Company announcement
No. 10/2022
Inside information

Orphazyme A/S
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Company Registration No. 32266355

Commencement of in-court restructuring of Orphazyme A/S

- *Will institute a reduction of approximately 50% of current work force*
- *Intends to delist American Depositary Shares from Nasdaq US*

Copenhagen, Denmark, March 10, 2022 – Orphazyme A/S (ORPHA.CO; ORPH) ("Orphazyme" or the "Company"), a late-stage biopharmaceutical company, announces that, following the receipt of the negative Trend Vote as announced on February 23, 2022 (please see company announcement no. 07/2022), and considering the Company's financial position, the Board of Directors of the Company has today decided to file a petition for an in-court restructuring of Orphazyme. As a part of the restructuring efforts, the Company will also institute a reduction of approximately 50% of the Company's current global workforce.

The aim of the in-court restructuring 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.

As part of the Company's efforts to reduce costs, Orphazyme recently closed its commercial operations in Germany, the UK, and the US. Specifically in Denmark, Orphazyme will tomorrow, March 11, 2022, initiate negotiations under the Danish Act on Collective Redundancies and the Act on Information and Consultation.

"*Following the very disappointing outcomes from the regulatory authorities in the US and EU, we are forced to consider some extremely difficult choices. Our employees have worked tirelessly with a focus on bringing arimoclomol as a potential new treatment option to patients with Niemann-Pick disease type C*", commented Orphazyme Chief Executive Officer, Anders Vadsholt. "*It is with a heavy heart that we are faced with the prospect of parting ways with valued colleagues, and I want to thank them for their hard work, their commitment to Orphazyme, and their outstanding dedication to showing up for patients in need.*"

The Company moreover announced that it plans to voluntarily delist American Depositary Shares (ADSs) representing its ordinary shares from the Nasdaq Global Select Market (Nasdaq). Orphazyme has given formal notice to Nasdaq of its intention to voluntarily delist the ADSs. Orphazyme intends to file a Form 25 with the Securities and Exchange Commission (SEC) on March 21, 2022, to initiate the delisting, which is expected to become effective on March 31, 2022. As soon thereafter as the Company is eligible, the Company intends to file a Form 15 with the SEC to suspend its reporting obligations under the Securities Exchange of 1934, as amended. The Company expects that the deregistration of the ADSs and the underlying ordinary shares will become effective 90 days after the filing of the Form 25 with the SEC.

For additional information, please contact

Orphazyme A/S

Georges Gemayel, Chairman of the Board of Directors

Bo Jesper Hansen, Deputy Chairman of the Board of Directors

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

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 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 U.S. and EU regulatory processes for the potential approval of arimoclomol by the FDA or EMA, the size of the Company's potential downsizing, the impact of the Company's restructuring process, and the Company's intention to delist the ADSs from Nasdaq and deregister the ADSs and ordinary shares with the SEC. 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," "could", 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.