## **PHARMING**

# PHARMING ANNOUNCES NEW DATA ON RUCONEST IN PRIMATE MODEL OF DELAYED GRAFT FUNCTION (DGF)

**Leiden, The Netherlands, May 23, 2013.** Biotech company Pharming Group NV ("Pharming" or "the Company") (NYSE Euronext: PHARM) announced today that recombinant human C1 esterase inhibitor (rhC1INH; RUCONEST®), an investigational drug in the U.S., has been shown to have a beneficial effect as a donor pre-treatment therapy in an animal model of kidney transplantation. The results of the study were presented at the American Transplant Congress in Seattle, Washington.

In the study, Dr. Luis Fernandez of the University of Wisconsin used a non-human primate model to evaluate the outcomes of kidney transplantation from brain dead donors. Kidneys that were treated with rhC1INH prior to transplantation had a significantly lower incidence of delayed graft function (DGF) when transplanted to the recipient animals. Dr. Fernandez and colleagues were also able to show how rhC1INH inhibited the complement cascade to confer this benefit.

DGF, a form of ischemia-reperfusion injury (IRI), is a serious and costly complication in the clinical transplantation setting. When DGF occurs, it necessitates the use of dialysis and leads to prolonged hospitalization, which results in adverse long term outcomes and significantly higher costs. As demonstrated with this animal model, donor pre-treatment with rhC1INH prior to transplantation could potentially represent a novel approach to addressing some of the limitations of current strategies to reduce the impact of DGF. Current interventions focus on activities that occur after the organ is harvested from the donor (e.g. cold storage or machine perfusion of the organ).

Pharming and its partners continue to be interested in the broad therapeutic areas of IRI and capillary leak syndromes and continue to review indications for potential future clinical development.

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

RUCONEST (INN conestat alfa) is a recombinant version of the human protein C1 esterase inhibitor, and is produced with Pharming's proprietary transgenic technology. RUCONEST is approved in Europe for the treatment of acute angioedema attacks in patients with HAE, a genetic disorder in which the patient is deficient in or lacks a functional plasma protein C1 esterase inhibitor, resulting in unpredictable and debilitating episodes of intense swelling. The swelling may occur in one or more anatomical areas, including 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. RUCONEST is an investigational drug in the U.S. and has been granted orphan drug designation by the FDA both for the treatment of acute attacks of HAE and for prophylactic treatment of HAE.

#### **About Pharming Group NV**

Pharming Group NV is developing innovative products for the treatment of unmet medical needs. RUCONEST® is a recombinant human C1 esterase 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. RUCONEST® is partnered with Santarus, Inc. (NASDAQ: SNTS) in North America and a BLA for RUCONEST was submitted to the FDA in April 2013. The product is also being evaluated for various follow-on indications. Pharming has a unique GMP compliant, validated platform for the production of recombinant human proteins that has proven capable of producing industrial volumes of high quality recombinant human protein in a more economical way compared to current cell based technologies. Pharming now plans to utilise this platform for the development of rhFVIII for the treatment of Haemophilia A. Additional information is available on the Pharming website, www.pharming.com.

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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