Pharming Group announces presentation of new leniolisib data at 2022 ASH Annual Meeting

Presentation to highlight data from the ongoing long-term open-label extension study of leniolisib, a PI3Kδ inhibitor under investigation for APDS, a rare primary immunodeficiency

Leiden, The Netherlands, November 23, 2022: Pharming Group N.V. ("Pharming" or "the Company") (EURONEXT Amsterdam: PHARM / Nasdaq: PHAR) announces today that new clinical data for leniolisib, an oral, selective phosphoinositide 3-kinase delta (PI3Kδ) inhibitor, an investigational treatment for activated phosphoinositide 3-kinase delta syndrome (APDS), a rare primary immunodeficiency, will be presented by V. Koneti Rao, MD, FRCPA, staff physician in the Primary Immune Deficiency Clinic at the National Institute of Health in Bethesda, Maryland, at the 64th American Society of Hematology (ASH) Annual Meeting and Exposition, in New Orleans, Louisiana taking place from Saturday, December 10 through Tuesday, December 13, 2022.

Information regarding Pharming's data presentation can be found below, and on the ASH conference website: https://www.hematology.org/meetings/annual-meeting.

Presentation title: Interim Analysis of Safety and Hematological Parameters of an Ongoing Long-Term Open-Label Extension Study of Investigational PI3Kδ Inhibitor Leniolisib for Patients with Activated PI3Kδ Delta Syndrome (APDS) through December 2021

Presentation type: oral

Session name: 203. Lymphocytes and Acquired or Congenital Immunodeficiency Disorders: Delineating Immunity from Mice to Humans

Abstract number: 608

Session date and time: Monday, December 12, 2022 from 10:30AM – 12:30PM CST

Presentation date and time: Monday, December 12, 2022 at 10:45AM CST

Location: Room: Ernest N. Morial Convention Center, 278-282

About Activated Phosphoinositide 3-Kinase δ Syndrome (APDS)

APDS is a rare primary immunodeficiency that affects approximately 1 to 2 people per million. APDS is caused by variants in either of two genes, *PIK3CD* or *PIK3R1*, that regulate maturation of white blood cells. Variants of these genes lead to hyperactivity of the PI3Kδ (phosphoinositide 3-kinase delta) pathway.[1,2] Balanced signaling in the PI3Kδ pathway is essential for physiological immune function. When this pathway is hyperactive, immune cells fail to mature and function properly, leading to immunodeficiency and dysregulation.[1,3] APDS is characterized by severe, recurrent sinopulmonary infections, lymphoproliferation, autoimmunity, and enteropathy.[4,5] Because these symptoms can be

associated with a variety of conditions, including other primary immunodeficiencies, people with APDS are frequently misdiagnosed and suffer a median 7-year diagnostic delay.[6] As APDS is a progressive disease, this delay may lead to an accumulation of damage over time, including permanent lung damage and lymphoma.[4-7] The only way to definitively diagnose this condition is through genetic testing.

About Leniolisib

Leniolisib is a small-molecule inhibitor of the delta isoform of the 110 kDa catalytic subunit of class IA PI3K. PI3Kδ is expressed predominately in hematopoietic cells and is essential to normal immune system function through conversion of phosphatidylinositol-4-5-trisphosphate (PIP2) to phosphatidylinositol-3-4-5-trisphosphate (PIP3). Leniolisib inhibits the production of PIP3 and PIP3 serves as an important cellular messenger activating AKT (via PDK1) and regulates a multitude of cell functions such as proliferation, differentiation, cytokine production, cell survival, angiogenesis, and metabolism. Unlike PI3Kα and PI3Kβ, which are ubiquitously expressed, PI3Kδ and PI3Kγ are expressed primarily in cells of hematopoietic origin. The central role of PI3Kδ in regulating numerous cellular functions of the adaptive immune system (B-cells and, to a lesser extent, T cells) as well as the innate immune system (neutrophils, mast cells, and macrophages) strongly indicates that PI3Kδ is a valid and potentially effective therapeutic target for immune diseases such as APDS. To date, leniolisib has been well tolerated during both the Phase 1 first-in-human trial in healthy subjects and the Phase II/III registration-enabling study in patients with APDS.

About Pharming Group N.V.

Pharming Group N.V. (EURONEXT Amsterdam: PHARM/Nasdaq: PHAR) is a global biopharmaceutical company dedicated to transforming the lives of patients with rare, debilitating, and life-threatening diseases. Pharming is commercializing and developing an innovative portfolio of protein replacement therapies and precision medicines, including small molecules, biologics, and gene therapies that are in early to late-stage development. Pharming is headquartered in Leiden, Netherlands, and has employees around the globe who serve patients in over 30 markets in North America, Europe, the Middle East, Africa, and Asia-Pacific.
For more information, visit www.pharming.com.

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 2021 Annual Report and the Annual Report on Form 20-F for the year ended December 31, 2021 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.

References

1. Lucas CL, et al. Nat Immunol. 2014;15:88-97.
2. Elkaim E, et al. J Allergy Clin Immunol. 2016;138(1):210-218.
3. Nunes-Santos C, Uzel G, Rosenzweig SD. J Allergy Clin Immunol. 2019;143(5):1676-1687.
4. Coulter TI, et al. J Allergy Clin Immunol. 2017;139(2):597-606.
5. Maccari ME, et al. Front Immunol. 2018;9:543.
6. Jamee M, et al. Clin Rev Allergy Immunol. 2019;May 21.
7. Condliffe AM, Chandra A. Front Immunol. 2018;9:338.

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