

**Company announcement**  
No. 07/2021

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

[www.orphazyme.com](http://www.orphazyme.com)

Company Registration No. 32266355

## Notice to convene Annual General Meeting

**Copenhagen, Denmark, March 3, 2021** – Orphazyme A/S (ORPHA.CO; ORPH) (the “Company”), a late-stage biopharmaceutical company pioneering the Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases, today announced that the Company’s Annual General Meeting will be held on:

**Thursday, March 25, 2021 at 5:00 PM (CET)**

at the Company’s address Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark.

In order to protect the health and safety of all, reduce the risk of COVID-19 spreading, and given the Danish government’s restrictions on assembly, we strongly recommend that shareholders refrain from attending the Annual General Meeting in person and instead exercise their shareholder rights by giving proxy to the Board of Directors or by voting by correspondence prior to the Annual General Meeting.

The notice to convene the Annual General Meeting, including Appendix 1: Candidates for the Board of Directors and Appendix 2: Revised Remuneration Policy, is attached.

Additional information about the Annual General Meeting is available on the Company’s website:

<https://orphazyme.gcs-web.com/annual-general-meeting-0>

**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, sIBM, 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 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.