Pharming announces results from clinical trials for the treatment of COVID-19 with RUCONEST®

Pharming-sponsored study in US met primary endpoint
RUCONEST®-treated patients having statistically significant lower disease severity scores (p=0.0056) as compared with patients treated with standard of care

Leiden, The Netherlands, 14 September 2021: Pharming Group N.V. ("Pharming" or "the Company") (Euronext Amsterdam: PHARM/NASDAQ: PHAR) announces the topline results from two randomized, open label, controlled, pilot clinical trials of patients hospitalized with COVID-19 treated with RUCONEST® (recombinant human C1 inhibitor) for the prevention of severe SARS-CoV-2 infection. The primary endpoint in both studies was disease severity on the 7-point WHO ordinal scale on Day 7.

In the US study, conducted under a Pharming IND, which had included 32 patients at the time of the interim analysis, patients treated with RUCONEST® plus standard of care had statistically significant lower WHO disease severity scores at Day 7 (mean 1.83, SD 0.65) as compared with those patients who received standard of care alone (mean 3.22, SD 1.86; p=0.0056). Data on secondary endpoints and biomarker evaluations were concordant with the primary endpoint findings.

In the investigator-led study, conducted in Switzerland, Brazil and Mexico and part of the National Research Program "COVID-19" (NRP 78) of the Swiss National Science Foundation (SNSF), which included 83 patients by the time of the interim analysis, no difference in the primary variable was observed between the treatment groups. However, there was a significant difference in disease severity at baseline, i.e., prior to treatment, between the groups. Specifically, patients in the RUCONEST® arm had statistically significant more severe disease than those patients in the standard of care arm (p=0.0324).

Although the two studies used a similar design and both enrolled patients who were being admitted to the hospital with severe pneumonia due to COVID-19 infection, different dosing regimens of RUCONEST® were used. In the Investigator-led study RUCONEST® was dosed in addition to the standard of care for three days, whereas in the US study it was four days. Also, there were differences in the patient populations enrolled and in the standard of care regimens administered.

The primary endpoint in both studies was disease severity on the 7-point WHO ordinal scale on Day 7. This endpoint has been suggested by WHO for clinical trials in patients with COVID-19 as it measures illness severity over time.

The trials were conducted following a compassionate use program, which saw encouraging results in patients who were administered RUCONEST® following hospitalization with COVID-19-related severe pneumonia. The results were first announced in April 2020 and subsequently published in Frontiers in Immunology in August 2020.

Anurag Relan MD, Pharming's Chief Medical Officer commented:

"These results indeed support our initial hypothesis on the need to control the hyperinflammatory process in patients with severe COVID-19 infection. It is unfortunate we cannot draw many conclusions from the investigator led study, due to the imbalance between the RUCONEST®-arm and the control group at the start of the trial. We, however, thank all of the principal investigators and their staff for conducting this important research in an extremely challenging environment. We will analyze the full results of these studies as we design future clinical trials with RUCONEST® for the treatment of COVID-19, as well as other serious conditions."

RUCONEST® was well tolerated and no drug-related serious adverse events were observed in either study.

Both studies have now concluded, and the results will be published in peer-review medical journals.

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 (rhC1INH) franchise. C1INH is a naturally occurring protein that down regulates the complement and contact cascades in order to control inflammation 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 (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.

In addition, we are investigating the clinical efficacy of rhC1INH in the treatment of further indications, including pre-eclampsia, acute kidney injury and severe pneumonia as a result of COVID-19 infections.

We are also 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 United States and Europe.

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

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 2020 Annual Report and the Annual Report on Form 20-F for the year ended December 31, 2020 filed with the U.S. Securities and Exchange Commission, 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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