

Company announcement

No. 14/2022

Orphazyme A/S in restructuringOle Maaløes Vej 3
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Company Registration No. 32266355

Updated Financial Calendar for 2022

Copenhagen, Denmark, March 29, 2022 – Orphazyme A/S in restructuring (ORPHA.CO; ORPH) (“Orphazyme” or the “Company”), a late-stage biopharmaceutical company, announces an update to the Company’s Financial Calendar for 2022, following that the Danish Business Authority upon request from the Company has approved to extend the deadline for the Company’s filing of its annual report for the financial year ended December 31, 2021, following the in-court restructuring proceedings of the Company (please see company announcements no. 10/2022 and 11/2022). The extended deadline allows the Company to submit its annual report for 2021 no later than 1 month after the completion of the ongoing in-court restructuring proceedings in accordance with section 140 of the Danish Financial Statements Act. The Company is also in dialogue with the Danish Financial Supervisory Authority regarding the publication of the Company’s annual report for 2021.

The Company intends to submit a No Action Request to the U.S. Securities and Exchange Commission (“SEC”) to obtain relief from filing the Company’s annual report on Form 20-F with the SEC for the financial year ended December 31, 2021. The current deadline for filing of the Form 20-F with the SEC is May 2, 2022, absent an extension under Rule 12b-25, which the Company expects to obtain, extending the deadline to May 17, 2022.

Orphazyme has thus until further notice updated its financial calendar for the remainder of 2022:

Deadline for submission of shareholder proposals to Annual General Meeting

No later than six weeks prior to the Annual General Meeting 2022

Annual Report 2021

No later than three weeks prior to the Annual General Meeting 2022

Annual General Meeting 2022

No later than 1 month after the completion of the ongoing in-court restructuring proceedings

Interim Report First Half 2022

August 25, 2022

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 and has operations in Switzerland. ADSs representing Orphazyme’s shares are listed on Nasdaq U.S. (ORPH) and its shares are listed on Nasdaq Copenhagen (ORPHA). Orphazyme has filed for voluntary delisting of its ADSs representing Orphazyme’s shares listed on Nasdaq U.S.

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. On February 23, 2022, the EMA Committee for Medicinal Products for Human Use (CHMP) issued a negative Trend Vote on the Marketing Authorization Application (MAA) for arimoclomol in NPC filed with the European Medicines Agency (EMA). The Company has subsequently decided to withdraw its MAA.

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 intention to file annual reports in Denmark and the United States and the 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 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.