

Company announcement

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Orphazyme publishes offering circular and the indicative offer price range for its IPO

Orphazyme, a Danish biotech company with a late stage orphan drug pipeline, today published an offering circular and the indicative price range in connection with its intended initial public offering ("IPO" or the "Offering") and subsequent admission to trading and official listing of its shares on Nasdaq Copenhagen.

The Offering in brief

- Indicative offer price range of DKK 64 to DKK 80 per share of nominal value DKK 1 each
- Implied market capitalisation of DKK 1.5bn to DKK 1.6bn after the issuance of new shares in the Offering (excluding potential exercise of the over-allotment option also consisting of new shares and assuming full exercise of pre-IPO warrants)
- The Offering comprises:
 - Issue of new shares with gross proceeds of up to DKK 690 million (hereof DKK 90 million pursuant to the over-allotment option)
 - Up to 10,781,250 new shares to be issued by the Company (assuming full exercise of the over-allotment option)
 - An over-allotment option of up to 1,406,250 new shares, which would generate up to DKK 90 million in gross proceeds if exercised in full. The over-allotment option may be exercised in whole or in part during a period of 30 calendar days after the first day of trading and official listing on Nasdaq Copenhagen
- SEB, Vækstfonden, BankInvest on behalf of certain clients, Handelsbanken and Spar Nord have committed, subject to certain conditions, to subscribe for a number of shares

in the Offering corresponding to DKK 230 million – equivalent to 38.3% of the Offering (excluding the overallotment option). A corresponding number of shares in the Offering will be reserved for these investors.

- The offer period starts 6 November 2017 and ends on 16 November at 12 a.m. (noon) CET, but may be closed in whole or in part at the earliest on 15 November 2017 at 00:01 a.m. CET
- Offer price to be announced and first day of trading and official listing on Nasdaq Copenhagen is expected on 17 November 2017 but may be moved forward accordingly if the offer period is closed before 16 November 2017
- The expected settlement date of the Offering is 21 November 2017, but may be moved forward accordingly if the offer period is closed before 16 November 2017

Orphazyme is currently owned by Novo Holdings A/S (33.1%), Coöperative Aescap Venture I U.A. (15.2%), Sunstone Life Science Ventures Fund II K/S (15.6%), Orpha Pooling B.V. (a joint venture between LSP V Coöperatieve U.A. and ALS Invest 2 B.V.) (21.0%), certain funds managed by Idinvest (7.2%) and Kurma Biofund II (6.6%) and other minority shareholders including members of senior management (1.4%).¹ None of the current larger shareholders of Orphazyme have expressed an intention to sell shares in connection with the IPO and will continue as shareholders in Orphazyme in addition to undertaking lock-up obligations in connection with the IPO. For a further description of the Offering, please refer to the offering circular published by Orphazyme on 6 November 2017.

Bo Jesper Hansen, Deputy Chairman, said:

“Orphazyme has undergone a significant transformation in the past years and now has a late stage orphan drug pipeline. We expect to have completed three phase II/III trials by the end of 2020 with the first potential marketing authorisation in 2020. We believe that Orphazyme will benefit from an enhanced public profile and look forward to support the Company going forward.”

Anders Hinsby, CEO of Orphazyme, said:

“We are excited about the broad investor interest we have received since the announcement of our intention to float and we are proud that a group of leading Danish institutional investors have decided to support our IPO. Today’s IPO launch is a key milestone for us and we look forward to welcome new shareholders in Orphazyme.”

Background and reasons for the Offering

The Offering is expected to support Orphazyme’s operational strategy, advance Orphazyme’s public and commercial profile, and provide Orphazyme with improved access to public capital markets and a diversified base of new Danish and international shareholders.

Net proceeds from the Offering together with Orphazyme’s current cash resources will be used to primarily fund four clinical trials. Orphazyme expects to have completed three potential registration trials by the end of 2020.

¹ The ownership stakes are stated as of the date hereof, i.e. before the completion of the Offering. If the offer price is set below the top end of the offer price range, a number of bonus shares will be issued in favour of the former preference shareholders of Orphazyme, as part of the 2017 Capital Structure Adjustment (as further described in the offering circular). Consequently, the ownership stakes set out above may be adjusted accordingly.

Other information about the Offering

The Offering consists of an initial public offering to retail and institutional investors in Denmark and private placements to institutional investors in certain other jurisdictions (excluding the United States). The Offering outside the United States will be made in compliance with Regulation S under the U.S. Securities Act of 1933, as amended.

Up to 93,750 offer shares will be reserved for certain members of the Board of Directors to subscribe for in connection with the Offering at the offer price. Up to 21,875 of the offer shares will be reserved for the participants in Orphazyme's long-term incentive programme to subscribe for at the offer price as an investment in connection with the Offering. Up to 93,750 offer shares will be reserved for Orphazyme's employees to subscribe for in connection with the Offering at the offer price.

The offer price will be determined through a book-building process and is expected to be announced, together with the final number of offer shares sold in the Offering through Nasdaq Copenhagen no later than 8:00 a.m. CET on 17 November 2017.

Payment for and settlement of the offer shares are expected to take place on or around 21 November 2017 by way of delivery of temporary purchase certificates under the temporary ISIN DK0060911055 against payment in immediately available funds in Danish kroner in book-entry form to investors' accounts with VP SECURITIES A/S ("VP Securities") and through the facilities of Euroclear Bank S.A./N.A., as operator of the Euroclear System ("Euroclear") and Clearstream Banking, S.A. ("Clearstream"). Subject to completion of the Offering and registration of the new offer shares with the Danish Business Authority, the temporary purchase certificates will automatically be exchanged in VP Securities for a corresponding number of shares in Orphazyme, which are expected to be delivered two business days after settlement under the permanent ISIN DK0060910917 in book-entry form to the holder of the temporary purchase certificates' account with VP Securities and through the facilities of Euroclear and Clearstream.

Prior to the Offering, there has been no public market for the temporary purchase certificates or Orphazyme's shares. Application has been made for the temporary purchase certificates to be admitted to trading on Nasdaq Copenhagen (the "Admission") under the symbol "ORPHA TEMP" and for the shares to be admitted to trading and official listing on Nasdaq Copenhagen under the symbol "ORPHA".

The admission to trading and official listing is subject to, among other things, Nasdaq Copenhagen approving the distribution of shares, the Offering not being withdrawn prior to settlement thereof and Orphazyme making an announcement to that effect.

The first day of trading and official listing on Nasdaq Copenhagen is expected to be 17 November 2017 subject to the Offering not being withdrawn prior to settlement and completion of the Offering or accelerated. The first day of trading of the temporary purchase certificates on Nasdaq Copenhagen is expected to be 17 November 2017 and the last day of trading of the temporary purchase certificates on Nasdaq Copenhagen is expected to be 21 September 2017. The first day of trading of the shares on Nasdaq Copenhagen under the permanent ISIN is expected to be 22 November 2017. In connection with the temporary purchase certificates being automatically exchanged for shares, the temporary purchase certificates will cease to exist.

If the Offering is terminated or withdrawn, the Offering and any associated arrangements will lapse, all submitted orders will be automatically cancelled, any monies received in respect of the Offering will be returned to the investors without interest (less any transaction costs) and the Admission on Nasdaq Copenhagen will be cancelled. Consequently, any trades in the temporary purchase certificates or the shares effected on or off the market before settlement of the Offering may subject investors to liability for not being able to deliver the temporary purchase certificates and/or the shares sold and investors who have sold or acquired temporary purchase certificates and/or shares on or off the market may incur a loss. All dealings in the temporary purchase certificates and/or offer shares prior to settlement of the Offering are for the account of, and at the sole risk of, the parties concerned.

A number of pre-IPO warrants may be exercised (or lapse if not exercised) in connection with the Offering. The maximum nominal capital increase resulting from the exercise of such warrants is limited to DKK 866,965 and the capital increase thereto is expected to take place in connection with settlement of the Offering, i.e. on or around 21 November 2017.

New Board of Directors

Anders Hedegaard and Catherine Moukheibir are expected to be elected as new members of the Board of Directors of Orphazyme prior to admission to trading and official listing on Nasdaq Copenhagen.

Anders Hedegaard is currently chief executive officer of GN Store Nord A/S and GN Hearing A/S and was previously CEO of Bavarian Nordic A/S. Anders Hedegaard holds a Master of Science in Chemical Engineering and Biochemistry from the Technical University of Denmark.

Catherine Moukheibir is currently chairman of the board of directors of MedDay Pharmaceuticals SA, member of the board of directors of Zealand Pharma A/S (publ) and Genkyotex SA (publ), as well as a member of the board of directors of Ablynx NV (publ) and Cerenis Therapeutics SA (publ). Catherine Moukheibir holds a Master in Economics and an MBA degree, both from Yale University.

Following the expected election of the new board members, the Board of Directors is expected to comprise Georges Gemayel (Chairman), Bo Jesper Hansen (Deputy Chairman), Anders Hedegaard, Catherine Moukheibir, Martijn Kleijwegt, Martin Bonde, Rémi Droller and Sten Verland.

Offering circular

The offering circular has been prepared in English (including a Danish translation of the summary). Special attention should be given to the risk factors described in the offering circular.

The offering circular will be made available to eligible investors at no cost at the registered office of Orphazyme, Ole Maaløes Vej 3, DK-2200 Copenhagen N, or on Orphazyme's website, www.orphazyme.com.

The offering circular can also be obtained upon request to Carnegie Investment Bank, filial af Carnegie Investment Bank AB (publ), Sverige, e-mail: prospekter@carnegie.dk or Danske Bank A/S, e-mail: prospekter@danskebank.dk.

About Orphazyme

Orphazyme is a Danish biotech company with a late stage orphan drug pipeline, developing new treatment options for orphan protein misfolding diseases. The Company was founded in 2009 based on early scientific discovery in heat shock proteins ("HSPs"). Since inception, the Company has translated scientific discovery into a late stage clinical development programme. The Company is headquartered in Copenhagen and currently has 30 employees.

The Company focuses on severe and mostly fatal diseases with a high unmet need, and with a particularly strong commitment to neuromuscular diseases and a group of severe genetic diseases called lysosomal storage diseases. The Company plans to pursue development of its lead candidate through to registration in the EU and the United States after which launch and commercialisation is expected to be undertaken by the Company.

The lead candidate arimoclomol is in development as a potential treatment for four orphan diseases; two neuromuscular diseases, sporadic Inclusion Body Myositis ("sIBM") and Amyotrophic Lateral Sclerosis ("ALS"), and two lysosomal storage diseases, Niemann Pick type C ("NPC") and Gaucher disease.

Bank syndicate

Carnegie and Danske Bank are acting as Joint Global Coordinators and Joint Bookrunners, and Oddo BHF SCA is acting as Co-Lead Manager for the proposed IPO.

Press conference

A press conference will be held today, 6 November, at Gorrissen Federspiel Advokatpartnerselskab, Axeltorv 2, 1609 Copenhagen V at 11:00-12:00 CET. At the press conference, Deputy Chairman Bo Jesper Hansen, CEO Anders Hinsby and CFO Anders Vadsholt will present the Offering, followed by a Q&A session and individual interviews. The press conference will be held in Danish.

For additional information, please contact**Orphazyme**

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This announcement is only being distributed to and is only directed at (i) persons who are outside the United Kingdom or (ii) to investment professionals falling within Article 19(5) of the U.K. Financial Services and Markets Act 2000 (Financial Promotion) Order 2005 (the “Order”) or (iii) high net worth entities falling within Article 49(2)(a) – (d) of the Order (the persons described in (i) through (iii) 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 document or any of its contents.

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Carnegie Investment Bank, filial af Carnegie Investment Bank AB (publ), Sverige and Danske Bank A/S (together the “Joint Global Coordinators and Joint Bookrunners”) and Oddo BHF SCA (“Co-Lead Manager” and together with the Joint Global Coordinators and Joint Bookrunners, the “Managers”) and their affiliates are acting exclusively for the Company and no-one else in connection with the potential offering. They will not regard any other person as their respective clients in relation to the potential offering and will not be responsible to anyone other than the Company for providing the protections afforded to their respective clients, nor for providing advice in relation to the potential offering, the contents of this announcement or any transaction, arrangement or other matter referred to herein.

In connection with the potential offering, the Managers and any of their affiliates, acting as investors for their own accounts, may subscribe for or purchase shares and in that capacity may retain, purchase, sell, offer to sell or otherwise deal for their own accounts in such shares and other securities of the Company or related instruments in connection with the potential offering or otherwise. Accordingly, references in the offering memorandum, if published to the shares being issued, offered, subscribed, acquired, placed or otherwise dealt in should be read as including any issue or offer to, or subscription, acquisition, placing or dealing by, such Managers and any of their affiliates acting as investors for their own accounts. The Managers do not intend to disclose the extent of any such investment or transactions otherwise than in accordance with any legal or regulatory obligations to do so.

In connection with the offering, Danske Bank A/S (the “Stabilising Manager”) (or persons acting on behalf of the Stabilising Manager) may over-allot securities or effect transactions with a view to supporting the market price of the securities at a level higher than that which might otherwise prevail. However, there is no assurance that the Stabilising Manager (or persons acting on behalf of the Stabilising Manager) will undertake stabilisation. Any stabilisation action may begin on or after the date of commencement of trading and official listing of the securities on Nasdaq Copenhagen A/S and, if begun, may be ended at any time, but it must end no later than 30 days after the date of commencement of trading and official listing of the securities.

Matters discussed in this announcement may constitute forward-looking statements. Forward-looking statements are statements that are not historical facts and that can be identified by words such as “believe”, “expect”, “anticipate”, “intends”, “estimate”, “will”, “may”, “continue”, “should”, and similar expressions. The forward-looking statements in this announcement are based upon various assumptions, many of which are based, in turn, upon further assumptions. Although the Company believes that these assumptions were reasonable when made, these assumptions are inherently subject to significant known and unknown risks, uncertainties, contingencies and other important factors that are difficult or impossible to predict and are beyond its control. Such risks, uncertainties, contingencies and other important factors could cause actual events to differ materially from the expectations expressed or implied in this release by such forward-looking statements. The information, opinions and forward-looking statements contained in this announcement speak only as at its date, and are subject to change without notice.