

PHARMING AND SALIX ANNOUNCE INITIATION OF CLINICAL STUDY OF RUCONEST® FOR PROPHYLAXIS OF HEREDITARY ANGIOEDEMA

LEIDEN, THE NETHERLANDS, RALEIGH, NC, August 28, 2014 - Pharming Group NV (EURONEXT: PHARM) and Salix Pharmaceuticals, Ltd. (NASDAQ:SLXP) today announced the initiation of a clinical study of Ruconest®, (C1 Esterase Inhibitor [Recombinant]) 50 IU/kg, for attack prophylaxis in patients with hereditary angioedema (HAE).

The Phase 2 randomized, double-blind, placebo-controlled, crossover study will evaluate the safety and efficacy of Ruconest® when used for the prophylaxis of angioedema attacks in patients with HAE. The study will enroll approximately 30 patients and compare a dosing regimen of Ruconest® given either once or twice weekly versus placebo over a total of 12 weeks. The initiation of the study follows the filing of an Investigational New Drug (IND) application with the U.S. Food and Drug Administration (FDA). The study will be conducted at sites in Europe and the United States.

"We expect the results of the study to be highly informative in determining the clinical benefit of Ruconest® to prevent HAE attacks," said Bruno Giannetti, MD PhD, Chief Operating Officer of Pharming.

This randomized-controlled study follows an open-label study with Ruconest® given once weekly, which was published in the peer reviewed journal *Allergy* in November 2012. The 25 patients included in the open-label study had a history of frequent HAE attacks (mean 0.9 attacks/week). During the 8 week Ruconest® treatment period, the mean frequency of HAE attacks was reduced by more than 50% to 0.4 attacks/ week. The repeated administrations were generally safe and well-tolerated. The results of that open-label study suggest that Ruconest® could be effective in providing long term prophylaxis in patients with frequent attacks.

Under the terms of the Pharming-Salix license agreement, the companies will equally share the development costs for Ruconest® for HAE prophylaxis and Pharming will receive an undisclosed milestone payment from Salix at FDA approval for this additional indication.

In July 2014, the US FDA approved Ruconest®, the first and only plasma-free, recombinant C1-INH, for the treatment of acute attacks in adult and adolescent patients with hereditary angioedema (HAE). Effectiveness was not established in HAE patients with laryngeal attacks. Salix currently plans on making RUCONEST available to patients during the fourth quarter of 2014.

RUCONEST® is manufactured by Pharming Group NV in the Netherlands. Salix has licensed exclusive rights from Pharming to commercialize RUCONEST® in North America and market RUCONEST® for the treatment of acute HAE attack symptoms.

Indication:

RUCONEST® is a C1 esterase inhibitor [recombinant] indicated for the treatment of acute attacks in adult and adolescent patients with hereditary angioedema (HAE). Effectiveness was not established in HAE patients with laryngeal attacks.

Important Safety Information:

RUCONEST® (C1 esterase inhibitor [recombinant]) is contraindicated in patients with a history of allergy to rabbits or rabbit-derived products, and patients with a history of life-threatening immediate hypersensitivity reactions to C1 esterase inhibitor preparations, including anaphylaxis.

Severe hypersensitivity reactions may occur. The signs and symptoms of hypersensitivity reactions may include hives, generalized urticaria, tightness of the chest, wheezing, hypotension, and/or anaphylaxis during or after injection of RUCONEST. Should symptoms occur, discontinue RUCONEST and administer appropriate treatment. Because hypersensitivity reactions may have symptoms similar to HAE attacks, treatment methods should be carefully considered.

Serious arterial and venous thromboembolic (TE) events have been reported at the recommended dose of plasma derived C1 esterase inhibitor products in patients with risk factors. Risk factors may include the presence of an indwelling venous catheter/access device, prior history of thrombosis, underlying atherosclerosis, use of oral contraceptives or certain androgens, morbid obesity, and immobility. Monitor patients with known risk factors for TE events during and after RUCONEST administration.

RUCONEST has not been studied in pregnant women; therefore, should only be used during pregnancy if clearly needed.

The most common adverse reactions (incidence $\geq 2\%$) were headache, nausea, and diarrhea. The serious adverse reaction in clinical studies of RUCONEST was anaphylaxis.

Please see complete Prescribing Information for RUCONEST.

About RUCONEST®

RUCONEST® (C1 Esterase Inhibitor [Recombinant]) 50 IU/kg is an injectable medicine that is used to treat acute angioedema attacks in adult and adolescent patients with hereditary angioedema (HAE). HAE is caused by a deficiency of the C1 esterase inhibitor protein, which is present in blood and helps control inflammation (swelling) and parts of the immune system. A shortage of C1 esterase inhibitor can lead to repeated attacks of swelling, pain in the abdomen, difficulty breathing and other symptoms. RUCONEST® contains C1 esterase inhibitor at 50 IU/kg.

When administered at the onset of HAE attack symptoms at the recommended dose, RUCONEST® works to return a patient's C1-INH levels to normal range and quickly relieve the symptoms of an HAE attack with a low recurrence of symptoms.

RUCONEST® was approved by the U.S. Food and Drug Administration (FDA) on July 17, 2014 and is the first and only plasma-free, recombinant C1-INH approval.

About HAE

RUCONEST has been granted Orphan Drug designation by the FDA for the treatment of acute angioedema attacks in patients with hereditary angioedema (HAE). With RUCONEST now approved by the FDA, Salix believes this designation should provide seven years of marketing exclusivity in the United States.

Hereditary angioedema (HAE) is a genetic condition occurring between 1 in 10,000 to 1 in 50,000 people. Those with HAE experience episodes of swelling in their extremities, face and abdomen, with potentially life-threatening swelling of the airway. When it occurs in the abdomen, this swelling can be accompanied by bouts of nausea, vomiting and severe pain. Swelling in the face or extremities can be painful, disfiguring, and disabling.

HAE patients have a defect in the gene that controls production of a protein found in the blood vessels, called C1 inhibitor or C1-INH. When a person's C1-INH levels are low, fluid from blood vessels can leak into nearby connective tissues, causing severe pain and swelling and, in rare cases, death from asphyxiation from airway swelling.

About Pharming Group NV

Pharming Group NV is developing innovative products for the treatment of unmet medical needs. RUCONEST® (conestat alfa) is a recombinant human C1 esterase inhibitor approved for the treatment of angioedema attacks in patients with HAE in the USA, Israel, all 27 EU countries plus Norway, Iceland and Liechtenstein. RUCONEST® is distributed in the EU by Swedish Orphan Biovitrum. RUCONEST® is partnered with Salix Pharmaceuticals Inc. (NASDAQ: SLXP) in North America. 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 utilize this platform for the development of rhFVIII for the treatment of Haemophilia A. Additional information is available on the Pharming website; www.pharming.com.

About Salix Pharmaceuticals

Salix Pharmaceuticals, Ltd., headquartered in Raleigh, North Carolina, develops and markets prescription pharmaceutical products and medical devices for the prevention and treatment of gastrointestinal diseases. Salix's strategy is to in-license late-stage or marketed proprietary therapeutic products, complete any required development and regulatory submission of these products, and commercialize them through the Company's 500-member specialty sales force.

Salix trades on the NASDAQ Global Select Market under the ticker symbol "SLXP".

PHARMING

For more information, please visit our website at www.salix.com or contact Salix at 919-862-1000. Follow us on Twitter (@SalixPharma) and Facebook (www.facebook.com/SalixPharma). Information on our Twitter feed, Facebook page and website is not incorporated in our filings with the SEC.

Salix Disclosure Notice

As previously announced on July 8, 2014, Salix, Cosmo Pharmaceuticals S.p.A. and Irish domiciled Cosmo Technologies Limited entered into an Agreement and Plan of Merger and Reorganization, pursuant to which a subsidiary of Cosmo Technologies Limited will merge with and into Salix, with Salix as the surviving entity, and Salix will become an indirect, wholly-owned subsidiary of Cosmo Technologies Limited, which will change its name to Salix Pharmaceuticals, plc.

Please Note: The statements provided herein that are not historical facts are or might constitute projections and other forward-looking statements regarding future events. Although we believe the expectations reflected in such forward-looking statements are based on reasonable assumptions, our expectations might not be attained. Forward-looking statements are just predictions and are subject to known and unknown risks and uncertainties that could cause actual events or results to differ materially from expected results. Factors that could cause actual events or results to differ materially from those described herein include, among others: uncertainties as to the ability to successfully complete the proposed transaction in accordance with its terms and in accordance with the expected schedule; the possibility that competing offers will be made; the possibility that various closing conditions for the proposed transaction may not be satisfied or waived, including that a governmental entity may prohibit or refuse to grant any approval required for the consummation of the proposed transaction; the unpredictability of the duration and results of regulatory review of New Drug Applications, Biologics License Agreements, and Investigational NDAs; generic and other competition in an increasingly global industry; litigation and the possible impairment of, or inability to obtain, intellectual property rights and the costs of obtaining such rights from third parties in an increasingly global industry; the cost, timing and results of clinical trials and other development activities involving pharmaceutical products; post-marketing approval regulation, including the ongoing Department of Justice investigation of Salix's marketing practices; market acceptance for approved products; revenue recognition and other critical accounting policies; the need to acquire new products; changes in tax laws or interpretations thereof; general economic and business conditions; and other factors. Readers are cautioned not to place undue reliance on the forward-looking statements included herein, which speak only as of the date hereof. Salix does not undertake to update any of these statements in light of new information or future events, except as required by law. The reader is referred to the documents that Salix files from time to time with the SEC.

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.

Contact

Sijmen de Vries, CEO: T: +31 71 524 7400

FTI Consulting

Julia Phillips/ John Dineen, T: +44 203 727 1136

Salix Pharmaceuticals Ltd.

Adam C. Derbyshire

Executive Vice President and Chief Financial Officer

T: +1 919-862-1000

G. Michael Freeman

Associate Vice President, Investor Relations and Corporate Communications

T: +1 919-862-1000