

Company announcement No. 23/2022 Inside information Orphazyme A/S in restructuring Ole Maaløes Vej 3 DK-2200 Copenhagen N

www.orphazyme.com Company Registration No. 32266355

Orphazyme announces update on in-court restructuring proceedings

Copenhagen, Denmark, April 29, 2022 – Orphazyme A/S in restructuring (ORPHA.CO; ORPH) ("Orphazyme" or the "Company"), a late-stage biopharmaceutical company, announces an update to the in-court restructuring proceedings of the Company (please see company announcement no. 10/2022 and 11/2022).

In accordance with the statutory restructuring plan referenced in company announcement nos. 15/2022, 17/2022 and 20/2022, Orphazyme has been pursuing potential opportunities for a sale of its assets and operations, a sale of the Company and/or a refinancing (potentially in combination with a compulsory composition).

Orphazyme has to date received certain non-binding offers (the "Offers") to purchase all of Orphazyme's assets and operations. The terms of the Offers received to date indicate that the vast majority of the consideration to be paid to Orphazyme would be used to settle outstanding recognised liabilities of Orphazyme's current creditors. Following negotiations, Orphazyme, with respect to the Offers received, expects to enter into exclusive negotiations with a selected potential buyer. Following completion of such transaction, Orphazyme would have no activities and would be dissolved, when possible.

There can be no guarantee that that any transaction will be completed as proposed, or at all, or at any particular time.

Further announcement(s) will be made by Orphazyme if a binding agreement is reached with respect to any offer.

For additional information, please contact

Orphazyme A/S in restructuring

Anders Vadsholt, Chief Executive Officer and Chief Financial Officer	+45 2898 9055
John Sommer Schmidt, Restructuring Administrator	+45 8620 7500

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 prospective sale to one or more third parties or refinancing, satisfaction of outstanding liabilities and timing thereof. 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 or judicial 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.