



Bellerophon Announces FDA Agreement on Phase 2b Study Design for INOpulse® in Pulmonary Hypertension Associated with Interstitial Lung Disease (PH-ILD) Company to Host Key Opinion Leader Event on PH-ILD in New York City

Warren, NJ, Aug. 3, 2017 —Bellerophon Therapeutics, Inc. (Nasdaq: BLPH), a clinical-stage biotherapeutics company, today announced agreement with the U.S. Food and Drug Administration (FDA) on the Phase 2 study design for INOpulse® in pulmonary hypertension associated with Interstitial Lung Disease (ILD).

The Company met with the FDA in June 2017 to present positive results from its recently completed Phase 2a study in idiopathic pulmonary fibrosis (IPF), and to review clinical plans for its Phase 2b trial, entitled iNO-PF, in IPF as well as other pulmonary fibrosing diseases within ILD. Subsequently, the Agency has accepted the Company's proposed Phase 2b study design as well as an Investigational New Drug (IND) application to assess the effect of INOpulse on patients at both low and high risk for pulmonary hypertension associated with pulmonary fibrosis.

The FDA recognized the dual mode of action of vasodilation and ventilation/perfusion matching of pulsed iNO therapy, which the Company believes can provide a clinically important benefit to a wide range of patients, including those that may not exhibit signs of pulmonary hypertension at rest.

“We are very pleased to have concordance with the FDA on our iNO-PF Phase 2b trial for INOpulse in ILD and to have a finalized plan to move forward with this important trial,” said Fabian Tenenbaum, Chief Executive Officer of Bellerophon Therapeutics. “The proprietary targeted delivery and the dual mode of action of INOpulse may allow it to be used in pulmonary fibrosing diseases where systemic vasodilators have proven to be ineffective. The lack of approved therapies for pulmonary hypertension associated with interstitial lung diseases represents a unique opportunity to develop a new therapy in this serious and significant unmet medical need.”

The iNO-PF Phase 2b study design is based on the results of the prior Phase 2a study for INOpulse in the treatment of Pulmonary Hypertension associated with Idiopathic Pulmonary Fibrosis (PH-IPF), presented at the American Thoracic Society (ATS) International Conference on May 21, 2017. This Phase 2a study met its primary endpoint, showing an average of 15.3% increase in blood vessel volume ($p < 0.001$). There was a significant association between ventilation and vasodilation, demonstrating the ability of INOpulse to provide targeted selective delivery to well ventilated sections of the lung. The study also showed consistent benefit in hemodynamics and exercise capacity, with a clinically meaningful reduction of 14% in systolic pulmonary arterial pressure (sPAP) and an average improvement of 75 meters in 6-Minute Walk Distance.

Planned Study Design

The iNO-PF trial is planned for 2018 and is designed to recruit 40 subjects diagnosed with pulmonary fibrosis, half of which are at intermediate to high risk of pulmonary hypertension as determined by echocardiography. Importantly, the FDA has agreed to a Phase 2b design that eliminates the need for right heart catheterization, an invasive procedure which can present significant challenges for potential study participants.

To support the progress of its program for INOpulse in the treatment of ILDs, Bellerophon has formed a Scientific Advisory Committee chaired by Dr. Steven D. Nathan (Inova Fairfax Hospital) as well as Dr. Ganesh Raghu (University of Washington), Dr. Kevin Flaherty (University of Michigan), Dr. Marilyn K. Glassberg Ceste (University of Miami), Dr. Jeffrey Swigris (National Jewish Health – Denver) and Dr. Lisa Lancaster (Vanderbilt University Medical Center).

KOL Investor Meeting, August 9th

Bellerophon will host a Key Opinion Leader lunch on the topic of Pulmonary Hypertension Associated with Interstitial Lung Disease (PH-ILD) on Wednesday, August 9 in New York City. The meeting will feature a presentation by Dr. Steven D. Nathan, who will discuss the current treatment landscape and unmet medical need for patients with PH-ILD. Dr. Nathan will be available to answer questions following the lunch.

Steven D. Nathan, MD, FCCP, is director of the Advanced Lung Disease Program and director of the Lung Transplant Program at Inova Fairfax Hospital. He also is Professor of Medicine at Virginia Commonwealth University Inova Campus. Dr. Nathan is board certified in pulmonary diseases, critical care medicine and internal medicine. The author of more than 380 publications, Dr. Nathan has written original research manuscripts, abstracts, reviews, book chapters and a book on idiopathic pulmonary fibrosis (IPF), which he co-edited. Dr. Nathan is a reviewer for multiple journals and is on the editorial board for the journal, Thorax. He has served on multiple committees, including the U.S. Food and Drug Administration advisory boards as well as steering committees for clinical trials in IPF and pulmonary hypertension, where he has also served as chair. He is also chairperson of Pilot for IPF, an international educational initiative for pulmonary fibrosis.

Bellerophon's management team will also provide an overview of the Company's ongoing clinical development program with their proprietary INOpulse delivery system as well as their clinical trial strategy to progress INOpulse for the treatment of PH-ILD.

This event is intended for institutional investors, sell-side analysts, investment bankers, and business development professionals only. Please RSVP in advance to Mac@LifeSciAdvisors.com if you plan to attend, as space is limited.

A live webcast of the event, with slides, will be available at <http://lifesci.rampard.com/20170809/reg.jsp> and posted after the event on the "Investors" page of the Company's website at www.bellerophon.com.

About Bellerophon

Bellerophon Therapeutics is a clinical-stage biotherapeutics company focused on developing innovative therapies at the intersection of drugs and devices that address significant unmet medical needs in the treatment of cardiopulmonary diseases. The Company is currently developing three product candidates under its INOpulse program, a proprietary pulsatile nitric oxide delivery system. The first is for the treatment of pulmonary arterial hypertension (PAH), for which the Company has commenced Phase 3 clinical trials in 2016. The second is for the treatment of pulmonary hypertension associated with chronic obstructive pulmonary disease (PH-COPD) and the third candidate is for the treatment of pulmonary hypertension associated with Interstitial Lung Disease (PH-ILD), both of which are in Phase 2 development. For more information, please visit www.bellerophon.com.

Forward-looking Statements

Any statements in this press release about Bellerophon's future expectations, plans and prospects, including statements about the clinical development of its product candidates, regulatory actions with respect to the Company's clinical trials and expectations regarding the sufficiency of the Company's cash balance to fund clinical trials, operating expenses and capital expenditures, and other statements containing the words "anticipate," "believe," "continue," "could," "estimate," "expect," "intend," "may," "plan," "potential," "predict," "project," "should," "target," "would," and similar expressions, constitute forward-looking statements within the meaning of The Private Securities Litigation Reform Act of 1995. Actual results may differ materially from those indicated by such forward-looking statements as a result of various important factors, including: the uncertainties inherent in the initiation of future clinical trials, availability and timing of data from ongoing and future clinical trials and the results of such trials, whether preliminary or interim results from a clinical trial will be predictive of the final results of that trial or whether results of early clinical trials will be indicative of the results of later clinical trials, expectations for regulatory approvals, the FDA's substantial discretion in the approval process, availability of funding sufficient for our foreseeable and unforeseeable operating expenses and capital expenditure requirements and other factors discussed in the "Risk Factors" section of the Company's most recent filings with the Securities and Exchange Commission. In addition, any forward-looking statements included in this press release represent Bellerophon's views only as of the date of this release and should not be relied upon as representing the Company's views as of any subsequent date. The Company specifically disclaims any obligation to update any forward-looking statements included in this press release.

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