

**Company announcement**

No. 04/2021

**Orphazyme A/S**

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Company Registration No. 32266355

## **Capital increase of 80,553 shares (equivalent to approximately 0.23% of the existing shares) in Orphazyme A/S as a result of an issue of bonus shares to KUMCRI and directed issuance of new shares**

**Copenhagen, Denmark, February 25, 2021** – Orphazyme A/S (ORPHA.CO; ORPH) (“the Company”), a late-stage biopharmaceutical company pioneering the Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases, today decided on a capital increase as a result of a directed issue of bonus shares to the University of Kansas Medical Center Research Institute, Inc. (“KUMCRI”) and a directed issue of new shares to Kim Stratton, former CEO of the Company. The capital increase was decided pursuant to the Board of Directors’ authorization laid down in Article 3.2 and 3.3 of the Articles of Association.

According to the license agreement entered into on October 31, 2017 between among others the Company and KUMCRI, the Company shall, following the end of each calendar year, issue or otherwise deliver to KUMCRI a total number of shares equal to half of the aggregate amount of cash funding that KUMCRI has reported to have spent under the grant from the US Food and Drug Administration received with respect to the Company’s on-going Phase 2/3 clinical trial on arimoclomol for the treatment of sIBM. On the basis of a financial report of expenditures provided by KUMCRI, the Company will issue 22,553 new shares each with a nominal value of DKK 1, equal to approximately 0.065% of its total share capital, in favor of KUMCRI. Accordingly, the Company’s share capital will be increased by nominally DKK 22,553 by way of an issue of bonus shares at par value by transfer of DKK 22,553 from the Company’s distributable reserves. The number of shares to be delivered to KUMCRI has been calculated based on the average closing price of the Company’s shares on Nasdaq Copenhagen during a 30-day period preceding January 29, 2021.

According to agreements entered into between Kim Stratton and the Company, Kim Stratton is entitled to receive a grant of 58,000 shares at a subscription price of DKK 1 per share as the conditions related to the grant have been met. Thus, the Company will issue 58,000 new shares each with a nominal value of DKK 1, and the share capital will be increased by nominally DKK 58,000.

Accordingly, the Company will issue a total of 80,553 shares, and the Company’s share capital will in total be increased by nominally DKK 80,553, equal to approximately 0,23% of the Company’s existing share capital.

The new shares are negotiable instruments and will in every respect carry the same rights as the existing shares from the time of registration of the capital increase with the Danish Business Authority (which is expected to occur today). The new shares are expected to be admitted to trading and official listing on Nasdaq Copenhagen in the week commencing March 1, 2021.

Following the share capital increase, the total nominal share capital will be DKK 34,948,387, divided into 34,948,387 shares each with a nominal value of DKK 1. Each share carries one vote at the Company’s general meetings and accordingly the total number of voting rights in the Company will be 34,948,387. Updated Articles of Association can be found at [www.orphazyme.com](http://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 pioneering the Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases. The company is harnessing amplification of Heat-Shock Proteins (or HSPs) in order to develop and commercialize novel therapeutics for diseases caused by protein misfolding, protein aggregation, and lysosomal dysfunction, including lysosomal storage diseases and neuromuscular degenerative diseases. Arimoclomol, the company's lead candidate, is in clinical development for four orphan diseases: Niemann-Pick disease Type C (NPC), Amyotrophic Lateral Sclerosis (ALS), Inclusion Body Myositis (IBM) and Gaucher disease. Orphazyme is headquartered in Denmark and has operations in the U.S. and Switzerland. Orphazyme's shares are listed on Nasdaq U.S. (ORPH) and 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 seven phase 1, four phase 2 and one pivotal phase 2/3 trial. Arimoclomol is in clinical development for NPC, Gaucher Disease, sIBM, and ALS. Arimoclomol has received orphan drug designation (ODD) for NPC, IBM, and ALS in the US and EU. Arimoclomol has received fast-track designation (FTD) from the U.S. Food and Drug Administration (FDA) for NPC, IBM and ALS. In addition, arimoclomol has received breakthrough therapy designation (BTD) and rare-pediatric disease designation (RPDD) from the FDA for NPC.

**Forward-looking statement**

This company announcement may contain certain forward-looking statements. 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. 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.