

Company announcement

No. 02/2019

Orphazyme A/S

Ole Maaløes Vej 3

DK-2200 Copenhagen N

www.orphazyme.com

Company Registration No. 32266355

Major shareholder announcement

Copenhagen, January 30, 2019 – Orphazyme A/S, a biopharmaceutical company dedicated to developing treatments for patients living with rare diseases, hereby announces the receipt of notification pursuant to Section 38 of the Danish Capital Markets Act from Novo Holdings A/S on behalf of Novo Nordisk Fonden that Novo Holdings A/S as of today's date holds a total of 2,984,368 shares in the Company, corresponding to 14.97% of the share capital and the voting rights in the Company.

Novo Holdings A/S is wholly-owned by Novo Nordisk Fonden.

For additional information, please contact**Orphazyme A/S**

Anders Vadsholt, CFO

+45 28 98 90 55

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.