

## **Amendment of proposals put forward at the Extraordinary General Meeting**

**Copenhagen, Denmark, February 14, 2022** – Orphazyme A/S (ORPHA.CO; ORPH) (“Orphazyme” or the “Company”), a late-stage biopharmaceutical company pioneering the Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases, today announced that the Board of Directors has decided to limit the scope of the proposed authorizations set forward under items 1.2, 1.3 and 1.4 of the agenda of the Extraordinary General Meeting to be held tomorrow, February 15, 2022.

This entails that the proposed authorizations to increase the share capital by issuance of shares, convertible bonds and warrants, as applicable, will be subject to such share capital increases taking place at or above market price and that share capital increases below market price will not be authorized as initially proposed.

The wording of the revised more limited authorizations proposed under agenda items 1.2, 1.3 and 1.4 are set out below:

**Article 3.7:**

*"In the period until 1 January 2027, the Board of Directors is authorised to increase the Company's share capital through one or more issues of new shares without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 20,000,000. The capital increase may be effected by cash payment or conversion of debt and shall take place at market price as determined by the Board of Directors."*

**Article 3.8:**

*"In the period until 1 January 2027, the Board of Directors is authorised to issue convertible bonds on one or more occasions without pre-emption rights for the existing shareholders and with a total principal amount of up to DKK 70,000,000 which are convertible into shares in the Company. The convertible bonds shall be effected by cash payment. The conversion price as determined by the Board of Directors may be above or at the market price at the time of issuance of the convertible bonds. The issuance of convertible bonds may be directed at qualified investors. The Board of Directors is authorised in the period until 1 January 2027 to increase the Company's share capital by up to nominally DKK 20,000,000 by conversion of convertible bonds issued pursuant to this Article 3.8 and to effect the associated capital increases."*

**Article 3.9:**

*"In the period until 1 January 2027, the Board of Directors is authorised to issue warrants on one or more occasions without pre-emption rights for the existing shareholders granting the holders right to subscribe for shares in the Company for a total amount of up to nominally DKK 20,000,000. The Board of Directors is entitled to determine the exercise price for the warrants upon issue given that the exercise price may be above or at the market price at the time of issuance. The Board of Directors is authorised in the period until 1 January 2027 to increase the Company's share capital by up to nominally DKK 20,000,000 resulting from the exercise of warrants pursuant to this Article 3.9 and to effect the associated capital increases."*

The amended authorizations proposed under agenda items 1.2, 1.3 and 1.4 shall be passed by at least two-thirds of the votes cast as well as at least two-thirds of the share capital represented at the Extraordinary General Meeting, cf. Article 7.2 of the Articles of Association.

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

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**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 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 arimoclomol**

Arimoclomol is an investigational drug candidate that amplifies the production of heat shock proteins (HSPs). HSPs can rescue defective misfolded proteins, clear protein aggregates, 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 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](http://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.