

# PHARMING AND SWEDISH ORPHAN BIOVITRUM ANNOUNCE FIRST SALES OF RUCONEST IN EUROPE

**Leiden, the Netherlands, and Stockholm, Sweden, December 28, 2010.** Pharming Group NV (NYSE Euronext: PHARM) and Swedish Orphan Biovitrum (STO: SOBI) announce the first European sales of Ruconest™ (Conestat alfa) in Denmark and Norway.

Ruconest<sup>™</sup> is the first recombinant C1 esterase inhibitor for acute treatment of angioedema attacks in patients with Hereditary Angioedema (HAE). Ruconest<sup>™</sup> is developed by the Dutch Biotech company Pharming and is marketed and distributed throughout the EU by Swedish Orphan Biovitrum.

On October 28<sup>-</sup> 2010, Ruconest<sup>™</sup> received European Marketing Authorization in the 27 EU countries plus Norway, Iceland and Liechtenstein. A number of these countries are now ready to sell Ruconest<sup>™</sup>.

"We are very pleased to provide Ruconest™ as the first recombinant C1 esterase inhibitor to European patients suffering from this severe, disabling, and life threatening disease", says Anders Edvell, Vice President, Marketing and Sales at Swedish Orphan Biovitrum. "The pharmacy purchasing price, estimated from the first approved prices in Europe, will be approximately €1800 per vial, which represents a competitive price per unit C1 inhibitor."

Sijmen de Vries, CEO of Pharming: "The European launch of Ruconest™ is a very important moment for all patients suffering from acute attacks of HAE. Enzyme replacement with human C1 inhibitor, the well established gold standard for HAE treatment; will from now on become available to all patients in all countries of the EU for the treatment of the various types of acute attacks of HAE."

## About HAE and C1 esterase inhibitor

HAE is a human genetic disorder in which the patient is deficient in or lacks a functional plasma protein C1 inhibitor, resulting in an overreaction of the immune system. The disease is characterized by unpredictable and debilitating episodes of intense swelling of the extremities, face, trunk, genitals, abdomen and upper airway, which may last up to five days when untreated. In addition to the life-threatening nature of the disease in case of laryngeal attacks, quality of life for individuals with the disease may be seriously impaired. Approximately one in 30,000 individuals (1:10,000 – 1:50,000) suffers from HAE with an average of approximately eight acute attacks per year.



### **About Ruconest**

Ruconest™ (INN conestat alfa), the first recombinant C1 esterase inhibitor, is a recombinant human protein developed through Pharming's proprietary technology in milk of transgenic rabbits. In the clinical data presented by Zuraw et al in Journal of Allergy and Clinical Immunology (October 2010), the combined results of two similar but independent, randomized, placebo-controlled studies were presented. Efficacy and safety in 70 HAE patients were evaluated in two doses of 50 and 100 U/Kg of rhC1INH for the treatment of acute angioedema attacks. Both doses of rhC1INH (50U/kg and 100U/kg) significantly reduced the time to beginning of relief of symptoms for all anatomical locations studied (abdominal, genitourinary, facial-laryngeal or peripheral) compared to placebo and resulted in a high response rate for rhC1INH (>90%). Adverse events occurred less frequently in the rhC1INH arm than in the placebo arm. No neutralizing antibodies against rhC1INH or host-related impurities were observed. The authors conclude that rhC1INH constitutes a highly effective alternative to plasma derived C1INH for the treatment of acute angioedema attacks in HAE patients.

# **About Swedish Orphan Biovitrum**

Swedish Orphan Biovitrum is a Swedish-based niche specialty pharmaceutical company with an international market presence. The company is focused on providing and developing specialist pharmaceuticals for rare disease patients with high medical needs. The portfolio consists of about 60 marketed products and an emerging late-stage clinical development pipeline. Our focus areas are: hemophilia, inflammation/autoimmune diseases, fat malabsorption, cancer and inherited metabolic disorders. Swedish Orphan Biovitrum had proforma revenues 2009e of about 2 BSEK and approximately 500 employees. The head office is located in Sweden and the share (STO: SOBI) is listed on NASDAQ OMX Stockholm. For more information please visit www.sobi.com.

## **About Pharming Group NV**

Pharming Group NV is developing innovative products for the treatment of unmet medical needs. Ruconest™ (Rhucin® in non-European territories) 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. The product is also under development for follow-on indications, i.e. antibody-mediated rejection (AMR) and delayed graft function (DGF) following kidney transplantation. The advanced technologies of the Company include innovative platforms for the production of protein therapeutics, technology and processes for the purification and formulation of these products. 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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