

**Company announcement**  
No. 52/2020

Orphazyme A/S  
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Company Registration No. 32266355

## **Orphazyme A/S commences a global offering, consisting of initial public offering of ADSs in the U.S. and concurrent private placement of ordinary shares in Europe**

- *Orphazyme A/S has today commenced an initial public offering of American Depositary Shares on the Nasdaq Global Market ("Nasdaq") in the United States under the symbol "ORPH", filed a preliminary prospectus relating to the global offering and commenced a concurrent private placement of ordinary shares in Europe*
- *The proposed global offering of up to 7,616,146 new ordinary shares consists of an initial public offering of ordinary shares in the form of ADSs offered in the U.S. offering and a concurrent private placement of ordinary shares offered in a European private placement*

**Copenhagen, Denmark, September 21, 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 commenced a global offering of up to 7,616,146 new ordinary shares, consisting of an initial public offering of American Depositary Shares ("ADSs") representing ordinary shares of the Company in the U.S. (the "U.S. Offering") and a concurrent private placement of ordinary shares in Europe (the "European Private Placement" and together with the U.S. Offering, the "Offering").

With reference to the company announcement no. 49/2020, dated September 4, 2020, regarding the initial public filing of registration statement on Form F-1 (the "F-1") with the U.S. Securities and Exchange Commission (the "SEC"), Orphazyme further announced that it has today filed an amendment to the registration statement, including a preliminary prospectus (the "U.S. Prospectus"), with the SEC and has commenced the Offering of up to 7,616,146 new ordinary shares, which may be in the form of ADSs (together the "Securities"). The total number of ordinary shares (including ordinary shares in the form of ADSs) in the U.S. Offering and the European Private Placement is subject to reallocation between these offerings to the extent permitted under applicable laws and regulations.

In addition, Orphazyme expects to grant BofA Securities, Cowen, and Guggenheim Securities, as representatives of the several underwriters in the Offering, acting severally and not jointly, an option to subscribe for and purchase additional ordinary shares, which may be in the form of ADSs, up to a number corresponding to 15% of the total number of ordinary shares in the Offering.

The Company has applied to list the ADSs on Nasdaq in the U.S. under the symbol "ORPH".

The Board has not yet determined whether to complete the Offering. Even if the Board determines to complete the Offering, the 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.

**Use of proceeds**

Orphazyme intends to use the net proceeds from the Offering together with its existing cash resources for the following purposes:

- to continue the regulatory approval process for and fund the commercial launch, if approved, of arimoclomol for the treatment of NPC;
- to advance the clinical development of arimoclomol for the treatment of ALS;
- to advance the clinical development of arimoclomol for the treatment of sIBM;
- to advance the clinical development of arimoclomol for the treatment of neurological Gaucher disease; and
- the remaining amounts for working capital and general corporate purposes, including to fund the development of our next generation of HSP amplifiers.

Orphazyme's expected use of the net proceeds from the Offering represents its current intentions based upon its present plans and business conditions. As of the date of this announcement, Orphazyme cannot predict with certainty all of the particular uses of the net proceeds of the Offering or the amounts that Orphazyme will actually spend on the uses set forth above. The amounts and timing of Orphazyme's actual use of net proceeds will vary based on numerous factors, including its ability to obtain additional financing, the relative success and cost of its research, preclinical and clinical development programs, and whether Orphazyme enters into collaborations with third parties in the future. As a result, the management of Orphazyme will have broad discretion in the application of the net proceeds, and investors will be relying on its judgment regarding the application of the net proceeds of the Offering.

Except for the expected use of the net proceeds from the Offering described above and number of securities offered in the Offering, the preliminary prospectus filed with the SEC does not contain any material information not already published by Orphazyme.

**ADSs**

ADSs are U.S. dollar-denominated negotiable instruments issued by a depositary bank that facilitate U.S. trading and investment in shares of non-U.S. companies. Each ADS represents one underlying share of Orphazyme.

**Share capital and authorization**

Subject to pricing of the Offering, Orphazyme's Board of Directors (the "Board") expects to exercise the authorization in accordance with article 3.1 of Orphazyme's Articles of Association granted by Orphazyme's extraordinary general meeting held on September 21, 2020, to increase Orphazyme's share capital by such amount of new shares offered in the Offering. Subject to the Offering being fully subscribed and based on the assumed offering size, the Offering would represent 28.1% of Orphazyme's current share capital, excluding the additional shares that the underwriters have an option to purchase, and 32.3% assuming full exercise by the underwriters of their option to purchase additional shares.

**Admission for trading and official listing**

Subject to completion of the Offering, it is expected that the new ordinary shares will be admitted to trading and official listing on Nasdaq Copenhagen A/S in the existing permanent ISIN code of the Company, DK0060910917. Any new ordinary shares will initially be issued in a temporary ISIN code, which will only be registered with VP Securities A/S and not admitted to trading and official listing on Nasdaq Copenhagen A/S or any other stock exchange. The temporary ISIN code in VP Securities A/S will be merged with the existing permanent ISIN code for the existing shares, DK0060910917, as soon as possible following registration of the share capital increase with the Danish Business Authority. Subject to completion and settlement of the Offering, the new ordinary shares are expected to be admitted to trading and official listing on Nasdaq Copenhagen A/S on or around September 30, 2020.

The Company expects to publish a listing prospectus (the "Listing Prospectus") in order to admit the new ordinary shares to trading and official listing on Nasdaq Copenhagen A/S.

**Expected timetable for the Offering**

<b>Date (CET)</b>	<b>Event</b>
September 21, 2020	Launch of the Offering and filing of preliminary prospectus with the SEC
September 24, 2020	Pricing of the Offering – announcement of subscription price
September 25, 2020	First day of trading of the ADSs on Nasdaq
September 28, 2020	Deadline for final filing of the U.S. Prospectus with the SEC and approval and publication of the Listing Prospectus
September 29, 2020	Settlement of the Offering and registration of the capital increase with the Danish Business Authority
September 30, 2020	Admission of the new ordinary shares to trading and official listing on Nasdaq Copenhagen
October 1, 2020	Merger of ISIN codes

**Important information**

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.

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.

This company announcement 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. 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.

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.

The proposed offering of ADSs will only be made by means of a prospectus. Copies of the preliminary prospectus relating to the offering 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](mailto: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](mailto: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](mailto:GSEquityProspectusDelivery@guggenheimpartners.com).

**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. 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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