Pharming announces the presentation of the results of the RUCONEST® Phase II study for prophylaxis of Hereditary Angioedema attacks

Leiden, The Netherlands, 14 November 2016: Pharming Group N.V. (the "Company") (EURONEXT: PHARM) today announced that the results of its "Randomized, Double-Blind, Placebo-Controlled Trial of Recombinant Human C1 Inhibitor for Prophylaxis of Hereditary Angioedema Attacks", were presented by Marco Cicardi, Professor of Internal Medicine University of Milan, Hospital L. Sacco Milan and co-prinicipal investigator for the study. The presentation was held during the American College of Allergy, Asthma and Immunology 2016 Scientific Meeting ("ACAAI") meeting in San Francisco yesterday afternoon. The presentation can be found on our website, here.

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About HAE

Hereditary Angioedema (HAE) is a rare genetic disorder. The condition is caused by a deficiency of the C1 esterase inhibitor protein, which is normally present in blood and helps control inflammation (swelling) and parts of the immune system. Because defective C1-Inhibitor does not adequately perform its regulatory function, a biochemical imbalance can occur and produce unwanted peptides that induce the capillaries to release fluids into surrounding tissue, thereby causing swelling or edema.

HAE 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 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 angioedema 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®

RUCONEST® (recombinant C1 esterase inhibitor) 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.

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 esterase inhibitor 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 esterase inhibitor approved by the U.S. Food and Drug Administration (FDA) and was approved in July 2014.

Recently RUCONEST® demonstrated very positive data for prophylaxis (prevention) of acute attacks of HAE in patients with HAE. If approved for this indication, RUCONEST® will have access to this additional market. RUCONEST® therefore, has the potential to be the only recombinant C1 esterase inhibitor product approved to target both the acute market (worth approximately $845 million) and the HAE prophylaxis market (separately worth approximately $700 million).

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

EMA:
http://www.ema.europa.eu/docs/en_GB/document_library/EPAR_-_Product_Information/human/001223/WC500098542.pdf

FDA:
http://www.fda.gov/downloads/BiologicsBloodVaccines/BloodBloodProducts/ApprovedProducts/LicensedProductsBLAs/FractionatedPlasmaProducts/UCM405634.pdf

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 and rest of the world. The product is available on a named-patient basis in other territories where it has not yet obtained marketing authorization.

RUCONEST® is commercialized by Pharming in Algeria, Andorra, Austria, Bahrain, Belgium, France, Germany, Ireland, Jordan, Kuwait, Lebanon, Luxembourg, Morocco, Netherlands, Oman, Portugal, Qatar, Syria, Spain, Switzerland, Tunisia, United Arab Emirates, United Kingdom and Yemen.

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 the United States by a subsidiary of Valeant Pharmaceuticals International, Inc. (NYSE: VRX/TSX: VRX), following Valeant's acquisition of Salix Pharmaceuticals, Ltd.

RUCONEST® is distributed in Argentina, Colombia, Costa Rica, the Dominican Republic, Panama and Venezuela by Cytobioteck, in South Korea by HyupJin Corporation and in Israel by Megapharm.

RUCONEST® is also being investigated in a Phase II clinical trial 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 Pompé 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.

Pharming has declared that the Netherlands is its "Home Member State" pursuant to the amended article 5:25a paragraph 2 of the Dutch Financial Supervision Act.

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 commercialize 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.

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