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Company announcement
No. 14/2020

Orphazyme A/S
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www.orphazyme.com
Company Registration No. 32266355

Orphazyme completes registration of share capital increases of an aggregated nominal value of DKK 7,032,937

Copenhagen, Denmark, February 11, 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 registration of share capital increases of an aggregated nominal value of DKK 7,032,937 has been completed.

Reference is made to company announcements no. 08/2020 and no. 12/2020.

The capital increases of Orphazyme A/S (the "Company") with an aggregated nominal value of DKK 7,032,937 (7,032,937 shares of a nominal value of DKK 1 each), representing approximately 35.16% of the share capital prior to the capital increases, have been completed and registered with the Danish Business Authority.

After registration of the capital increases, the share capital of the Company amounts to a nominal value of DKK 27,038,386 divided into 27,038,386 shares of DKK 1 each. Each share of DKK 1 carries one vote at the Company's general meetings and accordingly the total number of voting rights in the Company are 27,038,386.

The new shares subscribed for in the directed issue and private placement are expected to be admitted to trading and official listing under the permanent ISIN code DK0060910917 with effect from February 12, 2020.

The shares to be redelivered by the Company to each of Novo Holdings A/S and Orpha Pooling B.V. are expected to be admitted to trading and official listing under the permanent ISIN code DK0060910917 subsequent to 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.

Updated Articles of Association can be found at www.orphazyme.com.

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.

Forward-looking statements

This company announcement may contain certain forward-looking statements. 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 Danske Bank A/S and Guggenheim Securities, LLC. 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.

Danske Bank A/S and Guggenheim Securities, LLC 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 Danske Bank A/S and Guggenheim Securities, LLC, nor for providing advice in relation to the Orphazyme A/S' shares or the Offering.

In connection with any offering of the Offer Shares, Danske Bank A/S and Guggenheim Securities, LLC 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 Danske Bank A/S and Guggenheim Securities, LLC or their affiliates may enter into financing arrangements and swaps with investors in connection with which Danske Bank A/S and Guggenheim Securities, LLC (or their affiliates) may from time to time acquire, hold or dispose of Orphazyme A/S' shares. Danske Bank A/S and Guggenheim Securities, LLC 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.