

Company announcement No. 08/2021 Orphazyme A/S Ole Maaløes Vej 3 DK-2200 Copenhagen N

www.orphazyme.com Company Registration No. 32266355

Resolutions passed at the Annual General Meeting

Copenhagen, Denmark, March 25, 2021 – Orphazyme A/S (ORPHA.CO; ORPH) (the "Company"), a latestage biopharmaceutical company pioneering the Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases, today held its Annual General Meeting, at which the Annual General Meeting:

- Took note of the Board of Directors' report on the Company's activities in the past financial year;
- Adopted the Company's Annual Report 2020;
- Adopted that the loss related to the financial year 2020 is carried forward;
- Granted discharge of liability to the Board of Directors and the Executive Management in relation to the Annual Report 2020;
- Approved the remuneration report for 2020 in the advisory vote;
- Approved the remuneration of the Board of Directors for the current financial year;
- Re-elected Georges Gemayel, Bo Jesper Hansen, Anders Hedegaard, Carrolee Barlow, Catherine Moukheibir, Martijn Kleijwegt, Martin Bonde, and Rémi Droller as members of the Board of Directors;
- Elected Stephanie Okey as new member of the Board of Directors;
- Re-elected EY Godkendt Revisionspartnerselskab as the Company's auditor in accordance with the recommendation from the Audit Committee;
- Adopted an authorization to the Board of Directors to approve the acquisition of treasury shares in the period until March 25, 2026 with a total nominal value of up to 10% of the share capital of the Company subject to the Company's holding of treasury shares after such acquisition does not exceed 20% of the Company's share capital;
- Approved certain adjustments to the Company's Remuneration Policy, including (i) adjustments to allow for a share-based incentive program to the Executive Management and certain employees comprising restricted share units and performance share units, (ii) adjustment of the shareholding requirement for the Executive Management, (iii) adjustment of the claw back clause with the purpose of aligning the clause with the revised Danish Recommendations on Corporate Governance and (iv) adjustment of the situations under which accelerated vesting may occur;
- Adopted an amendment to Article 3.1 of the Articles of Association regarding renewal and extension of the existing authorization to the Board of Directors to increase the Company's share capital without preemption rights for existing shareholders by up to a nominal amount of DKK 6,989,767 and the deletion of Article 3.1.1; and
- Adopted an authorization to the Board of Directors included in a new Article 3.5 of the Articles of Association to increase the share capital in the period until March 25, 2026 without pre-emption rights for existing shareholders by up to a nominal amount of DKK 1,300,000 in connection with the issue of new shares to members of the Board of Directors, executives and/or employees of the Company subject to a total cap of nominally DKK 2,000,000 for both authorizations under the current Article 3.2 and the new Article 3.5 of the Articles of Association.

After the Company's Annual General Meeting was held, the Board of Directors constituted itself by appointing Georges Gemayel as Chairman and Bo Jesper Hansen as Deputy Chairman of the Board of Directors.

For additional information, please contact

Orphazyme A/S

Anders Vadsholt, Interim CEO and 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), Inclusion Body Myositis (IBM) 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, IBM, and ALS. Arimoclomol has received orphan drug designation (ODD) for NPC, IBM, 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, IBM, and ALS. In addition, arimoclomol has received breakthrough therapy designation (BTD) and repediatric 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 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 publicly, or