

Company announcement

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Orphazyme A/S

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Company Registration No. 32266355

Capital increase of 26,060 shares (equivalent to approximately 0.13% of the existing shares) in Orphazyme A/S as a result of an issue of bonus shares to KLSDC and UCLB

Copenhagen, Denmark, January 31, 2019 – Orphazyme A/S (ticker: ORPHA.CO), a biopharmaceutical company dedicated to developing treatments for patients living with rare diseases, today decided on a capital increase as a result of a directed issue of bonus shares to Kansas Life Sciences Development Company, Inc. (“KLSDC”) and UCL Business PLC (“UCLB”). The capital increase was decided pursuant to the Board of Directors’ authorization laid down in Article 3.6 of the Articles of Association.

According to the license agreement entered into on October 31, 2017 between among others Orphazyme A/S, KLSDC, and UCLB, Orphazyme A/S shall, following the end of each calendar year, issue or otherwise deliver to KLSDC and UCLB a total number of shares equal to the aggregate amount of cash funding that KLSDC and UCLB have reported to have spent under the grant from the US Food and Drug Administration received with respect to Orphazyme A/S’ on-going Phase II/III clinical trial on arimoclomol for the treatment of sIBM. On the basis of a financial report of expenditures provided by KLSDC and UCLB, Orphazyme A/S will issue 26,060 new shares each with a nominal value of DKK 1, equal to approximately 0.13% of its total share capital, in favor of KLSDC and UCLB. Accordingly, Orphazyme A/S’ share capital will be increased by nominally DKK 26,060 by way of an issue of bonus shares at par value by transfer of DKK 26,060 from Orphazyme’s distributable reserves. The number of shares to be delivered to KLSDC and UCLB has been calculated based on the average closing price of Orphazyme’s shares on Nasdaq Copenhagen during a 30-day period preceding the date of issuance. Reference is made to the prospectus published by Orphazyme A/S on November 6, 2017 for a further description of the license agreement.

The new shares are negotiable instruments and will in every respect carry the same rights as the existing shares from the time of registration of the capital increase with the Danish Business Authority (which is expected to occur today). The new shares are expected to be admitted to trading and official listing on Nasdaq Copenhagen on or around February 4, 2019.

Following the share capital increase, the total nominal share capital will be DKK 19,965,624, divided into 19,965,624 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.

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 and lysosomal dysfunction. 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 statement

This press release 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 press release 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.