

Company announcement No. 11/2022

Orphazyme A/S
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<u>www.orphazyme.com</u> Company Registration No. 32266355

Notice of in-court restructuring proceedings of Orphazyme A/S

Copenhagen, Denmark, March 18, 2022 – Orphazyme A/S (ORPHA.CO; ORPH) ("Orphazyme" or the "Company"), a late-stage biopharmaceutical company, announces that following the Company's filing of a petition for an in-court restructuring (in Danish *rekonstruktion*) of Orphazyme on March 10, 2022 (please see company announcement no. 10/2022), the Danish Maritime and Commercial High Court (the "Court") has commenced the restructuring proceedings of the Company on March 11, 2022 at 7:00 am (CET). The Court has appointed attorney John Sommer Schmidt as restructuring administrator.

The restructuring administrator has sent a notice of commencement of in-court restructuring proceedings of Orphazyme to all known creditors in accordance with section 11 b of the Danish Insolvency Act.

A meeting with creditors will be held on April 7, 2022, at 11:00 AM (CEST) at Gorrissen Federspiel, Axel Torv 2, 1609 Copenhagen V. All creditors are entitled to attend the meeting where the restructuring plan will be presented. The statutory restructuring plan required to be prepared under the Danish Insolvency Act is expected to be sent by the restructuring administrator to all known creditors on March 31, 2022.

The purpose of the restructuring process is to explore whether a basis can be established which allows for all or part of the Company's operations to continue, including a basis for injecting further capital, and/or a basis for a sale of all or parts of the Company's assets. At this stage it is uncertain whether a solution can be found, and a further update will be provided at the appropriate time.

For additional information, please contact

Orphazyme A/S

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

John Sommer Schmidt, Restructuring Administrator +45 8620 7500

About Orphazyme A/S

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).

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. A marketing authorization application (MAA) for arimoclomol in NPC has been filed with the European Medicines Agency (EMA) and is under review. On February 17, 2022, the EMA Committee for Medicinal Products for Human Use (CHMP) issued a negative Trend Vote on the MAA following an Oral Explanation, indicating that its current orientation was to not approve arimoclomol when it convenes by the end of March 2022, and Orphazyme considers it unlikely that this position will change before the formal vote is undertaken.

Forward-looking statemen

This company announcement may contain certain forward-looking statements under the U.S. Private Securities Litigation Reform Act of 1995 and otherwise. 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. These statements may include, without limitation, any statements preceded by, followed by, or including words such as "target," "believe," "expect," "aim," "intend," "may," "anticipate," "estimate," "plan," "project," "will," "can have," "likely," "should," "would," and other words and terms of similar meaning or the negative thereof. Forward-looking statements are subject to inherent risks and uncertainties beyond the Company's control that could cause the Company's actual results, performance, or achievements to be materially different from the expected results, performance, or achievements expressed or implied by such forward-looking statements, including the risks and uncertainties that are described in the Risk Factors section of the Company's Annual Report on Form 20-F for the year ended December 31, 2020 filed with the U.S. Securities and Exchange Commission (SEC) on March 2, 2021, the Company's Report on Form 6-K filed with the SEC on June 11, 2021, and other filings Orphazyme makes with the SEC from time to time. These documents are available on the "Investors & Media" section of Orphazyme's website at www.orphazyme.com. 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.