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**Company announcement**  
No. 08/2020

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

## **Orphazyme initiates directed issue and private placement of shares**

**Copenhagen, Denmark, February 6, 2020** – Orphazyme A/S (ORPHA.CO), a biopharmaceutical company pioneering Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases, today announces that the Board of Directors has resolved to initiate a directed issue and private placement of up to an aggregate of 7,032,937 new shares and existing shares, at market price as determined by the Board of Directors of the Company through a book-building process, corresponding to approximately 35.16% of the Company's currently registered share capital.

The directed issue and private placement is expected to cover the Company's financing needs until well into 2021, and support in particular the imminent US and European filings for approval of arimoclomol for the treatment of Niemann-Pick disease Type C (NPC), as well as the preparations for commercial launch.

Up to 3,961,264 new shares will be offered in a directed issue and private placement, along with a private placement of up to 3,071,673 existing shares at market price as determined by the Board of Directors of the Company through a book-building process, corresponding to approximately 35.16% of the Company's currently registered share capital.

As well as the development of arimoclomol in NPC, Orphazyme also intends to use the proceeds to cover completion of arimoclomol's clinical development in sporadic Inclusion Body Myositis (sIBM) and Amyotrophic Lateral Sclerosis (ALS) and to prepare regulatory filings in Europe and potentially the US in these indications, as well as for general corporate purposes.

Based on the positive data package announced in 2019, Orphazyme currently expects to submit an application for approval of arimoclomol for the treatment of NPC in H1 2020 in the US and in H2 2020 in the EU. Orphazyme expects to release results from the ongoing phase 2 trial for Gaucher Disease in H1 2020 and continue its ongoing phase 3 trial in ALS and phase 2/3 trial in sIBM. Orphazyme has been granted Fast Track and Breakthrough Therapy designation from the US Food and Drug Administration (FDA) for the development of arimoclomol in NPC, and Fast Track designation for the development of arimoclomol in sIBM.

### **Directed issue and private placement of up to 7,032,937 shares**

The Board of Directors of Orphazyme A/S (the "Company") has today resolved to make a directed issue and private placement of up to 3,961,264 new shares of a nominal value of DKK 1 each (the "New Shares") and private placement of up to 3,071,673 existing shares of a nominal value of DKK 1 each (the "Existing Shares" and together with the New Shares, the "Offer Shares") at market price as determined by the Board of Directors of the Company through a book-building process (the "Offering"). The New Shares will be issued without pre-emption rights for existing shareholders.

The offering of Existing Shares is facilitated by a share loan from Novo Holdings A/S and Orpha Pooling B.V. (the "Lending Shareholders") to the Company pursuant to a stock lending and subscription agreement with an obligation for the Company to redeliver new shares of an equivalent number as the Existing Shares borrowed by the Company from each of the Lending Shareholders (the "Replacement Shares"), which will be issued without pre-emption rights for existing shareholders. The Lending Shareholders do not participate in the Offering, and are only facilitating the loan of the Lending Shares for purposes of the Company's offering of Existing Shares in the Offering.

The resolution to issue the New Shares and the Replacement Shares is adopted pursuant to the authorization from the general meeting in accordance with Article 3.4 of the Company's Articles of Association.

The Offer Shares will be offered to institutional and professional investors in Denmark and internationally (the New Shares being issued without pre-emption rights to the Company's existing shareholders) pursuant to applicable exemptions from the obligation to publish a prospectus, including if outside the United States in "offshore transactions" (within the meaning of Regulation S under the United States Securities Act of 1933, as amended (the "Securities Act")) and if in the United States to qualified institutional buyers as defined by Rule 144A of the Securities Act or institutional accredited investors as defined by Regulation D under the Securities Act pursuant to an exemption from the registration requirements of the Securities Act.

The Company, the Board of Directors, the Executive Management and certain executive officers have undertaken lock-up obligations on customary terms and conditions according to which the Company, the Board of Directors, the Executive Management and certain executive officers, subject to certain customary exemptions, shall not sell, contract to sell or otherwise dispose of or issue any securities of the Company for a period of 90 days from the date of registration of the share capital increases related to the Offering.

#### **Subscription price**

The subscription price and gross proceeds will be determined by the Board of Directors of the Company through a book-building process. It is expected that the allocation, the subscription price and the number of Offer Shares will be announced on February 7, 2020, but the Offering may be closed earlier. If the Offering is oversubscribed, allocation of Offer Shares will take place on an individual basis.

#### **Share capital and the shares**

If all 7,032,937 New Shares and Replacement Shares are subscribed for and issued, the issued share capital of the Company will increase by approximately 35.16% from DKK 20,005,449 to DKK 27,038,386 divided into 27,038,386 shares of nominal value DKK 1 each.

The New Shares and the Replacement Shares will be issued in the name of the shareholder and be recorded in the Company's share register. The New Shares and the Replacement Shares will be negotiable instruments and will in every respect carry the same rights as the existing shares in the Company. The New Shares and Replacement Shares will carry the right to receive dividends (if and when declared) and other rights in the Company as from the time of registration of the capital increase with the Danish Business Authority, which is expected to take place around February 11, 2020.

The Existing Shares will also be registered in the name of the shareholder and be recorded in the Company's share register.

#### **Admission for trading and official listing**

The New Shares will be issued under the temporary ISIN code DK0061274289. No application for admission to trading and official listing has been, or will be, filed for the New Shares issued under the temporary ISIN code, and the temporary ISIN code will only be registered with VP Securities A/S. 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 increases with the Danish Business Authority. The New Shares are expected to be admitted to trading and official listing on Nasdaq Copenhagen A/S on February 12, 2020.

The Replacement Shares will be issued under the temporary ISIN code DK0061274362. No application for trading and official listing has, or will be, been filed for the Replacement Shares issued under the temporary ISIN code, and the temporary ISIN code will only be registered with VP Securities A/S. The temporary ISIN code in VP Securities A/S, under which the Replacement Shares will be issued, will later be merged with the existing permanent ISIN code, DK0060910917, in which the existing shares of the Company are issued.

The Existing Shares are already admitted to trading and official listing at Nasdaq Copenhagen A/S under the permanent ISIN code DK0060910917.

### Expected timetable

Date	Event
February 6, 2020	Decision by the Board of Directors to exercise its authorization to increase the share capital
February 7, 2020	Pricing and allocation – announcement of subscription price
February 11, 2020	Settlement and payment for the Offer Shares and Replacement Shares
February 11, 2020	Registration of the capital increases with the Danish Business Authority
February 12, 2020	New Shares to be admitted to trading and official listing on Nasdaq Copenhagen A/S

The admission to trading and official listing of the New Shares and the later admission to trading and official listing of the Replacement Shares on Nasdaq Copenhagen A/S is subject to the Offering not being withdrawn prior to the settlement of the Offering and the Company making an announcement to that effect, and in respect of the Replacement Shares only (which will only be delivered to the Lending Shareholders and not be sold as part of the Offering), the publication of a listing prospectus to be approved by the Danish Financial Supervisory Authority, which is expected to occur no later than by the end of March 2020.

### Managers

Guggenheim Securities, LLC and Danske Bank A/S are acting as Joint Global Coordinators and will receive subscription and purchase orders in connection with the Offering.

### For additional information, please contact

#### Orphazyme A/S

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### About Orphazyme A/S

Orphazyme is a biopharmaceutical company focused on bringing novel treatments to patients living with life-threatening or debilitating rare diseases. Our research focuses on developing therapies for diseases caused by misfolding of proteins, including lysosomal storage diseases. Arimoclomol, the company's lead candidate, is in clinical development for four orphan diseases: Niemann-Pick disease Type C, Gaucher disease, sporadic Inclusion Body Myositis, and Amyotrophic Lateral Sclerosis. The Denmark-based company is listed on Nasdaq Copenhagen (ORPHA.CO). For more information, please visit [www.orphazyme.com](http://www.orphazyme.com).

### Forward-looking statements

This company announcement may contain certain forward-looking statements, including with respect to the timing of the Company's clinical trials and the expected submission of application for approval of arimoclomol for the treatment of NPC in the U.S. 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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This announcement is not for publication or distribution, directly or indirectly, in or into the United States of America. This announcement and the information it contains do not constitute an offer of securities for sale nor a solicitation of any offer to buy any securities issued by Orphazyme A/S in the United States or any other jurisdiction where such offer or sale would be unlawful and the announcement and the information contained herein are not distribution or release, directly or indirectly, in or into such jurisdictions. The securities referred to herein have not been and will not be registered under the U.S. Securities Act of 1933, as amended (the "U.S. Securities Act"), and may not be offered or sold in the United States, except pursuant to registration or an applicable exemption from registration under the U.S. Securities Act and applicable state securities law. No public offering of securities is being made in the United States under the U.S. Securities Act.

With respect to the member states of the European Economic Area subject to Regulation 2017/1129/EU (together with any applicable implementing measures, the "Prospectus Regulation") (each a "Relevant Member State"), no action has been undertaken or will be undertaken to make an offer to the public of the securities referred to herein requiring the publication of a prospectus in any Relevant Member State. As a result, these securities may only be offered or sold in any Relevant Member State pursuant to an exemption under the Prospectus Regulation.

This announcement is only addressed to, and directed at, persons in Relevant Member States of the European Economic Area who are "qualified investors" within the meaning of Article 2(e) of the Prospectus Regulation ("Qualified Investors").

In the United Kingdom, this announcement is directed only at Qualified Investors (i) who are "investment professionals" falling within Article 19(5) of the Financial Services and Markets Act 2000 (Financial Promotion) Order 2005, as amended (the "Order"), or (ii) persons falling within Article 49(2)(a)-(d) ("high net worth companies, unincorporated associations etc.") of the Order all such persons together being referred to as ("Relevant Persons").

Under no circumstances should persons who are not Relevant Persons rely or act upon the contents of this announcement. Any investment or investment activity to which this announcement relates in the United Kingdom is available only to, and will be engaged only with, Relevant Persons.

Any investment decision in connection with the Offering must be made on the basis of all publicly available information relating to Orphazyme A/S and Orphazyme A/S' shares. Such information has not been independently verified by Guggenheim Securities, LLC and Danske Bank A/S. The information contained in this announcement is for background purposes only and does not purport to be full or complete. No reliance may be placed for any purpose on the information contained in this announcement or its accuracy or completeness. This announcement does not purport to identify or suggest the risks (direct or indirect) which may be associated with an investment in Orphazyme A/S and Orphazyme A/S' shares.

Guggenheim Securities, LLC and Danske Bank A/S are acting for Orphazyme A/S only in connection with the Offering and no one else, and will not be responsible to anyone other than Orphazyme A/S for providing the protections offered to clients of Guggenheim Securities, LLC and Danske Bank A/S, nor for providing advice in relation to the Orphazyme A/S' shares or the Offering.

In connection with any offering of the Offer Shares, Guggenheim Securities, LLC and Danske Bank A/S and any of their respective affiliates acting as an investor for their own account may take up as a proprietary position any Offer Shares and in that capacity may retain, purchase or sell for their own account such Offer Shares. In addition certain of Guggenheim Securities, LLC and Danske Bank A/S or their affiliates may enter into financing arrangements and swaps with investors in connection with which Guggenheim Securities, LLC and Danske Bank A/S (or their affiliates) may from time to time acquire, hold or dispose of Orphazyme A/S' shares. Guggenheim Securities, LLC and Danske Bank A/S do not intend to disclose the extent of any such investment or transactions otherwise than in accordance with any legal or regulatory obligation to do so.