

Company announcement No. 17/2022 Inside information Orphazyme A/S in restructuring Ole Maaløes Vej 3 DK-2200 Copenhagen N

www.orphazyme.com Company Registration No. 32266355

Correction: Proposal for a statutory restructuring plan

Copenhagen, Denmark, April 4, 2022 – Orphazyme A/S in restructuring (ORPHA.CO) ("Orphazyme" or the "Company"), a late-stage biopharmaceutical company, today announced that, in respect to company announcement no. 15/2022 dated March 31, 2022, the Company has been made aware of a typographical error in the header of the table included in Appendix 1B regarding the interim set-off balance as of March 11, 2022.

Accordingly, the following correction is made to the header of the table included in Appendix 1B:

Current wording: "Beløb i mio. DKK / DKK million".

Corrected wording: "Beløb i tusinder DKK / DKK thousand".

Please find attached the corrected Appendix 1B to the proposal for a statutory restructuring plan. No other changes or corrections have been made to the proposal for a statutory restructuring plan, including appendices.

For additional information, please contact

Orphazyme A/S in restructuring

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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 Company's restructuring process. 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.