

**Company announcement**

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Inside information

Orphazyme A/S

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Company Registration No. 32266355

**Kim Stratton, CEO, resigns from Orphazyme**

**Copenhagen, Denmark, December 10, 2020** – Orphazyme A/S (ORPHA.CO) (“Orphazyme”), a late-stage biopharmaceutical company pioneering the Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases, today announces that Kim Stratton has decided to resign from Orphazyme following a dialogue initiated by the board of directors. Orphazyme will initiate a search for a new CEO immediately. Meanwhile, in order to ensure continuity going forward, Georges Gemayel, Chairman of the Board of Directors, will temporarily assist the executive team of Orphazyme with the day-to-day operations until a new CEO is appointed.

*Chairman of the Board of Directors, George Gemayel, states that “We are fortunate that Kim Stratton was available at a critical juncture of Orphazyme’s history during which the company successfully raised new equity of approximately DKK 1.3 bn, completed a listing of ADS in the United States and filed an NDA with the FDA and an MAA with the EMA. With these important milestones and the establishment of our operations in the United States, Orphazyme is well-positioned for a potential commercial launch of our product candidate, arimoclomol in the United States and Europe. Until a new CEO is appointed, I look very much forward to working temporarily with Orphazyme’s executive team to ensure a seamless transition and to continue the important work lying ahead.”*

**For additional information, please contact****Orphazyme A/S**

Georges Gemayel  
Chairman of the Board of Directors

Bo Jesper Hansen  
Deputy Chairman of the Board of Directors

Anders Vadsholt, CFO +45 28 98 90 55

**About Orphazyme A/S**

Orphazyme is a late-stage biopharmaceutical company pioneering the Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases. The company is harnessing amplification of Heat-Shock Proteins (or HSPs) in order to develop and commercialize novel therapeutics for diseases caused by protein misfolding, protein aggregation, and lysosomal dysfunction, including lysosomal storage diseases and neuromuscular degenerative diseases. Arimoclomol, the company’s lead candidate, is in clinical development for four orphan diseases: Niemann-Pick disease Type C (NPC), Amyotrophic Lateral Sclerosis (ALS), sporadic Inclusion Body Myositis (sIBM) and Gaucher disease. Orphazyme is headquartered in Denmark and has operations in the U.S. and Switzerland. Orphazyme’s shares are listed on Nasdaq U.S. (ORPH) and Nasdaq Copenhagen (ORPHA).

**About arimoclomol**

Arimoclomol is an investigational drug candidate that amplifies the production of Heat-Shock Proteins (HSPs). HSPs can rescue defective misfolded proteins, clear protein aggregates, and improve the function of lysosomes. Arimoclomol is administered orally, crosses the blood-brain barrier, and has now been studied in seven phase 1, four phase 2 and one pivotal phase 2/3 trial. Arimoclomol is in clinical development for NPC, Gaucher Disease, sIBM, and ALS. Arimoclomol has received orphan drug designation (ODD) for NPC, sIBM, and ALS in the US and EU. Arimoclomol has received fast-track designation (FTD) from the U.S. Food and Drug Administration (FDA) for NPC, sIBM and ALS. In addition, arimoclomol has received breakthrough therapy designation (BTD) and rare-pediatric disease designation (RPDD) from the FDA for NPC.

**Forward-looking statement**

This company announcement may contain certain forward-looking statements. Although the Company believes its expectations are based on reasonable assumptions, all statements other than statements of historical fact included in this company announcement about future events are subject to (i) change without notice and (ii) factors beyond the Company’s control. These statements may include, without limitation, any statements preceded by, followed by, or including words such as “target,” “believe,” “expect,” “aim,” “intend,” “may,” “anticipate,” “estimate,” “plan,” “project,” “will,” “can have,” “likely,” “should,” “would,” “could”, and other words and terms of similar meaning or the negative thereof. Forward-looking statements are subject to inherent risks and uncertainties beyond the Company’s control that could cause the Company’s actual results, performance, or achievements to be materially different from the expected results, performance, or achievements expressed or implied by such forward-looking statements. Except as required by law, the Company assumes no obligation to update these forward-looking statements publicly, or to update the reasons actual results could differ materially from those anticipated in the forward-looking statements, even if new information becomes available in the future.