

**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, March 27, 2019 at 5:00 PM (CET), at the Company's address Ole Maaløes Vej 3, DK-2200 Copenhagen N, 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 2018
4. Distribution of profit or covering of loss according to the adopted annual report 2018
5. Resolution to grant discharge of liability to the Board of Directors and the Executive Management
6. Approval of remuneration of the Board of Directors for the current financial year
7. Election of members to the Board of Directors
8. Election of auditor
9. Authorization to acquire treasury shares
10. Any proposals from the Board of Directors
  - a. Approval of revised Remuneration Policy
11. 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 2018 be noted by the Annual General Meeting.

**Item 3: Presentation and adoption of the annual report 2018**

The Board of Directors proposes that the Company's annual report 2018 be adopted.

**Item 4: Distribution of profit or covering of loss according to the adopted annual report 2018**

The Board of Directors proposes that the loss related to the financial year 2018 amounting to DKK 230 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 2018.

**Item 6: 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 2019 be approved by the Annual General Meeting:

Members of the Board of Directors shall receive an annual base fee of:	EUR 31,500
The Chairman of the Board of Directors shall in addition receive:	EUR 24,900
The Deputy Chairman of the Board of Directors shall in addition receive:	EUR 12,450

The remuneration level for the Board of Directors is maintained at the same level as for 2018.

The members of the Board of Directors 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 Annual General Meeting approve the following supplementary fees for 2019 for committee work:

Members of the Audit Committee shall receive an annual supplementary fee of:	EUR 6,700
Members of the Remuneration Committee shall receive an annual supplementary fee of:	EUR 4,700
Members of the Nomination Committee shall receive an annual supplementary fee of:	EUR 3,100
The Chairman of the Audit Committee shall receive an annual supplementary fee of:	EUR 13,500
The Chairman of the Remuneration Committee shall receive an annual supplementary fee of:	EUR 9,000
The Chairman of the Nomination Committee shall receive an annual supplementary fee of:	EUR 6,500

The supplementary fees for committee work are maintained at the same level as for 2018.

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.

With reference to item 10a "Approval of revised Remuneration Policy", the members of the Board of Directors may receive share-based remuneration in the form of restricted share units through an annual grant. The value of restricted share units in each grant may correspond to up to 50% of each Board members' 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. Reference is made to the description of the revised Remuneration Policy under item 10a "Approval of revised Remuneration Policy".

**Item 7: 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, Martin Bonde, Rémi Droller, Sten Verland, Martijn Kleijwegt, Anders Hedegaard, and Catherine Moukheibir.

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 8: Election of auditor**

In accordance with the Audit Committee's recommendation the Board of Directors proposes re-election of ERNST & YOUNG 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 9: Authorization to acquire treasury shares**

No proposal has been made, as in connection with the IPO the Board of Directors was authorized in the period until November 2, 2022 to approve the acquisition of treasury shares, on one or more occasions, with a total nominal value of up to 10% of the share capital of the Company, for so long as the Company's holding of treasury shares after such acquisition does not exceed 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 the acquisition.

**Item 10a: Approval of revised Remuneration Policy**

The Board of Directors has adopted a revised Remuneration Policy for the Company applicable to the Company's Board of Directors and Executive Management.

The revised Remuneration Policy reflects that share-based remuneration can be offered to members of the Board of Directors in the form of restricted share units. The value of restricted share units in each grant may correspond to up 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. The vesting period is proposed to be one year in order to follow the election period for the Board members.

The exercise price for restricted share units granted to members of the Board of Directors will be equal to the market price at the time of grant and accordingly, members of the Board of Directors will only have a benefit of the proposed share-based incentive program if the share price increases after grant thus benefitting all shareholders.

Share-based incentives to Board of Directors are commonly used among competing international biotech and pharmaceutical companies and are proposed to be able to attract and retain qualified members of the Board of Directors and to remain competitive. The Company believes that share-based incentives to the members of the Board of Directors are beneficial to the shareholders' long-term interests as share-based incentives together with the base fee, reflects the objective of a motivated and lasting value creation for the shareholders.

The revised Remuneration Policy also updates the Company's current long-term incentive program for the Executive Management comprising Performance Shares and Matching Shares. The revised Remuneration Policy allows for the Company to continue with the current long-term incentive program for the Executive Management and thus for additional grants of Performance Shares and Matching Shares thereunder.

The Board of Directors proposes that the revised Remuneration Policy be approved by the Annual General Meeting. The revised version of the Remuneration Policy is available on the Company's website at:

<http://phx.corporate-ir.net/phoenix.zhtml?c=254662&p=irol-agm>

#### **Adoption requirements**

All 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 DKK 19,984,799, 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, March 20, 2019**.

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 Annual General Meeting and wish to attend the Annual General Meeting, must request an admission card no later than on **Friday, March 22, 2019**. The registration form is available on the Company's website,

<http://phx.corporate-ir.net/phoenix.zhtml?c=254662&p=irol-agm>

Admission cards may be requested as follows:

- Electronically via VP Investor Services A/S' website, [www.vp.dk/agm](http://www.vp.dk/agm); or
- By completing and forwarding the registration form by letter to VP Investor Services A/S, Weidekampsgade 14, DK-2300 Copenhagen S; or
- By emailing a scanned version to [vpinvestor@vp.dk](mailto:vpinvestor@vp.dk).

Starting this year, 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, <http://phx.corporate-ir.net/phoenix.zhtml?c=254662&p=irol-agm> and click on the link to the Company's Investor Portal;
2. Log on to the Investor Portal with your NemID or VPID; and
3. Enter your email address under "My Profile" and click "Save preferences".

The electronic admission card must be presented at the Annual 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 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**

Subject to having requested an admission card, shareholders may attend the Annual General Meeting in person or by proxy. Proxy forms must be received by VP Investor Services A/S no later than on **Friday, March 22, 2019**. The proxy form is available on the Company's website, <http://phx.corporate-ir.net/phoenix.zhtml?c=254662&p=irol-agm>

Proxies may be submitted as follows:

- Electronically via VP Investor Services A/S' website, [www.vp.dk/agm](http://www.vp.dk/agm); or
- By completing and forwarding the registration form by letter to VP Investor Services A/S, Weidekampsgade 14, DK-2300 Copenhagen S; or
- By emailing a scanned version to [vpinvestor@vp.dk](mailto:vpinvestor@vp.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 VP Investor Services A/S no later than on **Tuesday, March 26, 2019, 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, <http://phx.corporate-ir.net/phoenix.zhtml?c=254662&p=irol-agm>

Voting by correspondence may be submitted as follows:

- Electronically via VP Investor Services A/S' website, [www.vp.dk/agm](http://www.vp.dk/agm); or
- By completing and forwarding the registration form by letter to VP Investor Services A/S, Weidekampsgade 14, DK-2300 Copenhagen S; or
- By emailing a scanned version to [vpinvestor@vp.dk](mailto:vpinvestor@vp.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, <http://phx.corporate-ir.net/phoenix.zhtml?c=254662&p=irol-agm>

- The notice convening the Annual General Meeting, including the agenda and the complete proposals and Appendix 1: Candidates for the Board of Directors
- The 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 2018
- 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, March 27, 2019, 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](mailto: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, <http://phx.corporate-ir.net/phoenix.zhtml?c=254662&p=irol-agm>

**Language**

The Annual 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 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 N, March 5, 2019

The Board of Directors