**Pharming announces enrolment of first patient in US clinical trial for the treatment of COVID-19 with RUCONEST®**

**Highlights:**

* **First patient treated in second clinical trial globally investigating the use of RUCONEST® to prevent severe SARS-CoV-2 infections in hospitalised patients with confirmed COVID-19**
* **Planned expansion to multiple centres across the US**

*Leiden, The Netherlands*, 10 December 2020:Pharming Group N.V. (“Pharming” or “the Company”) (Euronext Amsterdam: PHARM) today announced that the first patient has been enrolled in a randomised, open label, parallel group, controlled, pilot clinical trial in up to 120 patients hospitalised with confirmed COVID-19 treated with RUCONEST® (recombinant human C1 inhibitor) for the prevention of severe SARS-CoV-2 infections at the Valley Hospital in Ridgewood, New Jersey in the United States.

Initially based at Valley Hospital in Ridgewood, New Jersey, this trial is planned to include patients at multiple centres in the US. This clinical trial follows an ongoing investigator-initiated, multinational, multicentre study, led by Dr. Michael Osthoff from the University Hospital Basel, into the use of RUCONEST® in the prevention of severe SARS-CoV-2 infections in patients hospitalised with related severe pneumonia. Pharming announced the enrolment of the first patient in that trial in August this year.

**RUCONEST® AND COVID-19**

RUCONEST® is a recombinant C1 esterase inhibitor (C1INH) approved for the treatment of hereditary angioedema (HAE) in Europe and the US. C1INH is a protein that naturally occurs in the human body. It regulates several inflammatory pathways in the body by inhibiting certain proteins that are part of the human immune system. In diseases like HAE, deficiency of functional C1 inhibitor leads to excessive activation of the complement system and other immunological and haemostatic pathways, giving cause to angioedema attacks. In HAE, these attacks are characterised by acute and painful swellings of soft tissues. Administration of C1 inhibitor can normalise the low C1INH levels and stop angioedema attacks.

Systemic hyperinflammation is a hallmark of more severe stages of COVID-19 leading to acute respiratory distress syndrome, mechanical ventilation and ultimately death. Treatment with RUCONEST® may; 1) dampen uncontrolled complement activation and collateral lung damage; 2) reduce capillary leakage and subsequent pulmonary edema by direct inhibition of the kallikrein-kinin system; and 3) reduce the generation of microthrombi by inhibiting MASP-1 induced clot formation and factor XII amplified thrombo-inflammation.

C1 inhibitor is an acute phase reactant, meaning that the body naturally increases production during inflammatory conditions, such as infections. Despite this, a relative deficiency may occur and complement activation continues unchecked, often leading to a cytokine storm, a dangerous biochemical process that worsens the complications of COVID-19 infection, such as organ failure and death.

These clinical studies in hospitalised patients with confirmed COVID-19 seek to identify if the administration of additional C1INH can control or stop the systemic hyperinflammation syndrome or cytokine storm. Headline data will be made publicly available following either an interim analysis or after all patients have been treated.

About Pharming Group N.V.

Pharming Group N.V. is a global, commercial stage biopharmaceutical company developing innovative protein replacement therapies and precision medicines for the treatment of rare diseases and unmet medical needs.

The flagship of our portfolio is our recombinant human C1 esterase inhibitor, or rhC1INH, franchise. C1INH is a naturally occurring protein that downregulates the complement cascade in order to control swelling in affected tissues.

Our lead product, RUCONEST® is the first and only plasma-free rhC1INH protein replacement therapy. It is approved for the treatment of acute hereditary angioedema, or HAE, attacks. We are commercializing RUCONEST® in the United States, the European Union and the United Kingdom through our own sales and marketing organization, and the rest of the world through our distribution network.

We are also developing rhC1INH for subsequent indications, including pre-eclampsia, acute kidney injury and we also investigating the clinical efficacy of rhC1INH in COVID-19.

In addition, we are studying our oral precision medicine, leniolisib (a phosphoinositide 3-kinase delta, or PI3K delta, inhibitor), for the treatment of activated PI3K delta syndrome, or APDS, in a registration enabling Phase 2/3 study in the US and Europe.

Furthermore, we are also leveraging our transgenic manufacturing technology to develop next-generation protein replacement therapies most notably for Pompe disease, which program is currently in the preclinical stage.

Forward-looking Statements

*This press release contains forward-looking statements, including with respect to timing and progress of Pharming’s preclinical studies and clinical trials of its product candidates, Pharming’s clinical and commercial prospects, Pharming’s ability to overcome the challenges posed by the COVID-19 pandemic to the conduct of its business, and Pharming’s expectations regarding its projected working capital requirements and cash resources, which statements are subject to a number of risks, uncertainties and assumptions, including, but not limited to the scope, progress and expansion of Pharming’s clinical trials and ramifications for the cost thereof; and clinical, scientific, regulatory and technical developments. In*

*light of these risks and uncertainties, and other risks and uncertainties that are described in Pharming’s 2019 Annual Report and its report for the six months ended 30 June 2020, the events and circumstances discussed in such forward-looking statements may not occur, and Pharming’s actual results could differ materially and adversely from those anticipated or implied thereby. Any forward-looking statements speak only as of the date of this press release and are based on information available to Pharming as of the date of this release.*

Inside Information

*This press release relates to the disclosure of information that qualifies, or may have qualified, as inside information within the meaning of Article 7(1) of the EU Market Abuse Regulation.*

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