

To the shareholders of Orphazyme A/S

The Board of Directors hereby convenes the Annual General Meeting of Orphazyme A/S, CVR no. 32 26 63 55 (the "Company"), to be held on Thursday, March 26, 2020 at 5:00 PM (CET), at the Company's address Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark.

Agenda

1. The Board of Directors' election of the Chairman of the meeting
2. The Board of Directors' report on the Company's activities in the past financial year
3. Presentation and adoption of the annual report 2019
4. Distribution of profit or covering of loss according to the adopted annual report 2019
5. Resolution to grant discharge of liability to the Board of Directors and the Executive Management
6. Approval of remuneration of the Board of Directors for the current financial year
7. Election of members to the Board of Directors
8. Election of auditor
9. Authorization to acquire treasury shares
10. Any proposals from the Board of Directors
 - a. Authorization to the Board of Directors to increase the share capital without pre-emption rights for existing shareholders
 - b. Amendments to Article 3 of the Articles of Association
 - c. Amendment to Article 5.1 of the Articles of Association
 - d. Amendments to Articles 2.4 and 6.2 of the Articles of Association
 - e. Approval of revised Remuneration Policy
11. Any other business

Items on the agenda, including complete proposals:**Item 1: The Board of Directors' election of the Chairman of the meeting**

The Board of Directors will elect the Chairman of the Annual General Meeting.

Item 2: The Board of Directors' report on the Company's activities in the past financial year

The Board of Directors proposes that the report on the Company's activities in 2019 be noted by the Annual General Meeting.

Item 3: Presentation and adoption of the annual report 2019

The Board of Directors proposes that the Company's annual report 2019 be adopted.

Item 4: Distribution of profit or covering of loss according to the adopted annual report 2019

The Board of Directors proposes that the loss related to the financial year 2019 amounting to approximately DKK 337.5 million be carried forward.

Item 5: Resolution to grant discharge of liability to the Board of Directors and the Executive Management

The Board of Directors proposes that the Annual General Meeting grant discharge of liability to the

members of the Board of Directors and the Executive Management for liability towards the Company in relation to the approved annual report 2019.

Item 6: Approval of remuneration of the Board of Directors for the current financial year

The Board of Directors proposes that the following remuneration level to the Board of Directors for 2020 be approved by the Annual General Meeting:

Members of the Board of Directors shall receive an annual base fee of: EUR 31,500

The Chairman of the Board of Directors shall in addition receive: EUR 24,900

The Deputy Chairman of the Board of Directors shall in addition receive: EUR 12,450

The remuneration level for the Board of Directors is maintained at the same level as for 2019.

The members of the Board of Directors receive annual supplementary fees for their work in the Company's Audit Committee, Remuneration Committee, and Nomination Committee. The Board of Directors proposes that the Annual General Meeting approve the following supplementary fees for 2020 for committee work:

Members of the Audit Committee shall receive an annual supplementary fee of: EUR 6,700

Members of the Remuneration Committee shall receive an annual supplementary fee of: EUR 4,700

Members of the Nomination Committee shall receive an annual supplementary fee of: EUR 3,100

The Chairman of the Audit Committee shall receive an annual supplementary fee of: EUR 13,500

The Chairman of the Remuneration Committee shall receive an annual supplementary fee of: EUR 9,000

The Chairman of the Nomination Committee shall receive an annual supplementary fee of: EUR 6,500

The supplementary fees for committee work are maintained at the same level as for 2019.

In addition, members of the Board of Directors may receive a fixed travel allowance as determined by the Board of Directors and be reimbursed for expenses in connection with board meetings (e.g. related to travel and accommodation). Further, the Company may pay or reimburse social security duties and similar taxes imposed by public authorities in relation to board fees.

Further, the members of the Board of Directors will receive share-based remuneration for 2020 in the form of restricted share units through an annual grant corresponding to 50% of each Board member's fixed annual base fee, such base fee to include additional base fees to the Chairman and Deputy Chairman but excluding any additional fees for committee membership.

Item 7: Election of members to the Board of Directors

Pursuant to Article 8.1 of the Articles of Association, the Board of Directors shall consist of not less than six and not more than nine members elected by the general meeting. The members of the Board of Directors elected by the general meeting are elected for a term of one year pursuant to Article 8.2 of the Articles of Association. Re-election of board members may take place.

The Board of Directors proposes re-election for a one-year term of the following members of the Board of Directors elected by the general meeting: Georges Gemayel, Bo Jesper Hansen, Anders Hedegaard, Catherine Moukheibir, Martijn Kleijwegt, Martin Bonde, Rémi Droller, and Sten Verland.

Please see **Appendix 1** for a description of the nominated candidates' qualifications and positions, including information about other executive/management functions held by the candidates in Danish and foreign companies.

Item 8: Election of auditor

In accordance with the Audit Committee's recommendation the Board of Directors proposes re-election of ERNST & YOUNG Godkendt Revisionspartnerselskab, CVR no. 30 70 02 28, as the Company's auditor. The Audit Committee's recommendation has not been influenced by third parties, nor has it been subject to any contractual obligation restricting the Annual General Meeting's choice of certain auditors or audit firms.

Item 9: Authorization to acquire treasury shares

With reference to company announcement no. 11/2020, the Board of Directors has partly exercised its authorization to acquire treasury shares granted on the Extraordinary General Meeting of the Company held on January 25, 2020.

In order to continuously provide the Company with the necessary flexibility to raise new capital and to facilitate the possibility for a transaction structure similar to the one announced in February 2020, the Board of Directors proposes to replace the current authorization to the Board of Directors with a new authorization to approve the acquisition of treasury shares in the period until March 26, 2025, 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 10a: Authorization to the Board of Directors to increase the share capital without pre-emption rights for existing shareholders

By decisions of February 6, 2020, the Board of Directors has partly exercised the authorization in Article 3.4 to increase the Company's share capital without pre-emption rights for existing shareholders following which a nominal value of DKK 7,032,937 of the authorization has been issued. The Board of Directors proposes that Articles 3.4 and 3.4.1 be deleted from the Articles of Association.

In order to continuously provide the Company with the necessary flexibility to raise new capital and to facilitate the possibility for a transaction structure similar to the transaction announced in February 2020, the Board of Directors further proposes that a new Article 3.4 be included in the Articles of Association with the following wording:

"In the period until 26 March 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 10,815,000. 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."

Item 10b: Amendments to Article 3 of the Articles of Association

The authorizations in Articles 3.1 – 3.3 to the Board of Directors to increase the share capital of the Company have expired. For this reason, the Board of Directors proposes that Articles 3.1 – 3.3 be deleted from the Articles of Association.

As a consequence of the above, the current Articles 3.4 – 3.8 will be renumbered to Articles 3.1 – 3.5 and the new Article 3.5 will be amended as follows:

"New shares issued pursuant to Articles 3.1, 3.2, 3.3 and 3.4 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 10c: Amendment to Article 5.1 of the Articles of Association

The Board of Directors proposes that Article 5.1 be amended as follows:

"The agenda for the annual general meeting shall include the following:

a. The Board of Directors' report on the Company's activities in the past financial year

- b. Presentation and adoption of the annual report
- c. Distribution of profit or covering of loss according to the adopted annual report
- d. Resolution to grant discharge of liability to the Board of Directors and the Executive Management
- e. Presentation of the remuneration report
- f. Approval of remuneration of the Board of Directors for the current financial year
- g. Election of members to the Board of Directors
- h. Election of auditor
- i. Authorisation to acquire treasury shares
- j. Any proposals from the Board of Directors or shareholders
- k. Any other business"

Item 10d: Amendments to Articles 2.4 and 6.2 of the Articles of Association

To reflect that the keeper of the Company's register of shareholders will be changed to Computershare A/S, CVR no. 27 08 88 99, the Board of Directors proposes that the current Article 2.4 of the Articles of Association be amended as follows:

"The register of shareholders is kept by Computershare A/S, CVR no. 27 08 88 99."

Further, to allow attendance at general meetings based on a simple notification rather than a request and receipt of an admission card, the Board of Directors proposes that the current Article 6.2 be amended as follows:

"A shareholder who is entitled to attend the general meeting pursuant to Article 6.1 and who wants to attend the general meeting shall notify the Company of his/her attendance not later than three days prior to the date of the general meeting."

Item 10e: Approval of revised Remuneration Policy

The Board of Directors proposes that the revised Remuneration Policy for the Company as adopted by the Board of Directors be approved by the Annual General Meeting.

The Remuneration Policy has been revised in order to ensure compliance with the new requirements under sections 139 and 139a of the Danish Companies Act.

The revised Remuneration Policy is attached as **Appendix 2** and is available on the Company's website, <https://orphazyme.qcs-web.com/annual-general-meeting-0>.

If the revised Remuneration Policy is approved, Article 11 of the Company's Articles of Association regarding overall guidelines on incentive pay will be deleted.

Adoption requirements

The proposals under item 10.a – 10.d 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.

All other proposed resolutions 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 27,038,386, 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 **Thursday, March 19, 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 Annual General Meeting and wish to attend the Annual General Meeting, must request an admission card no later than on **Friday, March 20, 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 vpinvestor@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 Annual 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 Annual 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

Proxy forms must be received by VP Securities A/S no later than on **Friday, March 20, 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 proxy form by letter to VP Securities A/S, Weidekampsgade 14, DK-2300 Copenhagen S; or
- By emailing a scanned version to vpinvestor@vp.dk.

How to vote by correspondence

Shareholders who are entitled to participate in the Annual General Meeting are also entitled to vote by correspondence. Votes by correspondence must be received by VP Securities A/S no later than on **Wednesday, March 25, 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 vpinvestor@vp.dk.

Additional information on the website

Until and including the day of the Annual General Meeting, the following information regarding the Annual General Meeting will be available on the Company's website, www.orphazyme.com:

- The notice convening the Annual General Meeting, including the agenda and the complete proposals as well as Appendix 1: Candidates for the Board of Directors and Appendix 2: Revised Remuneration Policy;

- The aggregated number of shares and voting rights as of the date of the notice to convene the Annual General Meeting;
- The Company's annual report for 2019;
- The proposed revised Articles of Association; and
- Other documents for the use of the Annual General Meeting, including the proxy/voting by correspondence form and the registration form.

Registration of admission cards at the Annual General Meeting on Thursday, March 26, 2020, will open at 4:00 PM (CET).

Questions from the shareholders

Shareholders may ask questions to the Board of Directors and the Executive Management at the Annual General Meeting. Questions regarding the agenda and documents concerning the Annual General Meeting may be submitted in writing prior to the Annual 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 annual general meeting, which is available on the Company's website, <https://orphazyme.qcs-web.com/annual-general-meeting-0>.

Language

The Annual 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 Annual 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, March 3, 2020

The Board of Directors