

Company announcement

No. 49/2020

Orphazyme A/S

Ole Maaløes Vej 3

DK-2200 Copenhagen N

www.orphazyme.com

Company Registration No. 32266355

Orphazyme files registration statement in the U.S. in connection with proposed global offering, consisting of initial public offering of ADSs in the U.S. and concurrent private placement of ordinary shares in Europe

Copenhagen, Denmark, September 4, 2020 – Orphazyme A/S (ORPHA.CO) (the "Company"), a late-stage biopharmaceutical company pioneering the Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases, today announced that it has filed a registration statement on Form F-1 (the "Registration Statement") with the U.S. Securities and Exchange Commission ("SEC") relating to a proposed registered initial public offering of the Company's ordinary shares, to be delivered in the form of American Depositary Shares ("ADSs"), in the United States, as well as a concurrent private placement of ordinary shares (together with the ADSs, the "Securities") to certain qualified investors in Europe. The number and price of the Securities to be sold in the offering has not yet been determined.

The Company has applied to list the ADSs on the Nasdaq Global Market in the United States under the symbol "ORPH". ADSs are U.S. dollar-denominated negotiable instruments issued by a depository bank that facilitate U.S. trading and investment in shares of non-U.S. companies. Each ADS represents the right to receive one ordinary share.

BofA Securities, Cowen, and Guggenheim Securities are acting as the global coordinators and joint book-running managers of the global offering. Danske Markets is acting as the lead manager of the global offering. In addition, BofA Securities, Cowen, Guggenheim Securities (in each case, or their affiliates), and Danske Bank are acting as joint book-running managers in the European private placement.

A registration statement on Form F-1 relating to the proposed sale of these Securities has been filed with the SEC but has not yet become effective. These Securities may not be sold, nor may offers to buy be accepted, prior to the time the Registration Statement becomes effective. The Form F-1 Registration Statement filed with the SEC does not contain any material information not already published by the Company.

This press release shall not constitute an offer to sell or the solicitation of an offer to buy these Securities, nor shall there be any sale of any Securities in any state or jurisdiction in which such offer, solicitation, or sale would be unlawful prior to registration or qualification under the securities laws of any such state or jurisdiction. Any potential offering is subject to market and other conditions, including board approval.

The Board of Directors has convened an Extraordinary General Meeting to be held on September 21, 2020, for the purpose of passing certain resolutions with respect to the proposed offering. The Board of Directors has not yet determined whether to proceed with an offering, but should the Extraordinary General Meeting authorize the Board of Directors to increase the Company's share capital, the Board of Directors will have the flexibility to determine the timing, number of Securities to be issued, and the price of the Securities. Even if the Board of Directors determines to proceed with the proposed offering, the offering may not be consummated.

The ordinary shares of the Company are currently listed on Nasdaq Copenhagen under the symbol "ORPHA". The Company's ordinary shares will remain listed for trading on Nasdaq Copenhagen in Denmark.

The proposed offering of ADSs will only be made by means of a prospectus. When the preliminary prospectus relating to the offering is available, copies may be obtained from BofA Securities, NC1-004-03-43, 200 North College Street, 3rd floor, Charlotte, NC 28255-0001, Attn: Prospectus Department, or by email at dg.prospectus_requests@bofa.com; Cowen, c/o Broadridge Financial Solutions, Attn: Prospectus Department, 1155 Long Island Avenue, Edgewood, NY 11717, by email at PostSaleManualRequests@broadridge.com or by telephone at (833) 297-2926; or Guggenheim Securities, Attention: Equity Syndicate Department, 330 Madison Avenue, 8th Floor, New York, NY 10017, by telephone at (212) 518-9544, or by email at GSEquityProspectusDelivery@guggenheimpartners.com.

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. 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, including lysosomal storage diseases and neuromuscular degenerative diseases. Arimoclomol, the company'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 shares are listed on Nasdaq Copenhagen (ORPHA.CO).

Forward-looking statement

This company announcement may contain certain forward-looking statements, including relating to the terms of the proposed offering, the Extraordinary General Meeting and the completion of the proposed offering. 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.