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**Company announcement**  
No. 54/2020

**Orphazyme A/S**  
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Company Registration No. 32266355

## **Orphazyme A/S provides update on previously announced global offering**

**Copenhagen, Denmark, September 25, 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 an update on its previously announced global offering. On September 24, 2020, the Company received a filing communication from the U.S. Food and Drug Administration (FDA) in connection with its new drug application (NDA) for arimoclomol for the treatment of Niemann-Pick disease Type C. The FDA letter follows acceptance on a Priority Review basis by the FDA of the Company’s NDA for arimoclomol in NPC and the FDA’s establishment of the Prescription Drug User Fee Act (“PDUFA”) target action date of March 17, 2021. The Company’s receipt of the filing communication does not impact the FDA’s acceptance of the Company’s NDA, the target PDUFA action date or the FDA’s Priority Review determination.

In accordance with applicable securities laws, the Company intends to amend the registration statement on Form F-1 on file with the U.S. Securities and Exchange Commission on September 25, 2020. The Company has also updated its proposed timing for the global offering referenced in the company announcement no. 51/2020, dated September 21, 2020, and now intends to price the global offering and announce the subscription price on September 28, 2020, subject to market conditions, and to settle the global offering on October 1, 2020. The Company also reported that the book for the global offering is fully covered as of September 24, 2020.

### **Important information**

The ordinary shares of the Company are currently listed on Nasdaq Copenhagen under the symbol “ORPHA”. In connection with the global offering, Nasdaq has approved the listing of the American Depositary Shares (“ADSs” and, together with the ordinary shares, the “Securities”) in the U.S. under the symbol “ORPH”.

A registration statement on Form F-1 relating to the proposed sale of the Securities has been filed with the SEC but has not yet become effective. The Securities may not be sold, nor may offers to buy be accepted, prior to the time the registration statement on Form F-1 becomes effective.

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The Board has not yet determined whether to complete the global offering. Even if the Board determines to complete the global offering, the global offering may not be consummated. Neither the timing, number of shares, number of ordinary underlying shares of Orphazyme, nor the price of the ADSs and thereby the price of the underlying shares has been finally determined. If consummated, the final price for the ordinary shares and ADSs will be determined following a book-building process.

This Company announcement shall not constitute an offer to sell or the solicitation of an offer to buy the 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. The offering is subject to market and other customary closing conditions, and there can be no assurance as to whether or when the offering may be completed.

**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. 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 statements**

This company announcement may contain certain forward-looking statements, including with respect to the terms, timing and completion of the proposed offering and the timing and approval for marketing by the FDA of arimoclomol for NPC. 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.

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