

Company announcement

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Orphazyme A/SOle Maaløes Vej 3
DK-2200 Copenhagen Nwww.orphazyme.com
Company Registration No. 32266355

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Completion of the Offering and registration of share capital increase

With reference to company announcements no. 2/2017 dated 6 November 2017 and no. 5/2017 dated 16 November 2017, Orphazyme A/S ("Orphazyme") today announces the completion of its Initial Public Offering (the "IPO" or "Offering") and admission to trading and official listing on Nasdaq Copenhagen A/S ("Nasdaq Copenhagen").

The Offering has today proceeded to completion and the admission to trading and official listing on Nasdaq Copenhagen today becomes unconditional. The offer shares allocated to investors in the form of temporary purchase certificates, as described in company announcement no. 5/2017 dated 16 November 2017, as well as the shares allocated to, among others, certain members of the Board of Directors, Executive Management and certain key employees in connection with exercise of their respective pre-IPO warrants, as described in the offering circular published by Orphazyme on 6 November 2017, have been settled.

As a result, Orphazyme's share capital has been increased by 8,338,092 new shares of a nominal value of DKK 1 each, which has been registered with the Danish Business Authority.

Following the share capital increase, the total nominal share capital of Orphazyme is DKK 19,928,184, divided into 19,928,184 shares each with a nominal value of DKK 1. Each share carries one vote at Orphazyme's general meetings. Updated Articles of Association can be found at www.orphazyme.com.

The offer shares are delivered in the form of temporary purchase certificates under the temporary ISIN (DK0060911055). On 22 November 2017, the temporary purchase certificates will automatically be exchanged in VP SECURITIES A/S for a corresponding number of shares in Orphazyme under the permanent ISIN (DK0060910917). In connection with the automatic exchange, the temporary purchase certificates will cease to exist.

The last day of trading of the temporary purchase certificates will be 20 November 2017, and the first day of trading of Orphazyme's shares will be 21 November 2017. Consequently, after the trading has closed on Nasdaq Copenhagen on 20 November 2017, all future trading in Orphazyme's shares on Nasdaq Copenhagen will continue and settle under the permanent ISIN (DK0060910917) and under the symbol "ORPHA".

Bank Syndicate

Carnegie and Danske Bank are acting as Joint Global Coordinators and Joint Bookrunners, and Oddo BHF SCA is acting as Co-Lead Manager for the IPO.

For additional information, please contact**Orphazyme**

Anders Hinsby, CEO

+45 31 44 31 39

About Orphazyme

Orphazyme is a Danish biotech company with a late stage orphan drug pipeline, developing new treatment options for orphan protein misfolding diseases. The Company was founded in 2009 based on early scientific discovery in heat shock proteins ("HSPs"). Since inception, the Company has translated scientific discovery into a late stage clinical development programme. The Company is headquartered in Copenhagen and currently has 30 employees.

The Company focuses on severe and mostly fatal diseases with a high unmet need, and with a particularly strong commitment to neuromuscular diseases and a group of severe genetic diseases called lysosomal storage diseases. The Company plans to pursue development of its lead candidate through to registration in the EU and the United States after which launch and commercialisation is expected to be undertaken by the Company.

The lead candidate arimoclomol is in development as a potential treatment for four orphan diseases; two neuromuscular diseases, sporadic Inclusion Body Myositis ("sIBM") and Amyotrophic Lateral Sclerosis ("ALS"), and two lysosomal storage diseases, Niemann Pick type C ("NPC") and Gaucher disease.

Important notice

This announcement is not an offer to sell or a solicitation of any offer to buy any securities issued by Orphazyme A/S (the "Company") in any jurisdiction where such offer or sale would be unlawful and the announcement and the information contained herein are not for distribution or release, directly or indirectly, in or into such jurisdictions.

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In any member state of the European Economic Area ("EEA Member State"), other than Denmark, that has implemented Directive 2003/71/EC as amended (together with any applicable implementing measures in any member State, the "Prospectus Directive"), this communication is only addressed to and is only directed at investors in that EEA Member State who fulfil the criteria for exemption from the obligation to publish an offering memorandum, including qualified investors, within the meaning of the Prospectus Directive as implemented in each such EEA Member State.

This announcement is only being distributed to and is only directed at (i) persons who are outside the United Kingdom or (ii) to investment professionals falling within Article 19(5) of the U.K. Financial Services and Markets Act 2000 (Financial Promotion) Order 2005 (the "Order") or (iii) high net worth entities falling within Article 49(2)(a) – (d) of the Order (the persons de-scribed in (i) through (iii) above together being referred to as "relevant persons"). The securities are only available to, and any invitation, offer or agreement to subscribe, purchase or otherwise acquire such securities will be engaged in only with, relevant persons. Any person who is not a relevant person should not act or rely on this announcement or any of its contents.

In connection with the offering, Danske Bank A/S (the "Stabilising Manager") (or persons acting on behalf of the Stabilising Manager) may over-allot securities or effect transactions with a view to supporting the market price of the

securities at a level higher than that which might otherwise prevail. However, there is no assurance that the Stabilising Manager (or persons acting on behalf of the Stabilising Manager) will undertake stabilisation. Any stabilisation action may begin on or after the date of commencement of trading and official listing of the securities on Nasdaq Copenhagen A/S and, if begun, may be ended at any time, but it must end no later than 30 days after the date of commencement of trading and official listing of the securities.