

Company announcement No. 37/2020 Inside information Orphazyme A/S Ole Maaløes Vej 3 DK-2200 Copenhagen N

www.orphazyme.com Company Registration No. 32266355

Orphazyme announces confidential submission of a draft registration statement for a potential registered public offering in the United States

Copenhagen, Denmark, July 1, 2020 – Orphazyme A/S (ORPHA.CO) (the "Company"), a late-stage biopharmaceutical company harnessing the amplification of the Heat-Shock Proteins in order to develop and commercialize novel therapeutics for the treatment of neurodegenerative orphan diseases, today announced that it has confidentially submitted a draft registration statement on Form F-1 to the U.S. Securities and Exchange Commission ("SEC") relating to a potential registered public offering of the Company's ordinary shares, in the form of American Depositary Shares ("ADSs"), in the United States. The draft registration statement is subject to ongoing review by the SEC, and the proposed listing of ADSs representing the Company's ordinary shares is subject to approval for listing on the Nasdaq Global Market. The Company's ordinary shares will continue to be admitted for trading on Nasdaq Copenhagen. Any potential offering is subject to market and other conditions, including board approval.

This company announcement does not constitute an offer to sell or the solicitation of an offer to buy any securities issued or to be issued by the Company. Any offers, solicitations or offers to buy, or any sales of securities issued or to be issued by the Company will be made in accordance with the registration requirements of the Securities Act of 1933, as amended (the "Securities Act"). This announcement is being issued in accordance with Rule 135 under the Securities Act.

For additional information, please contact

Orphazyme A/S

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About Orphazyme A/S

Orphazyme is a biopharmaceutical company pioneering the Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases. The company is focused on developing therapies for diseases caused by protein misfolding, protein aggregation, and lysosomal dysfunction. Arimoclomol, the company's lead candidate, is in clinical development for four orphan diseases: Niemann-Pick disease Type C (NPC), Gaucher Disease, sporadic Inclusion Body Myositis (sIBM), and Amyotrophic Lateral Sclerosis (ALS). 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, including the clinical development and potential benefits of arimoclomol for Gaucher disease, NPC, SIBM and ALS, 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," "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.