



## PHARMING

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## **PHARMING AND SANTARUS ANNOUNCE SUBMISSION OF RUCONEST BIOLOGICS LICENSE APPLICATION TO FDA**

**LEIDEN, The Netherlands and SAN DIEGO, Calif. (April 17, 2013)** –Pharming Group NV (NYSE Euronext: PHARM) and Santarus, Inc. (NASDAQ: SNTS) today announced the submission of a Biologics License Application (BLA) to the U.S. Food and Drug Administration (FDA) to obtain marketing approval for RUCONEST® (recombinant human C1 esterase inhibitor) 50 U/kg, an investigational drug for the treatment of acute angioedema attacks in patients with hereditary angioedema (HAE).

The safety and efficacy of RUCONEST for the treatment of HAE attacks were evaluated in a clinical program that included a Phase III randomized placebo-controlled study conducted under a Special Protocol Assessment agreement with the FDA. The pivotal Phase III clinical study showed statistically significant and clinically relevant improvement in the primary endpoint of time to beginning of relief of symptoms for RUCONEST compared with placebo. The RUCONEST clinical program also included two additional randomized placebo-controlled studies and four open label treatment studies. In total, the BLA dossier includes ten clinical studies covering 940 administrations in 236 subjects.

Santarus licensed certain exclusive rights from Pharming to commercialize RUCONEST in North America for the treatment of acute attacks of HAE as well as other potential future indications. Under the terms of the license agreement, a \$5 million milestone is payable to Pharming upon FDA acceptance for review of the BLA for RUCONEST.

### **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<sup>®</sup> 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<sup>®</sup> 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](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<sup>™</sup>](#) (budesonide) extended release tablets for the induction of remission in patients with active, mild to moderate ulcerative colitis and [ZEGERID<sup>®</sup>](#) (omeprazole/sodium bicarbonate) for the treatment of certain upper gastrointestinal disorders are promoted to gastroenterologists. [GLUMETZA<sup>®</sup>](#) (metformin hydrochloride extended release tablets) and [CYCLOSET<sup>®</sup>](#) (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).

Santarus' product development pipeline includes the investigational drug RUCONEST<sup>®</sup> (recombinant human C1 esterase inhibitor). A BLA for RUCONEST was submitted to the FDA in April 2013 for the treatment of acute angioedema attacks in patients with HAE. Santarus is also developing rifamycin SV MMX<sup>®</sup>, which is in Phase III clinical testing for 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 accept the RUCONEST BLA for review following submission and ultimately approve it; 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 accepting the RUCONEST BLA or 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.*

*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. This caution is made under the safe harbor provisions of Section 21E of the Private Securities Litigation Reform Act of 1995.*

*Santarus<sup>®</sup>, FENOGLIDE<sup>®</sup>, UCERIS<sup>™</sup>, and ZEGERID<sup>®</sup> are trademarks of Santarus, Inc. GLUMETZA<sup>®</sup> is a trademark of Biovail Laboratories International S.r.l. licensed exclusively in the United States to Depomed, Inc. CYCLOSET<sup>®</sup> is a trademark of VeroScience LLC. MMX<sup>®</sup> is a trademark of Cosmo Technologies Limited. RUCONEST<sup>®</sup> is a trademark of Pharming Group N.V.*

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