

## Pharming announces completion of acquisition of all North American commercialisation rights for RUCONEST® from Valeant

*Leiden, The Netherlands, 8 December 2016:* Pharming Group N.V. (“Pharming” or “the Company”) (EURONEXT: PHARM) announces today that it has completed a definitive agreement to acquire all North American commercialisation rights for its own product, RUCONEST® (recombinant human C1 esterase inhibitor), including all rights in the US, Mexico and Canada, from certain subsidiaries of Valeant Pharmaceuticals International, Inc. (“Valeant”) (NYSE/TSX: VRX). RUCONEST® is an orphan drug designated therapy developed by Pharming, already approved for the treatment of acute Hereditary Angioedema (“HAE”) attacks in patients in the US and EU. This transaction will accelerate Pharming’s development into a profitable specialty pharmaceutical company with its own independent commercial infrastructure, which will form the foundation for future growth.

- **Transformational acquisition of commercial rights to Pharming’s own product RUCONEST®**
- **Immediate and substantial positive impact on Pharming’s operational results and near-term profitability; annualised run rate of sales, increased from US\$35 million in Q3 2016 to more than US\$40 million, based on the average of the most recent two months of sales (October and November).**
- **US\$125 million deal value, with an upfront fee paid to Valeant of US\$60 million, and future self-funding sales milestone payments up to a further US\$65 million in total**
- **Funding through a combination of new equity, straight debt and new convertible bonds of €104 million before costs.**
- **Cash position after closing of deal and payment of all transaction costs strengthened to €34.3 million**
- **Additional new investment going into RUCONEST® sales force, medical science liaison personnel and marketing activities in the US and Europe to accelerate sales growth in both the US and Europe**

Since the US Food and Drug Administration (“FDA”) approval of RUCONEST® on 16 July, 2014, US net product sales have grown from US\$0.3 million in 2014 to an annualised run rate of approximately US\$35 million at the end of the third quarter of 2016 and recently, based on average sales over the last two months (October and November) to an annualised run rate of more than US\$40 million, within the US acute HAE market of around US\$850 million per year.

Recently, RUCONEST® has shown very good positive data in prophylaxis of HAE, meeting its primary endpoints for both once-weekly and twice-weekly dosing regimens in a Phase II clinical trial (announced on July 18, 2016). If approved in this indication, RUCONEST® will be able to enter this additional market, worth around US\$700 million per year. RUCONEST® therefore has the potential to be the only recombinant C1 esterase inhibitor approved to target both the acute market and the HAE prophylaxis market.

### **Structure of the Deal**

Under the terms of the agreement, Pharming has paid Valeant an upfront fee of US\$60 million upon Closing. In addition, over the coming years the Company will make one-time-only payments to Valeant on achievement of a small number of specific sales milestones events, totalling a maximum of US\$65 million. The specific details of these self-funding additional transaction terms are not disclosed for commercial reasons.

The transaction has been completed and Pharming is now responsible for selling RUCONEST® directly in the US.

### **Growth of sales force and supplementary marketing efforts crucial for success**

Helping to ensure a seamless transition, all of the dedicated RUCONEST® sales force who previously worked for Valeant or one of its subsidiaries, a total of 11 people, have accepted offers to join Pharming to maintain and boost the RUCONEST® sales effort in the US. The Company also plans to drive sales growth by increasing the size of the sales force, investing in medical science liaison personnel and supporting additional marketing activities, including patient advocacy programmes and the provision of significant unconditional support for the HAEA (the US HAE patients' association) and its programmes as well as other HAE centers of excellence in the US. In addition, Pharming is planning further investment in the acceleration of RUCONEST® sales efforts to drive growth in the European, Middle Eastern and African markets which Pharming assumed responsibility over in October 2016 from SOBI, as announced on 14 July 2016, and to make RUCONEST available in Canada and Mexico.

Valeant and Pharming will work closely on the transition for customers and HAE patients under a transition services agreement entered into today in connection with the closing of the transaction. This will enable Pharming to replace core functions currently undertaken by Valeant and its contractors in a smooth and timely manner without any risk for patients.

### **Funding the transaction and development investments**

The transaction was funded by a complex combination of new financial instruments, including a Rights Issue to existing shareholders. These instruments are as follows:

- A Rights Offer to existing shareholders to acquire one new share for every seven shares already held, at a price of €0.205 per new share, which raised €8.8 million, including a Rump Offer of shares represented by unexercised rights, to institutional shareholders.
- A US\$40 million, 42 months' straight debt facility on standard commercial terms (8.25% interest and one-off 9% final payment at maturity in June 2020). The syndicate includes one of the Company's current lenders, Silicon Valley Bank, and new lender Kreos Capital. The debt allows for twelve months of interest-only payments and is then paid off in 30 equal instalments.
- A five-year redeemable 8.5% convertible Eurobond facility (the Ordinary Bonds) for €12.5 million which will be convertible into shares at €0.284, a premium of 25% to the 20-day volume-weighted average price (VWAP) as at 18 November 2016, the business day prior to

publication of Pharming's prospectus on 21 November 2016. The issue was led by Kreos Capital and other EU and US institutional investors. These bonds have an 8.5% interest rate.

- An 18-month redeemable amortizing convertible bond (the Amortizing Bond) for €45 million, led by Hudson Bay Capital with other institutional investors. These bonds are convertible into shares at €0.289, a premium of 35% to the price on 5 December 2016, the day before closing of the bond issue. The bonds carry no interest rate and can be partly redeemed in cash at discretion of the Company. Further details of this instrument were announced on 7 December 2016.
- The costs of the whole financing programme are approximately €9.7 million, including legal costs, and the costs for the Valeant transaction will be approximately €1.0 million. The amount payable to repay the existing debt facility will be approximately €15.6 million. The net proceeds of all the financing transactions after payment of costs, the repayment of the existing debt facility and the payment of the upfront amount to Valeant will be €19.4 million. Following closing, the cash position will be approximately €34.3 million.
- A total of 88,025,158 warrants to subscribe for Pharming shares at €0.284 per share, a premium of 25% to the 20-day volume-weighted average price (VWAP) as at 18 November 2016, have been issued to institutional investors who subscribed for the different financing instruments.
- Only the 42,981,939 shares bought in the Rights Offer and associated rump offer have been issued in connection with the acquisition. Up to a further 199,864,273 shares are represented by the convertible bonds and will only be issued upon future conversion or in case of amortization in equity by the Company.

Pharming anticipates that this transaction, after taking full account of the costs of the transaction and the financing including interest, will be accretive to earnings within 2017 and will enable the Company to reach profitability, potentially as much as three years earlier than under the Valeant license.

**Sijmen de Vries, Pharming CEO, commented:**

*"This is a quantum leap forward for Pharming and marks a significant step for the Company taking control of its own destiny and providing a real prospect of reaching profitability soon. Now that we have taken full ownership of our key asset and by integrating the Valeant US sales team members and building that team, we will work with a single-minded focus, energy and investments to bring this treatment to all eligible HAE patients in the US. For well over a decade Pharming has been dedicated to the HAE market and has been working with HAE physicians and patients on the development of a safe and effective recombinant enzyme replacement therapy. I am delighted that our shareholders, including our new investors, have taken this opportunity to support our strategic efforts to accelerate profitability for Pharming, taking us in to a new phase as a specialty pharmaceutical company.*

*I would like to thank the Pharming team, including our advisers, for their help in bringing this highly complex transaction to completion. This is an exciting day for all of Pharming's employees and for our shareholders."*

Stifel Nicolaus Europe Limited acted solely as lead European Placement Agent and Roth Capital Partners, acted solely as lead US Placement Agent. Trout Capital LLC acted as a Co-Placement Agent and First Berlin Securities Brokerage GmbH is acted as a Placement Advisor.

Stifel Nicolaus Europe Limited acted as sole financial adviser to Pharming in connection with the proposed re-acquisition of Ruconest US rights from Valeant.

### **About Kreos Capital**

Kreos Capital is the leading provider of growth debt financing in Europe and Israel to high-growth companies. Since 1998, Kreos has completed over 450 transactions and committed more over EUR 1.7 billion in 15 different countries. Kreos is dedicated to supporting management teams and their equity investors with flexible loan structures for all stages of a growth company's development and to addressing the needs for growth capital, working capital, acquisition financings, lower mid-market buy-outs, roll-up strategies, bank re-financings as well as pre- and post-IPO financings. Kreos's most recent fund, Kreos V, was launched in January 2016 and has EUR 400 million of equity commitments from top-tier institutional investors. The Kreos global team has extensive debt financing, management and equity investing experience, covering the pan-European market from its locations in London, Tel Aviv and Stockholm.

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### **Commercial rights history**

Valeant acquired the North American license rights to RUCONEST® through its acquisition of Salix Pharmaceuticals, Inc. ("Salix") on 1 April, 2015. Prior to this, Salix had acquired the rights through its acquisition of Santarus, Inc. ("Santarus") on 3 January, 2014. Pharming originally entered into an agreement with Santarus for development and commercialisation of RUCONEST® in the US, Canada and Mexico on 10 September 2010.

### **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 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 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 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, United States of America 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 Argentina, Colombia, Costa Rica, the Dominican Republic, Panama and Venezuela by Cytobiotech, 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](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 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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