

Company announcement

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Orphazyme A/S
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www.orphazyme.com Company Registration No. 32266355

Orphazyme establishes a U.S. At-the-Market Offering Program with Cowen

Copenhagen – November 4, 2021 – Orphazyme A/S (ORPHA.CO; ORPH or the "Company"), a late-stage biopharmaceutical company, today announces that it has filed with the U.S. Securities and Exchange Commission (the "SEC") a prospectus supplement ("Prospectus") relating to a U.S. At-the-Market Offering Program, pursuant to which the Company may issue and sell American Depositary Shares ("ADSs") having an aggregate offering price of up to \$50,000,000, each ADS representing one ordinary share of the Company, to be sold in the United States at market price, from time to time, at its option, in "at the market" transactions on The Nasdaq Global Select Market (the "ATM Program"). The ADSs are intended to be sold pursuant to a Sales Agreement (the "Sales Agreement") with Cowen and Company, LLC ("Cowen"). The timing of any sales will depend on a variety of factors.

The ADSs intended to be sold under the Sales Agreement, if any, will be issued and sold by methods deemed to be an "at the market offering" as defined in Rule 415(a)(4) promulgated under the Securities Act of 1933, as amended, or in negotiated transactions, if authorized by the Company, in each case, pursuant to the Company's effective shelf registration statement on Form F-3. Any ADSs representing ordinary shares will be issued through capital increases without any pre-emption rights for existing shareholders subject to the Board of Directors deciding to increase the share capital in connection with each such issuance. The aggregate number of any such ADSs sold under the ATM Program is expected to be disclosed on a weekly basis or such longer period as may be agreed with Nasdaq Copenhagen. No assurance can be made that sales under the ATM Program will take place.

To the extent that ADSs are issued pursuant to the ATM Program, the Company expects to use the net proceeds from sales of any ADSs pursuant to the ATM Program to continue the regulatory approval process for and fund the commercial launch, if approved, of arimoclomol for the treatment of Niemann-Pick disease type C and for working capital and general corporate purposes.

A shelf registration statement on Form F-3 relating to this ATM Program was filed with the SEC on October 15, 2021 and declared effective by the SEC on October 22, 2021. Any sales under the ATM Program will be made pursuant to the Prospectus relating to the ATM Program. Before purchasing ADSs in the offerings, prospective investors should read the Prospectus, together with the documents incorporated by reference therein. A copy of the Prospectus may be obtained on the SEC's website at www.sec.gov. Alternatively, a copy of such Prospectus may be obtained from Cowen and Company, LLC, c/o Broadridge Financial Solutions, 1155 Long Island Avenue, Edgewood, NY, 11717, Attn: Prospectus Department, by email at PostSaleManualRequests@broadridge.com or by telephone at (833) 297-2926.

This company announcement does not constitute an offer to sell or a solicitation to buy the securities mentioned and no sale of such securities will be made in the United States, any state or province in which such offer, solicitation or sale would be unlawful until the securities are registered or their distribution is permitted under the securities laws of that state or province.

For additional information, please contact:

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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 The Nasdaq Global Select Market (ORPH) and its shares are listed on Nasdaq Copenhagen (ORPHA).



Forward-looking statements

This company announcement may contain certain forward-looking statements under the U.S. Private Securities Litigation Reform Act of 1995 and otherwise, including as to any potential sales under the ATM Program and the application of net proceeds therefrom. 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 November 4, 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 future.

Disclaimer

This announcement does not, and shall not, in any circumstances constitute a public offering nor an invitation to solicit the interest of the public in Denmark, the United States or in any other jurisdiction, in connection with any offer.

The distribution of this document may, in certain jurisdictions, be restricted by local legislation. Persons into whose possession this document comes are required to inform themselves about and to observe any such potential local restrictions.

This announcement is not an advertisement and not a prospectus within the meaning of Regulation (EU) 2017/1129 of the European Parliament and of the Council of 14 June 2017 (the "Prospectus Regulation").

With respect to the member States of the European Economic Area, including Denmark no action has been undertaken or will be undertaken to make an offer to the public of the securities referred to herein requiring a publication of a prospectus in any relevant member State. As a result, the securities may not and will not be offered in any relevant member State except in accordance with the exemptions set forth in Article 1(4) of the Prospectus Regulation or under any other circumstances which do not require the publication by the Company of a prospectus pursuant to Article 3 of the Prospectus Regulation and/or to applicable regulations of that relevant member State.

This announcement is only being distributed to and is only directed at (i) persons who are outside the United Kingdom or (ii) within the United Kingdom, to "qualified investors" (as defined in the UK Prospectus Regulation) who are (a) investment professionals falling within Article 19(5) of the UK Financial Services and Markets Act 2000 (Financial Promotion) Order 2005 (the "Order") or (b) high net worth entities falling within Article 49(2)(a) – (d) of the Order (the persons described in (i) and (ii) 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. The "UK Prospectus Regulation" means Regulation (EU) 2017/1129 as it forms part of domestic law in the United Kingdom by virtue of the European Union (Withdrawal) Act 2018.