

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, June 29, 2022 at 2:00 PM (CEST), at the offices of Gorrissen Federspiel, Axeltorv 2, DK-1609 Copenhagen V.

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 2021
4. Covering of loss according to the adopted annual report 2021
5. Resolution to grant discharge of liability to the Board of Directors and the Executive Management
6. Presentation of the remuneration report for 2021
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 Amendment of the Company's Articles of Association with the view to amend the Company's purpose
 - 11.2 Amendment of the Company's Articles of Association with the view to change the minimum and maximum number of members on the Board of Directors
 - 11.3 Amendment of the Company's Articles of Association with the view to amend the rules of signature
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 2021 be noted.

Item 3: Presentation and adoption of the annual report 2021

The Board of Directors proposes that the Company's annual report 2021 be adopted.

Item 4: Covering of loss according to the adopted annual report 2021

The Board of Directors proposes that the loss related to the financial year 2021 according to the adopted annual report for 2021.

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 2021.

Item 6: Presentation of the remuneration report for 2021

The Company has prepared a remuneration report for 2021, 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 2021.

The remuneration report for 2021 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 2022 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.

Item 8: Election of members to the Board of Directors

With reference to agenda item 11.2, it is proposed to amend Article 8.1 of the Company's Articles of Association with the view to change the minimum and maximum number of members on the Board of Directors as the Company no longer has any material ongoing operational business activities following the restructuring process and sale of substantially all of its assets and business activities.

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.

Accordingly and provided that the proposal under agenda item 11.2 is adopted, the Board of Directors proposes election of the following three members to the Board of Directors for a one-year term :

- Bo Jesper Hansen,
- Anders Fink Vadsholt, and
- John Sommer Schmidt.

Georges Gemayel and Andrew Mercieca will not stand for 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

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: Amendment of the Company's Articles of Association with the view to amend the Company's purpose

Following the sale of substantially all of the Company's assets and business activities, including those relating to the development and approval of arimoclomol, as announced by the Company through company announcement no. 24 on May 15, 2022 and company announcement no. 29 on May, 31 2022, the Board of Directors proposes that the objects of the Company as include in the Company's Articles of Association are amended.

Consequently, it is proposed that Article 1.2 of the Company's Articles of Association be worded as follows:

"The Company's objects are to engage in activities and investments related to biopharmaceutical research and development as well as to carry out any associated activities as deemed relevant by the Board of Directors. Furthermore, the Company may participate in partnerships or co-operate with other businesses."

Item 11.2: Amendment of the Company's Articles of Association with the view to change the minimum and maximum number of members on the Board of Directors

As the Company no longer has any material ongoing operational business activities following the restructuring process and sale of substantially all of its assets and business activities, the Board of Directors proposes that the Company's Articles of Association are amended with the view to reduce the number of members of the Board of Directors to three members.

Consequently, it is proposed that Article 8.1 of the Company's Articles of Association be worded as follows:

"The Board of Directors consists of not less than three and not more than six members elected by the general meeting."

Item 11.3: Amendment of the Company's Articles of Association with the view to amend the rules of signature

In light of the recent changes in the Company's management, as also described under agenda items 8 and 11.2, the Board of Directors proposes that the Company's rules of signature are amended.

Consequently, it is proposed that Article 10.1 of the Company's Articles of Association be worded as follows:

"The Company shall be bound (i) by the joint signatures of two members of the Board of Directors or (ii) by the joint signatures of a member of the Board of Directors and a member of the Executive Management."

Adoption requirements

The proposal under agenda item 11.1-11.3 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 Company's 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 Company's Articles of Association. The Remuneration Report as presented under agenda item 6 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, June 22, 2022**.

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, June 24, 2022 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 letter to Computershare A/S, Lottenborgvej 26D, floor 1, DK-2800 Kongens Lyngby, Denmark (to be received by the Company within the deadline); or
- By emailing 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 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, June 24, 2022**. 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 **Tuesday, June 28, 2022 at 3:00 PM (CEST)**. The voting form (which is the same form used for granting a proxy) is available on the Company's website, <https://orphazyme.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;

- 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 2021;
- The Company's remuneration report 2021;
- The proposed revised 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, June 29, 2022, will open at 1:00 PM (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 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 Gorrissen Federspiel, Axeltorv 2, DK-1609 Copenhagen V, which can be reached by car or public transport.

Copenhagen, June 7, 2022

The Board of Directors