



## PHARMING

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### PHARMING AND SANTARUS ANNOUNCE NEW DATA FROM OPEN-LABEL REPEAT TREATMENT STUDY WITH RUCONEST (RECOMBINANT HUMAN C1 ESTERASE INHIBITOR)

#### **RUCONEST improved symptoms of acute hereditary angioedema attacks with similar results for repeated treatments**

**LEIDEN, Netherlands and SAN DIEGO (November 8, 2013)** – Pharming Group NV (NYSE Euronext: PHARM) and Santarus, Inc. (NASDAQ: SNTS) announced that new data from an open-label extension of the pivotal Phase III clinical study with RUCONEST® (recombinant human C1 esterase inhibitor, or rhC1INH) will be featured in a poster presentation on November 9 & 10, 2013 at the 2013 American College of Allergy, Asthma & Immunology Annual Scientific Meeting at the Convention Center in Baltimore, Maryland. The poster is titled, *Efficacy and Safety of Recombinant Human C1 Esterase Inhibitor for Acute Attacks of Hereditary Angioedema: An Open-Label Study*.

RUCONEST was administered for the treatment of 224 repeat acute angioedema attacks in 44 patients with hereditary angioedema (HAE) following initial treatment in the pivotal randomized controlled clinical study. RUCONEST is an investigational drug in the U.S. and has been granted orphan drug designation by the U.S. Food and Drug Administration (FDA) for the treatment of acute attacks of HAE.

“The results from this clinical trial are consistent with findings from previous studies supporting that RUCONEST reduces time to symptom relief when used for repeat HAE attacks,” said H. Henry Li, M.D., Ph.D., Director of Chevy Chase Clinical Research, Institute for Asthma and Allergy, Chevy Chase, Maryland. “In addition, it is important clinically to evaluate the safety findings from these multiple exposures.”

The median time in minutes (95% confidence interval [CI]) to onset of symptom relief following treatment as measured by patient responses to a Treatment Effect Questionnaire (TEQ) for the first five repeat attacks ranged from 62.5 (48, 90) to 134.0 (75, 150) and across all attacks was a median of 75.0 (69, 89). The median (95% CI) time in minutes to minimal symptoms (first three attacks per patient) as measured by a TEQ ranged from 243 (76, 1440) to 304 (150, 719) and for all assessed attacks was 303 (211, 367).

#### **Exposure and Safety Information**

- Only one dose of RUCONEST was administered for 96% of the 224 attacks.
- 12 of 44 (27%) patients experienced at least one treatment emergent adverse event (TEAE) within 72 hours of completion of RUCONEST infusion.

- TEAEs occurring in 5% of patients were nasopharyngitis, cough, fibrin D-dimer increase, and headache
- The percentage of patients experiencing TEAEs did not increase with RUCONEST treatments for repeat attacks.
- There were no discontinuations due to adverse events, no thrombotic or anaphylactic events, and no neutralizing anti-C1INH antibodies observed with repeat RUCONEST treatment.

Santarus and Pharming are seeking U.S. marketing approval of RUCONEST for the treatment of acute angioedema attacks in patients with HAE. The Biologics License Application (BLA) filing for RUCONEST is under review by the U.S. Food and Drug Administration (FDA) with a response expected by April 16, 2014. RUCONEST is approved in Europe for the treatment of acute angioedema attacks in patients with HAE and is an investigational drug in the U.S. that has been granted orphan drug designation by the FDA.

### **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.

### **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 Biologics License Application for RUCONEST is under review by the U.S. Food and Drug Administration. 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. In July 2013, the Platform was partnered with Shanghai Institute for Pharmaceutical Industry (SIPI), a Sinopharm Company, for joint global development of new products. Pre-clinical development and manufacturing will take place at SIPI and are funded by SIPI. Pharming and SIPI initially plan 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](http://www.pharming.com).

### **About Santarus**

Santarus, Inc. is a specialty biopharmaceutical company focused on acquiring, developing and commercializing proprietary products that address the needs of patients treated by physician specialists. The company's current commercial efforts are focused on five products. [UCERIS®](#) (budesonide) extended release tablets for the induction of remission in patients with active, mild to moderate ulcerative colitis and [ZEGERID®](#) (omeprazole/sodium bicarbonate) for the treatment of certain upper gastrointestinal disorders are promoted to gastroenterologists. [GLUMETZA®](#) (metformin hydrochloride extended release tablets) and [CYCLOSET®](#) (bromocriptine mesylate) tablets, which are indicated as adjuncts to diet and exercise to improve glycemic control in adults with type 2

diabetes, and [FENOGLIDE](#)<sup>®</sup> (fenofibrate) tablets, which is indicated as an adjunct to diet to reduce high cholesterol, are promoted to endocrinologists and other physicians who treat patients with type 2 diabetes. Full prescribing and safety information for Santarus' products is available at [www.santarus.com](http://www.santarus.com) or by contacting Santarus at 1-888-778-0887.

Santarus' product development pipeline includes the investigational drug RUCONEST<sup>®</sup> (recombinant human C1 esterase inhibitor). A Biologics License Application for RUCONEST for the treatment of acute angioedema attacks in patients with hereditary angioedema is under review by the U.S. Food and Drug Administration with a response expected in April 2014. Santarus is also developing rifamycin SV MMX<sup>®</sup>, which is in Phase III clinical testing for the treatment of travelers' diarrhea. In addition, the company has completed a Phase I clinical program with SAN-300, an investigational monoclonal antibody. More information about Santarus is available at [www.santarus.com](http://www.santarus.com).

*Santarus and Pharming caution you that statements included in this press release that are not a description of historical facts are forward-looking statements. The inclusion of forward-looking statements should not be regarded as a representation by Santarus or Pharming that any of its plans or objectives will be achieved. Actual results may differ materially from those set forth in this release due to the risks and uncertainties inherent in Santarus and Pharming's businesses, including, without limitation: whether the FDA will approve the RUCONEST BLA in a timely manner or at all; whether the FDA will concur with the clinical interpretation of the Phase III study results or the conduct of the study; whether the FDA ultimately will require additional clinical studies or other development programs before approving RUCONEST; risks related to Santarus' dependence on Pharming for many functions related to RUCONEST, and Pharming's ability to continue to perform these functions based on its limited financial resources; risks related to the license and supply arrangements between Santarus and Pharming, including the potential for termination of the arrangements; other difficulties or delays in development, testing, manufacturing and marketing of, and obtaining and maintaining regulatory approvals for, Santarus and Pharming's products; and other risks detailed in prior press releases as well as in public periodic filings with the Securities and Exchange Commission, including Santarus' Quarterly Report on Form 10-Q for the quarter ended September 30, 2013.*

*You are cautioned not to place undue reliance on these forward-looking statements, which speak only as of the date hereof. All forward-looking statements are qualified in their entirety by this cautionary statement and neither Santarus nor Pharming undertakes any obligation to revise or update this news release to reflect events or circumstances after the date hereof, except as may be required by law. This caution is made under the safe harbor provisions of Section 21E of the Private Securities Litigation Reform Act of 1995.*

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