

# PHARMING

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## PHARMING COMPLETES RUCONEST® US PIVOTAL PHASE III STUDY

**Leiden, The Netherlands, September 27, 2012.** Biotech company Pharming Group NV (“Pharming” or “the Company”) (NYSE Euronext: PHARM) today announces that it has completed the clinical phase of the US pivotal Phase III clinical study (Study 1310) evaluating the investigational drug RUCONEST® (recombinant human C1 inhibitor) for the treatment of acute attacks of angioedema in patients with Hereditary Angioedema (HAE). All 75 randomized patients have now reached the final time points according to protocol.

Over the coming weeks, as usual in the conduct of clinical trials, the trial database will be finalized and locked. The results will be analyzed and subsequently announced. Positive results of the study will trigger a US\$ 10 million milestone payment to Pharming from its US partner Santarus Inc.

### **RUCONEST® Phase III Study (Study 1310)**

Pharming is conducting a Phase III clinical study with RUCONEST® under a Special Protocol Assessment (SPA) that is intended to support the submission of a Biologics License Application (BLA) to the U.S. Food and Drug Administration (FDA). RUCONEST® is being evaluated for the treatment of acute attacks of angioedema in patients with HAE in an international, multicenter, randomized, placebo-controlled Phase III study at a dosage strength of 50 U/kg, with a primary endpoint of time to beginning of relief of symptoms. Santarus (NASDAQ: SNTS) has licensed certain exclusive rights from Pharming to commercialize RUCONEST® in North America for the treatment of acute attacks of HAE and other future indications. Under the terms of the license agreement, a \$10 million milestone is payable to Pharming upon successful achievement of the primary endpoint of the Phase III clinical study.

### **About RUCONEST® and Hereditary Angioedema**

RUCONEST® (INN conestat alfa) is a recombinant version of the human protein C1 inhibitor (C1INH). RUCONEST® is produced through Pharming's proprietary technology in milk of transgenic rabbits and is approved in Europe for the treatment of acute angioedema attacks in patients with HAE. RUCONEST® is an investigational drug in the U.S. and has been granted orphan drug designation for the treatment of acute attacks of HAE, a genetic disorder in which the patient is deficient in or lacks a functional plasma protein C1 inhibitor, resulting in unpredictable and debilitating episodes of intense swelling of the extremities, face, trunk, genitals, abdomen and upper airway. The frequency and severity of HAE attacks vary and are most serious when they involve laryngeal edema, which can close the upper airway and cause death by asphyxiation. According to the U.S. Hereditary Angioedema Association, epidemiological estimates for HAE range from one in 10,000 to one in 50,000 individuals.

### **About Pharming Group NV**

Pharming Group NV is developing innovative products for the treatment of unmet medical needs. RUCONEST® is a recombinant human C1 inhibitor approved for the treatment of angioedema attacks in patients with HAE in all 27 EU countries plus Norway, Iceland and Liechtenstein, and is distributed in the EU by Swedish Orphan Biovitrum (OMX: SOBI). RUCONEST® is partnered with Santarus, Inc (NASDAQ: SNTS) in North America where the drug is undergoing Phase III clinical development. The product is also being evaluated for follow-on indications in the areas of transplantation and reperfusion injury. The advanced technologies of the Company include innovative and validated platforms for the production of protein therapeutics, technology and processes for the purification and formulation of these products. A feasibility study, using the validated transgenic rabbit platform, aimed at the development of recombinant Factor VIII for the treatment of Haemophilia A is underway with partner, Renova Life, Inc. Additional information is available on the Pharming website, [www.pharming.com](http://www.pharming.com). To download the Pharming Group Investor Relations App, click [here](#).

*This press release contains forward looking statements that involve known and unknown risks, uncertainties and other factors, which may cause the actual results, performance or achievements of the Company to be materially different from the results, performance or achievements expressed or implied by these forward looking statements.*

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