

Company announcement No. 63/2020 Orphazyme A/S Ole Maaløes Vej 3 DK-2200 Copenhagen N

www.orphazyme.com Company Registration No. 32266355

Major shareholder announcement

Copenhagen, Denmark, October 5, 2020 – Orphazyme A/S (ORPHA.CO; ORPH) ("Orphazyme"), 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 October 2, 2020, Danske Bank A/S holds a total of 1,148,770 shares in Orphazyme, corresponding to 3.31% of the share capital and that Danske Bank A/S as of October 2, 2020 controls 4.91% of the voting rights in the Orphazyme.

Danske Bank A/S' shareholding consists of a 2.57% indirect and 0.74% 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 Orphazyme consists of a 4.17% indirect and 0.74% direct control through Danica Pension Livsforsikringsaktieselskab, Danica Pension Försäkringsaktiebolag, Investeringsforeningen Danske Invest SICAV.

For additional information, please contact

Orphazyme A/S

Anders Vadsholt, CFO

+45 28 98 90 55

About Orphazyme A/S

Orphazyme is a late-stage biopharmaceutical company pioneering the Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases. Orphazyme 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, Orphazyme's lead candidate, is in clinical development for four orphan diseases: Niemann-Pick disease Type C (NPC), Amyotrophic Lateral Sclerosis (ALS), sporadic Inclusion Body Myositis (sIBM) and Gaucher disease. Orphazyme is headquartered in Denmark and has operations in the U.S. and Switzerland. Orphazyme's ordinary shares are listed on Nasdaq Copenhagen (ORPHA.CO).

Forward-looking statements

This company announcement may contain certain forward-looking statements, including with respect to the terms, timing and completion of the proposed offering. Although Orphazyme 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 Orphazyme'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," would," would," and other words and terms of similar meaning or the negative thereof. Forward-looking statements are subject to inherent risks and uncertainties beyond Orphazyme's control that could cause Orphazyme's actual results, performance, or achievements to be materially different from the expected results, performance, or achievements. Except as required by law, Orphazyme assumes no obligation to update these forward-looking statements, even if new information becomes available in the future.

----00000-----