

Company announcement

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Orphazyme A/S

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Company Registration No. 32266355

Orphazyme strengthens balance sheet with EUR 9 million financing from Kreos Capital

- *Loan agreement to provide EUR 9 million upon closing*
- *Strengthens balance sheet ahead of anticipated filing of arimoclomol for Niemann Pick Type-C (NPC)*

Copenhagen, Denmark, August 27, 2019 – Orphazyme A/S (ticker: ORPHA.CO), a biopharmaceutical company dedicated to developing treatments for patients living with rare diseases, today announces that it has entered into a structured debt facility (Loan Agreement) with Kreos Capital VI (UK) Limited (Kreos) to secure funding of EUR 9 million. Orphazyme plans to draw down the loan on September 1, 2019. Proceeds from the loan will be used to fund the on-going preparations for filing of arimoclomol in the US and Europe, commercial planning, and for general corporate purposes.

Anders Fink Vadsholt, CFO of Orphazyme said, "Engaging in this loan facility diversifies our financing strategy and allows us, with only limited dilution, to bolster our cash position as we move closer to significant value inflection points, including the anticipated filing of arimoclomol for NPC in the US and Europe during H1 2020."

Aris Constantinides, General Partner at Kreos, commented, "This is an important time for Orphazyme as the company prepares to file its lead program with regulatory authorities in the US and Europe. We are very pleased to support Orphazyme in its mission to bring innovative new medicines to patients with rare diseases."

About the financing

Term	42 months
Repayment schedule	The first twelve (12) months are interest only with the remaining thirty (30) months requiring equal instalments comprising principal and interest
Interest rate	Annual fixed interest rate of 9.75%
Representations, warranties and general undertakings	Usual representation and warranties are included in the loan agreement. The group is subject to general undertakings such as restrictions on financial indebtedness, security and disposals outside its ordinary course of business. No financial covenants are included
Security	First priority security over certain assets, including intellectual property
Fees	The company shall pay various customary fees to Kreos for the establishment of the loan upon execution of the loan agreement, and in connection with drawn down and a facilitation fee payable at Kreos' discretion prior to the earlier of 8 years or delisting.

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.

About Kreos Capital

Kreos Capital is the leading provider of growth-debt financing to high-growth companies in Europe and Israel with revenues up to EUR 300 million. Since 1998, as the pioneer growth debt provider across Europe and Israel, Kreos has completed 540 transactions and committed more than EUR 2.3 billion in 15 different countries. Kreos is dedicated to supporting management teams and their equity investors with flexible loan structures for all stages of a growth company's development and to address the needs for growth capital, working capital, acquisition financings, lower mid-market buy-outs, roll-up strategies, banks re-financings, as well as pre- and post-IPO financings. Kreos' most recent fund, EUR 700 million Kreos VI, was launched in January 2019. The Kreos global team has extensive debt financing, management, and equity investing experience, covering the markets in Europe and Israel from its locations in London, Tel-Aviv, and Stockholm.

Forward-looking statement

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.