

Company announcement

No. 46/2020

Orphazyme A/S

Ole Maaløes Vej 3

DK-2200 Copenhagen N

www.orphazyme.com

Company Registration No. 32266355

Notice to convene Extraordinary General Meeting in Orphazyme A/S

Copenhagen, Denmark, August 28, 2020 – Orphazyme A/S (ORPHA.CO), a late-stage biopharmaceutical company pioneering the amplification of the Heat-Shock Proteins in order to develop and commercialize novel therapeutics for the treatment of neurodegenerative orphan diseases, today announced:

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 September 21, 2020 at 10:00 AM (CEST), at the Company's address Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark.

To reduce the risk of COVID-19 spreading, we recommend that shareholders refrain from attending the Extraordinary General Meeting in person and instead cast their votes by correspondence or proxy. The Company will also reduce its number of representatives at the Extraordinary General Meeting to reduce the risk of infection.

Agenda

1. Approval of amendment to an authorization to the Board of Directors to increase the share capital of the Company without pre-emption rights for existing shareholders
2. Approval of an authorization to the Board of Directors to decide whether a general meeting shall be held fully by electronic means
3. Approval of amendment to Article 6.6 of the Articles of Association regarding exercise of voting rights
4. Approval of an authorization to the Board of Directors to let the Company indemnify the Executive Management and employees in relation to certain claims related to the offer, sale and listing of the Company's securities in the United States to be included in the Articles of Association
5. Approval of indemnification of the Board of Directors related to the offer, sale and listing of the Company's securities in the United States
6. Approval of adjustments to the Company's Remuneration Policy
7. Approval of remuneration of the Board of Directors for 2020
8. Approval of share-based remuneration for the Board of Directors
9. Election of new member of the Board of Directors
10. Approval of authorization of the Chairman of the meeting

Items on the agenda, including complete proposals:

In light of the contemplated listing of the Company in the United States and the offering of American Depositary Shares, the Board of Directors proposes a number of items to be approved by the Extraordinary General Meeting including in relation to the authorization to undertake the offering, U.S. specific risk management and the remuneration level and structure. The complete proposals are set out below.

In general, the Board of Directors believes that the offering of American Depositary Shares and the U.S. listing will benefit the Company and its shareholders as it would provide additional capital to fund the Company's product candidate, arimoclomol for the treatment of NPC through regulatory approval, support the Company's further clinical development of its product candidate for other indications, including ALS, sIBM and neurological Gaucher disease and establish a public market for the American Depositary Shares and facilitate future access to the public capital markets.

Item 1: Approval of amendment to an authorization to the Board of Directors to increase the share capital of the Company without pre-emption rights for existing shareholders

For the purposes of ensuring flexibility and facilitate the contemplated capital increase related to the offering of American Depositary Shares and the U.S. listing, the Board of Directors proposes to amend Article 3.1 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 and shall be effected by cash payment, debt conversion or contribution in kind."

Item 2: Approval of an authorization to the Board of Directors to decide whether a general meeting shall be held fully by electronic means

In light of the ongoing COVID-19 pandemic including the prohibition imposed on larger gatherings as well as to ensure flexibility with respect to future general meetings of the Company, the Board of Directors proposes to adopt a new Article 4.3 of the Articles of Association with the following wording to authorize the Board of Directors to decide that general meetings may be held completely or partially by electronic means:

"If the Board of Directors finds it appropriate, and if the general meeting can be conducted in a technically safe manner, the Board of Directors may decide that the general meeting shall be held fully or partially as an electronic general meeting. If so decided, shareholders will be able to attend, express their opinion and vote at the general meeting by electronic means. Detailed information on the procedures for electronic attendance and participation will be made available on the company's website and in the relevant notices convening the general meetings, and written information on the subject will also be sent to shareholders registered in the Company's register of shareholders if so requested."

Accordingly, the current Articles 4.3-4.7 will be re-numbered to Articles 4.4-4.8.

Item 3: Approval of amendment to Article 6.6 of the Articles of Association regarding exercise of voting rights

The Board of Directors proposes that the current Article 6.6 of the Articles of Association be amended as follows:

"Each share of the nominal value of DKK 1 shall carry 1 vote. A person registered as a holder of shares of the Company in VP Securities A/S and acting in a professional capacity on behalf of other natural or legal persons, including holders of American Depositary Shares representing shares of the Company, may exercise voting rights attached to any such shares in a manner that is not identical to the exercise of the voting rights attached to other shares of the Company held by such person"

Item 4: Approval of an authorization to the Board of Directors to let the Company indemnify the Executive Management and employees in relation to certain claims related to the offering of American Depositary Shares and the listing of the Company in the United States to be included in the Articles of Association

The Board of Directors proposes that the Extraordinary General Meeting authorizes the Board of Directors to resolve to let the Company indemnify the Executive Management and employees of the Company and its subsidiaries in relation to certain claims related to the offer, sale and listing of the Company's securities in the United States and in respect of the Company's reports filed with or furnished to the U.S. Securities and Exchange Commission thereafter.

It is the view of the Board of Directors that the proposal is necessary in order to attract, motivate and retain qualified members to the Executive Management and employees. The Board of Directors has concluded that the planned offer, sale and listing of the Company's securities in the United States entails a significantly increased risk exposure for the Executive Management and for certain employees of the Company and its subsidiaries, which significantly exceeds the level of exposure that would normally be expected as a member of the Executive Management or as an employee of a Danish listed company. On this basis, the Board of Directors finds that it is necessary and in the Company's interest to be able to indemnify members of the Executive Management and employees of the Company and its subsidiaries for certain claims made personally against such persons in connection with their services to the Company.

If the proposal is adopted, a new article 9.2 will be included in the Articles of Association of the Company with the following wording:

"The Board of Directors are authorized to resolve to let the Company indemnify members of the Executive Management and employees of the Company or its subsidiaries for certain claims against these individuals in connection with their services to the Company.

The Company's indemnification covers claims and reasonable legal costs arising from the offer, sale and listing of the Company's securities in the United States and/or the Company's subsequent status as a listed company in the United States, including in respect of the Company's reports filed with or furnished to the U.S. Securities and Exchange Commission thereafter.

Notwithstanding the foregoing, the Company's indemnification shall not cover the following claims:

- (i) claims against a person pursuant to Danish law raised before the Danish Courts, except claims arising from the offer, sale and listing of the Company's securities in the United States and/or the Company's subsequent status as a listed company in the United States, including in respect of the Company's reports filed with or furnished to the U.S. Securities and Exchange Commission thereafter;*
- (ii) claims against a person for damages and legal costs related to criminal and/or grossly negligent or willful acts or omissions committed by the indemnified person;*
- (iii) claims against an indemnified person, which is attributable to the gaining or purported gaining of any profit or advantage to which the indemnified person or any related natural or legal person was not legally entitled;*
- (iv) claims covered by insurance. To the extent the insurer refuses to provide cover for other reasons than those referenced in items (i) and (ii) above, the Company's indemnification will cover such claims, provided, however, that the Company shall in such event be entitled at any time to represent the insured in respect of the insurer and shall automatically by subrogation enter into any and all rights under said insurance policy;*
- (v) claims brought against the indemnified person by the Company or any subsidiary of the Company; or*
- (vi) any sum payable to a regulatory authority by way of a penalty in respect of the indemnified person's personal non-compliance with any requirement of a regulatory nature howsoever arising.*

The indemnification is limited to a maximum amount per claim per person equivalent to the gross proceeds obtained by the Company in connection with the offering of American Depositary Shares in the United States.

The indemnification shall remain in force for a period of five years after the resignation of the indemnified person from the Company or its subsidiaries, if the claims made within such period are related to such person's services to the Company."

Item 5: Approval of indemnification of the Board of Directors related to the offering of American Depositary Shares and the listing of the Company in the United States

The Board of Directors proposes that the Extraordinary General Meeting approve that the Board of Directors shall be indemnified by the Company in relation to certain claims related to the offer, sale and listing of the Company's securities in the United States and in respect of the Company's reports filed with or furnished to the U.S. Securities and Exchange Commission thereafter.

It is the view of the Board of Directors that the proposal is necessary in order to attract, motivate and retain qualified members to the Board of Directors. The Board of Directors has concluded that the planned offer, sale and listing of the Company's securities in the United States entails a significantly increased risk exposure for the Board of Directors, which significantly exceeds the level of exposure that would normally be expected as a member of the Board of Directors in a Danish listed company. On this basis, the Board of Directors finds that it is necessary and in the Company's interest to indemnify members of the Board of Directors for certain claims made personally against such board members in connection with their services to the Company. The indemnification will become effective as of the date of approval by the Extraordinary General Meeting.

The Company's indemnification covers claims and reasonable legal costs arising from the offer, sale and listing of the Company in the United States and/or the Company's subsequent status as a listed company in the United States, including in respect of the Company's reports filed with or furnished to the U.S. Securities and Exchange Commission thereafter.

Notwithstanding the foregoing, the Company's indemnification shall not cover the following claims:

- (i) claims against a person pursuant to Danish law raised before the Danish Courts, except claims arising from the offer, sale and listing of the Company's securities in the United States and/or the Company's subsequent status as a listed company in the United States, including in respect of the Company's reports filed with or furnished to the U.S. Securities and Exchange Commission thereafter;
- (ii) claims against a person for damages and legal costs related to criminal and/or grossly negligent or willful acts or omissions committed by the member of the Board of Directors;
- (iii) claims against a person, which is attributable to the gaining or purported gaining of any profit or advantage to which the member of the Board of Directors or any related natural or legal person was not legally entitled;
- (iv) claims covered by insurance. To the extent the insurer refuses to provide cover for other reasons than those referenced in items (i) and (ii) above, the Company's indemnification will cover such claims, provided, however, that the Company shall in such event be entitled at any time to represent the insured in respect of the insurer and shall automatically by subrogation enter into any and all rights under said insurance policy;
- (v) claims brought against the member of the Board of Directors by the Company or any subsidiary of the Company; or
- (vi) any sum payable to a regulatory authority by way of a penalty in respect of the member of the Board of Director's personal non-compliance with any requirement of a regulatory nature howsoever arising.

The indemnification is limited to a maximum amount per claim per person equivalent to the gross proceeds obtained by the Company in connection with the offering of American Depositary Shares in the United States.

The indemnification shall remain in force for a period of five years after the resignation of the member of the Board of Directors from the Board of Directors, if the claims made within such period are related to such person's service as member of the Board of Directors.

Item 6: Approval of adjustments to the Company's Remuneration Policy

The Board of Directors proposes the following adjustments to the Company's Remuneration Policy to be approved by the Extraordinary General Meeting in light of the Company's contemplated listing in the United States and as result of an external benchmark analysis of comparable peer companies in order to ensure that the level and structure of remuneration for the Board of Directors and Executive Management remain attractive in order to attract, motivate and retain qualified professionals:

- Change to the exercise price for the restricted share units program for the Board of Directors and the cap on value for new board members in order to achieve a program which more closely resembles the structure of share-based incentive programs used by the Company's peers, as concluded on the basis of a benchmark analysis conducted on behalf of the Board of Directors.
- Change to the cap related to Performance Shares.
- The Company may in certain cases indemnify members of the Executive Management and members of the Board of Directors for claims not covered by the D&O insurance.
- The Matching Shares and Performance Shares will vest on an accelerated basis in connection with a public takeover.
- Minor technical amendments.

The Board of Directors' proposed and updated Remuneration Policy is set out in Appendix 1.

Item 7: Approval of remuneration of the Board of Directors for 2020

In light of the U.S. listing, the results of an external benchmark analysis of comparable peer companies, and with the view of continuously attracting, motivating and retaining qualified members to the Board of Directors in line with the principles set out in the Remuneration Policy to be approved under item 6 above, the Board of Directors proposes that the following remuneration level for 2020 be approved by the Extraordinary General Meeting with effect from July 1, 2020 and on a proportionate basis for the remainder of the current financial year:

Members of the Board of Directors shall receive an annual base fee of:	EUR 36,225 (previously EUR 31,500)
The Chairman of the Board of Directors shall in addition receive an annual fee of:	EUR 45,555 (previously EUR 24,900)
The Deputy Chairman of the Board of Directors shall in addition receive annual fee of:	EUR 14,318 (previously EUR 12,450)

The members of the Board of Directors will 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 Extraordinary General Meeting approve the following supplementary fees for 2020 be approved by the Extraordinary General Meeting with effect from July 1, 2020 and on a proportionate basis for the remainder of the current financial year:

Members of the Audit Committee shall receive an annual supplementary fee of:	EUR 7,035 (previously EUR 6,700)
Members of the Remuneration Committee shall receive an annual supplementary fee of:	EUR 4,935 (previously EUR 4,700)
Members of the Nomination Committee shall receive an annual supplementary fee of:	EUR 3,255 (previously EUR 3,100)
The Chairman of the Audit Committee shall receive an annual supplementary fee of:	EUR 14,175 (previously EUR 13,500)
The Chairman of the Remuneration Committee shall receive an annual supplementary fee of:	EUR 9,450 (previously EUR 9,000)
The Chairman of the Nomination Committee shall receive an annual supplementary fee of:	EUR 6,825 (previously EUR 6,500)

In addition and as approved by the Annual General Meeting held on March 26, 2020, 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: Approval of share-based remuneration for the Board of Directors

In light of the U.S. listing, the results of an external benchmark analysis of comparable peer companies, and with the view of continuously attracting, motivating and retaining qualified members to the Board of Directors, the Board of Directors proposes that the Extraordinary General Meeting approves a new grant of share-based remuneration for 2020 in the form of restricted share units corresponding to 50% of each Board member's fixed annual base fee and up to 100% of any new 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. The new grant of restricted share units for 2020 will be made pursuant to and in accordance with the principles set out in the revised Remuneration Policy approved by the Extraordinary General Meeting under item 6 above.

Exercise of the new grant of restricted share units will be contingent on certain conditions including (i) the initiation or completion of an offering and listing of American Depositary Shares in the United States and (ii) that the individual member of the Board of Directors does not exercise the restricted share units granted in March 2020 and approved at the Annual General Meeting on March 26, 2020. Accordingly, the value of the grants of share-based remuneration to the Board of Directors made in 2020 does not exceed 50% of the Board member's fixed annual base fee, including additional base fees to the Chairman and Deputy Chairman but excluding any additional fees for committee membership, and up to 100% of any new Board member's fixed annual base fee as an initial on-boarding grant, such base fee to include additional base fees to the Chairman and Deputy Chairman but excluding any additional fees for committee membership. The taxation of the individual board members on the basis the share-based remuneration is of no concern to the Company, save that the Company may provide a tax gross-up for any additional taxation imposed as a result of non-exercise of restricted share units granted in 2020.

Item 9: Election of new member of the Board of Directors

The Board of Directors proposes to elect Carolee Barlow as new member of the Board of Directors. Please see Appendix 2 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 10: Approval of authorization of 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.

Adoption requirements

The proposals under item 1-4 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 item 5-10 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 DKK 27,079,630, 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 **Monday, September 14, 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 Extraordinary General Meeting and wish to attend the Extraordinary General Meeting, must request an admission card no later than on **Thursday, September 17, 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 Computershare A/S' website, computershare.com/dk; or
- By completing and forwarding the registration form by letter to Computershare A/S, Lottenborgvej 26D, DK-2800 Kongens Lyngby; or
- By emailing a scanned version to gf@computershare.dk; or
- By contacting Computershare A/S by phone on +45 45 46 09 99.

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 ID; and
3. Enter your email address under "My Profile" and click "Save preferences".

The electronic admission card must be presented at the Extraordinary 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 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

Subject to having requested an admission card, shareholders may attend the Extraordinary General Meeting in person or by proxy. Proxy forms must be received by Computershare A/S no later than on **Thursday, September 17, 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 Computershare A/S' website, computershare.com/dk; or
- By completing and forwarding the registration form by letter to Computershare A/S, Lottenborgvej 26D, DK-2800 Kongens Lyngby; or
- By emailing a scanned version to gf@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 **Friday, September 18, 2020 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 Computershare A/S' website, computershare.com/dk; or
- By completing and forwarding the registration form by letter to Computershare A/S, Lottenborgvej 26D, DK-2800 Kongens Lyngby; or
- By emailing a scanned version to gf@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, <https://orphazyme.gcs-web.com/annual-general-meeting-0>:

- The notice convening the Extraordinary General Meeting, including the agenda and the complete proposals
- The proposed revised Articles of Association
- The aggregate number of shares and voting rights as of the date of the notice to convene the Extraordinary General Meeting
- 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 September 21, 2020, will open at 9:00 AM (CEST).

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.gcs-web.com/annual-general-meeting-0>.

Language

The Extraordinary 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 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 N, August 28, 2020.

The Board of Directors

For additional information, please contact**Orphazyme A/S**

Anders Vadsholt, CFO

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About Orphazyme A/S

Orphazyme is a biopharmaceutical company pioneering the Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases. The company is focused on developing therapies for diseases caused by protein misfolding, protein aggregation, and lysosomal dysfunction. Arimoclomol, the company's lead candidate, is in clinical development for four orphan diseases: Niemann-Pick disease Type C (NPC), Gaucher Disease, sporadic Inclusion Body Myositis (sIBM), and Amyotrophic Lateral Sclerosis (ALS). The Denmark-based company is listed on Nasdaq Copenhagen (ORPHA.CO). For more information, please visit www.orphazyme.com.

Forward-looking statement

This company announcement may contain certain forward-looking statements. Although the Company believes its expectations are based on reasonable assumptions, all statements other than statements of historical fact included in this company announcement about future events, including the clinical development and potential benefits of arimoclomol for Gaucher disease, NPC, sIBM and ALS, are subject to (i) change without notice and (ii) factors beyond the Company's control. These statements may include, without limitation, any statements preceded by, followed by, or including words such as "target," "believe," "expect," "aim," "intend," "may," "anticipate," "estimate," "plan," "project," "will," "can have," "likely," "should," "would," "could", and other words and terms of similar meaning or the negative thereof . Forward-looking statements are subject to inherent risks and uncertainties beyond the Company's control that could cause the Company's actual results, performance, or achievements to be materially different from the expected results, performance, or achievements expressed or implied by such forward-looking statements. Except as required by law, the Company assumes no obligation to update these forward-looking statements publicly, or to update the reasons actual results could differ materially from those anticipated in the forward-looking statements, even if new information becomes available in the future.