



**Bellerophon Therapeutics Completes Enrollment of its Phase 2 Trial of INOpulse®
For Treatment of Pulmonary Hypertension Associated with Chronic Obstructive Pulmonary Disease
(PH-COPD)**

Hampton, NJ, June 26, 2014 – Bellerophon Therapeutics, LLC, a clinical stage biotherapeutics company, today announced that it has completed enrollment of its 159-patient Phase 2 clinical trial of INOpulse for the treatment of pulmonary hypertension associated with chronic obstructive pulmonary disease (PH-COPD). PH-COPD is a serious complication of COPD that can significantly affect the health status of patients with this condition. Current standard of care is primarily limited to oxygen therapy, pulmonary rehabilitation and lung transplant. This follows the previously announced completion of enrollment for Bellerophon’s 80-patient Phase 2 clinical trial of INOpulse for the treatment of pulmonary arterial hypertension (PAH).

Bellerophon’s INOpulse device delivers brief, controlled pulses of inhaled nitric oxide, which is a selective, short-acting pulmonary vasodilator. INOpulse is portable, which is intended to allow for treatment of ambulatory patients on daily basis outside the hospital.

This Phase 2 trial, conducted at 43 sites in the United States, is a randomized, double-blind, placebo-controlled short-term dose response trial of INOpulse for patients with PH-COPD. The trial assesses acute hemodynamic responses in patients to inform the dose selection for the next phase of clinical development of INOpulse for PH-COPD. The trial is expected to be completed in the third quarter of 2014.

About Inhaled Nitric Oxide

Nitric oxide is naturally produced and released by the inner lining of the blood vessels and results in smooth muscle relaxation. In particular, nitric oxide controls muscle tone in blood vessels and thus is an important factor in regulating blood pressure. As the muscles of the blood vessels relax, blood flow increases, helping the heart to deliver more blood to the body. We are evaluating the effect of nitric oxide when administered by inhalation, as a pulsed dose, to the alveoli of the lungs. We expect inhaled nitric oxide, delivered using the INOpulse device, to act in a similar manner to naturally produced nitric oxide with minimal effects on blood pressure outside of the lungs, an important safety consideration.

About PH-COPD

Chronic obstructive pulmonary disease (COPD) is a disease characterized by progressive and persistent airflow limitations. Patients with more severe COPD frequently have hypoxemia and are treated with long-term oxygen therapy. Despite treatment with oxygen, hypoxemia in these patients may progress and contribute to the development of pulmonary hypertension (PH). In addition, COPD patients can have deficiency in endogenous nitric oxide production in their lungs, which can worsen their PH. This PH puts pressure on the right side of the heart, making it less able to cope with stressors and potentially leading to progressive cardiac dilation, heart failure and death.



PH is a serious complication of COPD that can significantly affect the health status of patients with this condition. Data published in literature suggests that even small elevations in mean pulmonary artery pressure in patients with advanced COPD can impact hospitalization, patient-assessed functional outcomes and mortality. PH is a well-known predictor of increased morbidity and mortality in COPD patients and is associated with poor quality of life, worse clinical outcomes and shorter survival time with a 4-year survival rate of only 50% compared to 80% for COPD patients with similar respiratory disease severity who do not have pulmonary hypertension. There are no currently approved drug therapies for this condition, and current standard of care is primarily limited to oxygen therapy, pulmonary rehabilitation and lung transplant.

About Bellerophon

Bellerophon Therapeutics LLC, is a privately-held, 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 and cardiac diseases. Two of the company's product candidates are based on its proprietary pulsatile nitric oxide delivery device, INOpulse, and are in Phase 2 clinical trials – one for the treatment of PAH and a second for the treatment of PH-COPD. The company's third product candidate, bioabsorbable cardiac matrix (BCM), is an injectable device currently undergoing a feasibility clinical trial, which is a CE mark registration trial in the European Union and is comparable to a Phase 2 trial in U.S. drug development, for the prevention of cardiac remodeling and subsequent congestive heart failure following acute myocardial infarction (heart attack).

Bellerophon acquired exclusive worldwide rights to develop and commercialize the INOpulse programs in PAH, PH-COPD and pulmonary hypertension associated with idiopathic pulmonary fibrosis (PH-IPF) from Ikaria, Inc. in February 2014 as part of Ikaria's spin-out of certain of its research and development assets and subsidiaries. Bellerophon has an exclusive worldwide license to BCM from BioLineRx Ltd.

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