

Company announcement

No. 03/2022

Inside information

Orphazyme A/S

Ole Maaløes Vej 3

DK-2200 Copenhagen N

www.orphazyme.com

Company Registration No. 32266355

Changes to the Executive Management of Orphazyme – Anders Vadsholt to be appointed CEO

Copenhagen, Denmark, January 31, 2022 – Orphazyme A/S (ORPHA.CO; ORPH) (“Orphazyme” or the “Company”), a late-stage biopharmaceutical company pioneering the Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases, today announces that Chief Executive Officer (CEO) Christophe Bourdon will resign from his position as CEO at Orphazyme A/S to take on the role as CEO of LEO Pharma A/S. Effective April 1, 2022, Chief Financial Officer (CFO), Anders Vadsholt, will be appointed CEO of Orphazyme.

Chairman of the Board of Directors, Georges Gemayel, said: “*Christophe Bourdon’s leadership has skillfully navigated Orphazyme through turbulent times, resulting in the company identifying a pathway to continue to pursue the approval of arimoclomol for the treatment of Niemann-Pick disease type C (NPC) in both Europe and the US. I would like to thank Christophe for his significant contribution to the Company and wish him all the best in his future endeavors. The Board is very pleased to be able to appoint Anders Vadsholt as the Company’s new CEO. Having held the position of CFO since 2016, Anders brings great understanding of the goals and values at Orphazyme and his appointment as CEO ensures continuity in leadership at the Company for the important journey ahead – especially, leading the team during the important next steps for Orphazyme as we work to deliver on our commitment to NPC patients.*”

CFO, Anders Vadsholt, said: “*I have enjoyed a strong partnership with Christophe during the past year as we, together with our tenacious and dedicated Orphazyme team, worked towards getting arimoclomol approved in Europe and the US. I look forward to continuing to pursue that goal as I believe arimoclomol, if approved, has the potential to make a major difference in the lives of thousands of patients suffering from NPC.*”

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

Georges Gemayel, Chairman of the Board of Directors

Bo Jesper Hansen, Deputy Chairman of the Board of Directors

Anders Vadsholt, Chief Financial Officer

+45 2898 9055

About Orphazyme A/S

Orphazyme is a late-stage biopharmaceutical company developing arimoclomol for Niemann-Pick disease type C (NPC). Orphazyme is headquartered in Denmark and has operations in the U.S. and Switzerland. ADSs representing Orphazyme’s shares are listed on Nasdaq U.S. (ORPH) and its shares are listed on Nasdaq Copenhagen (ORPHA).

About arimoclomol

Arimoclomol is an investigational drug candidate that amplifies the production of heat shock proteins (HSPs). HSPs can rescue defective misfolded proteins, clear protein aggregates, and improve the function of lysosomes. Arimoclomol is administered orally, and has now been studied in 10 Phase 1, four Phase 2, and three pivotal Phase 2/3 trials. Arimoclomol has received Orphan Drug Designation (ODD) for NPC in the US and EU. Arimoclomol has received Fast-Track Designation (FTD), Breakthrough Therapy Designation (BTD), and Rare Pediatric Disease Designation (RPDD) from the U.S. Food and Drug Administration (FDA) for NPC. On June 17, 2021, Orphazyme received a Complete Response Letter from the FDA regarding its New Drug Application for arimoclomol for the treatment of NPC. A marketing authorization application (MAA) for arimoclomol in NPC has been filed with the European Medicines Agency and is under review.

Forward-looking statement

This company announcement may contain certain forward-looking statements under the U.S. Private Securities Litigation Reform Act of 1995 and otherwise, including the U.S. and EU regulatory processes for the potential approval of arimoclomol. 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, including the risks and uncertainties that are described in the Risk Factors section of the Company's Annual Report on Form 20-F for the year ended December 31, 2020 filed with the U.S. Securities and Exchange Commission (SEC) on March 2, 2021, the Company's Report on Form 6-K filed with the SEC on June 11, 2021, and other filings Orphazyme makes with the SEC from time to time. These documents are available on the "Investors & Media" section of Orphazyme's website at www.orphazyme.com. 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.