

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 25, 2021 at 5:00 PM (CET), at the Company's address Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark.

In order to protect the health and safety of all, reduce the risk of COVID-19 spreading, and given the Danish government's restrictions on assembly, we strongly recommend that shareholders refrain from attending the Annual General Meeting in person and instead exercise their shareholder rights by giving proxy to the Board of Directors or by voting by correspondence prior to the Annual General Meeting.

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 2020
4. Allocation of loss according to the adopted annual report 2020
5. Resolution to grant discharge of liability to the Board of Directors and the Executive Management
6. Presentation of the Company's remuneration report for 2020
7. Approval of remuneration of the Board of Directors for the current financial year
8. Election of members to the Board of Directors
9. Election of auditor
10. Authorization to acquire treasury shares
11. Any proposals from the Board of Directors
 - 11.1 Approval of adjustments to the Company's Remuneration Policy
 - 11.2 Amendments to Article 3.1 of the Articles of Association
 - 11.3 New Article 3.5 of the Articles of Association
12. 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 2020 be noted by the Annual General Meeting.

Item 3: Presentation and adoption of the annual report 2020

The Board of Directors proposes that the Company's annual report 2020 be adopted.

Item 4: Allocation of loss according to the adopted annual report 2020

The Board of Directors proposes that the loss related to the financial year 2020 amounting to approximately DKK 633.2 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 2020.

Item 6: Presentation of the Company's remuneration report for 2020

The Company has prepared a remuneration report for 2020, which according to applicable legislation is presented to the Annual General Meeting for an advisory vote.

The remuneration report has been prepared in accordance with the requirements of section 139b of the Danish Companies Act and includes information on remuneration awarded or due to the members of the Board of Directors and the Executive Management of the Company concerning the financial year 2020.

The remuneration report for 2020 is available on the Company's website, <https://orphazyme.gcs-web.com/corporate-governance/governance-documents>.

The Board of Directors proposes that the Annual General Meeting approves the Company's remuneration report for 2020 in the advisory vote.

Item 7: 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 2021 be approved by the Annual General Meeting:

Members of the Board of Directors shall receive an annual base fee of: EUR 36,225

The Chairman of the Board of Directors shall in addition receive: EUR 45,555

The Deputy Chairman of the Board of Directors shall in addition receive: EUR 14,318

The remuneration level for the Board of Directors is maintained at the same level as for 2020 as approved by the Extraordinary General Meeting on September 21, 2020.

The members of the Board of Directors receive annual supplementary fees for their work in the Company's Audit Committee, Remuneration Committee, Nomination Committee, and Science Committee. The Board of Directors proposes that the Annual General Meeting approve the following supplementary fees for 2021 for committee work:

Members of the Audit Committee shall receive an annual supplementary fee of: EUR 7,035

Members of the Remuneration Committee shall receive an annual supplementary fee of: EUR 4,935

Members of the Nomination Committee shall receive an annual supplementary fee of: EUR 3,255

Members of the Science Committee shall receive an annual supplementary fee of: EUR 3,255

The Chairman of the Audit Committee shall receive an annual supplementary fee of: EUR 14,175

The Chairman of the Remuneration Committee shall receive an annual supplementary fee of: EUR 9,450

The Chairman of the Nomination Committee shall receive an annual supplementary fee of: EUR 6,825

The Chairman of the Science Committee shall receive an annual supplementary fee of: EUR 6,825

The supplementary fees for committee work are maintained at the same level as for 2020 as approved by the Extraordinary General Meeting on September 21, 2020.

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 2021 in the form of restricted share units ("RSUs") 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.

As announced in company announcement no. 72/2020 of December 10, 2020, the Company's CEO had resigned and the Company initiated a search for a new CEO. The Company has, as announced in company announcement no. 05/2021 of March 1, 2021, appointed Christophe Bourdon as the Company's CEO, effective as of April 1, 2021. In order to ensure continuity going forward, the Company's Chairman of the Board of Directors, Georges Gemayel, is assisting the Executive Management of the Company in an interim period with ad-hoc tasks, until the appointment of Christophe Bourdon as CEO is effective. As a result hereof the Board of Directors proposes that the Annual General Meeting approves an ad-hoc fee to the Chairman of the Board of Directors for 2021 of EUR 88,605 and an additional grant of RSUs equal to 100% of the annual grant of RSUs for 2021. The remuneration for the ad hoc tasks of the Chairman are maintained at the same level as for 2020.

Item 8: 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, Carrolee Barlow, Catherine Moukheibir, Martijn Kleijwegt, Martin Bonde and Rémi Droller. In addition, the Board of Directors proposes that the general meeting elect Stephanie Okey as new member of the Board of Directors. The current board member Sten Verland is not seeking re-election.

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 9: Election of auditor

In accordance with the Audit Committee's recommendation, the Board of Directors proposes re-election of EY 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 10: Authorization to acquire treasury shares

In order to continuously provide the Company with the necessary flexibility to raise new capital through alternative transaction structures, the Board of Directors proposes to renew the current authorization to the Board of Directors to acquire treasury shares in the period until March 25, 2026, on one or more occasions, with a total nominal value of up to 10% 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 11: Any proposals from the Board of Directors**Item 11.1: Approval of adjustments to the Company's Remuneration Policy**

The Board of Directors proposes that the Annual General Meeting approves certain adjustments to the Company's Remunerations Policy as adopted by the Board of Directors.

The revised Remuneration Policy includes the following material changes:

- Updated to allow for a share-based incentive program to the Executive Management and certain employees comprising restricted share units (RSUs) and performance share units (PSUs), including related pre-defined performance criteria to be determined by the Board of Directors based on corporate goals, e.g. targets related to development in the Company's share price, organizational development as well as pre-clinical and clinical development;
- Adjustment of the shareholding requirement for the Executive Management so that the shareholding may be built up over a period of 36 months;
- Adjustment of the claw back clause with the purpose of aligning the clause with the revised Recommendations on Corporate Governance; and
- Adjustment of the situations under which accelerated vesting may occur.

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

Item 11.2: Amendments to Article 3.1 of the Articles of Association

By decisions of September 28, 2020, the Board of Directors has partly exercised the authorization in Article 3.1 of the Articles of Association to increase the Company's share capital without pre-emption rights for existing shareholders following which a nominal value of DKK 7,616,146 of the authorization has been issued.

In order to continuously provide the Company with the necessary flexibility to raise new capital, the Board of Directors proposes that the authorization in Article 3.1 be reset and extended until March 25, 2026. Consequently, the Board of Directors proposes that Article 3.1.1 be deleted from the Articles of Association and that Article 3.1 of the Articles of Association be amended as follows:

"In the period until 25 March 2026, 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 6,989,767. The capital increase shall take place at market price as determined by the Board of Directors and shall be effected by cash payment, debt conversion or contribution in kind."

Item 11.3: New Article 3.5 of the Articles of Association

The Board of Directors proposes to adopt a new Article 3.5 of the Articles of Association in order to authorize the Board of Directors to increase the Company's share capital without pre-emption rights for the Company's existing shareholders in connection with the issue of new shares to members of the Board of Directors, executives and/or employees of the Company. The capital increase may take place below market price.

The Board of Directors proposes that a new Article 3.5 be included as follows:

"In the period until 25 March 2026, the Board of Directors is authorised to increase the Company's share capital in one or more issues without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 1,300,000 in connection with the issue of new shares to members of the Board of Directors, executives and/or employees of the Company. The new shares shall be issued against cash payment at a subscription price to be determined by the Board of Directors, which may be below the market price."

Further, the Board of Directors proposes a total cap of nominally DKK 2,000,000 for the capital increase authorizations set out in Article 3.2 and the new Article 3.5. Consequently, the following new Article 3.6 is proposed:

"The authorisations granted to the Board of Directors pursuant to Articles 3.2 and 3.5 can in the aggregate only be exercised to increase the share capital by a maximum nominal amount of DKK 2,000,000."

If the new Article 3.5 and Article 3.6 is approved, the current Article 3.5 will be renumbered to Article 3.7, and the new Article 3.7 will be amended as follows:

"New shares issued pursuant to Articles 3.1, 3.2, 3.3, 3.4 and 3.5 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."

Adoption requirements

The proposals under item 11.2-11.3 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 Annual 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. The Remuneration Report is subject to an advisory vote.

Share capital and shareholders' participation and voting rights

The Company's nominal share capital is 34,948,837, 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 18, 2021**.

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.

Notification of participation

Shareholders, who are entitled to attend the Annual General Meeting and wish to attend the Annual General Meeting must notify the Company of participation no later than on **Friday, March 19, 2021 at 11:59 PM (CET)**. The registration form is available on the Company's website, <https://orphazyme.gcs-web.com/annual-general-meeting-0>.

Notification of participation may be provided as follows:

- Electronically via the Company's shareholder portal at <https://portal.computershare.dk/portal/index.asp?page=login&asident=22035&lan=en>;
- Electronically via Computershare A/S' website, www.computershare.com/dk;
- By completing and forwarding the registration form by letter to Computershare A/S, Lottenborgvej 26D, floor 1, DK-2800 Kongens Lyngby, Denmark (to be received by the Company within the deadline);
- By emailing a scanned version to gf@computershare.dk; or
- By contacting Computershare A/S by telephone on (+45) 45 46 09 99.

A shareholder or a proxy may participate in the Annual General Meeting together with an adviser, provided that notification of the adviser's participation has been provided.

A confirmation of registration will be sent by email to the email address provided by the shareholder upon registration of participation. The electronic admission card must be presented at the Annual General Meeting either via smartphone/tablet or as a printed version. 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 Computershare A/S no later than on **Friday, March 19, 2021**. 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 the Company's shareholder portal at <https://portal.computershare.dk/portal/index.asp?page=login&asident=22035&lan=en>;
- Electronically via Computershare A/S' website, www.computershare.com/dk;
- By completing and forwarding the proxy form by letter to Computershare A/S, Lottenborgvej 26D, floor 1, DK-2800 Kongens Lyngby, Denmark; or
- By emailing a scanned version to gf@computershare.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 Computershare A/S no later than on **Wednesday, March 24, 2021, 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 the Company's shareholder portal at <https://portal.computershare.dk/portal/index.asp?page=login&asident=22035&lan=en>;
- Electronically via Computershare A/S' website, www.computershare.com/dk;
- By completing and forwarding the vote by correspondence form by letter to Computershare A/S, Lottenborgvej 26D, floor 1, DK-2800 Kongens Lyngby, Denmark; or
- By emailing a scanned version to gf@computershare.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 2020;
- The Company's remuneration report 2020;
- 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 25, 2021, 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.gcs-web.com/annual-general-meeting-0>.

Language

The Annual General Meeting will be held in English in accordance with Article 4.7 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, March 3, 2021

The Board of Directors