

Company announcement

No. 18/2020

Orphazyme A/S

Ole Maaløes Vej 3

DK-2200 Copenhagen N

www.orphazyme.com

Company Registration No. 32266355

Major shareholder announcement

Copenhagen, Denmark, February 12, 2020 – Orphazyme A/S (ORPHA.CO), a biopharmaceutical company pioneering 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 February 11, 2020, Consonance Capman GP LLC indirectly holds 1,900,000 shares and voting rights in Orphazyme, corresponding to 7.03% 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, established in 2005 and located in New York City, USA, 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.

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

For additional information, please contact**Orphazyme A/S**

Anders Vadsholt, CFO

+45 28 98 90 55

About Orphazyme A/S

Orphazyme is a biopharmaceutical company focused on bringing novel treatments to patients living with life-threatening or debilitating rare diseases. Our research focuses on developing therapies for diseases caused by misfolding of proteins, including lysosomal storage diseases. Arimoclomol, the company's lead candidate, is in clinical development for four orphan diseases: Niemann-Pick disease Type C, Gaucher disease, sporadic Inclusion Body Myositis, and Amyotrophic Lateral Sclerosis. The Denmark-based company is listed on Nasdaq Copenhagen (ORPHA.CO). For more information, please visit www.orphazyme.com.

Forward-looking statement

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.