

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 Tuesday, February 15, 2022 at 5:00 PM (CET), at the Company's address Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark.

In order to continuously provide the Company with the necessary flexibility to raise new capital to continue funding of its operations and protract a long-term capital structure, the Board of Directors proposes a number of authorizations to be approved by the Extraordinary General Meeting, including authorizations to issue shares, convertible bonds and warrants.

In general, the Board of Directors believes that the proposed authorizations will benefit the Company and its shareholders as they would provide flexibility to raise additional capital to fund the Company's product candidate, arimoclomol, for the treatment of NPC through the US and EU regulatory approval processes.

Covid-19

We expect the current Covid-19 health risks and government precautions to continue on the date of the Extraordinary General Meeting.

Shareholders wishing to attend the Extraordinary General Meeting in person are encouraged not to bring advisors or other authorized guests to limit the number of physical attendees at the Extraordinary General Meeting. Further, meeting participants will be asked to present a valid corona passport and meeting participants having or showing symptoms of virus infection are kindly requested not to attend the meeting.

Shareholders who do not attend the Extraordinary General Meeting are encouraged to vote by submitting a postal vote or a proxy to the Board of Directors, as described below. Shareholders may submit written questions to the Company's management before the Extraordinary General Meeting.

The above precautions are based on rules and recommendations issued by the Danish Health Authorities effective on the date hereof. If applicable rules and recommendations are updated before the Extraordinary General Meeting, additional precautions may be taken.

Agenda

1. Authorizations to the Board of Directors
 - 1.1 Approval of a new authorization to the Board of Directors to issue shares at market price without pre-emption rights for existing shareholders
 - 1.2 Approval of a new authorization to the Board of Directors to issue shares below market price without pre-emption rights for existing shareholders
 - 1.3 Approval of a new authorization to the Board of Directors to issue convertible bonds above, at or below market price without pre-emption rights for existing shareholders and to effect the associated capital increases
 - 1.4 Approval of new authorization to the Board of Directors to issue warrants above, at or below market price without pre-emption rights for existing shareholders and to effect the associated capital increases
 - 1.5 Approval of amendment of the Articles of Association
2. Election of new member of the Board of Directors
3. Approval of authorization to the Chairman of the meeting

Adoption requirements

The proposals under agenda item 1.1 and 1.5 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 proposals under agenda item 1.2-1.4 on the agenda shall be passed by at least nine-tenths of the votes cast as well as at least nine-tenths of the share capital represented at the Extraordinary General Meeting, cf. Article 7.2 of the Articles of Association and the Danish Company's Act section 107(2)(i).

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 is 34,952,241, divided into shares of DKK 1 each. 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 **Tuesday, February 8, 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 Extraordinary General Meeting and wish to attend the Extraordinary General Meeting must notify the Company of participation no later than on **Friday, February 11, 2022 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 proxy@computershare.dk; or
- By contacting Computershare A/S by telephone on (+45) 45 46 09 99.

A shareholder or a proxy is entitled to participate in the Extraordinary General Meeting together with an adviser, provided that notification of the adviser's participation has been provided, however shareholders are encouraged not to bring advisers due to Covid-19 in order to limit the number of participants.

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 Extraordinary 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 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

Proxy forms must be received by Computershare A/S no later than on **Friday, February 11, 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 proxy@computershare.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 Computershare A/S no later than on **Monday, February 14, 2022 at 3:00 PM (CET)**. The vote by correspondence 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 proxy@computershare.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, www.orphazyme.com:

- The notice convening the Extraordinary General Meeting, including the agenda and the complete proposals as well as Appendix 1: Candidate for the Board of Directors;
- The aggregate number of shares and voting rights as of the date of the notice to convene the Extraordinary General Meeting;
- The Company's Articles of Association;
- 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 Tuesday, February 15, 2022 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 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 4.7 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, January 24, 2022

The Board of Directors

Extraordinary General Meeting of Orphazyme A/S, CVR no. 32 26 63 55 (the "Company"), to be held on Tuesday, February 15, 2022 at 5:00 PM (CET), at the Company's address Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark.

Items on the agenda, including complete proposals:

Item 1: Authorizations to the Board of Directors

Item 1.1: Approval of a new authorization to the Board of Directors to issue shares at market price without pre-emption rights for existing shareholders

It is proposed to grant the Board of Directors an authorization in the period until 1 January 2027 to increase the Company's share capital through on one or more issues of new shares at market price without pre-emption rights for the Company's existing shareholders by up to a total of nominal amount of DKK 20,000,000.

The Board of Directors proposes that a new Article 3.6 of the Articles of Association be included as follows:

"In the period until 1 January 2027, 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 20,000,000. 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."

Further, the Board of Directors proposes a total cap of nominally DKK 40,000,000 for the capital increase authorizations set out in the current Article 3.1 of the Articles of Association, the new Article 3.6 as proposed in this agenda item 1.1 and the new Article 3.7 as proposed under agenda item 1.2. Consequently, the following new Article 3.10 is proposed:

"The authorisations granted to the Board of Directors pursuant to Articles 3.1, 3.6 and 3.7 can in the aggregate only be exercised to increase the share capital by a maximum nominal amount of DKK 40,000,000."

The authorization is furthermore subject to the terms as set out in the current Article 3.7 of the Articles of Association cited below under agenda item 1.5.

Item 1.2: Approval of a new authorization to the Board of Directors to issue shares below market price without pre-emption rights for existing shareholders

It is proposed to grant the Board of Directors an authorization in the period until 1 January 2027 to increase the Company's share capital through on one or more issues of new shares below market price without pre-emption rights for the Company's existing shareholders by up to a total of nominal amount of DKK 20,000,000.

The Board of Directors proposes that a new Article 3.7 of the Articles of Association be included as follows:

"In the period until 1 January 2027, the Board of Directors is authorised to increase the Company's share capital through 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 20,000,000. 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."

Further, the authorization is subject to the general limitation proposed as a new Article 3.10 of the Article of Association cited above under agenda item 1.1.

The authorization is furthermore subject to the terms as set out in the current Article 3.7 of the Articles of Association cited below under agenda item 1.5.

Item 1.3: Approval of a new authorization to the Board of Directors to issue convertible bonds above, at or below market price without pre-emption rights for existing shareholders and to effect the associated capital increases

It is proposed to grant the Board of Directors an authorization in the period until 1 January 2027 to issue convertible bonds on one or more occasions with a total principal amount of up to DKK 70,000,000 without pre-emption rights for the Company's existing shareholders which are convertible into shares in the Company.

As a consequence of the above, it is also proposed to authorize the Board of Directors in the period until 1 January 2027 to make the related increases of the share capital as a result of conversion of such convertible bonds by a nominal amount of up to DKK 20,000,000 and to effect the associated capital increases.

The Board of Directors proposes that a new Article 3.8 of the Articles of Association be included as follows:

"In the period until 1 January 2027, the Board of Directors is authorised to issue convertible bonds on one or more occasions without pre-emption rights for the existing shareholders and with a total principal amount of up to DKK 70,000,000 which are convertible into shares in the Company. The convertible bonds shall be effected by cash payment. The conversion price as determined by the Board of Directors may be above, at or below the market price at the time of issuance of the convertible bonds. The issuance of convertible bonds may be directed at qualified investors. The Board of Directors is authorised in the period until 1 January 2027 to increase the Company's share capital by up to nominally DKK 20,000,000 by conversion of convertible bonds issued pursuant to this Article 3.8 and to effect the associated capital increases."

All other terms and conditions governing the convertible bonds are determined by the Board of Directors at the time of issuance. If the proposal in this agenda item 1.3 to authorize the Board of Directors to issue convertible bonds is adopted and the Board of Directors decides to issue convertible bonds pursuant to the proposed Article 3.8, the terms and conditions related to the convertible bonds are to be incorporated as an appendix to the Company's Articles of Association.

The authorization is furthermore subject to the terms as set out in the current Article 3.7 of the Articles of Association cited below under agenda item 1.5.

Item 1.4: Approval of new authorization to the Board of Directors to issue warrants above, at or below market price without pre-emption rights for existing shareholders and to effect the associated capital increases

It is proposed to grant the Board of Directors an authorization in the period until 1 January 2027 to issue warrants on one or more occasions without pre-emption rights for the Company's existing shareholders granting the warrant holders rights to subscribe for a total of nominally DKK 20,000,000.

As a consequence of the above, it is also proposed to authorize the Board of Directors in the period until 1 January 2027 to make the related increase of the share capital resulting from the exercise of warrants by a nominal amount of up to DKK 20,000,000 and to effect the associated capital increases.

The Board of Directors proposes that a new Article 3.9 on the Articles of Association be included as follows:

"In the period until 1 January 2027, the Board of Directors is authorised to issue warrants on one or more occasions without pre-emption rights for the existing shareholders granting the holders right to subscribe for shares in the Company for a total amount of up to nominally DKK 20,000,000. The Board of Directors is entitled to determine the exercise price for the warrants upon issue given that the exercise price may be above, at or below the market price at the time of issuance. The Board of Directors is authorised in the period until 1 January 2027 to increase the Company's share capital by up to nominally DKK 20,000,000 resulting from the exercise of warrants pursuant to this Article 3.9 and to effect the associated capital increases."

All other terms and conditions governing the warrants are determined by the Board of Directors at the time of issuance. If the proposal in this agenda item 1.4 to authorize the Board of Directors to issue warrants is adopted and the Board of Directors decides to issue warrants pursuant to the proposed Article 3.9, the terms and conditions related to the warrants are to be incorporated as an appendix to the Company's Articles of Association.

The authorization is furthermore subject to the terms as set out in the current Article 3.7 of the Articles of Association cited below under agenda item 1.5.

Item 1.5: Approval of amendment of the Articles of Association

If the authorizations proposed under agenda items 1.1-1.4 are adopted by the general meeting, it is proposed to renumber the current Article 3.6 of the Articles of Association and renumber and amend the current Article 3.7 of the Articles of Association as a consequence hereof.

The Board of Directors proposes that the current Article 3.7 of the Articles of Association will be renumbered and amended as follows:

"New shares issued pursuant to Articles 3.1, 3.2, 3.3, 3.4, 3.5, 3.6, 3.7, 3.8 and 3.9 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: Election of new member of the Board of Directors

The Board of Directors proposes to elect Andrew Mercieca as new member of the Board of Directors. Please see Appendix 1 for a description of the nominated candidate's qualifications and positions, including information about other executive/management functions held by the candidate in Danish and foreign companies.

Item 3: Approval of authorization to 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.