

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 Wednesday, May 17, 2023 at 11:00 AM (CEST), at the offices of Gorrissen Federspiel, Axeltorv 2, DK-1609 Copenhagen V, 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 for 2022
4. Allocation of profit according to the adopted annual report for 2022
5. Resolution to grant discharge of liability to the Board of Directors and the Executive Management
6. Presentation of the remuneration report for 2022 for an advisory vote
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 Update of the Company's remuneration policy
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 2022 be noted.

Item 3: Presentation and adoption of the annual report for 2022

The Board of Directors proposes that the Company's annual report 2022 be adopted.

Item 4: Allocation of profit according to the adopted annual report for 2022

The Board of Directors proposes that the profit related to the financial year 2022 according to the adopted annual report 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 for 2022.

Item 6: Presentation of the remuneration report for 2022 for an advisory vote

The Company has prepared a remuneration report for 2022, which according to applicable legislation is presented to the Annual General Meeting for an advisory vote.

The remuneration report 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 2022.

The remuneration report for 2022 is available on the Company's website, <https://orphazyme.gcs-web.com/corporate-governance/governance-documents>.

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 2023 be approved by the Annual General Meeting:

Members of the Board of Directors shall receive an annual base fee of: DKK 450,000

The Chairman of the Board of Directors shall in addition receive: DKK 300,000

The members of the Board of Directors will not receive any supplementary fees for their work in the Company's Audit Committee, Remuneration Committee and Nomination Committee.

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.

In addition to the fixed fees, the members of the Board of Directors will receive share-based remuneration for 2023 in the form of restricted share units ("RSUs") through a grant corresponding to 50% of each Board member's fixed annual base fee, such base fee to include additional base fees to the Chairman. The RSUs will be subject to the terms of conditions as set out in the remuneration policy.

The Board of Directors notes that Anders Fink Vadsholt will not be granted Board RSUs due to his position as CEO of the Company.

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 three and not more than six 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 Company's 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: Bo Jesper Hansen, John Sommer Schmidt, and Anders Fink Vadsholt.

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

The Board of Directors proposes to renew the current authorization to the Board of Directors to acquire treasury shares in the period until June 29, 2027, 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 not exceeding 10% 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: Update of the Company's remuneration policy

The Board of Directors proposes to update the Company's remuneration policy.

The updated remuneration policy reflects the changed practice in board remuneration and includes the option of awarding a cash-based transformation bonus to the CEO linked to the development in the Company's total market cap on Nasdaq Copenhagen for the remainder of 2023 and for 2024. The transformation bonus provides for a bonus ranging from DKK 1.5 million up to DKK 8.5 million based on the Company's market cap between DKK 30 million and DKK 300 million or above.

The Board of Directors notes that the transformation bonus to the CEO will replace all grants under both the short-term and the long-term incentive program in 2023.

Adoption requirements

All proposed resolutions on the agenda shall be passed by a simple majority of votes cast pursuant to Article 7.1 of the Company's Articles of Association, except the remuneration report presented under agenda item 6, which is subject to an advisory vote.

Share capital and shareholders' participation and voting rights

The Company's nominal share capital is 35,312,241, 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 **Wednesday, May 10, 2023**.

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, May 12, 2023 at 11:59 PM (CEST)**. 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 ordinary mail to Computershare A/S, Lottenborgvej 26D, floor 1, DK-2800 Kongens Lyngby, Denmark (to be received by the Company within the deadline); or
- By e-mailing a scanned version to gf@computershare.dk.

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 e-mail to the e-mail 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, May 12, 2023 at 11:59 PM (CEST)**. 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 ordinary mail to Computershare A/S, Lottenborgvej 26D, floor 1, DK-2800 Kongens Lyngby, Denmark; or

- By e-mailing 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 **Tuesday, May 16, 2023 at 03:00 PM (CEST)**. The voting form (which is the same form used for granting a proxy) is available on the Company's website, <https://orphazyme.qcs-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 ordinary post to Computershare A/S, Lottenborgvej 26D, floor 1, DK-2800 Kongens Lyngby, Denmark; or
- By e-mailing 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: Updated 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 2022;
- The Company's remuneration report for 2022;
- The current Articles of Association of the Company; 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 Wednesday, May 17, 2023, will open at 10:30 AM (CEST).

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 e-mail 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 4.7 of the Company's Articles of Association.

How to get there

The Annual General Meeting will be held at Gorrissen Federspiel, Axeltorv 2, DK-1609 Copenhagen V, Denmark, which can be reached by car or public transportation.

Copenhagen, April 25, 2023

The Board of Directors