



Bellerophon Therapeutics Completes Enrollment for its Phase 2 Trial of INOpulse® For Treatment of Pulmonary Arterial Hypertension (PAH)

Hampton, NJ, June 17, 2014 – Bellerophon Therapeutics, LLC, a clinical stage biotherapeutics company, today announced that it has completed enrollment of its 80-patient Phase 2 clinical trial of INOpulse for the treatment of pulmonary arterial hypertension (PAH). PAH is a life-threatening, progressive disorder characterized by abnormal constriction of the arteries of the lung, leading to increased blood pressure in the lungs and abnormal strain on the heart’s right ventricle, eventually leading to heart failure.

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

This Phase 2 study, being conducted at 52 sites in the United States and Canada, is a randomized, placebo-controlled trial of INOpulse as an add-on therapy for use in patients whose disease is progressing despite treatment with other PAH medications. The trial will determine the safety, tolerability and efficacy in this population of two different doses of INOpulse for PAH. The primary endpoint is a change in pulmonary vascular resistance at 16 weeks from baseline. Secondary endpoints include change in mean pulmonary arterial pressure and cardiac index as well as change in six-minute walk distance. The trial is expected to be completed by the end of 2014.

“The U.S. Food and Drug Administration has granted orphan drug designation for nitric oxide for the treatment of PAH, which will give us seven years of exclusivity in the U.S. if INOpulse is the first inhaled nitric oxide therapy approved in this indication,” stated Daniel Tassé, interim chief executive officer of Bellerophon. “There is presently no cure for PAH and, despite several approved therapies, the mortality rate remains high. The completion of enrollment in this important Phase 2 trial is therefore a key milestone for Bellerophon, and we look forward to the continued development of this potential new first-in-class therapy.”

About Inhaled Nitric Oxide

Nitric Oxide is naturally produced and released by portions 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 PAH

PAH is a life-threatening, progressive disorder characterized by abnormally high blood pressure, or hypertension, in the pulmonary artery, the blood vessel that carries blood from the heart to the lungs. PAH occurs when most of the very small arteries throughout the lungs narrow in diameter, increasing the resistance to blood flow through the lungs. To overcome the increased resistance, pressure



increases in the pulmonary artery and the right ventricle -- the heart chamber that pumps blood into the pulmonary artery. In addition, PAH may cause changes to the blood vessel lining that hinders the natural production of nitric oxide.

Prior to the availability of current treatments, primary PAH patients had an average survival of less than three years. There are a number of drugs approved for the treatment of PAH that work by reducing vascular resistance. However, despite the availability of multiple therapies for this indication, the mortality rate for PAH patients remains high, with estimates of median survival ranging from three to five years. Patients with PAH also report severe impairment of health-related quality of life, including poor general and emotional health and impaired physical functioning. The most common symptoms of PAH are shortness of breath during exertion and fainting spells. Additional symptoms can include dizziness, swelling of the ankles or legs, chest pain and a racing pulse. PAH is classified as an orphan disease by the FDA, indicating that there are fewer than 200,000 patients who have been diagnosed with the condition in the United States..

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 pulmonary hypertension associated with chronic obstructive pulmonary disease (PH-COPD). The company's third product candidate, bioabsorbable cardiac matrix (BCM), is an injectable device currently undergoing a feasibility clinical trial, 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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