

**Company announcement**

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**Orphazyme A/S**Ole Maaløes Vej 3  
DK-2200 Copenhagen N

www.orphazyme.com

Company Registration No. 32266355

**New long-term share-based incentive program**

**Copenhagen, Denmark, April 22, 2021** – Orphazyme A/S [ORPHA.CO (DK); ORPH (US)], a late-stage biopharmaceutical company pioneering the heat shock protein response for the treatment of neurodegenerative rare diseases, has today introduced a new long-term incentive program (the "LTIP").

The LTIP is designed and structured around the concept of retaining members of the Executive Management and other employees of the Group, while also creating an incentive for a positive share price development and corporate performance for the benefit of the Company's shareholders.

The LTIP grants comprise Restricted Share Units ("RSUs") and Performance Share Units ("PSUs") which entitle the participants, subject to vesting occurring, to be allocated a number of shares in the Company, equivalent to the number of vested RSUs and/or PSUs, against payment of the par value of each share.

The RSUs will have a total vesting period of three years calculated from January 1 or July 1 in the grant year and with one third of the granted RSUs vesting on each January 1 or July 1 in the following three financial years. Vesting of RSUs is not conditional upon achieving any financial or non-financial targets. However, vesting is conditional upon (i) the participant remaining employed with a group member throughout the total vesting period for RSUs or the participant becoming a good leaver during the total vesting period for RSUs, in which case the participant will be entitled to keep any vested RSUs and receive a pro rata allocation, and (ii) the participant having complied in all respects with the general terms and conditions as determined by the Board of Directors. The vested RSUs can only be exercised within four months after the expiration of the total vesting period for RSUs. However, the RSU delivery period may be extended to the next open trading window in certain circumstances.

The PSUs will have a total vesting period of three years calculated from January 1 or July 1 in the grant year and with the granted PSUs vesting, in whole or in part, on January 1 or July 1 in the third year following the date of the grant. Vesting of PSUs is conditional upon (i) an increase in the quoted share price of the Company's shares, (ii) the participant remaining employed with a group member throughout the vesting period for PSUs or the participant becoming a good leaver during the vesting period for PSUs, in which case the participant may be entitled to keep a proportion of the PSUs, and (iii) the participant having complied in all respects with the general terms and conditions as determined by the Board of Directors. Any vested PSUs can only be exercised within four months after the expiration of the vesting period for PSUs. However, the PSU delivery period may be extended to the next open trading window in certain circumstances.

Based on the current number of participants in the LTIP, the program and other share-based retention grants are expected to comprise up to 950,000 shares in total. The theoretical fair value of each RSU has been estimated at DKK 58.04 and the theoretical fair value of each PSU under the LTIP has been estimated at DKK 20.02.

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

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 Niemann-Pick disease type C (NPC), Amyotrophic Lateral Sclerosis (ALS), 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 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, and ALS. Arimoclomol has received orphan drug designation (ODD) for NPC 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 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, including in respect of the anticipated commercialization of arimoclomol. 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, including the risk that applicable regulatory authorities fail to approve arimoclomol on the anticipated timeline or at all. 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.