

1 Name and objects

- 1.1 The Company's name is Orphazyme A/S.
- 1.2 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, within its line of business, participate in partnerships or co-operate with other businesses.

2 Share capital and shares

- 2.1 The Company's nominal share capital is DKK 35,312,000 divided into shares of DKK 1,000 each or multiples thereof.
- 2.2 The share capital has been fully paid up.
- 2.3 The shares shall be issued in the name of the holder and shall be recorded in the name of the holder in the Company's register of shareholders.
- 2.4 The register of shareholders is kept by Computershare A/S, CVR no. 27 08 88 99.
- 2.5 The shares are negotiable instruments. No restrictions shall apply to the transferability of the shares.
- 2.6 No shares shall carry special rights.
- 2.7 No shareholder shall be under an obligation to have his/her shares redeemed in whole or in part by the Company or by any third party.
- 2.8 The shares are registered with and issued in dematerialised form through VP SECURITIES A/S, CVR no. 21 59 93 36. Dividend is paid out through VP SECURITIES A/S and is deposited at the registered dividend accounts at VP SECURITIES A/S. Rights concerning the shares shall be notified to VP SECURITIES A/S in accordance with applicable rules.

3 Increase of share capital

- 3.1 In the period until 25 March 2026, 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 6,989,767. 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.
- 3.1.1 By decision of February 11, 2022, the Board of Directors has partly exercised the authorisation in Article 3.1 to increase the Company's share capital following which a nominal value of DKK 360,000 of the authorisation has been issued.
- 3.2 In the period until 2 November 2022, the Board of Directors is authorised to increase the Company's share capital in one or more issues without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 1,300,000 in connection with the issue of new shares to members of the Board of Directors, executives and/or employees of the Company. The new shares shall be issued against cash payment at a subscription price to be determined by the Board of Directors, which may be below the market price.
- 3.2.1 By decision of 4 March 2019, 27 March 2020, 29 July 2020, 1 February 2021 and 25 February 2021, the Board of Directors has partly exercised the authorisation in Article 3.2 to increase the Company's share capital following which a nominal value of DKK 294,331 of the authorisation has been issued.

- 3.3 In the period until 2 November 2022, the Board of Directors is authorised to increase the Company's share capital in one or more issues without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 15,750,000 in connection with directed issues of bonus shares, and/or directed issues of new shares effected by cash payment, to Kansas Life Sciences Development Inc. and UCL Business PLC (or entities designated by them), respectively. The capital increase shall take place at par value (i.e. below market price). The value of such new shares to be issued can in any case not exceed a maximum of USD 2.5 million with a fixed exchange rate of DKK 6.30 per 1 USD based on the average closing price of the Company's shares on Nasdaq Copenhagen A/S for the 30 days immediately prior to the date of issuance.
- 3.3.1 By decision of 29 January 2018, 31 January 2019, 31 January 2020 and 25 February 2021, the Board of Directors has partly exercised the authorisation in Article 3.3 to increase the Company's share capital following which a nominal value of DKK 80,643 of the authorisation has been issued.
- 3.4 In the period until 25 January 2025, the Board of Directors is authorised to increase the Company's share capital through one or more issues of new shares with pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 25,000,000. 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.
- 3.5 In the period until 25 March 2026, the Board of Directors is authorised to increase the Company's share capital in one or more issues without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 1,300,000 in connection with the issue of new shares to members of the Board of Directors, executives and/or employees of the Company. The new shares shall be issued against cash payment at a subscription price to be determined by the Board of Directors, which may be below the market price.
- 3.6 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.
- 3.7 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 market price as determined by the Board of Directors.
- 3.8 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 or at 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.

- 3.9 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 or at 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.
- 3.10 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.
- 3.11 The authorisations granted to the Board of Directors pursuant to Articles 3.2 and 3.5 can in the aggregate only be exercised to increase the share capital by a maximum nominal amount of DKK 2,000,000.
- 3.12 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.

4 General meeting, venue and notice

- 4.1 The general meetings of the Company shall be held in the Capital Region of Denmark.
- 4.2 The annual general meeting of the Company shall be held each year in due time for the audited and approved annual report to be received by the relevant authorities before the applicable statutory time limit. The Company shall no later than eight weeks before the contemplated date of the annual general meeting publish the date of the general meeting and the deadline for submitting requests for specific proposals to be included on the agenda.
- 4.3 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.
- 4.4 Extraordinary general meetings shall be held when determined by the Board of Directors or requested by the Company's auditor. Furthermore, an extraordinary general meeting shall be held when requested by shareholders possessing no less than five per cent of the share capital. Such request shall be submitted in writing to the Board of Directors and be accompanied by a specific proposal for the business to be transacted. The Board of Directors convenes an extraordinary general meeting no later than two weeks after such request has been made.
- 4.5 General meetings shall be convened by the Board of Directors with at least three weeks' and not more than five weeks' notice. The notice shall be published on the Company's website. Furthermore, a notice of the general meeting shall be sent to all shareholders recorded in the Company's register of shareholders who have so requested.

- 4.6 For a period of three weeks prior to the general meeting, including the date of the general meeting, the following information shall be available on the Company's website:
- a. The notice convening the general meeting
 - b. The aggregate number of shares and voting rights as at the date of the notice
 - c. The documents to be presented at the general meeting
 - d. The agenda and the complete proposals as well as, for annual general meetings, the audited annual report
 - e. The forms to be used for voting by proxy or by postal vote
- 4.7 General meetings shall be held in English. The Board of Directors may decide to offer simultaneous interpretation into Danish. Documents prepared in connection with or following a general meeting shall be in English and to the extent required by law or if decided by the Board of Directors, in Danish.
- 4.8 The general meeting shall be presided over by a chairman elected by the Board of Directors who shall ensure that the general meeting is conducted in a proper and efficient manner.

5 Agenda for the annual general meeting

- 5.1 The agenda for the annual general meeting shall include the following:
- a. The Board of Directors' report on the Company's activities in the past financial year
 - b. Presentation and adoption of the annual report
 - c. Distribution of profit or covering of loss according to the adopted annual report
 - d. Resolution to grant discharge of liability to the Board of Directors and the Executive Management
 - e. Presentation of the remuneration report
 - f. Approval of remuneration of the Board of Directors for the current financial year
 - g. Election of members to the Board of Directors
 - h. Election of auditor
 - i. Authorisation to acquire treasury shares
 - j. Any proposals from the Board of Directors or shareholders
 - k. Any other business
- 5.2 Every shareholder shall be entitled to have a specific subject considered at the annual general meeting. Such proposals must be submitted in writing to the Board of Directors not later than six weeks prior to the annual general meeting.

6 Shareholders' attendance and voting rights at the general meeting

- 6.1 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 one week prior to the general meeting. The shares held by each shareholder at the record date are calculated based on the registration of the number of shares held by that 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 have not yet been registered.
- 6.2 A shareholder who is entitled to attend the general meeting pursuant to Article 6.1 and who wants to attend the general meeting shall notify the Company of his/her attendance not later than three days prior to the date of the general meeting.
- 6.3 A shareholder may attend in person or by proxy, and the shareholder or the proxy may attend together with an adviser.
- 6.4 The right to vote may be exercised by a written and dated instrument of proxy in accordance with applicable laws.
- 6.5 A shareholder who is entitled to participate in the general meeting pursuant to Article 6.1 may vote by postal vote in accordance with the provisions of the Danish Companies Act. Such postal votes shall be received by the Company not later than the business day before the general meeting. Postal votes cannot be withdrawn.
- 6.6 Each share of the nominal value of DKK 1,000 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.

7 Resolutions at general meetings

- 7.1 Resolutions by the general meeting shall be passed by a simple majority of votes cast unless otherwise prescribed by law or by these Articles of Association.
- 7.2 Adoption of changes to these Articles of Association, dissolution of the Company, merger or demerger requires that the decision is adopted with at least 2/3 of the votes cast as well as the share capital represented at the general meeting, unless applicable laws prescribe stricter or less strict adoption requirements or applicable laws confer specific authority to the Board of Directors or other bodies.

8 Board of Directors

- 8.1 The Board of Directors consists of not less than three and not more than six members elected by the general meeting.
- 8.2 The members of the Board of Directors elected by the general meeting are elected for a term of one year. Re-election of board members may take place.
- 8.3 The Board of Directors elects a Chairman and, if so decided by the Board of Directors, a Deputy Chairman among its members. If the Chairman of the Board of Directors resigns during a term of election, the Deputy Chairman (if elected) shall take up the position as Chairman until a new Chairman is elected among the members of the Board of Directors.
- 8.4 Any employee representatives on the Board of Directors and their alternates, if any, are elected in accordance with applicable law thereon in force from time to time.

- 8.5 Resolutions of the Board of Directors are passed by simple majority. In the event of equal votes, the Chairman or, in his/her absence, the Deputy Chairman shall have a casting vote.
- 8.6 The Board of Directors forms a quorum when more than half of its members are represented, including the Chairman or the Deputy Chairman.

9 Executive Management

- 9.1 The Board of Directors appoints an Executive Management consisting of one to three members to be in charge of the day-to-day management of the Company.
- 9.2 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.

10 Rules of signature

10.1 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.

11 Electronic communication

11.1 All communication from the Company to the individual shareholders, including notices convening general meetings, may take place electronically by posting on the Company's website or by email. General notices shall be published on the Company's website and in such other manner as may be prescribed by applicable laws. The Company may as an alternative choose to send notices, etc., by ordinary post.

11.2 Communication from a shareholder to the Company may take place by email or by ordinary post.

11.3 Each shareholder is responsible for ensuring that the Company has the correct email address at all times. The Company is not obliged to verify such contact information or to send notices in any other way.

11.4 The Company's website, www.orphazyme.com, contains information about system requirements and electronic communication procedures.

11.5 Company announcements shall be prepared in English and, if decided by the Board of Directors, in Danish.

12 Annual report

12.1 The Company's annual accounts shall be audited by a state-authorized public accountant elected by the general meeting for a one-year term. Re-election may take place to the extent permitted under applicable law.

12.2 Annual reports shall be prepared in English and, if decided by the Board of Directors, in Danish.

13 Financial year

13.1 The Company's financial year is the calendar year.

As adopted at the Company's extraordinary general meeting held on 2 November 2017 and subsequently amended by the Board of Directors on 6 November 2017, on 20 November 2017, on 29 January 2018, on 31 January 2019, on 4 March 2019, on the Company's extraordinary general meeting held on 25 January 2020, by the Board of Directors on 31 January 2020 and on 6 February 2020, on the Company's annual general meeting held on 26 March 2020 and by the Board of Directors on 27 March 2020 and on 29 July 2020, on the Company's extraordinary general meeting held on 21 September 2020, and by the Board of Directors on 28 September 2020, on 1 February 2021, on 25 February 2021 and on 24 March 2021, on the Company's annual general meeting held on 25 March 2021, by the Board of Directors on 11 February 2022, on the Company's extraordinary general meeting held on 15 February 2022, on the Company's annual general meeting held on 29 June 2022 and on the Company's extraordinary general meeting held on 2 November 2023