

Notice to convene Extraordinary General Meeting in Orphazyme A/S

Copenhagen, Denmark, January 3, 2020 – Orphazyme A/S (ORPHA.CO), a biopharmaceutical company pioneering Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases, today announces:

To the shareholders of Orphazyme A/S

The Board of Directors hereby convenes an Extraordinary General Meeting of Orphazyme A/S, CVR no. 32 26 63 55 (the "Company"), to be held on Saturday, January 25, 2020 at 2:00 PM (CET), at the Company's address Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark.

Purpose

The purpose of the Extraordinary General Meeting is to provide the Board of Directors of the Company with authorizations to raise new capital to fund the next important journey and value creation process in Orphazyme, based on the strong data arimoclomol already made available to the market.

Orphazyme expects to receive approval for NPC in H2 2020 in U.S. through the fast track approval process, with the aim to bring the product to patients shortly thereafter. Also, in 2020, Orphazyme will release results from the ongoing Phase II trial for Gaucher Disease. In addition to these significant events during next year, the Company will continue its ongoing Phase III trial in ALS and Phase II/III trial in sIBM.

Agenda

1. Authorizations to the Board of Directors to increase the share capital of the Company
 - a. Amendment to Article 3.4 of the Articles of Association regarding authorization to the Board of Directors to increase the share capital without pre-emption rights for existing shareholders
 - b. Authorization to the Board of Directors to increase the share capital with pre-emption rights for existing shareholders
2. Authorization to the Board of Directors to acquire treasury shares
3. Authorization of the Chairman of the meeting

Items on the agenda, including complete proposals:

Item 1: Authorizations to the Board of Directors to increase the share capital of the Company

- a. **Amendment to Article 3.4 of the Articles of Association regarding authorization to the Board of Directors to increase the share capital without pre-emption rights for existing shareholders**

The Board of Directors proposes that the current Article 3.4 of the Articles of Association be amended as follows:

"In the period until 25 January 2025, the Board of Directors is authorised to increase the Company's share capital in one or more issues of new shares without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 8,000,000. The capital increase shall take place at market price as determined by the Board of Directors through a book-building process and shall be effected by cash payment."

b. Authorization to the Board of Directors to increase the share capital with pre-emption rights for existing shareholders

The Board of Directors proposes to adopt a new Article 3.7 in the Articles of Association with the following wording:

"In the period until 25 January 2025, the Board of Directors is authorised to increase the Company's share capital through one or more issues of new shares with pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 25,000,000. The capital increase may be effected by cash payment or conversion of debt and shall take place at subscription price as determined by the Board of Directors which may be below the market price."

As a consequence of the above, the current Article 3.7 will then be Article 3.8 of the Articles of Association and be amended as follows:

"New shares issued pursuant to Articles 3.1, 3.2, 3.3, 3.4, 3.5, 3.6 and 3.7 shall be fully paid up, issued in the name of the holder and shall be recorded in the name of the holder in the Company's register of shareholders, shall be negotiable instruments and shall in every respect carry the same rights as the existing shares. The Board of Directors is authorised to lay down the terms and conditions for capital increases pursuant to the above authorisations and to make any such amendments to the Company's Articles of Association as may be required as a result of the Board of Directors' exercise of said authorisations."

Item 2: Authorization to the Board of Directors to acquire treasury shares

The Board of Directors proposes to increase and extend the current authorization to the Board of Directors in the period until 25 January 2025 to approve the acquisition of treasury shares, on one or more occasions, with a total nominal value of up to 20% of the share capital of the Company from time to time, subject to the Company's holding of treasury shares after such acquisition does not exceed 20% of the Company's share capital. The consideration may not deviate more than 10% from the official price quoted on Nasdaq Copenhagen at the time of acquisition.

Item 3: Authorization of the Chairman of the meeting

The Board of Directors proposes that the Chairman of the Extraordinary General Meeting is, with a right of substitution, authorized to file the resolutions passed by the general meeting with the Danish Business Authority and to make any such changes and additions as the Danish Business Authority may require as a condition for registering or approving the resolutions passed by the general meeting.

Adoption requirements

The proposal under item 1.a-1.b on the agenda shall be passed by at least two-thirds of the votes cast as well as at least two-thirds of the share capital represented at the Extraordinary General Meeting, cf. Article 7.2 of the Articles of Association.

The proposal under item 2 and 3 on the agenda shall be passed by a simple majority of votes cast, cf. Article 7.1 of the Articles of Association.

Share capital and shareholders' participation and voting rights

The Company's nominal share capital is DKK 19,984,799, divided into shares of DKK 1 each or multiples thereof. Each share of the nominal value of DKK 1 carries one vote.

The right of a shareholder to attend and vote at a general meeting is determined by the shares held by the shareholder at the record date. The record date is **Saturday, January 18, 2020**.

The shares held by each shareholder at the record date are calculated based on the registration of the number of shares held by the shareholder in the Company's register of shareholders as well as any notification of ownership received by the Company for the purpose of registration in the Company's register of shareholders, but which has not yet been registered.

How to obtain an admission card

Shareholders, who are entitled to attend the Extraordinary General Meeting and wish to attend the Extraordinary General Meeting, must request an admission card no later than on **Tuesday, January 21, 2020**. The registration form is available on the Company's website, <https://orphazyme.gcs-web.com/annual-general-meeting-0>.

Admission cards may be requested as follows:

- Electronically via VP Securities A/S' website, www.vp.dk/gf; or
- By completing and forwarding the registration form by letter to VP Securities A/S, Weidekampsgade 14, DK-2300 Copenhagen S; or
- By emailing a scanned version to ypinvestor@vp.dk; or
- By contacting VP Securities A/S by phone on +45 4358 8866.

The Company will distribute electronic admission cards by email to shareholders. Please register the email address to which the electronic admission card should be sent as follows:

1. Go to the Company's website, <https://orphazyme.gcs-web.com/annual-general-meeting-0> and click on the link to the Company's Investor Portal;
2. Log on to the Investor Portal with your NemID or VPID; and
3. Enter your email address under "My Profile" and click "Save preferences".

The electronic admission card must be presented at the Extraordinary General Meeting either via smartphone/tablet or as a printed version of the electronic admission card. Further, it will be possible for the shareholder to collect the admission card at the venue for the Extraordinary General Meeting upon presentation of appropriate identification. Ballot papers will be handed out at the registration desk at the venue.

How to submit a proxy

Subject to having requested an admission card, shareholders may attend the Extraordinary General Meeting in person or by proxy. Proxy forms must be received by VP Securities A/S no later than on **Tuesday, January 21, 2020**. The proxy form is available on the Company's website, <https://orphazyme.gcs-web.com/annual-general-meeting-0>.

Proxies may be submitted as follows:

- Electronically via VP Securities A/S' website, www.vp.dk/gf; or
- By completing and forwarding the registration form by letter to VP Securities A/S, Weidekampsgade 14, DK-2300 Copenhagen S; or
- By emailing a scanned version to ypinvestor@vp.dk.

How to vote by correspondence

Shareholders who are entitled to participate in the Extraordinary General Meeting are also entitled to vote by correspondence. Votes by correspondence must be received by VP Securities A/S no later than on **Friday, January 24, 2020, at 3:00 PM (CET)**. The voting form (which is the same form used for granting a proxy) is available on the Company's website, <https://orphazyme.gcs-web.com/annual-general-meeting-0>.

Voting by correspondence may be submitted as follows:

- Electronically via VP Securities A/S' website, www.vp.dk/gf; or
- By completing and forwarding the registration form by letter to VP Securities A/S, Weidekampsgade 14, DK-2300 Copenhagen S; or
- By emailing a scanned version to ypinvestor@vp.dk.

Additional information on the website

Until and including the day of the Extraordinary General Meeting, the following information regarding the Extraordinary General Meeting will be available on the Company's website, <https://orphazyme.gcs-web.com/annual-general-meeting-0>:

- The notice convening the Extraordinary General Meeting, including the agenda and the complete proposals
- The proposed revised Articles of Association
- The aggregate number of shares and voting rights as of the date of the notice to convene the Extraordinary General Meeting
- Other documents for the use of the Extraordinary General Meeting, including the proxy/voting by correspondence form and the registration form

Registration of admission cards at the Extraordinary General Meeting on Saturday, January 25, 2020, will open at 1:00 PM (CET).

Questions from the shareholders

Shareholders may ask questions to the Board of Directors and the Executive Management at the Extraordinary General Meeting. Questions regarding the agenda and documents concerning the Extraordinary General Meeting may be submitted in writing prior to the Extraordinary General Meeting by ordinary mail to Orphazyme A/S, Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark, or by email to contact@orphazyme.com.

Personal data

With regards to collection and processing of personal data, reference is made to the Company's information sheet on data protection in connection with the Extraordinary General Meeting, which is available on the Company's website, <https://orphazyme.qcs-web.com/annual-general-meeting-0>.

Language

The Extraordinary General Meeting will be held in English in accordance with Article 4.6 of the Company's Articles of Association.

How to get there

The Extraordinary General Meeting will be held at the Company's address Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark, which can be reached by car or public transport.

Copenhagen N, January 3, 2020

The Board of Directors

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

Anders Vadsholt, CFO

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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.

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.