

Company Announcement No 17/2023 11 October 2023 Lyskjær 8A DK-2730 Herlev www.orphazyme.com CVR: 32266355

Notice convening the Extraordinary General Meeting of Orphazyme A/S

The Board of Directors hereby convenes an Extraordinary General Meeting to be held at the Company's address at Lyskjær 8A, 2730 Herlev on Thursday November 2nd 2023 at 10:00 AM (CEST).

Agenda:

- 1. Proposal submitted by the Board of Directors to change the nominal value of the Company's shares through a reverse share split at a consolidation ratio of 1.000:1
- 2. Proposal submitted by the Board of Directors to complete a reduction of the Company's share capital as part of the implementation of the reverse share split
- 3. Proposal submitted by the Board of Directors to elect KPMG Statsautoriseret Revisionspartnerselskab (CVR no. 25 57 81 98) as independent auditor.
- 4. Authorization to the chairman of the meeting

Complete proposals:

Re item 1 on the agenda:

The Board of Directors proposes to change the nominal value of the Company's shares through a reverse share split at a consolidation ratio of 1.000:1.

The purpose of the reverse share split is to optimize the Company's cost structure by reducing the number of Shareholders. 82% of the Company's current Shareholders owns less than 1,000 shares (less value than DKK 1,000) each, and the issuer costs for the Company associated herewith amounts to more than DKK 400,000 per year. Reference is made to company announcement 16/2023 where the proposal is further accounted for.

The reverse share split implies that every 1,000 of the Company's shares each with a nominal value of DKK 1.00 will be consolidated into 1 new share with a nominal value of DKK 1,000. Shareholdings below 1,000 shares and excess shares following the share consolidation will be redeemed by the Company as described below.

Following the General Meeting's approval of the reverse share split, a mandatory four-week redemption period will initiate. During the redemption period trading in the Company's shares will continue in the ordinary course and Shareholders will have the opportunity to buy and sell existing shares with a nominal value of DKK 1,00 each, including for the purpose of owning enough shares to match the above-mentioned conversion ratios before the redemption and share consolidation takes effect.

After expiration of the four-week redemption period, any Shareholder's holdings of shares below 1,000 shares, as well as any excess shares (calculated on an "after consolidation" basis), will be redeemed by the Company in accordance with the principles of the Danish Companies Act §§ 70 - 72 at a price of DKK 0,94 per share with a nominal value of DKK 1.00. The redemption price is equivalent to the market price determined as the volume weighted average price of the Company's shares over the last 5 trading days during the period 4.-10. October 2023.

The cash payment will be made to each Shareholder's account associated with the respective share deposit.

After the Company's redemption, the redeemed shares with a nominal value of DKK 1.00 each will be consolidated into shares with a nominal value of DKK 1,000 each. The Company will hold these shares with a nominal value of DKK 1,000 as part of the Company's treasury shareholdings until the next General Meeting in the Company, where the Board of Directors will make a proposal pertaining to the handing of the treasury shares aimed at maximizing value for the Shareholders. The excess number of redeemed shares with a nominal value of DKK 1.00 each, that is insufficient to match the consolidation-ratio, will be canceled through a capital reduction, cf. item 2 on the agenda.

Following the completion of the share consolidation, the new shares with a nominal value of DKK 1,000 each will be issued under a new permanent ISIN code in VP SECURITIES A/S ("Euronext Securities") and will be admitted to trading and officially listed on Nasdaq Copenhagen. The expected dates for redemption and issuance in Euronext Securities, as well as the admission to trading and official listing of the new permanent ISIN code on Nasdaq Copenhagen, will be announced in connection with the publication of the statutory four-week redemption notice. After the completion of the share consolidation, all Shareholders will receive an updated statement from Euronext Securities, indicating the new number of shares in their share deposit account each with a nominal value of DKK 1,000.

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The adoption of the Board of Directors proposal will lead to the following amendments to Section 2.1 and Section 6.6. of the articles of association, effective from the expiry of the statutory four-week redemption period:

Section 2.1. will be amended to the following:

"2.1. Selskabets aktiekapital udgør nominelt kr. 35.312.241 fordelt i aktier à nominelt kr. 1.000 eller multipla heraf. /

2.1 The Company's nominal share capital is DKK 35,312,241 divided into shares of DKK 1,000 each or multiples thereof."

and

Section 6.6. will be amended to the following:

"6.6. Hvert aktiebeløb på nominelt kr. 1.000 giver én stemme. En person, der er registeret som indehaver af aktier i selskabet i VP Securities A/S, og som handler i erhvervsmæssig sammenhæng på vegne af andre fysiske eller juridiske personer, herunder indehavere af American Depositary Shares, der repræsenterer aktier i selskabet, kan udøve stemmerettigheder tilknyttet disse aktier på en måde, der ikke er identisk med udøvelsen af stemmerettighederne, der er tilknyttet andre aktier i selskabet, som en sådan person er indehaver af. /

6.6. Each share with a nominal value of DKK 1,000 entitles the holder to one vote. A person registered as a holder of shares in the company with VP Securities A/S, and who trades in a professional capacity on behalf of other natural or legal persons, including holders of American Depositary Shares representing shares in the company, may exercise voting rights associated with these shares in a manner that is not identical to the exercise of voting rights associated with other shares in the company that such a person holds."

Re item 2 on the agenda:

The Board of Directors proposes to complete a reduction of the Company's share capital by a nominal amount of DKK 241 by canceling treasury shares (241 shares with a nominal value of DKK 1.00 each) as part of the implementation of the reverse stock split. The proposal is made with the aim of ensuring that the Company's share capital, after the aforementioned share consolidation, is divisible by DKK 1,000. Consequently, after the share consolidation, there will be a whole number of shares with a nominal value of DKK 1,000.

The treasury shares will be repurchased from the Company's Shareholders for a total amount of DKK 227, in accordance with Section 188, paragraph 1, no. 2 of the Companies Act.

The adoption of the proposal will lead to the following amendment to Section 2.1 of the articles of association, effective from the implementation of the capital reduction and after the expiry of the statutory four-week creditor notification period:

Section 2.1. will be amended to the following:

"2.1. Selskabets aktiekapital udgør nominelt kr. 35.312.000 fordelt i aktier à nominelt kr. 1.000 eller multipla heraf. /

2.1 The Company's nominal share capital is DKK 35,312,000 divided into shares of DKK 1,000 each or multiples thereof."

As the Board of Directors' proposal is submitted as part of the implementation of the aforementioned reverse share split, the proposal shall lapse if the General Meeting does not approve the reverse share split, as described under agenda item 1.

Re item 3 on the agenda:

The Board of Directors proposes that the General Meeting elects KPMG Statsautoriseret Revisionspartnerselskab (CVR no. 25 57 81 98) as new auditor for the Company with immediate effect.

The proposal is motivated by a reduction of complexity in the business. Following the expected final settlement of the US Security class action case, the Company has no commercial activity left and therefore the Company's auditor competences are no more related to a global group with US representation and dual listing but more related to a small business with limited complexity and activity where the choice of auditor should reflect the new situation for the Company.

Re item 4 on the agenda:

The Board of Directors proposes that the General Meeting authorizes the chairman of the General Meeting (with the right of substitution) to apply for registration with the Danish Business Authority (Erhvervsstyrelsen) of the resolutions passed and to make any alteration of or addition to the resolutions passed and the Company's articles of association as may be required to obtain registration or may be suggested as expedient.

Adoption criteria:

Proposal 1, must be passed by at least 9/10 of the votes cast as well as the share capital represented at the General Meeting. Proposal 2, must be passed by at least 2/3 of the votes cast as well as the share capital represented at the General Meeting. Passing of proposals 3 and 4 may be adopted by a simple majority of votes.

Information about capital and voting rights

At the date of the convening notice, the Company's share capital amounts to nominally DKK 35.312.241, divided into shares in the denomination of DKK 1,00 which each confers one vote upon the holder.

Record date:

A Shareholder's right to attend a General Meeting and to vote shall be determined in proportion to the shares held by such Shareholder as of the record date. The shares held by each Shareholder as of the record date shall be determined based on the entry in the shareholders' register regarding the Shareholder's ownership and any notifications to the Company from the Shareholder regarding his ownership not yet recorded in the shareholders' register.

The record date shall be 26 October 2023.

Admission and voting cards, etc.:

Shareholders, proxy holders and any accompanying advisers must present an admission card in order to be admitted to the General Meeting.

Admission cards may be requested as follows:

By submitting a completed, dated and signed registration form to the Company's email contact@orphazyme.com

Admission confirmations will be distributed by e-mail.

Requests for admission cards must reach our e-mail by 27 October 2023, at 11:59 PM (CEST).

Any Shareholder is entitled to attend by proxy and to attend accompanied by an adviser. A proxy may also attend together with an adviser.

The following documents are available on the company's website www.orphazyme.com:

- The convening notice, including the agenda and complete proposals;
- Any other documents to be submitted at the general meeting;
- The forms to be used for voting by proxy or by correspondence; and
- The total number of shares and voting rights as of the convening date.

Questions:

Until one week prior to the General Meeting, Shareholders may submit questions in writing to the Company's Executive Board about decisions to be made at the General Meeting. Shareholders who wish to exercise their right to ask questions may send such questions by letter to the Company. The reply may be made in writing and the answer may be made available on the Company's website, www.orphazyme.com. The Board of directors may refrain from answering if the person asking the question is not represented at the General Meeting.

In addition, Shareholders may ask questions to the Company's Executive Board at the General Meeting regarding the said matters.

Voting by proxy:

Shareholders may vote at the general meeting by a proxy holder presenting a duly signed and dated proxy form. Electronic proxy forms must be issued and filed no later than by 27 October 2023, at 11:59 PM (CEST).

Vote by correspondence:

Shareholders also have the option of voting by correspondence. The relevant vote by correspondence forms to be used for this purpose may be obtained via the Company's website, www.orphazyme.com. A vote sent by ordinary mail must reach the Company by 1 November 2023, at 4:00 PM (CEST). A vote by correspondence cannot be revoked.

Herlev, October 11th, 2023

Yours sincerely,

The Board of Directors

For additional information, please contact:

Orphazyme A/S

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