Company announcement
No. 16/2021

Orphazyme A/S
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Company Registration No. 32266355

Major shareholder announcement

Copenhagen, Denmark, June 15, 2021 – Orphazyme A/S (ORPHA.CO; ORPH) ("the Company"), a late-stage biopharmaceutical company pioneering the heat shock protein response for the treatment of rare diseases, hereby announces the receipt of notification pursuant to Section 38 of the Danish Capital Markets Act from Sunstone Life Science Ventures A/S and Sunstone Life Science Ventures Fund II K/S, that as of June 11, 2021, Sunstone Life Science Ventures Fund II K/S holds less than 5% of the Company's share capital and that Sunstone Life Science Ventures A/S as of June 11, 2021, controls less than 5% of the voting rights in the Company.

Sunstone Life Science Ventures Fund II K/S is managed by its general partner Sunstone LSV General Partner II ApS. Sunstone Life Science Ventures A/S control all voting rights of Sunstone LSV General Partner II ApS.

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 rare diseases. The company is harnessing amplification of heat shock proteins (or HSPs) to develop and commercialize novel therapeutics for diseases caused by protein misfolding, protein aggregation, and lysosomal dysfunction, including lysosomal storage diseases. Arimoclomol, the company's lead candidate, is in clinical development for Niemann-Pick disease type C (NPC) 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.CO).

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 is in clinical development for NPC and Gaucher disease. Arimoclomol has received orphan drug designation (ODD) for NPC in the US and EU. Arimoclomol has received fast-track designation (FTD) from the U.S. Food and Drug Administration (FDA) for NPC. In addition, arimoclomol has received breakthrough therapy designation (BTD) and rare-pediatric disease designation (RPDD) from the FDA for NPC. Arimoclomol is an investigational treatment and has not been approved by the FDA.

Forward-looking statement
This company announcement may contain certain forward-looking statements, including in respect of the expected PDUFA action date of June 17, 2021 for arimoclomol for the treatment of NPC, the potential U.S. approval of arimoclomol in June and the opinion from the Committee for Medicinal Products for Human Use (CHMP) later this year. 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.