

## PHARMING AND HYUPJIN ANNOUNCE RECEIPT OF THE SOUTH KOREAN MARKETING AUTHORISATION FOR RUCONEST®

**Leiden, The Netherlands, and Seoul, South Korea, 21 December 2015:** Pharming Group N.V. ("Pharming" or "the Company") (Euronext Amsterdam: PHARM) and its partner, HyupJin Corporation, a Seoul based South Korean specialty pharma company, announce that HyupJin has received the marketing authorisation for RUCONEST® (recombinant human C1 inhibitor) in South Korea. RUCONEST is approved for the treatment of acute angioedema attacks in adult patients with hereditary angioedema HAE. Effectiveness was not established in HAE patients with laryngeal and oro-pharyngeal attacks. HyupJin will now seek reimbursement for RUCONEST in South Korea.

Sijmen de Vries, CEO of Pharming, commented: "HyupJin is a well-established and experienced company with a proven track record in South Korea. We look forward to them now extending the commercialisation of RUCONEST by providing HAE patients in South Korea with a new, safe and effective treatment to treat their HAE attacks."

Choon Duk Kim, General Manager of Hyupjin Corporation, said: "RUCONEST is the first approved recombinant C1 esterase inhibitor worldwide and this approval of RUCONEST will now enable us to provide the Korean HAE patients access to this most innovative way to treat their disease. We fully appreciate all the supports from Pharming Group for the approval of RUCONEST in South Korea and we look forward to getting started."

### About RUCONEST®

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

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 may help to return a patient's C1-INH levels to normal range and relieve the symptoms of an HAE attack with a low recurrence of symptoms within 24 hours.

RUCONEST is the only recombinant C1-INH approval from the U.S. Food and Drug Administration (FDA) and was approved in July 2014.

Under the Biologics Price Competition and Innovation Act of 2009, RUCONEST was granted Market exclusivity in the USA until July 2026

RUCONEST is designated as an orphan drug by the FDA for the treatment of acute attacks of angioedema caused by hereditary or acquired C1-INH deficiency.

### About HAE

Hereditary Angioedema (HAE) is a rare genetic disorder. It is characterised by spontaneous and recurrent episodes of swelling (edema attacks) of the skin in different parts of the body, as well as in the airways and internal organs. Edema of the skin usually affects the extremities, the face, and the genitals. Patients suffering from this kind of edema often withdraw from their social lives because of the disfiguration, discomfort and pain these symptoms may cause. Almost all HAE patients suffer from bouts of severe abdominal pain, nausea, vomiting and diarrhea caused by swelling of the intestinal wall.

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Edema of the throat, nose or tongue is particularly dangerous and potentially life-threatening and can lead to obstruction of the airway passages. Although there is currently no known cure for HAE, it is possible to treat the symptoms associated with edema attacks. HAE affects about 1 in 10,000 to 1 in 50,000 people worldwide. Experts believe that a lot of patients are still seeking the right diagnosis: although HAE is (in principle) easy to diagnose, it is frequently identified very late or not discovered at all. The reason HAE is often misdiagnosed is because the symptoms are similar to those of many other common conditions such as allergies or appendicitis. By the time it is diagnosed correctly, the patient has often been through a long lasting ordeal.

## **About Pharming Group N.V.**

Pharming Group N.V. 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 US, Israel, South Korea, all 28 EU countries plus Norway, Iceland and Liechtenstein.

RUCONEST is commercialised by Pharming in Austria, Germany and the Netherlands. RUCONEST is distributed by Swedish Orphan Biovitrum AB (publ) (SS: SOBI) in the other EU countries and in Azerbaijan, Belarus, Georgia, Iceland, Kazakhstan, Liechtenstein, Norway, Russia, Serbia and Ukraine.

RUCONEST is partnered with Salix Pharmaceuticals, Ltd. ("Salix") in North America. Salix is part of Valeant Pharmaceuticals International, Inc. (NYSE: VRX/TSX: VRX).

RUCONEST is also being investigated in a randomised Phase II clinical trial for prophylaxis of HAE and evaluated for various additional 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. Leads for Enzyme Replacement Therapy (ERT) in Pompe, Fabry's and Gaucher's diseases are under early evaluation. The platform is partnered with Shanghai Institute of 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 recombinant human Factor VIII for the treatment of Haemophilia A.

For more information, please visit <http://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 the Company to be materially different from the results, performance or achievements expressed or implied by these forward looking statements.*

## **About Hyupjin Corporation**

Headquartered in Seoul, Korea, Hyupjin Corporation develops and distributes healthcare products from prominent companies in the USA and Europe since 1975. With 30 years of experience in the medical market, Hyupjin Corporation has tight relationships with healthcare professionals and has built extensive know-how of the medical community. Hyupjin's product portfolio mainly consists of oncology and immunology products, but also OTCs, dietary supplements and medical devices. Hyupjin has made a consistent effort for the treatment of incurable diseases along with improvement of patient's quality of life. For more information, please visit [http://www.hyupjincorp.com/index\\_en.php](http://www.hyupjincorp.com/index_en.php).

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