

Pharming announces abstract presented at the American Academy of Allergy, Asthma & Immunology (AAAAI) Annual Meeting and sponsorship of the AAAAI Foundation Michael M. Frank, MD, FAAAAI Lectureship

Leiden, The Netherlands, 25 February 2019, Pharming Group N.V. (“Pharming” or “the Company”) (Euronext Amsterdam: PHARM) announced today that an abstract was presented this week at the American Academy of Allergy, Asthma & Immunology (AAAAI) Annual Meeting in San Francisco, CA.

Additionally, Pharming is pleased to announce sponsorship of the AAAAI Foundation Michael M. Frank, MD, FAAAAI Lectureship.

The presentation *Recombinant Human C1 Esterase Inhibitor as Short-Term Prophylaxis for Dental Procedures in Patients with Hereditary Angioedema: A Case Series* by Anna Valerieva, MD, PhD *et al.* described a retrospective study in patients diagnosed with hereditary angioedema (HAE) from Europe and the United States who received RUCONEST® (C1 esterase inhibitor [recombinant]) prior to dental procedures to prevent an HAE attack. The authors concluded that short-term prophylaxis with RUCONEST®, administered within approximately 60 minutes before the procedure, was efficacious and safe in adults and reduced the risk of an attack post-procedure. The presentation may be viewed at <https://synchronymed.com/smc-assets/AAAAI2019Poster111.pdf>

Pharming also announces sponsorship of one of the AAAAI Foundation 5-year named lectureships which provide funding for research that leads to the prevention and cure of asthma, allergic and immunologic diseases.

This year, the AAAAI Foundation is pleased to honor the life work of Dr. Michael Frank with the creation of the *Michael M. Frank, MD FAAAAI Lectureship*.

“Michael Frank has devoted his career to help improve the lives of HAE patients which makes him a perfect recipient of this lectureship”, stated Jonathan Bernstein, MD, FAAAAI—Lectureships and Major Gifts Chair AAAAI Foundation. “We are grateful to Pharming for their support in helping to establish the Michael Frank lectureship”.

The abovementioned presentation will be made available following the congress on Pharming’s website www.pharming.com.

About HAE

Hereditary Angioedema (HAE) is a rare genetic disorder. It is characterized 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.

Edema of the throat, nose or tongue can be particularly dangerous as this can lead to obstruction of the airway passages and be potentially life threatening. 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 RUCONEST®

US INDICATION

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.

IMPORTANT SAFETY INFORMATION

- RUCONEST® (C1 esterase inhibitor [recombinant]) is contraindicated in:
 - Patients with a history of allergy to rabbits or rabbit-derived products.
 - Patients with a history of life-threatening immediate hypersensitivity reactions to C1 esterase inhibitor preparations, including anaphylaxis.
- **Hypersensitivity:** Severe hypersensitivity reactions may occur. 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.
- **Thromboembolic Events:** 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.
- **Intravenous Use:** RUCONEST is for intravenous use after reconstitution only. No more than 2 doses should be administered within a 24-hour period.
- **Pregnancy and Nursing:** RUCONEST has not been studied in pregnant women; therefore, it should only be used during pregnancy if clearly needed. Advise patients to notify their physician if they are breastfeeding or plan to breastfeed.
- **Adverse reactions:** The serious adverse reaction in clinical studies of RUCONEST was anaphylaxis.
- **Common adverse reactions:** The most common adverse reactions (incidence $\geq 2\%$) were headache, nausea, and diarrhoea.

Please see Full Prescribing Information for RUCONEST® as applicable for various jurisdictions:

[FDA: RUCONEST®\[1\]](#) / [EMA: RUCONEST®\[2\]](#)

About Pharming Group N.V.

Pharming is a specialty pharmaceutical company developing innovative products for the safe, effective treatment of rare diseases and unmet medical needs. Pharming’s lead product, RUCONEST® (conestat alfa) is a recombinant human C1 esterase inhibitor approved for the treatment of acute Hereditary

Angioedema (“HAE”) attacks in patients in Europe, the US, Israel and South Korea. The product is available on a named-patient basis in other territories where it has not yet obtained marketing authorization.

RUCONEST® is distributed by Pharming in Austria, France, Germany, Luxembourg, the Netherlands, the United Kingdom and the United States of America. Pharming holds commercialisation rights in Algeria, Andorra, Bahrain, Belgium, Ireland, Jordan, Kuwait, Lebanon, Morocco, Oman, Portugal, Qatar, Syria, Spain, Switzerland, Tunisia, United Arab Emirates and Yemen. In some of these countries distribution is made in association with the HAEi Global Access Program (GAP).

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 distributed in Colombia, Costa Rica, the Dominican Republic, Panama, and Venezuela by Cytobioteck, in South Korea by HyupJin Corporation and in Israel by Kamada.

RUCONEST® is also being examined for approval for the treatment of HAE in young children (2-13 years of age) and evaluated for various additional follow-on indications.

Pharming’s technology platform includes a unique, GMP-compliant, validated process for the production of pure recombinant human proteins that has proven capable of producing industrial quantities of high quality recombinant human proteins in a more economical and less immunogenetic way compared with current cell-line based methods. Leads for enzyme replacement therapy (“ERT”) for Pompe and Fabry’s diseases are being optimized at present, with additional programs not involving ERT also being explored at an early stage at present.

Pharming has a long-term partnership with the China State Institute of Pharmaceutical Industry (“CSIPI”), a Sinopharm company, for joint global development of new products, starting with recombinant human Factor VIII for the treatment of Haemophilia A. Pre-clinical development and manufacturing will take place to global standards at CSIPI and are funded by CSIPI. Clinical development will be shared between the partners with each partner taking the costs for their territories under the partnership.

Additional information is available on the Pharming website: www.pharming.com

Forward-looking Statements

This press release of Pharming Group N.V. and its subsidiaries (“Pharming”, the “Company” or the “Group”) may contain forward-looking statements including without limitation those regarding Pharming’s financial projections, market expectations, developments, partnerships, plans, strategies and capital expenditures.

The Company cautions that such forward-looking statements may involve certain risks and uncertainties, and actual results may differ. Risks and uncertainties include without limitation the effect of competitive, political and economic factors, legal claims, the Company’s ability to protect intellectual property, fluctuations in exchange and interest rates, changes in taxation laws or rates, changes in legislation or accountancy practices and the Company’s ability to identify, develop and successfully commercialise new products, markets or technologies.

As a result, the Company’s actual performance, position and financial results and statements may differ materially from the plans, goals and expectations set forth in such forward-looking statements. The Company assumes no obligation to update any forward-looking statements or information, which should be taken as of their respective dates of issue, unless required by laws or regulations.

For further public information, contact

Pharming Group NV

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

Bruno Giannetti, COO: T: +31 71 524 7400

FTI Consulting, London, UK:

Victoria Foster Mitchell, T: +44 203 727 1136

LifeSpring Life Sciences Communication, Amsterdam, The Netherlands:

Leon Melens, Tel: +31 6 53 81 64 27