

**Company announcement**

No. 26/2022

Inside information

**Orphazyme A/S in restructuring**

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Company Registration No. 32266355

## Restructuring Proposal

**Copenhagen, Denmark, May 18, 2022** – Orphazyme A/S in restructuring (ORPHA.CO; ORPH) (“Orphazyme” or the “Company”), a late-stage biopharmaceutical company, announces that the enclosed restructuring proposal (the “Restructuring Proposal”) will be submitted to the Danish Maritime and Commercial High Court (the “Court”) and be sent to the Company’s known creditors following the agreement with KemPharm Denmark A/S to sell substantially all of its assets and business activities (the “Assets”), which is conditional upon the approval by the creditors and the affirmation of the Restructuring Proposal by the Court (please see company announcement no. 24/2022).

The Restructuring Proposal will be presented and voted on at a meeting with creditors to be held on Monday, May 30, 2022, at 1:00 PM (CEST) at the Maritime and Commercial High Court, Amaliegade 35, 2., 1256 Copenhagen C, Denmark. All creditors are entitled to attend the meeting.

If the Restructuring Proposal is adopted by the creditors and affirmed by the Court, the restructuring proceedings will be closed by completing the transfer of the Assets to KemPharm Denmark A/S, and Orphazyme will pay its debts to the creditors in accordance with the Restructuring Proposal. If the Restructuring Proposal is not adopted and affirmed, the restructuring proceedings will automatically cease and bankruptcy proceedings will be initiated against Orphazyme.

Please see the enclosed Restructuring Proposal including appendices.

**For additional information, please contact****Orphazyme A/S in restructuring**

Anders Vadsholt, Chief Executive Officer and Chief Financial Officer +45 2898 9055

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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 has requested a type B-meeting to be held early Q3 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 and the Company’s sale of substantially all of its assets and business activities to KemPharm Denmark A/S. 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.