value for Akebia," stated John P. Butler, President and Chief Executive Officer of Akebia.

Mr. Butler added, "Our alliance with Otsuka, one of the world's innovative pharmaceutical leaders, also allows us to prepare an optimal launch of vadadustat, as we will equally share commercial responsibility. Otsuka brings a well-established commercial presence and infrastructure in the U.S., and we share a strong commitment to improving patients' lives by delivering important new therapeutic options. This deal also underscores the confidence that we and others have placed in the underlying value of vadadustat and in our ability to bring innovative therapies to patients."

In addition to the collaboration with Otsuka, Akebia has established a collaboration with Mitsubishi Tanabe Pharma Corporation for the development and commercialization of vadadustat in Japan, Taiwan, South Korea, Indonesia, India and other countries in Asia. For other geographies, including the European Union, Akebia continues advancing discussions with multiple parties regarding a potential collaboration.

Conference Call and Webcast
Akebia management will host a conference call to review the details of the transaction beginning at 8:30 a.m. Eastern Time today, Tuesday, December 20, 2016. A live audio webcast of the presentation will be available on the company's website at http://ir.akebia.com/events.cfm. An archived presentation will be available for 90 days.

To access the conference call, follow these instructions:

Dial: (877) 458-0977 (U.S.); (484) 653-6724 (international)
Conference ID: 39235865

About Vadadustat
Vadadustat is an oral hypoxia-inducible factor (HIF) stabilizer currently in development for the treatment of anemia related to chronic kidney disease. Vadadustat exploits the same mechanism of action used by the body to adapt naturally to lower oxygen availability associated with a moderate increase in altitude. At higher altitudes, the body responds to lower oxygen availability with increased production of HIF, which coordinates the interdependent processes of iron mobilization and erythropoietin production to increase red blood cell production and, ultimately, improve oxygen delivery.

About Anemia Associated with CKD
Approximately 30 million people in the U.S. have chronic kidney disease (CKD), with an estimated 1.8 million of these patients suffering from anemia. Anemia results from the body's inability to coordinate red blood cell production in response to lower oxygen levels due to the progressive loss of kidney function with inadequate erythropoietin production. Left untreated, anemia significantly accelerates patients' overall deterioration of health with increased morbidity and mortality. Anemia is currently treated with injectable recombinant erythropoiesis stimulating agents, which are associated with inconsistent hemoglobin responses and well-documented safety risks.[4]

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 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_2TECT studies for non-dialysis patients with anemia secondary to 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.

About Otsuka
Otsuka Pharmaceutical is a global healthcare company with the corporate philosophy: "Otsuka - people creating new products for better health worldwide." Otsuka researches, develops, manufactures and markets innovative and original products, with a focus on pharmaceutical products for the treatment of diseases and nutraceutical products for the maintenance of everyday health.

Otsuka Pharmaceutical Development & Commercialization, Inc. (OPDC), based in Princeton, N.J., discovers and develops new compounds that address unanswered medical needs and advance human health, with a focus on neuroscience, oncology, and cardio-renal treatments. For more information about Otsuka in the U.S., visit www.otsuka-us.com and on Twitter at @OtsukaUS.

Otsuka Pharmaceutical is a subsidiary of Otsuka Holdings Co., Ltd., headquartered in Tokyo, Japan, with 2015 consolidated sales of $11.9 billion. Otsuka welcomes you to visit its global website at https://www.otsuka.co.jp/en.

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 indications and benefits of vadadustat, the satisfaction of Akebia's funding needs for the PRO_2TECT and $INNO_2$VATE Phase 3 programs, the potential commercialization of vadadustat if approved by the FDA, anticipated contributions from Otsuka pursuant to the Collaboration and License Agreement, and the progress toward securing a collaboration for other geographies. 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 ability of Akebia to successfully complete the clinical development of vadadustat; 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; the timing and content of decisions made by the FDA and other regulatory authorities; the rate of enrollment in clinical studies of vadadustat; the actual time it

takes to initiate and complete clinical studies; Akebia's ability to satisfy its obligations under the Collaboration and License Agreement; early termination of the Collaboration and License Agreement by Akebia or Otsuka; Akebia's ability to negotiate commercially reasonable terms with an additional collaboration partner; 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. Other risks and uncertainties include those identified under the heading "Risk Factors" in Akebia's Annual Report on Form 10-Q for the quarter ended September 30, 2016, 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.

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[1]Stages 1-4: JAMA 2007 Coresh et al. (Prevalence of CKD in the US). NHANES 1988-94 and 1999-2004.
Stage 5: USRDS 2013 report (ESRD).
Iseki K and Kohagura. Anemia as a risk factor for chronic kidney disease K. Kidney Int Suppl. 2007;107:S4-9.

[2]Culleton B, Manns B, Zhang J, Tonelli M, Klarenbach S, et al. Impact of anemia on hospitalization and mortality in older adults. Blood 2006;107(10):3841-3846.
Portolés J, Gorriz J, Rubio E, de Alvaro F, García F, et al. The development of anemia is associated to poor prognosisinNKF/KDOQI stage 3 chronic kidney disease. BMC nephrology 2013;14(1):2.

[3]Global sales of injectable erythropoiesis-stimulating agents as reported in public filings.

[4]Singh AK. What is causing the mortality in treating the anemia of chronic kidney disease: erythropoietin dose or hemoglobin level? Curr Opin Nephrol Hypertens 2010;19:420-424

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