

Company announcement
No. 18/2022

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

www.orphazyme.com

Company Registration No. 32266355

Notice received from the Depositary to holders of ADSs regarding termination of Deposit Agreement

NOTICE TO HOLDERS OF AMERICAN DEPOSITARY SHARES ("ADSs")
EVIDENCED BY
AMERICAN DEPOSITARY RECEIPTS ("ADRs")
REPRESENTING DEPOSITED COMMON SHARES OF:
ORPHAZYME A/S IN RESTRUCTURING
ONE ADS REPRESENTS ONE COMMON SHARE
CUSIP: 687305102 AND UNDERLYING ISIN: DK0060910917

Copenhagen, Denmark, April 6, 2022 – Orphazyme A/S in restructuring (ORPHA.CO) ("Orphazyme" or the "Company"), a late-stage biopharmaceutical company, has today received a notice from The Bank of New York Mellon, as depositary (the "Depositary"), that a notification has been sent to holders of American Depositary Shares ("ADSs") evidenced by American Depositary Receipts ("ADRs") representing deposited common shares of the Company, regarding termination of the Deposit Agreement dated September 28, 2020 among the Company, the Depositary, and Owners and ADR Holders (the "Deposit Agreement").

Owners and beneficial owners of the above ADRs ("ADR Holders") have on April 5, 2022 been notified by the Depositary that the Depositary will terminate the Deposit Agreement. As a result, the existing ADR facility will be terminated effective at 5:00 PM (Eastern Time) on July 6, 2022.

Under the terms of the Deposit Agreement, ADR Holders have until at least July 11, 2022 to surrender their Orphazyme ADRs for delivery of the underlying shares. If ADR Holders surrender ADRs for delivery of the underlying shares, ADR Holders must pay a cable fee of \$17.50, a cancellation fee of up to \$0.05 per ADRs surrendered and any applicable U.S. or local taxes or governmental charges. Payment should be made payable to the Depositary.

Subsequent to July 11, 2022 under the terms of the Deposit Agreement, the Depositary may attempt to sell the underlying shares. If the Depositary has sold such shares, ADR Holders must surrender their ADRs to obtain payment of the sale proceeds, net of the expenses of sale, any applicable U.S. or local taxes or government charges and a cancellation fee of up to \$0.05 per ADRs.

For additional information, please contact

Orphazyme A/S in restructuring

Anders Vadsholt, Chief Executive Officer and Chief Financial Officer

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About Orphazyme

Orphazyme is a late-stage biopharmaceutical company developing arimoclomol for Niemann-Pick disease type C (NPC). Orphazyme is headquartered in Denmark. Orphazyme's 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 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. The Company plans to request a Type C Meeting with the FDA in Q2 2022.

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 forward-looking statements about the termination of the ADS program and the treatment of the ordinary shares underlying the ADSs. 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, including pursuant to regulatory intervention. 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.