

**THIS PRESS RELEASE AND THE INFORMATION CONTAINED HEREIN, IS RESTRICTED AND IS NOT FOR RELEASE, DISTRIBUTION OR PUBLICATION IN WHOLE OR IN PART, DIRECTLY OR INDIRECTLY, IN OR INTO THE UNITED STATES, AUSTRALIA, CANADA OR JAPAN OR ANY OTHER JURISDICTION IN WHICH THE SAME WOULD BE UNLAWFUL. PLEASE SEE THE IMPORTANT NOTICE AT THE END OF THIS PRESS RELEASE.**

## **PHARMING CONVENE AN EXTRAORDINARY GENERAL MEETING OF SHAREHOLDERS**

Leiden, The Netherlands, 23 August 2016: Pharming Group N.V. (the “Company”) (Euronext: PHARM) announces today that it will hold an Extraordinary General Meeting (EGM) of shareholders to discuss the Company’s definitive agreement to acquire all North American commercialisation rights to its product RUCONEST® (recombinant human C1 esterase inhibitor), including all rights in the US, Mexico and Canada, from Valeant Pharmaceuticals International, Inc. (“Valeant”) (NYSE/TSX: VRX) (the “Transaction, announced on 09 August 2016”), and to seek approval to increase the Company’s authorized share capital. The EGM will be held at Holiday Inn Leiden, Haagse Schouwweg 10, 2332 KG Leiden, the Netherlands on Wednesday 05 October 2016 at 14:00 CET.

- The acquisition is anticipated to be accretive to earnings from day one on a consolidated level and (based on the expected range of the equity issue) will also be accretive on a per share level immediately.
- The anticipated additional sales revenues, less the current costs and the increase in the cost base required to accelerate RUCONEST® in the USA, are expected to enable the Company to transform and reach profitability during 2017.
- To finance the Transaction and (mainly) investments in commercialization, the Company plans to raise between €73m and €90m as a combination of straight debt and equity. Out of this, the Company seeks to raise between €35m and €60m in equity financing.
- Out of the current Authorized Capital of 650 million, the Company holds 184 million shares in treasury.
- To ensure that sufficient share capital is available, the Company seeks approval from shareholders for an increase in the authorized share capital of 150 million shares, such that up to a total of 334 million shares become available to enable the Transaction to close, although the Company expects that not all of this increase may be needed for issuance.
- Based on preliminary discussions, the Company expects to need between 120 and 240 million shares to complete the equity financing.

The Board of Management believe that the time is now right for Pharming to take back these rights and accelerate the sales of RUCONEST® with additional investment in sales force, medical science liaison personnel, involvement of patient advocacy groups, support for the patient associations and centers of excellence and increased key opinion leader involvement in the legitimate development of RUCONEST® going forward.

## THE TRANSACTION AND USE OF FUNDS

In order to be able to pay the US\$60 million upfront payment (approximately €54 million) to Valeant and (i) to invest in the additional commercialization efforts in the USA, (ii) to invest in additional commercialization efforts in the EU, (iii) to ensure the availability of sufficient working capital and (iv) pay for the costs of the acquisition and financing transactions, the Company seeks to raise a total amount of between €73 million and €90 million in a mixture of straight debt and new equity.

The equity part of this total amount is to be offered to existing shareholders first in a rights issue, to avoid concerns over dilution of existing shareholders by new shareholders (“The Rights Offering”). To the extent that shareholders do not wish to take up their rights and subscribe for the new shares, the rights shares not taken up will be offered to institutional investors in the EU and the USA at the same price.

We expect the approximate size of the equity component of the fundraising to be between €35m and €60m (the “Financing”). The final amount will depend on, inter alia, the Company’s stock price at the time of pricing and announcement of the Rights Offering, the size and terms of the debt offers received, the percentage uptake of the rights shares by existing shareholders and the appetite amongst new equity investors for any balance. A full prospectus including additional information and financial statements as at 31 August 2016 will be prepared and published following the EGM.

The pricing of the Rights Offering will be determined on the basis of a discount to the Volume Weighted Average Price (“VWAP”) of the Pharming shares over a period to be defined, which period will close no later than the date of the EGM (Wednesday 5 October).

At the moment, out of our authorized share capital of 650 million shares, 184 million shares are available for issue for the Financing. On the basis of Pharming’s recent share price, it is possible that Management may not have sufficient headroom with these shares alone to complete the financing:

Accordingly, the Board of Management is proposing that the Authorised Share Capital be increased by 150 million new shares to ensure to enable the Financing to succeed.

**For clarity, this requested increase in the authorized capital does NOT mean that all of the increase or available headroom shares will be issued.**

It is possible and expected on the basis of preliminary discussions with debt providers and institutional equity investors that Pharming could complete the Transaction by issuing significantly less stock than the full headroom allocation within the increased authorized

capital, and the Board of Management is committed to and will do all things necessary to minimize the actual number of shares issued, so as to preserve the rights and value for existing shareholders.

Based on the preliminary discussions, the Board of Management expects to issue a number of shares in the range of between 120 million and 240 million shares to complete the Financing.

The Notice to Convene, the Explanatory Notes and a Form of Proxy can be found on our website [www.pharming.com](http://www.pharming.com)

Board of Management  
Pharming Group N.V.  
Leiden  
23 August 2016

-O-

### About RUCONEST®

RUCONEST® (recombinant C1 esterase inhibitor ) is an orphan drug indicated for the treatment of hereditary angioedema (HAE). RUCONEST contains C1 esterase inhibitor for delivery at 50 IU/kg

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..

When administered at the onset of HAE attack symptoms at the recommended dose, RUCONEST helps to return a patient's C1 esterase inhibitor levels to normal range and to 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 the European Medicines Agency (EMA) and was approved in July 2014 by the FDA and in October 2010 by the EMA.

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

### Important Safety Information for RUCONEST®

RUCONEST® is a recombinant C1 esterase inhibitor 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 (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 implanted 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, it 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 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 disfigurement, 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 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 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 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 distributed in North America, Canada and Mexico by 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 Cytobiotech.

RUCONEST® is distributed in South Korea by HyupJin Corporation and in Israel by Megapharm.

RUCONEST® achieved primary and secondary endpoints in a Phase II randomized, double blind placebo-controlled clinical trial for prophylactic treatment of HAE and is being evaluated for other indications as well. The Phase II study was fully recruited shortly after the year-end, and is expected to report preliminary results around the middle of 2016.

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 Pompe and Fabry's diseases are being optimized at present, with additional programs not involving ERT also being explored at an early stage.

Pharming has a long term partnership with the Shanghai Institute of Pharmaceutical Industry ("SIPI"), a Sinopharm company, for joint global development of new products, starting with RUCONEST® and recombinant human Factor VIII for the treatment of Haemophilia A. Pre-clinical development and manufacturing will take place to global standards at SIPI and are funded by SIPI. 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](http://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.*

**Contact**

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

Robin Wright, CFO : T: +31 71 524 7432

**FTI Consulting**

Julia Phillips/ Victoria Foster Mitchell, T: +44 203 727 1136

**Important Notice**

This press release is not, and does not form part of, an offer to buy, sell or exchange, nor is it a solicitation of an offer to buy, sell or exchange, securities in or into the United States, Australia, Canada or Japan. No securities have been, or will be, registered under the U.S. Securities Act of 1933, as amended, or will be offered, sold, transferred or delivered, directly or indirectly, in or into the United States unless the relevant registration in the United States has been made, or such offer, sale, transfer or delivery is carried out pursuant to an exemption from, or in a transaction not subject to, registration, in accordance with the U.S. Securities Act of 1933, as amended. There will be no public offer of securities in the United States.

The securities referenced herein have not been approved or disapproved by the United States Securities and Exchange Commission or any other securities commission or regulatory authority in the United States, nor have any of the foregoing authorities passed upon or endorsed the merits of the proposed Financing. Any representation to the contrary is a criminal offence in the United States.