Pharming reports encouraging results from use of RUCONEST® in COVID-19 patients

- Five hospitalised patients with confirmed COVID-19 infections administered RUCONEST® under compassionate use program to treat the related severe pneumonia
- Multinational, randomized controlled, investigator-initiated study with up to 150 patients planned

Leiden, The Netherlands, 21 April, 2020: Pharming Group N.V. ("Pharming" or "the Company") (Euronext Amsterdam: PHARM) today announced encouraging results from five patients with confirmed COVID-19 (SARS-CoV-2) infections hospitalised with related severe pneumonia that were treated with RUCONEST® (recombinant human C1 inhibitor) under a compassionate use program at the University Hospital Basel, Switzerland.

Four male patients and one female patient (between 53-85 years of age) with COVID-19 and suffering from related severe pneumonia, who did not improve despite standard treatment, including hydroxychloroquine and lopinavir/ritonavir, were administered RUCONEST® at an initial dose of 8400 U, followed by 4200 U every 12 hours for three additional doses. No allergic reactions or drug related adverse events were reported.

Following treatment with RUCONEST®, fever resolved in four of the five patients within 48 hours, and laboratory markers of inflammation decreased significantly (CRP, IL-6). Soon thereafter, the patients were discharged from the hospital as fully recovered. One patient had increased oxygen requirement and was temporarily transferred to the ICU for intubation, but over the subsequent days recovered and was released from the ICU.

Following these initial results, a multinational, randomized, controlled, investigator-initiated clinical trial with up to 150 patients with confirmed COVID-19 infections, requiring hospitalisation due to significant COVID-19 related symptoms is planned. The study will be led by Dr. Michael Osthoff, University Hospital Basel, Switzerland.

Dr. Michael Osthoff, University Hospital Basel, Switzerland and the treating physician, said:
"Although this is an uncontrolled, small treatment experience, the results demonstrate the potential effectiveness of using RUCONEST® as an anti-inflammatory approach to inhibit the complement and contact systems after SARS-CoV-2 infection. We are now in the midst of planning a multinational, randomized controlled trial in up to 150 patients to further understand the safety and efficacy of this approach in preventing deterioration in COVID-19 patients."

Prof. Bruno Giannetti, Pharming's Chief Medical Officer, commented:
"Some of the dangerous biochemical processes occurring during the worsening of a COVID-19 infection towards life-threatening pneumonia are likely triggered by complement activation as part of a systemic hyperinflammatory syndrome, otherwise known as a 'cytokine storm'. C1 inhibitor has numerous anti-

inflammatory properties, including inhibition of the complement and contact systems. A compassionate treatment in a few patients suffering from COVID-19 pneumonia was, therefore, scientifically sound and these preliminary results are very encouraging. Amongst others, we need to better identify the best time point to start RUCONEST® treatment and the optimal dosing regimen. The planned multinational study under the leadership of the University of Basel is aimed at providing this information."

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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 authorisation.

RUCONEST® is commercialised by Pharming in the US and in Europe, and the Company holds all other commercialisation rights in other countries not specified below. In some of these other countries distribution is made in association with the HAEi Global Access Program (GAP). 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 Kamada.
RUCONEST® is also being evaluated for various additional indications. Pharming's technology platform includes a unique production process that has proven capable of producing industrial quantities of pure high quality recombinant human proteins in a more economical and less immunogenic way compared with current cell-line based methods.

Leads for enzyme replacement therapy ("ERT") for Pompe and Fabry's diseases are also being produced and optimised respectively at present.

Pharming has recently in-licensed leniolisib from Novartis, a small molecule and selective PI3Kδ inhibitor, which is in a registrational study for activated PI3K-delta syndrome (APDS), a rare form of Primary Immunodeficiency.

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. Preclinical development and manufacturing will take place to global standards at CSIPI and its affiliates 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.

Forward-looking Statements

This press release of Pharming Group N.V. and its subsidiaries ("Pharming", the "Company") 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.

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