

Company announcement
No. 02/2022

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

www.orphazyme.com

Company Registration No. 32266355

Notice to convene Extraordinary General Meeting

Copenhagen, Denmark, January 24, 2022 – 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 an Extraordinary General Meeting of the Company will be held on:

Tuesday, February 15, 2022 at 5:00 PM (CET)

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

In order to continuously provide the Company with the necessary flexibility to raise new capital to continue funding of its operations and protract a long-term capital structure, the Board of Directors proposes a number of authorizations to be approved by the Extraordinary General Meeting, including authorizations to issue shares, convertible bonds and warrants.

In general, the Board of Directors believes that the proposed authorizations will benefit the Company and its shareholders as they would provide flexibility to raise additional capital to fund the Company’s product candidate, arimoclomol, for the treatment of NPC during the US and EU regulatory approval processes.

Covid-19

We expect the current Covid-19 health risks and government precautions to continue on the date of the Extraordinary General Meeting.

Shareholders wishing to attend the Extraordinary General Meeting in person are encouraged not to bring advisors or other authorized guests to limit the number of physical attendees at the Extraordinary General Meeting. Further, meeting participants will be asked to present a valid corona passport and meeting participants having or showing symptoms of virus infection are kindly requested not to attend the meeting.

Shareholders who do not attend the Extraordinary General Meeting are encouraged to vote by submitting a postal vote or a proxy to the Board of Directors, as described below. Shareholders may submit written questions to the Company’s management before the Extraordinary General Meeting.

The above precautions are based on rules and recommendations issued by the Danish Health Authorities effective on the date hereof. If applicable rules and recommendations are updated before the Extraordinary General Meeting, additional precautions may be taken.

The notice to convene the Extraordinary General Meeting is attached as well as Appendix 1: Candidate for the Board of Directors.

Additional information about the Extraordinary 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, CFO

+45 28 98 90 55

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).

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.

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, including the U.S. and EU regulatory processes for the potential approval of arimoclomol and the potential raising of additional capital. 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.