

Company announcement
No. 01/2021

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

Major shareholder announcement

Copenhagen, Denmark, January 18, 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, hereby announces the receipt of notification pursuant to Section 38 of the Danish Capital Markets Act from Danske Bank A/S, that as of January 15, 2021, Danske Bank A/S holds a total of 34,697,703 shares in the Company, corresponding to 3.49% of the Company’s share capital and that Danske Bank A/S as of January 15, 2021 controls 4.95% of the voting rights in the Company.

Danske Bank A/S’ shareholding consists of a 2.24% indirect and 1.25% direct ownership through Danica Pension Livsforsikringsaktieselskab, Danica Pension Försäkringsaktiebolag, Investeringsforeningen Danske Invest and Danske Invest SICAV. Danske Bank A/S’ control of voting rights in the Company consists of a 3.70% indirect and 1.25% direct control through Danica Pension Livsforsikringsaktieselskab, Danica Pension Försäkringsaktiebolag, Investeringsforeningen Danske Invest and Danske Invest SICAV.

For additional information, please contact

Orphazyme A/S

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About Orphazyme A/S

The Company 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. The Company is headquartered in Denmark and has operations in the U.S. and Switzerland. The Company’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, crosses the blood-brain barrier, 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, IBM, 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 statements

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.