

Company announcement

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Company Registration No. 32266355

Modified long-term share-based incentive program for 2021

Copenhagen, Denmark, October 7, 2021 – Orphazyme A/S (ORPHA.CO (DK); ORPH (US)), a late-stage biopharmaceutical company, has today introduced a modified long-term incentive program for 2021 (the "LTIP") with the view to 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 modified 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 terms of the modified LTIP will be the same as the LTIP that was implemented in April 2021, however, the number of RSUs and PSUs and the applicable performance target for the PSUs will be reset and calculated based on a share price equal to DKK 31.94 per share, corresponding to the volume weighted average share price of the Company's shares as quoted on Nasdaq Copenhagen during the ten (10) trading days from September 1, 2021. The exercise of the RSUs and PSUs to be granted under the modified LTIP is conditional upon the participant not exercising his or her RSUs or PSUs granted in April 2021, which will subsequently lapse and no longer be exercisable.

In connection with the modified LTIP, the members of Executive Management will receive an extraordinary grant of RSUs and PSUs corresponding to 100% of the grant under the modified LTIP and on the same terms as the modified LTIP, and the sign-on RSUs granted to the CEO in April 2021 will also be reset after the same principles as the modified LTIP but with immediate vesting upon grant.

The modified LTIP including the other share-based retention grants to the Executive Management are expected to comprise up to 675,000 shares in total. The theoretical fair value of each RSU has been estimated at DKK 25.88 and the theoretical fair value of each PSU has been estimated at DKK 14.94.

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

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About Niemann-Pick disease type C

Niemann-Pick disease type C (NPC) is a rare, genetic, progressively debilitating, and often fatal neurodegenerative disease. It belongs to a family known as lysosomal storage diseases and is caused by mutations leading to defective NPC protein. As a consequence, lipids that are normally cleared by the lysosome accumulate in tissues and organs, including the brain, and drive the disease pathology. We estimate the incidence of NPC to be one in 100,000 live births and the number of NPC patients in the United States and in Europe to be approximately 1,800 individuals. There are no approved treatments for NPC in the U.S.

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 10 Phase 1, four Phase 2, and three pivotal Phase 2/3 trials. Arimoclomol has received Orphan Drug Designation (ODD) for NPC in the US and EU. Arimoclomol has received Fast-Track Designation (FTD), Breakthrough Therapy Designation (BTD), and Rare Pediatric Disease Designation (RPDD) from the U.S. Food and Drug Administration (FDA) for NPC. On June 17, 2021, Orphazyme received a Complete Response Letter from the FDA regarding its New Drug Application for arimoclomol for the treatment of NPC. A marketing authorization application (MAA) for arimoclomol in NPC has been filed with the European Medicines Agency and is under review.

About Orphazyme A/S

Orphazyme is a late-stage biopharmaceutical company developing arimoclomol for Niemann-Pick disease type C (NPC). Orphazyme is headquartered in Denmark and has operations in the U.S. and Switzerland. ADSs representing Orphazyme's shares are listed on Nasdaq U.S. (ORPH) and its shares are listed on Nasdaq Copenhagen (ORPHA).

Forward-looking statement

This company announcement may contain certain forward-looking statements under the U.S. Private Securities Litigation Reform Act of 1995 and otherwise. 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 risks and uncertainties that are described in the Risk Factors section of the Company's Annual Report on Form 20-F for the year ended December 31, 2020 filed with the U.S. Securities and Exchange Commission (SEC) on March 2, 2021, the Company's Report on Form 6-K filed with the SEC on June 11, 2021, and other filings Orphazyme makes with the SEC from time to time. These documents are available on the "Investors & Media" section of Orphazyme's website at www.orphazyme.com. 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.