

Company announcement

No. 06/2022

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Company Registration No. 32266355

Capital increase of 360,000 shares in Orphazyme A/S as a result of the utilization of the U.S. At-the-Market Offering Program

Copenhagen, Denmark, February 18, 2022 – Orphazyme A/S (ORPHA.CO (DK); ORPH (US)) (the “Company”), a late-stage biopharmaceutical company, announces that the Company has issued new shares in the period from Friday, February 11, 2022 to Thursday, February 17, 2022 as a result of the utilization of the Company’s U.S. At-the-Market Offering Program with Cowen and Company, LLC (“Cowen”). On February 11, 2022, a total of 360,000 ordinary shares of nominally DKK 1 each, represented by American Depositary Shares (“ADSs”), have been issued by the Company and sold in the market by Cowen as the sales agent at market price as determined by the Company’s Board of Directors in accordance with the authorization in article 3.1 of the Company’s Articles of Association.

After registration of the share capital increase relating to the share issuance, the Company’s share capital amounts to a nominal value of DKK 35,312,241 divided into 35,312,241 ordinary shares of DKK 1 per share. Each share carries one vote at the Company’s general meetings and accordingly the total number of voting rights in the Company is 35,312,241. The new ordinary shares rank *pari passu* with the Company’s existing ordinary shares and carry the same dividend and other rights. The ADSs do not carry the same rights as the Company’s ordinary shares and are not entitled to receive a dividend or vote as ordinary shares, except to the extent provided for through the depositary as record holder of the ordinary shares underlying the ADSs as set forth in the deposit agreement governing the ADSs.

The new ordinary shares are expected to be admitted to trading and official listing on Nasdaq Copenhagen under the permanent ISIN code DK0060910917 by the end of February 2022.

The Company continues to explore ways to strengthen its balance sheet.

Updated Articles of Association can be found at www.orphazyme.com.

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

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About Orphazyme A/S

Orphazyme is a late-stage biopharmaceutical company developing arimoclolomol for Niemann-Pick disease type C (NPC). Orphazyme is headquartered in Denmark and has operations in the U.S. and Switzerland. ADSs representing Orphazyme’s shares are listed on Nasdaq U.S. (ORPH) and its shares are listed on Nasdaq Copenhagen (ORPHA).

About arimoclolomol

Arimoclolomol 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. Arimoclolomol is administered orally, and has now been studied in 10 Phase 1, four Phase 2, and three pivotal Phase 2/3 trials. Arimoclolomol has received Orphan Drug Designation (ODD) for NPC in the US and EU. Arimoclolomol 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 arimoclolomol for the treatment of NPC. A marketing authorization application (MAA) for arimoclolomol in NPC has been filed with the European Medicines Agency and is under review.

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. 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.