

Company announcement No. 55/2020

Orphazyme A/S Ole Maaløes Vej 3 DK-2200 Copenhagen N

www.orphazyme.com Company Registration No. 32266355

Orphazyme prices its global offering, consisting of an initial public offering of American Depositary Shares in the U.S. and a concurrent private placement of ordinary shares in Europe

- As announced on September 21, 2020, Orphazyme commenced an initial public offering of American Depositary Shares ("ADSs") in the United States, and commenced a concurrent private placement of ordinary shares in Europe
- Today, September 29, 2020 (CET), Orphazyme announces the pricing of the global offering at a price per new ordinary share of DKK 70.1844, corresponding to a price of USD 11.00 per ADS

 The aggregate gross proceeds from the global offering will amount to approximately DKK 534,534,637 (USD 83,777,606 using a DKK/USD exchange rate of 6.3804) (assuming no exercise of the option to purchase additional shares) and DKK 614,714,770 (USD 96,344,237 using a DKK/USD exchange rate of 6.3804) (assuming full exercise of the option to purchase additional shares). Orphazyme intends to use the net proceeds as set forth below

Copenhagen, Denmark, September 29, 2020 (CET) – With reference to the company announcement no. 52/2020, dated September 21, 2020, pursuant to which Orphazyme A/S (ORPHA.CO) ("Orphazyme") announced the filing of a F-1 registration statement, including a preliminary prospectus, and commencement of the global offering, and with reference to the company announcement no. 54/2020, pursuant to which Orphazyme provided an update on the previously announced global offering, Orphazyme, a late-stage biopharmaceutical company pioneering the Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases, today announces the pricing of the global offering, consisting of an initial public offering of 3,966,146 ADSs on the Nasdaq Global Select Market ("Nasdaq") at the public offering price of USD 11.00 per ADS (equivalent to a subscription price of DKK 70.1844 per ordinary share using a DKK/USD exchange rate of 6.3804), each such ADS representing one ordinary share of Orphazyme, and a concurrent private placement of 3,650,000 ordinary shares in Europe at a subscription price of DKK 70.1844 (together with the initial public offering in the United States, the "global offering").

In respect of the global offering, the Board of Directors of Orphazyme has exercised the authorization in article 3.1 of Orphazyme's Articles of Association to increase the share capital by issuing 7,616,146 new ordinary shares of nominal value DKK 1 per share at a subscription price of DKK 70.1844 per new ordinary share.

In addition, Orphazyme has granted BofA Securities, Cowen and Guggenheim Securities, as representatives of the several underwriters in the global offering, acting severally and not jointly, an option to subscribe for and purchase up to 1,142,421 additional ordinary shares, which may be in the form of ADSs or ordinary shares, corresponding to 15% of the total number of ordinary shares in the global offering. The Board of Directors of Orphazyme has exercised the authorization in article 3.1 of Orphazyme's Articles of Association to increase the share capital by up to 1,142,421 ordinary shares of nominal value DKK 1 per share.



The ADSs are expected to begin trading on Nasdaq in the U.S. under the symbol "ORPH" on September 29, 2020. Orphazyme expects to submit an application for the admittance of the new ordinary shares to trading and official listing on Nasdaq Copenhagen A/S under the symbol "ORPHA" on September 29, 2020.

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 Form F-1 registration statement, including the preliminary prospectus and the F-6 registration statement related to the global offering have been declared effective by the U.S. Securities and Exchange Commission. The proposed offering of ADSs will only be made by means of a prospectus. A copy of the preliminary prospectus and, when available, the final prospectus may be obtained at no cost by visiting the SEC website at www.sec.gov. Copies of the preliminary prospectus and the final prospectus, when available, relating to the global offering may also 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.

Use of proceeds

The aggregate gross proceeds from the issue will amount to approximately DKK 534,534,637 (USD 83,777,606 using a DKK/USD exchange rate of 6.3804) (assuming no exercise of the option to purchase additional shares) and DKK 614,714,770 (USD 96,344,237 using a DKK/USD exchange rate of 6.3804) (assuming full exercise of the option to purchase additional shares).

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.

Capital increase

Subject to completion and settlement of the global offering, Orphazyme's share capital will consist of 34,697,703 ordinary shares of nominal value DKK 1 per share, assuming the option to purchase additional shares is not exercised (or 35,840,124 ordinary shares of nominal value DKK 1 per share if the option to purchase additional shares is exercised in full). The global offering represents 28.1% of Orphazyme's registered share capital before the registration of the share capital increase related to the global offering, and excluding the additional ordinary shares that the underwriters have an option to subscribe for and purchase, and 32.3% assuming full exercise by the underwriters of their option to subscribe for and purchase additional ordinary shares (which may be in the form of ADSs). The global offering will represent 22% of Orphazyme's registered share capital after the registration of the share capital increase related to the global offering, assuming the option to purchase additional shares is not exercised, and 24.4% assuming full exercise by the underwriters of their option to subscribe for may be in the form of ADSs).

Expected timetable for the Global Offering

Date (CET) September 29, 2020 September 30, 2020	Event First day of trading of the ADSs on Nasdaq Deadline for final filing of the U.S. Prospectus with the SEC and approval and
October 1, 2020	publication of the Listing Prospectus Settlement of the global offering and registration of the capital increase with the Danish Business Authority
October 2, 2020	Admission of the new ordinary shares to trading and official listing on Nasdaq Copenhagen
October 5, 2020	Merger of ISIN codes on Nasdaq Copenhagen



Admission for trading and official listing

Subject to completion of the global 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, DK0061411378, 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 ordinary 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 global offering, the new ordinary shares are expected to be admitted to trading and official listing on Nasdaq Copenhagen A/S on or around October 2, 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.

Important information

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

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 global offering is subject to market and other customary closing conditions, and there can be no assurance as to whether or when the global 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. Orphazyme is a late-stage biopharmaceutical company pioneering the Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases. Orphazyme 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, Orphazyme'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 ordinary 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 Orphazyme 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 Orphazyme'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 Orphazyme's control that could cause Orphazyme's actual results, performance, or achievements to be materially different from the expected results, performance, or achievements. Except as required by law, Orphazyme 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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