PHARMING

PHARMING AND SALIX ANNOUNCE FDA APPROVAL OF RUCONEST® FOR THE TREATMENT OF ACUTE ANGIOEDEMA ATTACKS IN PATIENTS WITH HEREDITARY ANGIOEDEMA (HAE)

RUCONEST® (C1 ESTERASE INHIBITOR [RECOMBINANT]) 50 IU/kg IS THE FIRST AND ONLY RECOMBINANT TREATMENT OPTION FOR ADULT AND ADOLESCENT PATIENTS SUFFERING FROM HEREDITARY ANGIOEDEMA

LEIDEN, THE NETHERLANDS, RALEIGH (NC), July 17, 2014 - Pharming Group NV (EURONEXT: PHARM) and Salix Pharmaceuticals, Ltd. (NASDAQ:SLXP), today announced that the Food and Drug Administration has approved RUCONEST® (C1 Esterase Inhibitor [Recombinant]) 50 IU/kg for the treatment of acute angioedema attacks in adult and adolescent patients with hereditary angioedema (HAE). Because of the limited number of patients with laryngeal attacks, effectiveness was not established in HAE patients with laryngeal attacks.

“We are pleased that RUCONEST® provides the HAE community with another FDA-approved option for treating painful and debilitating HAE attacks,” said Anthony Castaldo, President of the Hereditary Angioedema Association (US HAEA), a non-profit patient services and research organization with a membership of over 5,000 HAE patients in the United States.

RUCONEST® is a recombinant C1 esterase inhibitor that can be administered by the patient after receiving training by a healthcare provider. HAE attacks stem from a deficiency of the C1 inhibitor protein in the blood. HAE is a rare inherited genetic condition that is often not properly diagnosed until later in a patient’s life as the symptoms of an attack can mirror someone experiencing an allergic reaction. Severe, painful swelling can occur at any time, which means most people suffering from HAE deal with the constant fear of when their next attack might surface and how that might impair their lives and those around them.

“Results in the pivotal clinical trial demonstrate RUCONEST® is a safe and effective option for the treatment of acute hereditary angioedema attacks,” said Dr. Marc Riedl of the US HAEA Angioedema Center at the University of California – San Diego and primary investigator of the phase III study. “At the US HAEA Angioedema Center, we strive to better the lives of those suffering from hereditary angioedema and part of that is ensuring patients have access to advanced treatments that have been proven to work in clinical trials. RUCONEST is an important addition to those treatment options.”

Sijmen de Vries, CEO of Pharming, said: “The approval of RUCONEST® in the US is a very significant milestone for Pharming. For many years we have strived to make RUCONEST® - the first recombinant replacement therapy for C1Inhibitor deficiency - available to the HAE patient community in the US, because we were aware of the great value and benefit this product adds to patients’ lives. Today we are proud to have achieved this goal in the US.”

"RUCONEST® is a much needed treatment option for patients suffering from acute attacks of hereditary angioedema. Until now, there hasn't been an FDA approved recombinant C1 esterase inhibitor option to treat symptoms of HAE," said Carolyn J. Logan, President and Chief Executive Officer of Salix. "The unpredictability of HAE can make patients feel uncertain about when their next attack might strike, which is why it is important to have a medicine that can be administered by the patient that resolves an attack. Salix is proud to make RUCONEST® available."

The FDA approval of the Biologics License Agreements (BLA) for RUCONEST® for treatment of acute angioedema attacks in patients with HAE is based on a randomized, double-blind, placebo-controlled, phase III trial (RCT) which included an open-label extension (OLE) phase and is supported by the results of two additional RCTs and two additional OLE studies. The pivotal RCT and OLE studies analyzed the results from 44 subjects who experienced 170 HAE attacks. The primary efficacy endpoint was the time to beginning of symptom relief, assessed using patient-reported responses to two questions about the change in overall severity of their HAE attack symptoms after the start of treatment. These were assessed at regular time points for each of the affected anatomical locations for up to 24 hours. To achieve the primary endpoint, a patient had to have a positive response to both questions along with persistence of improvement at the next assessment time (i.e., the same or better response).
A statistically significant difference in the time to beginning of symptom relief was observed in the intent-to-treat population (n=75) between RUCONEST and placebo (p=0.031, log-rank test); the median time to beginning of symptom relief was 90 minutes for RUCONEST patients (n=44) and 152 minutes for placebo patients (n=31).

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.

Salix currently plans on making RUCONEST® accessible to patients later in 2014.

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 ≥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® is the first and only plasma-free, recombinant C1-INH approval from the U.S. Food and Drug Administration (FDA) and was approved in July 2014.

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.

Pharming Disclosure Notice:

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 Pharming to be materially different from the results, performance or achievements expressed or implied by these forward looking statements.

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 markets XIFAXAN® (rifaximin) tablets 200 mg and 550 mg, MOVIPREP® (PEG 3350, sodium sulfate, sodium chloride, potassium chloride, sodium ascorbate and ascorbic acid for oral solution, 100 g/7.5 g/2.691 g/1.015 g/5.9 g/4.7 g), OSMOPREP® (sodium phosphate monobasic monohydrate, USP, and sodium phosphate dibasic anhydrous, USP) Tablets, APRISO® (mesalamine) extended-release capsules 0.375 g, UCERIS® (budesonide) extended release tablets, for oral use, GIAZO® (balsalazide disodium) tablets, COLAZAL® (balsalazide disodium) Capsules, GLUMETZA® (metformin hydrochloride extended-release tablets) 500 mg and 1000 mg,

ZEGERID® (omeprazole/sodium bicarbonate) Powder for Oral Suspension, ZEGERID® (omeprazole/sodium bicarbonate) Capsules, METOZOLV® ODT (metoclopramide hydrochloride), RELISTOR® (methylnaltrexone bromide) Subcutaneous Injection, FULYZAQ® (crofelemer) delayed-release tablets, SOLESTA®, DEFLUX®, PEPCID® (famotidine) for Oral Suspension, DIURIL® (chlorothiazide) Oral Suspension, AZASAN® (azathioprine) Tablets, USP, 75/100 mg, ANUSOL-HC® 2.5% (Hydrocortisone Cream, USP), ANUSOL-HC® 25 mg Suppository (Hydrocortisone Acetate), PROCTOCORT® Cream (Hydrocortisone Cream, USP) 1% and PROCTOCORT® Suppository (Hydrocortisone Acetate Rectal Suppositories) 30 mg, CYCLOSET® (bromocriptine mesylate) tablets, FENOGLIDE® (fenofibrate) tablets. UCERIS (budesonide) rectal foam, RELSITOR®, encapsulated bowel prep and rifaximin for additional indications are under development.

For full prescribing information and important safety information on Salix products, including BOXED WARNINGS for OSMOPREP, AZASAN, GLUMETZA and METOZOLV, please visit www.salix.com where the Company promptly posts press releases, SEC filings and other important information or contact the Company at 919 862-1000.

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

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