



Bellerophon to Present Positive Clinical Data on INOpulse® at the American Thoracic Society 113th International Conference

Data Show that INOpulse was Associated with Clinically Meaningful Improvements in Hemodynamics and Exercise Capacity in Difficult-to-Treat PH-IPF Patients

Phase 2b Study in PH-IPF Population Planned

Warren, NJ, May 1, 2017 — Bellerophon Therapeutics, Inc. (Nasdaq: BLPH), a clinical-stage biotherapeutics company, today announced that it will present new clinical data from its Phase 2 study evaluating the use of INOpulse® in idiopathic pulmonary fibrosis patients with pulmonary hypertension (PH-IPF) at the American Thoracic Society (ATS) 113th International Conference, on Sunday, May 21st in Washington DC.

This proof of concept study (n=4) was conducted at University Hospital Antwerp led by Prof. W. De Backer MD, Director in the Department of Pulmonary Medicine. It was designed to assess the potential for pulsed inhaled nitric oxide (iNO) to improve hemodynamics and exercise capacity in PH-IPF patients. PH-IPF is a particularly difficult condition to manage as the treatments used in other pulmonary hypertension populations failed to show a benefit for IPF patients, limiting their applicability and creating a critical need for effective and safe long-term treatment options.

The study was divided into an acute and a chronic phase. The acute phase was designed to identify the optimum iNO dose and evaluate the impact on hemodynamic measures. In the chronic phase, the impact of iNO on exercise capacity was evaluated using the 6 minute walk test after 4 weeks of treatment.

Key findings

- Clinically important improvements were seen acutely and at 4 weeks in both hemodynamics and exercise capacity in all patients.
- Hemodynamics, as determined by reduction in systolic pulmonary arterial pressure (sPAP) showed improvement in all patients with an average reduction of 14% compared to baseline.
- Dose titration suggested that the iNO 30 dose can safely provide clinically relevant reduction in sPAP.
- The 6 minute walk distance increased on average 75 meters from baseline after 4 weeks of chronic use of INOpulse therapy.
- Improved oxygenation during the 6 minute walk test provides supportive evidence of targeted vasodilation and improved ventilation and perfusion (V/Q) matching.
- The investigators also evaluated the effect of INOpulse on two composite endpoints which combine oxygen saturation with walk distance: Distance Saturation Product (DSP) and Integral

Distance Saturation Product (IDSP). These composite endpoints showed consistent improvement and may prove to be valuable as predictors of disease progression and outcomes.

- Detailed study data, including for the composite endpoints and respiratory imaging, will be presented in the poster at the ATS meeting.

“The results from this study suggest that iNO allows selective vasodilation to the well-functioning parts of the lung to improve hemodynamic measures as well as exercise capacity,” said Prof. W. De Backer MD, Director in the Department of Pulmonary Medicine, University Hospital and University of Antwerp. “This is a potentially important advantage for pulsed iNO therapy since the currently available vasodilators that are used to treat pulmonary hypertension act systemically and have been shown to cause issues, such as worsening hypoxemia, when used on PH-IPF patients.”

“Development of pulmonary hypertension co-morbidity in IPF patients greatly impacts quality of life and survival. There are no approved treatments for this condition resulting in a profound unmet clinical need,” said Fabian Tenenbaum, Chief Executive Officer of Bellerophon Therapeutics. “We are pleased to present this study at ATS, which builds on our current understanding of pulsed iNO therapy to improve pulmonary hemodynamics and exercise capacity in PH-IPF patients. The next step in this program will be to conduct a larger, controlled Phase 2b study to inform the design of a pivotal Phase 3 trial.”

Details on this Phase 2 proof of concept study will be available at ATS in the following poster presentation.

Title: *Unraveling the mode of action of pulsed inhaled NO in severe IPF using Functional Respiratory Imaging (FRI)*

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Date: Sunday, May 21
Time: 9:15am - 4:15pm Eastern Time
Session: PH-IPF: P913

About the American Thoracic Society

The American Thoracic Society (ATS) improves global health by advancing research, patient care, and public health in pulmonary disease, critical illness, and sleep disorders. Founded in 1905 to combat TB, the ATS has grown to tackle asthma, COPD, lung cancer, sepsis, acute respiratory distress, and sleep apnea, among other diseases. The ATS 113th International Conference will take place at the Walter E. Washington Convention Center in Washington DC, May 19-24, 2017. More than 14,000 professionals are expected to attend. Additional information on the ATS 113th International Conference can be found at <http://conference.thoracic.org>.

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 Idiopathic Pulmonary Fibrosis (PH-IPF), both of which are in Phase 2 development. For more information, please visit www.bellerophon.com.

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