

Company announcement No. 27/2022

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

www.orphazyme.com

Company Registration No. 32266355

Changes to the Board of Directors

Copenhagen, Denmark, May 23, 2022 – Orphazyme A/S in restructuring (ORPHA.CO; ORPH) ("Orphazyme" or the "Company"), a late-stage biopharmaceutical company, has previously announced its entry into an agreement with KemPharm Denmark A/S to sell substantially all of its assets and business activities (please see company announcement no. 24/2022) and submission of the restructuring proposal to the Danish Maritime and Commercial High Court (the "Court") and the Company's known creditors (please see company announcement no. 26/2022).

Orphazyme today announces that Stephanie Okey, Carrolee Barlow and Martin Bonde will step down from their positions as members of the Board of Directors as of May 23, 2022 with the view to reduce the number of members of the Board of Directors to three members, as Orphazyme will no longer have any ongoing operational business activities once the restructuring proposal is adopted by the creditors and affirmed by the Court and the transaction with KemPharm Denmark A/S is completed on or around June 1, 2022.

"I would like to thank Stephanie Okey, Carrolee Barlow and Martin Bonde for their many contributions to Orphazyme and we wish them all the best for the future," said Chairman of the Board of Directors, Georges Gemayel.

Until the upcoming Annual General Meeting expected to be held on June 29, 2022, the Board of Directors will consist of Georges Gemayel, Chairman of the Board of Directors, Bo Jesper Hansen, Deputy Chairman of the Board of Directors, and Andrew Mercieca.

Additionally, the Company's board committees will continue with the three board members for the Audit Committee and the Remuneration Committee, with Andrew Mercieca as chairman of the Audit Committee and Bo Jesper Hansen as chairman of the Remuneration Committee, and with two board members for the Nomination Committee, with Bo Jesper Hansen as chairman of the Nomination Committee. The Science Committee will be discontinued.

For the Annual General Meeting of Orphazyme expected to be held on June 29, 2022, it is the intention of the Board of Directors to propose re-election of the current Deputy Chairman of the Board of Directors, Bo Jesper Hansen, election of the current CEO and CFO, Anders Fink Vadsholt, as a new member of the Board of Directors, and election of the Company's current restructuring administrator, John Sommer Schmidt, as a new member of the Board of Directors.

The current Chairman of the Board of Directors, Georges Gemayel, and member of the Board of Directors, Andrew Mercieca, will not be seeking re-election at the upcoming Annual General Meeting.

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

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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, the Company's sale of substantially all of its assets and business activities to KemPharm Denmark A/S and the Company's annual general meeting 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