

Copenhagen, Denmark, February 7, 2020 – Orphazyme A/S (ORPHA.CO), a biopharmaceutical company pioneering Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases, today announces the completion of an offering of 7,032,937 new shares and existing shares in a directed issue and private placement raising gross proceeds of approximately DKK 745,000,000.

Kim Stratton, Chief Executive Officer, said: “I would like to thank our investors, both existing and new, for their confidence in Orphazyme as the company enters a very exciting phase in its development. This funding provides a significant cash runway for Orphazyme as we progress our lead product arimoclomol towards market, and is expected to cover our ambitious clinical development plans well into 2021. In particular, the financing will support our imminent US and European filings of arimoclomol for Niemann-Pick disease Type C (NPC) and preparations for commercial launch.”

Orphazyme also intends to use the proceeds to cover completion of arimoclomol’s clinical development in sporadic Inclusion Body Myositis (sIBM) and Amyotrophic Lateral Sclerosis (ALS) and to prepare regulatory filings in Europe and potentially the US in these indications, as well as for general corporate purposes.

Based on the positive data package announced in 2019, Orphazyme currently expects to submit an application for approval of arimoclomol for the treatment of NPC in H1 2020 in the US and H2 2020 in the EU. Orphazyme expects to release results from the ongoing phase 2 trial for Gaucher Disease in H1 2020 and continue its ongoing phase 3 trial in ALS and phase 2/3 trial in sIBM. Orphazyme has been granted Fast Track and Breakthrough Therapy designation from the US Food and Drug Administration (FDA) for the development of arimoclomol in NPC, and Fast Track designation for the development of arimoclomol in sIBM.

The directed issue and private placement of 3,961,264 new shares of a nominal value of DKK 1 each (the “New Shares”) and private placement of 3,071,673 existing shares of a nominal value of DKK 1 each (the “Existing Shares”) and together with the New Shares, the “Offer Shares”) (the “Offering”) in Orphazyme A/S (the “Company”) has been successfully executed. Reference is made to company announcement no. 08/2020.

The Offer Shares were subscribed for and sold with significant demand from both domestic and international investors in the EU and the US.

The offering of Existing Shares was facilitated by a share loan from Novo Holdings A/S and Orpha Pooling B.V. (the “Lending Shareholders”) to the Company pursuant to a stock lending and subscription agreement with an obligation for the Company to redeliver new shares of an equivalent number as the Existing Shares borrowed by the Company from each of the Lending Shareholders (the “Replacement Shares”). The Lending Shareholders do not participate in the Offering, and are only facilitating the loan of the Lending Shares for purposes of the Company’s offering of Existing Shares in the Offering.

The investors have subscribed for or purchased 3,961,264 New Shares and 3,071,673 Existing Shares at an offer price of DKK 106 per Offer Share, as determined by the Board of Directors through a book-building process and accordingly 3,071,673 Replacement Shares will be subscribed for at an offer price of DKK 106 per Replacement Share, raising aggregate gross proceeds for Orphazyme of approximately DKK 745,000,000 (EUR 100 million/USD 110 million).

As the Offering was oversubscribed by Danish and international institutional and professional investors, the shares have been allocated on an individual basis.

Capital increases

Subject to settlement of the capital increases, the share capital of the Company will consist of 27,038,386 shares with a nominal value of DKK 1 each.

The nominal value of the New Shares and Replacement Shares represents approximately 35.16% of the Company’s registered share capital of DKK 20,005,449 before the capital increases and will account for approximately 26.01% of the Company’s registered share capital of DKK 27,038,386 upon completion of the capital increases.

Expected timetable

Date	Event
February 11, 2020	Settlement and payment for the Offer Shares and Replacement Shares

February 11, 2020 Registration of the capital increases with the Danish Business Authority
February 12, 2020 New Shares to be admitted to trading and official listing on Nasdaq Copenhagen A/S

The New Shares will be issued in the temporary ISIN code DK0061274289, the Existing Shares will be purchased in the permanent ISIN code DK0060910917, and the Replacement Shares for the Lending Shareholders will be issued in the temporary ISIN code DK0061274362.

Settlement for investors, who have subscribed for New Shares will take place in the temporary ISIN code DK0061274289, settlement for investors, who have purchased Existing Shares will take place in the permanent ISIN code DK0060910917, and settlement of the Replacement Shares will take place in the temporary ISIN code DK0061274362.

The admission to trading and official listing of the New Shares and the later admission to trading and official listing of the Replacement Shares on Nasdaq Copenhagen A/S is subject to the Offering not being withdrawn prior to the settlement of the Offering and the Company making an announcement to that effect, and in respect of the Replacement Shares only (which will only be delivered to the Lending Shareholders and not be sold as part of the Offering), the publication of a listing prospectus to be approved by the Danish Financial Supervisory Authority, which is expected to occur no later than by the end of March 2020.