

**Company announcement**  
No. 62/2020

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

## Major shareholder announcement

**Copenhagen, Denmark, October 5, 2020** – Orphazyme A/S (ORPHA.CO) (“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 Consonance Capman GP LLC that as of October 1, 2020, Consonance Capman GP LLC has reduced its indirect holdings of shares and voting rights in Orphazyme and now indirectly holds 1,339,909 shares and voting rights in Orphazyme, corresponding to 3.86% of the total share capital and voting rights of Orphazyme.

The shares and voting rights are directly held by (i) Consonance Capital Master Account LP which is controlled by Consonance Capital Management LP which in turn is controlled by Consonance Capman GP LLC, (ii) Consonance Capital Opportunity Master Fund LP which is controlled by Consonance Capital Management LP which in turn is controlled by Consonance Capman GP LLC and (iii) P Consonance Opportunities Ltd which is controlled by Consonance Capital Opportunities Fund Mgmt. LP which in turn is controlled by Consonance Capman GP LLC.

Consonance Capital is a healthcare investment firm that invests broadly across the healthcare landscape. Public investments are made under the direction of Consonance Capital Management (“CCM”), a long/short public equity manager. CCM is a focused life sciences manager that invests in innovative therapies and products. Consonance Capital was established in 2005 and is located in New York City.

CCM is the investment manager of the Orphazyme shares recently acquired by its funds.

### 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. 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,” “could”, 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 expressed or implied by such forward-looking statements. Except as required by law, Orphazyme 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.

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