# **PHARMING**

# PHARMING UPDATES ON ONGOING CALL OF SECOND TRANCHE OF €10 MILLION EQUITY WORKING CAPITAL FACILITY

**Leiden, The Netherlands, September 7, 2012.** Biotech company Pharming Group NV ("Pharming" or "the Company") (NYSE Euronext: PHARM) today announces that the investors have increased the ongoing call under the facility by 9,408,000 shares from 16,000,000 to 25,408,000.

Pharming also re- confirms that short selling is prohibited under this facility and also wishes to re- emphasize that both the timing and the amount of any tranche is determined by Pharming. Under the initial call of 16,000,000 shares made by the Company as announced on September 5, 2012, the individual investors have an option to purchase that is limited to up to 600% of these 16,000,000 shares during the 15 trading days of this call.

Following the issue of the 9,408,000 shares, the number of outstanding shares increases from 823,817,129 shares to 833,225,129 shares.

#### **RUCONEST® Phase III Study**

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 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. The study is expected to be completed by the end of the third quarter of 2012.

## 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 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, <a href="https://www.pharming.com">www.pharming.com</a>. To download the Pharming Group Investor Relations App, click <a href="https://www.pharming.com">here.</a>.

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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