

Company announcement

No. 19/2021

Orphazyme A/S
Ole Maaløes Vej 3
DK-2200 Copenhagen N

www.orphazyme.com Company Registration No. 32266355

Major shareholder announcement

Copenhagen, Denmark, June 24, 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 Aescap Venture Management B.V, that as of June 16, 2021, Aescap Venture Management B.V indirectly holds shares corresponding to less than 5% of the total share capital and voting rights in the Company.

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) in order to develop and commercialize novel therapeutics for diseases caused by protein misfolding, protein aggregation, and lysosomal dysfunction. Arimoclomol, the company's lead candidate, is in clinical development for rare diseases including 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).