

PROSPECTUS SUPPLEMENT
(To Prospectus dated October 22, 2021)

Up to \$50,000,000



Orphazyme A/S

Ordinary Shares

(including Ordinary Shares represented by American Depositary Shares)

We have entered into a sales agreement with Cowen and Company, LLC, or Cowen, relating to American Depositary Shares, or ADSs, representing ordinary shares of Orphazyme A/S offered by this prospectus supplement and the accompanying prospectus. Each ADS represents the right to receive one ordinary share. In accordance with the terms of the sales agreement, we may offer and sell ADSs having an aggregate offering price of up to \$50,000,000 from time to time through Cowen acting as our agent.

ADSs representing our ordinary shares are listed on The Nasdaq Global Select Market under the symbol "ORPH." Our ordinary shares are listed on Nasdaq Copenhagen A/S, or Nasdaq Copenhagen, under the symbol "ORPHA." On November 2, 2021, the last reported sale price of ADSs, as reported on The Nasdaq Global Select Market, was \$4.09 per ADS. The market prices of the ADSs have recently experienced extreme volatility in price and trading volume. From June 1, 2021 to November 2, 2021, the sale price of ADSs on The Nasdaq Global Select Market ranged from as low as \$3.78 to as high as \$77.77 and daily trading volume ranged from 13,400 shares to 168,470,900 shares. Investors that purchase ADSs in this offering may lose a significant portion of their investments if the price of our common stock subsequently declines. Please see the section of this prospectus supplement titled "Risk Factors."

Sales of the ADSs, if any, under this prospectus supplement and the accompanying prospectus may be made in sales deemed to be an "at the market offering" as defined in Rule 415(a)(4) promulgated under the Securities Act of 1933, as amended, or the Securities Act. Cowen is not required to sell any specific amount of securities, but will act as our sales agent using commercially reasonable efforts consistent with its normal trading and sales practices, on mutually agreed terms between Cowen and us. There is no arrangement for funds to be received in any escrow, trust or similar arrangement.

The compensation to Cowen for sales of ADSs sold pursuant to the sales agreement will be an amount equal to 3.0% of the gross proceeds of any ADSs sold under the sales agreement. See "Plan of Distribution" beginning on page S-41 for additional information regarding the compensation to be paid to Cowen. In connection with the sale of the ADSs on our behalf, Cowen will be deemed to be an "underwriter" within the meaning of the Securities Act and the compensation of Cowen will be deemed to be underwriting commissions or discounts. We have also agreed to provide indemnification and contribution to Cowen with respect to certain liabilities, including liabilities under the Securities Act or the Exchange Act of 1934, as amended, or the Exchange Act.

We are an "emerging growth company" as that term is used in the Jumpstart Our Business Startups Act of 2012 and, as such, have elected to comply with certain reduced public company reporting requirements for this prospectus supplement and future filings. See "Prospectus Supplement Summary—Implications of Being an Emerging Growth Company" and "Prospectus Supplement Summary—Implications of Being a Foreign Private Issuer" for additional information.

Investing in these securities involves a high degree of risk. You should review carefully the risks and uncertainties described under the heading "[Risk Factors](#)" on page S-7 of this prospectus supplement, on page 3 of the accompanying prospectus and under similar headings in the other documents that are incorporated by reference into this prospectus supplement and the accompanying prospectus.

None of the Securities and Exchange Commission, any state securities commission, the Danish Financial Supervisory Authority, nor any other foreign securities commission has approved or disapproved of these securities or determined if this prospectus supplement and the accompanying prospectus are truthful or complete. Any representation to the contrary is a criminal offense.

Cowen

The date of this prospectus supplement is November 4, 2021.

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ABOUT THIS PROSPECTUS SUPPLEMENT

This prospectus supplement and the accompanying prospectus is part of a registration statement on Form F-3 that we filed with the U.S. Securities and Exchange Commission, or the SEC, using a “shelf” registration process. Under the shelf registration process, we may offer ordinary shares (including ordinary shares represented by ADSs) having an aggregate offering price of up to \$75,000,000. Under this prospectus supplement and the accompanying prospectus, we may offer shares of our ordinary shares (including ordinary shares represented by ADSs) having an aggregate offering price of up to \$50,000,000 from time to time at prices and on terms to be determined by market conditions at the time of offering. This prospectus supplement describes the specific terms of this offering of ADSs and also adds to and updates the documents incorporated by reference into this prospectus supplement.

This prospectus supplement, the accompanying prospectus and the documents incorporated by reference herein and therein include important information about us, the securities being offered and other information you should know before investing in our securities. To the extent there is a conflict between the information contained in this prospectus supplement, on the one hand and the accompanying prospectus that was filed with the SEC before the date of this prospectus supplement, on the other hand, you should rely on the information in this prospectus supplement. If any statement in one of these documents is inconsistent with a statement in another document having a later date (for example, a document incorporated by reference in this prospectus supplement), the statement in the document having the later date modifies or supersedes the earlier statement. Any statement so modified or superseded will not be deemed, except as so modified or superseded, to constitute a part of this prospectus supplement.

You should rely only on the information contained in or incorporated by reference in this prospectus supplement and the accompanying prospectus and in any free writing prospectus supplement that we authorized for use in connection with this offering. We have not, and the sales agent has not, authorized anyone to provide you with different information. If anyone provides you with different or inconsistent information, you should not rely on it. We are not, and the sales agent is not, making an offer to sell these securities in any jurisdiction where the offer or sale is not permitted. You should assume that the information appearing in this prospectus supplement and the accompanying prospectus, the documents incorporated by reference in this prospectus supplement and the accompanying prospectus and in any free writing prospectus supplement that we have authorized for use in connection with this offering, is accurate only as of the date of those respective documents. Our business, financial condition, results of operations and prospects may have changed since those dates. You should read this prospectus supplement and the accompanying prospectus, the documents incorporated by reference in this prospectus supplement and the accompanying prospectus, and any free writing prospectus that we have authorized for use in connection with this offering, in their entirety before making an investment decision.

We further note that the representations, warranties and covenants made by us in any agreement that is filed as an exhibit to any document that is incorporated by reference in this prospectus supplement and the accompanying prospectus were made solely for the benefit of the parties to such agreement, including, in some cases, for the purpose of allocating risk among the parties to such agreements, and should not be deemed to be a representation, warranty or covenant to you. Moreover, such representations, warranties or covenants were accurate only as of the date when made. Accordingly, such representations, warranties and covenants should not be relied on as accurately representing the current state of our affairs.

Unless otherwise indicated or the context otherwise requires, all references in this prospectus supplement and the accompanying prospectus to the terms “Orphazyme,” “the Company,” “we,” “us” and “our” refer to Orphazyme A/S and its wholly owned subsidiaries. In this prospectus supplement and the accompanying prospectus, any reference to any provision of any legislation shall include any amendment, modification, re-enactment or extension thereof. Words importing the singular shall include the plural and vice versa, and words importing the masculine gender shall include the feminine or neutral gender. All references to “shares” in

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this prospectus supplement and the accompanying prospectus refer to ordinary shares of Orphazyme A/S with a nominal value of DKK 1 per share.

For investors outside the United States: We have not done anything that would permit the offering or possession or distribution of this prospectus supplement and the accompanying prospectus in any jurisdiction where action for that purpose is required, other than in the United States. Persons outside the United States who come into possession of this prospectus supplement and the accompanying prospectus must inform themselves about, and observe any restrictions relating to, the offering of the securities described herein and the distribution of this prospectus supplement and the accompanying prospectus outside the United States.

We are incorporated in Denmark, and many of our outstanding securities are owned by non-U.S. residents. Under the rules of the SEC, we are currently eligible for treatment as a “foreign private issuer.” As a foreign private issuer, we are not required to file periodic reports and financial statements with the SEC as frequently or as promptly as domestic registrants whose securities are registered under the Exchange Act.

TRADEMARKS

This prospectus supplement, the accompanying prospectus and the information incorporated by reference herein and therein include trademarks, tradenames and service marks, certain of which belong to us and others that are the property of other organizations. Solely for convenience, trademarks and tradenames referred to in this prospectus supplement appear without the ® and ™ symbols, but the absence of those references is not intended to indicate, in any way, that we will not assert our rights or that the applicable owner will not assert its rights to these trademarks and tradenames to the fullest extent under applicable law. We do not intend our use or display of other parties’ trademarks, trade names or service marks to imply, and such use or display should not be construed to imply, a relationship with, or endorsement or sponsorship of us by, these other parties.

MARKET AND INDUSTRY DATA

This prospectus supplement includes statistical and other industry and market data that we obtained from industry publications and research, surveys and studies conducted by third parties, as well estimates by our management based on such data. Management estimates are derived from publicly available information, our knowledge of our industry and assumptions based on such information and knowledge, which we believe to be reasonable. The market data and estimates used in this prospectus supplement involve a number of assumptions and limitations, and you are cautioned not to give undue weight to such data and estimates. We believe that the information from these industry publications, surveys and studies is reliable. The industry in which we operate is subject to a high degree of uncertainty and risk due to a variety of important factors, including those described in the sections titled “Risk Factors” and “Special Note Regarding Forward-Looking Statements” in this prospectus supplement and the accompanying prospectus and under similar sections contained in other documents that are incorporated by reference herein. These and other factors could cause results to differ materially from those expressed in the estimates made by the independent parties and by us.

PRESENTATION OF FINANCIAL INFORMATION

We maintain our books and records in Danish kroner and we prepare our audited consolidated financial statements in accordance with International Financial Reporting Standards, or IFRS, as issued by the International Accounting Standards Board, or IASB. None of the consolidated financial statements incorporated by reference into this prospectus supplement were prepared in accordance with accounting principles generally accepted in the United States, or U.S. GAAP. All references in this prospectus supplement to “\$” are to U.S. dollars, to “DKK” are to Danish kroner and to “€” are to the Euro.

PROSPECTUS SUPPLEMENT SUMMARY

This summary does not contain all of the information that may be important to you in making your investment decision. In addition to this summary, we urge you to read the entire prospectus supplement and the accompanying prospectus carefully, especially the risks of investing in the ordinary shares and ADSs discussed under “Risk Factors” beginning on page S-7 of this prospectus supplement, the accompanying prospectus and under similar headings in the other documents that are incorporated by reference into this prospectus supplement and the accompanying prospectus before deciding whether to invest in the ordinary shares and ADSs. You should also carefully read the information incorporated by reference into this prospectus supplement and the accompanying prospectus, including our financial statements, and the exhibits to the registration statement of which this prospectus supplement is a part.

Overview

We are a late-stage biopharmaceutical company committed to developing a new therapeutic option for people living with Niemann-Pick disease type C, or NPC, a rare, genetic, progressively debilitating and often fatal neurodegenerative disease. In November 2020, we submitted a marketing authorization application, or MAA, for our investigational product candidate, arimoclomol, in NPC to the European Medicines Agency, or EMA, and expect an opinion from the Committee for Human Medicinal Products, or CHMP, in the first quarter of 2022. We had also previously submitted a new drug application, or NDA, to the U.S Food and Drug Administration, or FDA, for arimoclomol in NPC. In June 2021, we received a complete response letter, or CRL, from the FDA following its review of the NDA, and in October 2021, we held a Type-A meeting with the FDA to assess a path forward for arimoclomol for NPC in the U.S. In the meeting, the FDA recommended that we submit additional data, information, and analyses to address certain topics in the CRL and engage in further interactions with the FDA to identify a pathway to resubmission of the NDA. The FDA also concurred with our proposal to remove the cognition domain from the NPCCSS endpoint, with the result that the primary endpoint is permitted to be recalculated using the 4-domain NPCCSS, subject to the submission of additional requested information, including performance-based validity evidence for selected domains inclusive of ambulation, speech and fine motor skills. The FDA did not agree with our proposal to rescore the swallow domain as additional information is required for consideration, such as documentation related to clarity and standardization in collection of patient experience data. The FDA also requested additional information to discuss our proposal to use a while-on-treatment estimand of the log transformed ratio to address the timing of both the treatment comparison and potential intercurrent events. We intend to work with the FDA to provide such information as requested by the FDA, but the FDA may still require us to conduct additional studies or clinical trials.

Arimoclomol is an orally- or naso/gastrically-administered small molecule that crosses the blood-brain barrier and is designed to selectively amplify the natural role of endogenous HSPs, which protect against cellular toxicity caused by protein misfolding, aggregation and lysosomal dysfunction. In our Phase 2/3 clinical trial of arimoclomol in NPC, we have observed evidence of slowing of disease progression. We also believe that arimoclomol has been well tolerated in clinical trials including more than 500 human subjects for various indications.

Risks Associated With Our Business

Our business is subject to numerous risks and uncertainties, including those highlighted in the section titled “Risk Factors” of this prospectus supplement immediately following this prospectus supplement summary, the accompanying prospectus and in the other documents incorporated by reference into this prospectus supplement and the accompanying prospectus. Some of these risks are:

- We have not received approval for any product candidate for commercial sale and, as a result, we have never generated any significant revenue and have incurred significant financial losses, and expect to continue to incur significant financial losses in the future, which makes it difficult to assess our future viability.

- We will require additional capital in the future, which may not be available to us on commercially favorable terms, or at all.
- Our business, operations and clinical development plans and timelines could be adversely affected by the effects of health epidemics, including the ongoing COVID-19 pandemic, on the manufacturing, clinical trial and other business activities performed by us or by third parties with whom we conduct business, including our contract manufacturers, contract research organizations, or CROs, shippers and others.
- Clinical trials being conducted to test our product candidate, arimoclomol, may not obtain the desired safety and efficacy results or may be delayed or more costly than anticipated. In addition, our completed clinical trials have been small, each with less than 100 persons; in larger clinical trials, additional risks, including safety risks or lack of efficacy, may materialize.
- As we have focused our efforts on the development of arimoclomol, we are currently highly dependent on obtaining and maintaining regulatory approval for arimoclomol and the potential success of this one product candidate.
- Because we are developing arimoclomol for the treatment of diseases in which there is little clinical experience, the FDA or other regulatory authorities may not consider the endpoints of our clinical trials to predict or provide clinically meaningful results.
- Arimoclomol may be shown to cause undesirable side effects or other adverse events that could delay or prevent its regulatory approval, limit its commercial profile or result in significant negative consequences following regulatory approval, if such approval is granted.
- Fast track, orphan drug, and breakthrough therapy designation by the FDA and EMA may not actually lead to a faster development or regulatory review or approval process and does not assure or increase the likelihood of FDA or EMA approval of arimoclomol. For instance, we received a complete response letter from the FDA following its review of our NDA for arimoclomol in NPC.
- Even if arimoclomol receives marketing approval, we may not be successful in our commercialization efforts and arimoclomol may fail to achieve the degree of market acceptance by physicians, patients, healthcare payors and others in the medical community necessary for commercial success.
- If we are unable to achieve and maintain adequate levels of coverage or reimbursement for arimoclomol, if commercialized, or any future product candidates we may seek to commercialize, or if patients are left with significant out-of-pocket costs, our commercial success may be severely hindered.
- We are currently dependent on third parties for manufacturing arimoclomol. If such third-party manufacturers do not deliver their manufactured products in time, this could have a material adverse effect on our business.
- Our business operations and current and future relationships with healthcare professionals, principal investigators, consultants, customers and third-party payors in the United States and elsewhere may be subject, directly or indirectly, to applicable anti-kickback, fraud and abuse, false claims, physician payment transparency, health information privacy and security and other healthcare laws and regulations, which could expose us to substantial penalties.
- If we are unable to obtain and maintain our marketing and distribution rights for arimoclomol, as well as patent protection for our technology and current or future product candidates, if we are unable to obtain or maintain orphan drug designation, if we are unable to benefit from orphan drug exclusivity, or if the scope of the marketing and distribution rights or patent protection obtained is not sufficiently broad, we may not be able to compete effectively in our markets.
- The trading price of our equity securities may be volatile due to factors beyond our control..

Implications of Being an Emerging Growth Company

As a company with less than \$1.07 billion in revenue during our last fiscal year, we qualify as an “emerging growth company,” as defined in the U.S. Jumpstart Our Business Startups Act of 2012, or JOBS Act. An emerging growth company may take advantage of specified reduced reporting and other burdens that are otherwise applicable generally to public companies. These provisions include an exemption from the auditor attestation requirement in the assessment of our internal control over financial reporting pursuant to the Sarbanes-Oxley Act of 2002, or Sarbanes-Oxley Act.

We may choose to take advantage of some but not all of these provisions, and therefore the information that we provide holders of ordinary shares and ADSs may be different than the information you might receive from other public companies in which you hold equity. In addition, Section 107 of the JOBS Act also provides that an emerging growth company can take advantage of an extended transition period for complying with new or revised accounting standards applicable to public companies in the United States. As a public company in Denmark, we are unable to take advantage of the extended transition period.

We may take advantage of these provisions for up to five years from the initial public offering of our ADSs or such earlier time that we are no longer an emerging growth company. We will cease to be an emerging growth company upon the earliest of the following:

- the last day of the first fiscal year in which our annual revenues were at least \$1.07 billion;
- the last day of the fiscal year following the fifth anniversary of the initial public offering of ADSs;
- the date on which we have issued more than \$1 billion of non-convertible debt securities over a three-year period; and
- the last day of the fiscal year during which we meet the following conditions: (i) the worldwide market value of our common equity securities held by non-affiliates as of our most recently completed second fiscal quarter is at least \$700 million, (ii) we have been subject to U.S. public company reporting requirements for at least 12 months and (iii) we have filed at least one annual report as a U.S. public company.

Implications of Being a Foreign Private Issuer

We are also considered a “foreign private issuer” under U.S. securities laws. In our capacity as a foreign private issuer, we are exempt from certain rules under the U.S. Securities Exchange Act of 1934, as amended, or the Exchange Act, that impose certain disclosure obligations and procedural requirements for proxy solicitations under Section 14 of the Exchange Act. In addition, our officers, directors and principal shareholders are exempt from the reporting and “short-swing” profit recovery provisions of Section 16 of the Exchange Act and the rules under the Exchange Act with respect to their purchases and sales of our securities. Moreover, we are not required to file periodic reports and financial statements with the SEC as frequently or as promptly as U.S. companies whose securities are registered under the Exchange Act. In addition, we are not required to comply with Regulation FD, which restricts the selective disclosure of material information.

Both foreign private issuers and emerging growth companies are also exempt from certain more stringent executive compensation disclosure rules for public companies in the United States under the Dodd-Frank Wall Street Reform and Consumer Protection Act of 2010. Even if we no longer qualify as an emerging growth company, so long as we remain a foreign private issuer, we will continue to be exempt from such compensation disclosures.

We may take advantage of these exemptions until such time as we are no longer a foreign private issuer. We will remain a foreign private issuer until such time that more than 50% of our outstanding voting securities are

held by U.S. residents and any of the following three circumstances applies: (1) the majority of our executive officers or directors are U.S. citizens or residents; (2) more than 50% of our assets are located in the United States; or (3) our business is administered principally in the United States.

Corporate History and Information

We were incorporated on June 19, 2009 as a private limited liability company under Danish law and later converted into a Danish public limited liability company on October 20, 2017. We are registered with the Danish Business Authority (Erhvervsstyrelsen) in Copenhagen, Denmark under company registration number (CVR) no. 32266355. We were publicly listed on Nasdaq Copenhagen, Denmark, in November 2017 under the symbol “ORPHA” and on The Nasdaq Global Select Market, United States, in September 2020 under the symbol “ORPH.”

Our headquarters and principal executive offices are located at Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark, and our telephone number is +45 39 17 82 72. Our website address is www.orphazyme.com. The information contained on, or accessible through, our website is not incorporated by reference into this prospectus supplement or the accompanying prospectus, and you should not consider any information contained in, or that can be accessed through, our website as part of this prospectus supplement or the accompanying prospectus or in deciding whether to purchase ordinary shares, including ordinary shares in the form of ADSs. We have included our website address as an inactive textual reference only.

THE OFFERING

Securities offered by us	ADSs representing our ordinary shares having an aggregate offering price of up to \$50,000,000.
Plan of Distribution	“At the market offering” that may be made from time to time through our sales agent, Cowen. See “Plan of Distribution” on page S-41.
ADSs	<p>Each ADS represents one ordinary share, nominal value DKK 1 per share. The offered ADSs may be evidenced by American Depositary Receipts, or ADRs. The depositary will hold ordinary shares underlying your ADSs. As an ADS holder, you will not be treated as one of our shareholders, you will not have shareholder rights and you may not be able to exercise your right to vote the shares underlying your ADSs. You will have the contractual rights of an ADS holder, as provided in the deposit agreement among us, the depositary and holders and beneficial owners of ADSs from time to time. ADS holders may only exercise voting rights with respect to the shares underlying the ADSs in accordance with the provisions of the deposit agreement, which will provide that a holder may vote the shares underlying any ADSs for any particular matter to be voted on by our shareholders either by withdrawing the shares underlying the ADSs or by instructing the depositary how to vote those shares. Our articles of association permit differentiated voting, allowing the depositary to vote the shares registered in its name that underlie the ADSs in a manner that is not identical. As a result, the depositary will be able to vote such shares in a manner to reflect the preferences of the ADS holders, thereby effectively permitting pass-through voting by ADS holders who indicate their preference to the depositary in accordance with and subject to the depositary’s procedures. The depositary will try, to the extent practical, to vote the shares underlying the ADSs as instructed by the ADS holders.</p> <p>You may surrender your ADSs to the depositary for cancellation in exchange for ordinary shares. The depositary will charge you fees for any cancellation.</p> <p>We may amend or terminate the deposit agreement without your consent. If you continue to hold your ADSs after an amendment to the deposit agreement, you agree to be bound by the deposit agreement as amended.</p> <p>To better understand the terms of the ADSs, you should carefully read the “Description of American Depositary Shares” section of this prospectus supplement and the deposit agreement incorporated by reference into the registration statement of which this prospectus supplement forms a part.</p>
ADS Depositary	The Bank of New York Mellon.

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Use of Proceeds	<p>We intend to use the net proceeds from the offering, if any, together with our existing cash, to continue the regulatory approval process for and fund the commercial launch, if approved, of arimoclomol for the treatment of NPC and for working capital and general corporate purposes.</p> <p>See “Use of Proceeds” for a more complete description of the intended use of proceeds from this offering.</p>
Dividend Policy	<p>We do not expect to pay dividends in the foreseeable future. If, however, we declare dividends on our ordinary shares, the depositary will distribute the cash dividends and other distributions it receives on our ordinary shares after deducting its fees and expenses in accordance with the terms set forth in the deposit agreement.</p>
Risk Factors	<p>See “Risk Factors” and the other information included in this prospectus supplement, the accompanying prospectus and in the documents incorporated by reference into this prospectus supplement and the accompanying prospectus for a discussion of factors you should carefully consider before deciding to invest in the ADSs.</p>
Nasdaq Global Select Market symbol	“ORPH.”

The number of ordinary shares issued and outstanding is based on 34,952,241 of our ordinary shares outstanding as of June 30, 2021, and excludes:

- subject to certain vesting criteria being satisfied, up to 695,723 ordinary shares that may be issued to cover the delivery of ordinary shares to participants of our long-term incentive program, or the LTIP, as of June 30, 2021;
- up to 705,976 ordinary shares underlying unvested or unexercised restricted share units, or RSUs, as of June 30, 2021; and
- bonus shares that we have agreed to issue pursuant to a license agreement with the University of Kansas and UCL Business PLC, as described further in the documents incorporated by reference into this prospectus supplement.

RISK FACTORS

Investing in our ordinary shares and the ADSs involves a high degree of risk. Before you invest in the ordinary shares or ADSs, you should consider carefully the risks described below and discussed in our Annual Report on Form 20-F for the year ended December 31, 2020 filed on March 2, 2021 and reports on Form 6-K, which are incorporated by reference in this prospectus supplement and the accompanying prospectus, together with other information in this prospectus supplement and the accompanying prospectus and the information and documents incorporated by reference in this prospectus supplement and the accompanying prospectus, and any free writing prospectus that we have authorized for use in connection with this offering before you make a decision to invest in the ADSs. If any of the following risks actually occurs, our business, results of operations, cash flows, financial condition and/or prospects could suffer materially. In such event, the trading price of the ordinary shares and ADSs could decline, which would cause you to lose all or part of your investment. Please also see “Special Note Regarding Forward-Looking Statements in this prospectus supplement and the accompanying prospectus.”

Risks Related to Development of Our Product Candidates

As we have focused our efforts on the development of arimoclomol, we are currently highly dependent on obtaining and maintaining regulatory approval for arimoclomol and the potential success of this one product candidate.

To date, we have focused substantially all of our efforts on the development of arimoclomol. We are currently conducting preclinical studies and clinical trials based on the arimoclomol molecule. In September 2020, the FDA accepted our NDA for arimoclomol for NPC with priority review and set a target action date of March 17, 2021 under the PDUFA to complete its review of our NDA. On September 24, 2020, we received a filing communication from the FDA in connection with our NDA for arimoclomol for the treatment of NPC in which the FDA summarized six potential review issues, including the FDA’s continuing evaluation of the integrity of data from our Phase 2/3 trial for NPC; the effect of the high degree of concomitant miglustat use in our Phase 2/3 trial for NPC on its ability to determine the of safety and efficacy of arimoclomol which could have potential implications for labeling/recommended dosing and post-marketing studies; the proposed primary hypothetical treatment effect used in our Phase 2/3 trial for NPC to estimate the treatment benefit effect; the meaningfulness of one metric utilized to evaluate patient progress in our Phase 2/3 trial for NPC; a potential QT safety signal; and differences among the formulations of arimoclomol used in our Phase 2/3 trial of NPC as compared to the formulation of arimoclomol to be marketed. The filing communication constituted preliminary notice from the FDA of potential review issues as part of its ordinary course review of our NDA and was not necessarily indicative of deficiencies identified during the review. On December 27, 2020, we were notified by FDA that the PDUFA target action date for our NDA had been extended by three months, to June 17, 2021. On June 17, 2021, the FDA issued a Complete Response Letter, or CRL, from the FDA following its review of the NDA. The FDA issued the CRL based on needing additional qualitative and quantitative evidence to further substantiate the validity and interpretation of the 5-domain NPC Clinical Severity Scale (NPCCSS) and, in particular, the swallow domain, in the context of the FDA’s preferred and recommended statistical approach. Further, the FDA noted in the CRL that additional data is needed to supplement confirmatory evidence beyond the single Phase 2/3 trial for NPC to address the weak and contradictory evidence, problematic hypothetical estimand and lack of statistical evidence on the 5-domain NPCCSS to support the benefit-risk assessment of the NDA. On October 13, 2021, the Company held a Type A meeting with the FDA to discuss the key topics in the CRL. Although the FDA stated that the additional rationale and reanalysis we presented to the FDA at this meeting were insufficient to address the deficiencies outlined in the CRL, the FDA recommended that we submit additional data, information and analyses to address the deficiencies noted in the CRL and engage in further interactions with the FDA to identify a pathway to resubmission of the NDA. With regard to the confirmatory evidence, the FDA recommended that we submit additional information, such as in vivo or PD biomarker data as well as PK data, that demonstrates arimoclomol target engagement and/or modulation of pathological processes relevant to NPC and assess the relationship between arimoclomol exposure and PD biomarker response. The

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FDA concurred with our proposal to remove the cognition domain from the NPCCSS endpoint, with the result that the primary endpoint is permitted to be recalculated using the 4-domain NPCCSS, subject to the submission of additional requested information, including performance-based validity evidence for selected domains inclusive of ambulation, speech and fine motor skills. The FDA did not agree with our proposal to rescore the swallow domain because, in the FDA's view, our submission lacked previously requested evidence, protocols and supporting materials to establish how the scores were initially assigned, and the FDA has requested additional information for consideration, such as documentation related to clarity and standardization in collection of patient experience data. The FDA has also requested additional information to discuss our proposal to use a while-on-treatment estimand of the log transformed ratio to address the timing of both the treatment comparison and potential intercurrent events. We intend to work with the FDA to provide such information as requested by the FDA, but the FDA may still require us to conduct additional studies or clinical trials. We are considering the optimal path forward, which could entail a material extension of the time and cost required for resubmission of the NDA. We cannot assure you that the aforementioned issues or any additional issues that may arise in the course of subsequent discussions with the FDA will not result in a delay of any potential approval of arimoclomol for the treatment of NPC by the FDA or a determination by the FDA not to approve the product candidate for marketing in the United States. Further, as part of its review process, the FDA may request additional information and data, require us to make modifications to ongoing NPC-related clinical trials, manufacturing or other processes, run additional studies or clinical trials or incur significant additional expenditures in order to obtain such approval. If arimoclomol does not obtain approval for the indications we are currently exploring, we will have spent substantial time and financial resources without receiving a return on investment. As a result, if arimoclomol does not become a success, this will have a material adverse effect on our business, results of operations, cash flows, financial condition and/or prospects.

Risks Related to this Offering

Investors in the offering will experience immediate and substantial dilution in the book value of their investment.

The price per ADS being offered may be higher than the net tangible book value per ADS outstanding prior to this offering. Accordingly, if you invest in the ordinary shares or ADSs in the offering, you will incur immediate substantial dilution of \$2.51 per ADS (based on the net tangible book value per share underlying the ADSs), based on an assumed offering price of \$4.09 per ADS, and our pro forma net tangible book value as of June 30, 2021. For a further description of the dilution that you will experience immediately after the offering, see "Dilution."

The trading price of our equity securities may be volatile due to factors beyond our control, and purchasers of the ordinary shares or ADSs could incur substantial losses.

The market prices of the ordinary shares or ADSs and shares may be volatile. The stock market in general and the market for biotechnology companies in particular have experienced extreme volatility that has often been unrelated to the operating performance of particular companies. As a result of this volatility, investors may not be able to sell their ordinary shares or ADSs or shares at or above the price originally paid for the security. The market price for the ordinary shares or ADSs and shares may be influenced by many factors, including:

- actual or anticipated fluctuations in our financial condition and operating results;
- the release of new data from the clinical trials of arimoclomol;
- announcements related to the timing, scope and likelihood of regulatory filings, submissions and marketing approvals of our product candidate and to meet existing or future regulatory standards or comply with post-approval requirements;

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- actual or anticipated changes in our growth rate relative to our competitors;
- competition from existing products or new products that may emerge;
- announcements by us or our competitors of significant acquisitions, strategic partnerships, joint ventures, collaborations or capital commitments;
- failure to meet or exceed financial estimates and projections of the investment community or that we provide to the public;
- issuance of new or updated research or reports by securities analysts;
- commentary by investors on the prospects for our business, the ordinary shares or ADSs on the internet, including blogs, articles and message boards, and/or social media and resulting in trading of our ordinary shares or the ADSs;
- unusual trading in our ordinary shares or ADSs or securities derivative thereof, including pursuant to naked, or uncovered, short positions, or “short squeezes;”
- fluctuations in the valuation of companies perceived by investors to be comparable to us;
- currency fluctuations;
- ordinary share price and volume fluctuations attributable to inconsistent trading volume levels of the ADSs;
- additions or departures of key management or scientific personnel;
- disputes or other developments related to proprietary rights, including patents, litigation matters and our ability to obtain patent protection for our technologies;
- class action lawsuits or litigation related to our product candidates, business and operations;
- changes to coverage policies or reimbursement levels by commercial third party payors and government payors and any announcements relating to coverage policies or reimbursement levels;
- announcement or expectation of additional debt or equity financing efforts;
- uncertainty caused by and the unprecedented nature of the ongoing COVID-19 pandemic;
- issuances or sales of the ordinary shares or ADSs by us, our insiders or our other shareholders; and
- general economic and market conditions.

These and other market and industry factors may cause the market price and demand for the ordinary shares or ADSs to fluctuate substantially, regardless of our actual operating performance. This volatility may limit or prevent investors from readily selling their shares of common stock and may otherwise negatively affect the liquidity of our common stock. For example, ADSs on The Nasdaq Global Select Market and shares on Nasdaq Copenhagen in June 2021 experienced extreme volatility in price and trading volume. From June 1, 2021 to November 2, 2021, the sale price of ADSs on The Nasdaq Global Select Market ranged from as low as \$3.78 to as high as \$77.77 and daily trading volume ranged from 13,400 shares to 168,470,900 shares. Investors who purchase ADS or ordinary shares may lose a significant portion of their investments if the price of such securities subsequently declines.

Information available in public media that is published by third parties, including blogs, articles, message boards and social and other media may include statements not attributable to us and may not be reliable or accurate.

We have received, and may continue to receive, a high degree of media coverage that is published or otherwise disseminated by third parties, including blogs, articles, message boards and social and other media.

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This includes coverage that is not attributable to statements made by our directors, officers or employees. You should read carefully, evaluate and rely only on the information contained in this prospectus supplement, the accompanying prospectus and the documents incorporated by reference herein and therein in determining whether to purchase our ADSs or ordinary shares. Information provided by third parties may not be reliable or accurate and could materially impact the trading price of our ADSs and/or ordinary shares, which could cause losses to your investments in our securities.

Future sales or issuances of our common stock in the public markets, or the perception of such sales, could depress the trading price of our common stock.

The sale of a substantial number of shares of our common stock or other equity-related securities in the public markets, or the perception that such sales could occur, could depress the market price of our common stock and impair our ability to raise capital through the sale of additional equity securities. We may sell large quantities of our common stock at any time pursuant to this prospectus supplement and the accompanying prospectus or in one or more separate offerings. We cannot predict the effect that future sales of common stock or other equity-related securities would have on the market price of our common stock.

We have broad discretion over the use of the net proceeds from this offering and may use them in ways with which you do not agree and in ways that may not enhance our operating results or the price of the ordinary shares or ADSs.

Our board of directors and management will have broad discretion over the application of the net proceeds that we receive from this offering. We may spend or invest these proceeds in ways with which our shareholders and holders of ADSs disagree or that do not yield a favorable return, if at all. We intend to use the net proceeds from this offering, together with our existing cash resources as described in "Use of Proceeds." However, our use of these proceeds may differ substantially from our current plans. Failure by our management to apply these funds effectively could harm our business, results of operations, cash flows, financial condition and/or prospects. Pending their use, we may invest the net proceeds from this offering in a manner that does not produce income or that loses value.

The actual number of shares we will issue under the sales agreement, at any one time or in total, is uncertain.

Subject to certain limitations in the sales agreement and compliance with applicable law, we have the discretion to deliver a placement notice to Cowen at any time throughout the term of the sales agreement. The number of shares that are sold by Cowen after delivering a placement notice will fluctuate based on the market price of our common stock during the sales period and limits we set with Cowen. Because the price per share of each share sold will fluctuate based on the market price of our common stock during the sales period, it is not possible at this stage to predict the number of shares that will be ultimately issued.

The common stock offered hereby will be sold in "at the market offerings," and investors who buy shares at different times will likely pay different prices.

Investors who purchase shares in this offering at different times will likely pay different prices, and so may experience different outcomes in their investment results. We will have discretion, subject to market demand, to vary the timing, prices, and numbers of shares sold, and there is no minimum or maximum sales price. Investors may experience a decline in the value of their shares as a result of share sales made at prices lower than the prices they paid.

U.S. Holders may suffer adverse tax consequences if we are characterized as a passive foreign investment company, or PFIC.

Based on our current estimates (and not final audited financials) of the composition of our income and valuation of our assets, including goodwill, we do not believe we were a PFIC for our taxable year ending

June 30, 2021. There can be no assurance that the United States Internal Revenue Service, or IRS, will agree with our conclusion and that the IRS would not successfully challenge our position. The determination of whether we are a PFIC is a fact-intensive determination made on an annual basis and the applicable law is subject to varying interpretation. Furthermore, because there are uncertainties in the application of the relevant rules, it is possible that the IRS may challenge our classification of certain income and assets as non-passive or our valuation of our tangible and intangible assets, each of which may result in us being treated as a PFIC for our taxable year ending June 30, 2021 or us becoming a PFIC for the current taxable year or any future taxable years. Our PFIC status may change from year to year and we have not yet made any determination as to our expected PFIC status for the current year and our status may depend, in part, on how quickly we use our cash. Accordingly, there can be no assurance that we will not be considered a PFIC in the current year or for any future taxable year. Our U.S. counsel expresses no opinion with respect to our PFIC status for our taxable year ending June 30, 2021, and the current or any future taxable year. Under the U.S. Internal Revenue Code of 1986, as amended, or Code, we will be a PFIC for any taxable year in which either (i) 75% or more of our gross income consists of “passive income,” or (ii) 50% or more of the quarterly weighted-average value of our assets, including cash, consists of assets that produce, or are held for the production of, “passive income.” Passive income generally includes interest, dividends, rents, certain non-active royalties and capital gains. Whether we will be a PFIC in any year depends on the composition of our income and assets, and the relative fair market value of our assets from time to time, which we expect may vary substantially over time. In addition, for purposes of the above calculations, a non-U.S. corporation that directly or indirectly owns at least 25% by value of the shares of another corporation is treated as if it held its proportionate share of the assets and received directly its proportionate share of the income of such other corporation.

If we are a PFIC for any taxable year during which a U.S. Holder (as defined below under “Material U.S. Federal Income Tax Considerations”) holds ordinary shares or ADSs, we will continue to be treated as a PFIC with respect to such U.S. Holder in all succeeding years during which the U.S. Holder owns the ordinary shares or ADSs regardless of whether we continue to meet the PFIC test described above, unless the U.S. Holder makes a specified election once we cease to be a PFIC. If we are classified as a PFIC for any taxable year during which a U.S. Holder holds ordinary shares or ADSs, the U.S. Holder may be subject to adverse tax consequences regardless of whether we continue to qualify as a PFIC, including ineligibility for any preferred tax rates on capital gains or on actual or deemed dividends, interest charges on certain taxes treated as deferred, and additional reporting requirements.

A U.S. Holder may in certain circumstances mitigate the adverse tax consequences of the PFIC rules by filing an election to treat the PFIC as a QEF, or, if shares of the PFIC are “marketable stock” for purposes of the PFIC rules, by making a mark-to-market election with respect to the shares of the PFIC. However, in the event that we are or become a PFIC, we do not intend to comply with the reporting requirements necessary to permit U.S. Holders to elect to treat us as a QEF. Furthermore, if a U.S. Holder were to make a mark-to-market election with respect to its ordinary shares or ADSs, the U.S. Holder would be required to include annually in its U.S. federal taxable income (taxable at ordinary income rates) an amount reflecting any year end increase in the value of its ordinary shares or ADSs. For further discussion of the PFIC rules and the adverse U.S. federal income tax consequences in the event we are classified as a PFIC, see the section titled “Material U.S. Federal Income Tax Considerations.” The U.S. federal income tax rules relating to PFICs are very complex. Prospective U.S. Holders are strongly urged to consult their own tax advisors with respect to the impact of PFIC status on the purchase, ownership and disposition of ordinary shares or ADSs, the consequences to them of an investment in a PFIC, any elections available with respect to the ordinary shares or ADSs and the IRS information reporting obligations with respect to the purchase, ownership and disposition of ordinary shares or ADSs of a PFIC.

The intended tax effects of our corporate structure depend on the application of the tax laws of various jurisdictions and on how we operate our business.

During the ordinary course of business, there are many transactions and calculations for which the ultimate tax determination is uncertain. For example, our effective tax rates could be adversely affected by changes in

foreign currency exchange rates or by changes in the relevant tax, accounting and other laws, regulations, principles and interpretations. As we intend to operate in numerous countries and tax jurisdictions, the application of tax laws can be subject to diverging and sometimes conflicting interpretations by tax authorities of these jurisdictions. It is not uncommon for tax authorities in different countries to have conflicting views, for instance, with respect to, among other things, the manner in which the arm's length standard is applied for transfer pricing purposes, or with respect to the valuation of intellectual property. In addition, it is uncertain whether we will be able to fully utilize our net operating losses as an income tax benefit for future periods. To the extent that our ability to use our net operating losses is restricted, this may result in us paying more tax and could therefore reduce our post-tax profits. In addition, tax laws are subject to change as new laws are passed and new interpretations of the law are promulgated by taxing authorities or sustained by judicial bodies. We are unable to predict what tax law changes may be proposed or enacted in the future or what effect such changes would have on our business, but such changes, to the extent they are brought into tax legislation, regulations, policies or practices, could affect our financial position and increase the complexity, burden and cost of tax compliance.

SPECIAL NOTE REGARDING FORWARD-LOOKING STATEMENTS

This prospectus supplement and the accompanying prospectus, including the documents incorporated by reference herein and therein, and any free writing prospectus that we have authorized for use in connection with this offering contain forward-looking statements within the meaning of Section 27A of the Securities Act of 1933, as amended, or the Securities Act, and Section 21E of the Securities Exchange Act of 1934, as amended, or the Exchange Act, that reflect our current expectations and views of future events. Known and unknown risks, uncertainties and other factors, including those listed under “Risk Factors” in this prospectus supplement and the accompanying prospectus and in any related free writing prospectuses and under similar headings in documents that are incorporated by reference into this prospectus supplement and the accompanying prospectus, including our most recent Annual Report on Form 20-F and reports on Form 6-K as updated by our subsequent filings, may cause our actual results, performance or achievements to be materially different from those expressed or implied by the forward-looking statements.

You can identify some of these forward-looking statements by words or phrases, such as “may,” “will,” “expect,” “anticipate,” “aim,” “estimate,” “intend,” “plan,” “believe,” “is/are likely to,” “potential,” “continue” or other similar expressions. We have based these forward-looking statements largely on our current expectations and projections about future events that we believe may affect our financial condition, results of operations, business strategy and financial needs. These forward-looking statements include statements relating to:

- the ability of our clinical trials to demonstrate acceptable safety and efficacy of our product candidate, and other positive results;
- the timing, progress and results of clinical trials for our product candidate, including statements regarding the timing of initiation and completion of studies or trials and related preparatory work, the period during which the results of the trials will become available, and our research and development programs;
- the timing, scope and likelihood of regulatory filings, submissions and approvals, including the MAA and NDA process for arimoclomol for the treatment of NPC and the potential regulatory approval of arimoclomol;
- our ability to obtain marketing approvals of our product candidate and to meet existing or future regulatory standards or comply with post-approval requirements;
- our expectations regarding our ability to fund our operating expenses and capital expenditure requirements;
- our payments of future milestone payments to our licensing partners, and the expected timing of such payments;
- our expectations regarding the potential market size and the size of the patient populations for our product candidate, if approved for commercial use;
- our expectations regarding the potential advantages of our product candidate over existing therapies;
- the impact of the ongoing COVID-19 pandemic on our business and operations;
- our expectations regarding the outcome and impact of class action lawsuits and any other litigation on our business and operations;
- our potential to enter into new collaborations;
- our expectations with regard to our ability to identify and develop additional product candidates or product candidates for other indications or technologies with significant commercial potential that are consistent with our commercial objectives;
- our expectations with regard to the willingness and ability of our current and future licensing and collaboration partners to pursue the development of our product candidate;

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- the commercialization and market acceptance of our product candidate;
- our marketing and manufacturing capabilities;
- the pricing of and reimbursement for our product candidate;
- the implementation of our business model and strategic plans for our business and product candidate;
- our ability to operate our businesses without infringing the intellectual property rights and proprietary technology of third parties;
- the scope of protection we are able to establish and maintain for intellectual property rights covering our product candidate;
- our analysis of our actual or potential patent infringement claims and the rights of our collaboration partners with respect to such claims;
- estimates of our expenses, future revenue, capital requirements, our needs for additional financing and our ability to obtain additional capital;
- regulatory development in the United States, Europe and other jurisdictions;
- our exposure to scrutiny as a public company in the United States;
- our ability to effectively manage our anticipated growth;
- our ability to attract and retain qualified employees and key personnel;
- our use of proceeds, if any, from this offering;
- our financial performance;
- our expectations regarding the time during which we will be an emerging growth company under the JOBS Act and qualify as a foreign private issuer; and
- developments and projections relating to our competitors and our industry, including competing therapies.

These forward-looking statements involve various risks and uncertainties. Although we believe that our expectations expressed in these forward-looking statements are reasonable, our expectations may later be found to be incorrect. Our actual results could be materially different from our expectations. We discuss many of these risks in greater detail in the documents incorporated by reference herein, including under the heading “Risk Factors.” These forward-looking statements represent our estimates and assumptions only as of the dates of this prospectus supplement and the documents incorporated by reference herein and therein, and any free writing prospectus, as applicable, regardless of the time of delivery of this prospectus supplement or any sale of the ADSs and, except as required by law, we undertake no obligation to update or revise publicly any forward-looking statements, whether as a result of new information, future events or otherwise after the date of this prospectus supplement. For all forward-looking statements, we claim the protection of the safe harbor for forward-looking statements contained in the Private Securities Litigation Reform Act of 1995.

USE OF PROCEEDS

We may offer and sell up to \$50,000,000 of our ordinary shares, including ordinary shares represented by ADSs, from time to time. The amount of proceeds from this offering will depend upon the number of ADSs sold and the market price at which they are sold. There can be no assurance that we will be able to sell any ADSs under or fully use the sales agreement with Cowen as a source of financing. We intend to use the net proceeds from this offering, if any, together with our existing cash, to continue the regulatory approval process for and fund the commercial launch, if approved, of arimoclomol for the treatment of NPC and for working capital and general corporate purposes.

Our management will have broad discretion over the use of the net proceeds from the offering. The amounts and timing of our expenditures will depend upon numerous factors, including the results of our research and development efforts, the timing, cost and success of preclinical studies and ongoing clinical trials or clinical trials we may commence in the future, the timing of regulatory submissions, our ability to obtain additional financing, the amount of cash obtained through our existing collaborations and future collaborations, if any, and any unforeseen cash needs.

DIVIDEND POLICY

We have never declared or paid any cash dividends on our ordinary shares and we do not anticipate paying any cash dividends on our ordinary shares in the foreseeable future. We intend to retain all available funds and any future earnings to fund the development and expansion of our business. In addition, our loan facility agreement with Kreos Capital VI (UK) Limited prohibits us to agree, make or agree to make any distribution by way of dividend or otherwise without the written consent of the lender thereunder. Any future determination related to our dividend policy and the declaration of any dividends will be made at the discretion of our board of directors and will depend on a number of factors, including our results of operations, financial condition, future prospects, contractual restrictions, restrictions imposed by applicable law and other factors our board of directors deems relevant.

If we pay any dividends on our ordinary shares, we will pay those dividends, which are payable in respect of the ordinary shares underlying the ADSs to the depositary, as the registered holder of such ordinary shares, and the depositary then will pay such amounts to our ADS holders in proportion to the ordinary shares underlying the ADSs held by such ADS holders, subject to the terms of the deposit agreement, including the fees and expenses payable thereunder. See “Description of American Depositary Shares.” Cash dividends on our ordinary shares, if any, will be paid in U.S. dollars.

Legal and Regulatory Requirements

In accordance with the DCA, dividends, if any, are declared with respect to a financial year at the annual general meeting of shareholders in the following year, where the statutory annual report (which includes the audited financial statements) for that financial year is approved. Any resolution to distribute interim dividends within six months of the date of the statement of financial position as set out in our latest adopted annual report must be accompanied by the statement of financial position from our latest annual report or an interim statement of financial position which must be reviewed by our auditor. If the decision to distribute interim dividends is passed more than six months after the date of the statement of financial position as set out in our latest adopted annual report, an interim statement of financial position must be prepared and reviewed by our auditor. The statement of financial position or the interim statement of financial position, as applicable, must show that sufficient funds are available for distribution. Dividends may not exceed the amount recommended by the board of directors for approval by the general meeting of shareholders. Moreover, dividends and interim dividends may only be made out of distributable reserves and may not exceed what is considered sound and adequate with regard to our financial condition or be to the detriment of our creditors and such other factors as the board of directors may deem relevant.

In accordance with the DCA, share buybacks, if any, may only be carried out by the board of directors using funds that could have been distributed as dividends at the latest annual general meeting of shareholders. Any share buyback must be conducted in accordance with an authorization obtained at a general meeting of our shareholders. The authorization must be granted for a defined period of time not exceeding five years. In addition, the authorization must specify the maximum permitted value of treasury shares as well as the minimum and maximum amount that we may pay as consideration for such shares. A decision by our board of directors to engage in share buybacks, if any, will be made in accordance with the factors applicable to dividend payments set forth above.

See “Taxation—Material Danish Income Tax Considerations” for a description of Danish withholding taxes and certain other Danish considerations relevant to the purchase or holding of ordinary shares and ADSs and “Taxation—Material U.S. Federal Income Tax Consequences for U.S. Holders” for a description of U.S. federal income tax considerations relevant to the purchase or holding of ordinary shares and ADSs.

DILUTION

If you invest in our ADSs, your interest will be immediately diluted to the extent of the difference between the offering price per share of our ADSs you pay in this offering and our net tangible book value per share of our ADSs immediately after this offering. Dilution results from the fact that the offering price is substantially in excess of the net book value per ADS. Our historical net tangible book value as of June 30, 2021 was \$26.2 million, or DKK 4.71 per ordinary share (equivalent to \$0.75 per ADS). Historical net tangible book value per share is determined by dividing our total consolidated tangible assets, less the amount of our total consolidated liabilities, by the number of ordinary shares outstanding as of June 30, 2021. Dilution in net tangible book value per share represents the difference between the amount per share paid by purchasers of ADSs in this offering and the as adjusted net tangible book value per share of our ordinary shares immediately after giving effect to this offering.

After giving effect to the sale of the ADSs in the aggregate amount of \$50.0 million in this offering at an assumed offering price of \$4.09 per ADS, the last reported sale price of the ADSs on The Nasdaq Global Select Market on November 2, 2021, and after deducting commissions and estimated aggregate offering expenses payable by us, our as adjusted net tangible book value as of June 30, 2021 would have been \$74.4 million, or \$1.58 per ADS. This represents an immediate increase in net tangible book value of \$0.83 per ADS to existing shareholders and holders of ADSs and an immediate dilution in net tangible book value of \$2.51 per ADS to new investors purchasing the ADSs in this offering.

The following table illustrates this dilution on a per ADS basis. The information is illustrative only and will adjust based on the actual prices at which ADSs are sold, the actual number of ADSs sold and other terms of the offering determined at the time our ADSs are sold pursuant to this prospectus supplement and the accompanying prospectus. The ADSs sold in this offering, if any, will be sold from time to time at various prices.

Assumed initial offering price per ADS	\$4.09
Historical net tangible book value per ADS as of June 30, 2021	\$0.75
Increase in net tangible book value per ADS attributable to this offering	<u>0.83</u>
As adjusted net tangible book value per ADS as of June 30, 2021	<u>1.58</u>
Dilution per ADS to investors participating in this offering	<u>\$2.51</u>

The foregoing tables and calculations are on 34,952,241 of our ordinary shares outstanding as of June 30, 2021, and excludes:

- subject to certain vesting criteria being satisfied, up to 695,723 ordinary shares that may be issued to cover the delivery of ordinary shares to participants of the LTIP as of June 30, 2021;
- up to 705,976 ordinary shares underlying unvested or unexercised RSUs as of June 30, 2021; and
- bonus shares that we have agreed to issue pursuant to a license agreement with the University of Kansas and UCL Business PLC, as described further in the documents incorporated by reference into this prospectus supplement.

To the extent that we issue additional ADSs or ordinary shares in the future, there will be further dilution to shareholders and the holders of ADSs. In addition, we may choose to raise additional capital because of market conditions or strategic considerations, even if we believe that we have sufficient funds for our current or future operating plans. If we raise additional capital through the sale of equity or convertible debt securities, the issuance of these securities could result in further dilution to our shareholders and the holders of ADSs.

DESCRIPTION OF SHARE CAPITAL AND ARTICLES OF ASSOCIATION

The following describes our issued share capital, summarizes the material provisions of our articles of association and highlights certain differences in corporate law in the Kingdom of Denmark and Delaware corporate law, the law under which many publicly listed companies in the United States are incorporated. Please note that this summary is not intended to be exhaustive. For further information, please refer to the full version of our articles of association, which are included as an exhibit to the registration statement of which this prospectus supplement forms a part.

Introduction

Set forth below is a summary of certain information concerning our share capital as well, as a description of certain provisions of our articles of association and relevant provisions of the DCA. The summary includes certain references to, and descriptions of, material provisions of our articles of association and Danish law in force as of the date of this prospectus supplement. The summary below contains only material information concerning our share capital and corporate status and does not purport to be complete and is qualified in its entirety by reference to our articles of association and applicable Danish law. Further, please note that as an ADS holder you will not be treated as one of our shareholders and will not have any shareholder rights.

General

We were incorporated on June 19, 2009 as a private limited liability company under Danish law and later converted into a Danish public limited liability company on October 20, 2017. We are registered with the Danish Business Authority (Erhvervsstyrelsen) in Copenhagen, Denmark under company registration number (CVR) no. 32266355. Our ADSs have been listed on The Nasdaq Global Select Market under the symbol “ORPH” since September 2020. Our ordinary shares have been listed in Denmark on Nasdaq Copenhagen under the symbol “ORPHA” since November 2017. Our company has been established with the objectives of engaging in medical research, production and sale of such products and related business.

Our headquarters and principal executive offices are located at Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark, and our telephone number is +45 39 17 82 72. Our website address is www.orphazyme.com. The information contained on, or accessible through, our website is not incorporated by reference into this prospectus supplement or the accompanying prospectus, and you should not consider any information contained in, or that can be accessed through, our website as part of this prospectus supplement or the accompanying prospectus or in deciding whether to purchase ordinary shares, including ordinary shares in the form of ADSs. We have included our website address as an inactive textual reference only.

Development of Share Capital

Since November 2017, we had one class of shares (prior to this date we had multiple classes of shares). As of June 30, 2021, our registered, issued and fully paid outstanding share capital was DKK 34,952,241 distributed into 34,952,241 shares of nominal value DKK 1 each, including 21,067,199 ADSs, each representing one ordinary share.

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The development of our share capital since December 31, 2016 and up to and including the date of this prospectus supplement is set forth in the table below.

Date of approval	Capital Increase, No. of Shares	Gross Proceeds, DKK000s	Share Capital, No. of Shares after change	Issued Share DKK Capital after change
Share capital at December 31, 2016			A-shares: 125,000 B-shares: 2,050,208 C-shares: 1,185,333	3,360,541
2017				
Capital increase by cash contribution, January 26, 2017			A-shares: 125,000 B-shares: 2,050,208 C-shares: 1,719,340	3,894,548
	534,007	48,060		
Capital increase by cash contribution, January 26, 2017			A-shares: 125,000 B-shares: 2,050,208 C-shares: 2,491,362	4,666,570
	772,022	69,482		
Capital increase by cash contribution, June 29, 2017			A-shares: 125,000 B-shares: 2,050,208 C-shares: 2,927,002	5,102,210
	435,640	39,208		
Conversion into a public limited liability company, October 20, 2017			A-shares: 125,000 B-shares: 2,050,208 C-shares: 2,927,002	5,102,210
	—	—		
Consolidation of share classes, November 2, 2017	—	—	5,102,210	5,102,210
Issuance of bonus shares, November 2, 2017	6,487,882	—	11,590,092	11,590,092
Initial public offering, November 6, 2017	7,500,000	600,000	19,090,092	19,090,092
Exercise of warrants, November 20, 2017	838,092	1,161	19,928,184	19,928,184
2018				
Issuance of bonus shares, January 29, 2018	11,380	—	19,939,564	19,939,564
2019				
Issuance of bonus shares, January 31, 2019	26,060	—	19,965,624	19,965,624
Issuance of matching shares, March 4, 2019	19,175	19	19,984,799	19,984,799
2020				
Issuance of bonus shares, January 31, 2020	20,650	—	20,005,449	20,005,449
Capital increase by cash contribution, February 6, 2020	7,032,937	745,491	27,038,386	27,038,386
Exercise of restricted share units, March 27, 2020(1)	4,616	282	27,043,002	27,043,002
Exercise of restricted share units, March 27, 2020(1)	1,927	118	27,044,929	27,044,929

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Date of approval	Capital Increase, No. of Shares	Gross Proceeds, DKK000s	Share Capital, No. of Shares after change	Issued Share DKK Capital after change
Exercise of restricted share units, March 27, 2020(1)	3,451	211	27,048,380	27,048,380
Vesting and exercise of matching shares, July 29, 2020	31,250	31	27,079,630	27,079,630
Exercise of restricted share units, March 27, 2020(1)	1,927	118	27,081,557	27,081,557
U.S. initial public offering, September 28, 2020	7,616,146	534,534	34,697,703	34,697,703
2021				
Vesting and exercise of matching shares, including US IPO bonus grants to certain participants, February 1, 2021	170,131	170	34,867,834	34,867,834
Issuance of bonus shares, February 25, 2021	22,553	23	34,890,387	34,890,387
Directed issuance of new shares, February 25, 2021	58,000	58	34,948,387	34,948,387
Exercise of restricted share units, March 24, 2021	3,854	236	34,952,241	34,952,241

- (1) Share issue was approved by our board of directors on March 27, 2020 and subsequently registered with the Danish Business Authority following exercise by the respective directors.

Authorizations to Our Board of Directors

Our board of directors is authorized to increase our share capital as follows:

- In accordance with article 3.1 of our articles of association, our board of directors is, until March 25, 2026, authorized 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.
- In accordance with article 3.2 of our articles of association, our board of directors is, until November 2, 2022, authorized to increase our share capital in one or more issues without pre-emption rights for our existing shareholders by up to a nominal amount of DKK 1,300,000 in connection with the issue of new shares to members of our board of directors, our executives and/or our employees. 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.
- In accordance with article 3.3 of our articles of association, our board of directors is, until November 2, 2022, authorized to increase our share capital in one or more issues of new shares without preemption rights for our existing shareholders by up to a nominal amount of DKK 15,750,000 in connection with 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, which will be below market price. The value of such new shares to be issued can in any case not exceed a maximum of \$2.5 million with a fixed exchange rate of DKK 6.30 per 1 USD based on the average closing price of the our ordinary shares on Nasdaq Copenhagen for the 30 days immediately prior to the date of issuance.
- In accordance with article 3.4 of our articles of association, our board of directors is, until January 25, 2025, authorized to increase our share capital in one or more issues of new shares with preemption rights for our 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.

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- In accordance with article 3.5 of our articles of association, our board of directors is, until March 25, 2026, authorized to increase our share capital in one or more issues of new shares without pre-emption rights for our existing shareholders by up to a nominal amount of DKK 1,300,000 in connection with the issue of new shares to members of our board of directors, our executives and/or our employees. The new shares shall be issued against cash payment at a subscription price to be determined by our board of directors, which may be below the market price.
- In accordance with article 3.6 of our articles of association, the authorizations granted to our board of directors pursuant to articles 3.2 and 3.5 can in the aggregate only be exercised to increase our share capital by a maximum nominal amount of DKK 2,000,000.

As of the date of this prospectus supplement, our board of directors partially exercised the authorization in article 3.2 of our articles of association to increase our share capital following which a nominal value of DKK 294,331 of the authorization has been issued. In addition, our board of directors has partly exercised the authorization in article 3.3 to increase our share capital following which a nominal value of DKK 80,643 of the authorization has been issued.

Further, our board of directors is authorized on behalf of the company until March 25, 2026 to acquire our own shares for a total nominal value of up to 10% of our share capital for the time being, so long as the company's holding of treasury shares after such acquisition does not exceed 20% of the company's share capital. The price paid for such shares may not deviate by more than 10% from the share price quoted on Nasdaq Copenhagen at the time of acquisition.

As of June 30, 2021, the total number of additional shares our board of directors is authorized to issue was 49,364,793.

Our ADSs are listed on The Nasdaq Global Select Market under the symbol "ORPH" and our ordinary shares are listed in Denmark on Nasdaq Copenhagen under the symbol "ORPHA."

Pre-emptive Rights

If our shareholders at a general meeting resolve to increase our share capital by a cash contribution, section 162 of the DCA will apply. Under that section, shareholders have a pre-emptive right to subscribe for new shares in proportion to their existing shareholdings. However, the pre-emptive right may be derogated from by a majority comprising at least two-thirds of the votes cast, as well as at least two-thirds of the share capital represented at the general meeting, provided the share capital increase takes place at market price or nine-tenths of the votes cast, as well as at least nine-tenths of the share capital represented at the general meeting if the share capital increase takes place below market price, unless (i) such capital increase is directed at certain but not all shareholders (in which case all shareholders must consent); or (ii) such capital increase is directed at our employees whereby a majority comprising at least two-thirds of the votes cast, as well as at least two-thirds of the share capital represented at the general meeting is required. Further, the pre-emptive rights may be derogated from by an exercise of the board of directors of a valid authorization in our articles of association, provided that the share capital increase takes place at or above market price.

Shareholders' Register

We are obliged to maintain a shareholders' register (Ejerbog). The shareholders' register is maintained by Computershare A/S, Lottenborgvej 26 D, 1., DK-2800 Kgs. Lyngby, Denmark, our Danish share registrar and transfer agent. It is mandatory that the shareholders' register is maintained within the European Union and that it is available to public authorities.

Pursuant to the DCA, public and private limited liability companies are required to register with the Danish Business Authority information regarding shareholders who own at least 5% of the share capital or the voting rights. Pursuant to this provision, we file registrations with the Danish Public Shareholders' Register of the

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Danish Business Authority. Shareholders that exceed or fall below the ownership threshold must notify us, and we will subsequently file the information with the Danish Business Authority. Reporting is further required upon passing or falling below thresholds of 5%, 10%, 15%, 20%, 25%, 50%, 90%, and 100% as well as one-third and two-thirds of the votes or the share capital. This also applies to beneficial holders of our shares, such as holders of the ADSs.

Articles of Association and Danish Corporate Law

General Meetings and Voting Rights

Our general meetings shall be held in the Capital Region of Denmark. Our annual general meeting 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. Not later than eight weeks before the contemplated date of the annual general meeting, we shall publish the date of the general meeting and the deadline for submitting requests for specific proposals to be included in the agenda.

Extraordinary general meetings shall be held when determined by our board of directors or requested by our auditor. Furthermore, our board of directors shall convene an extraordinary general meeting within two weeks of receipt of a written request from shareholders representing no less than 5% of the share capital containing specific proposals for the business to be transacted at such extraordinary general meeting.

General meetings shall be convened by our board of directors at least three weeks' and not more than five weeks' notice. The notice shall be published on our website. Furthermore, a notice of the general meeting shall be sent electronically to all shareholders recorded in our register of shareholders who have requested such notice.

In accordance with Danish law, the notice shall specify the time and place of the general meeting and the agenda containing the business to be transacted at the general meeting. If a proposal to amend our articles of association is to be considered at the general meeting, the main contents of the proposal shall be specified in the notice. Our general meetings shall be held in English. Our 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 our board of directors, in Danish.

Every shareholder is entitled to have specific business transacted at the general meeting, provided that the shareholder submits a written request to that effect to our board of directors not later than six weeks before the date of the general meeting.

The right of a shareholder to attend a general meeting and to vote is determined by the shares held by the shareholder at the record date. The record date is one week before the general meeting. The shares held by each shareholder are determined at the record date based on the number of shares held by that shareholder as registered in our register of shareholders and any notification of ownership received by us for the purpose of registration in our register of shareholders, but which have not yet been registered.

At the general meeting each share of the nominal value of DKK 1 shall carry one vote. Our articles of association permit a person registered as a holder of our shares in VP Securities A/S and acting in a professional capacity to exercise on behalf of other natural or legal persons, including holders of ADSs representing our ordinary shares, voting rights attached to any such shares in a manner that is not identical to the exercise of the voting rights attached to our other shares held by such person.

A shareholder who is entitled to attend the general meeting pursuant to our articles of association and who wants to attend the general meeting shall notify us of his/her attendance no later than three days prior to the date of the general meeting. A shareholder may, subject to having notified us of his/her attendance in accordance with our articles of association, attend in person or by proxy, and the shareholder or the proxy may attend together with an adviser.

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The right to vote may be exercised by a written and dated instrument of proxy in accordance with applicable laws. Our board of directors may be appointed as proxy. A shareholder who is entitled to participate in the general meeting according to our articles of association may vote by postal vote in accordance with the DCA. Such postal votes shall be received by us no later than the business day before the general meeting. Postal votes cannot be withdrawn. In accordance with Danish law, the notice shall specify the time and place of the general meeting and the agenda containing the business to be transacted at the general meeting. If a proposal to amend our articles of association is to be considered at the general meeting, the main contents of the proposal shall be specified in the notice.

Our articles of association permit our board to decide to hold general meetings partially or fully by electronic means in accordance with our articles of association and applicable Danish law.

Resolutions by the General Meetings and Amendments to the Articles of Association

Resolutions at general meetings shall be passed by a simple majority of votes cast, unless otherwise prescribed by law or by our articles of association. Adoption of changes to our articles of association, our dissolution, merger or demerger requires that the resolution is adopted by 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 our board of directors or other bodies. The provisions in our articles of association relating to a change of the rights of shareholders or a change to the capital are not more stringent than required by the DCA.

Redemption and Conversion Provisions

Except as provided for in the DCA, no shareholder is under an obligation to have its shares redeemed in whole or in part by us or by any third party, and none of the shares carry any redemption or conversion rights or any other special rights.

Dissolution and Liquidation

In the event of dissolution and liquidation, our shareholders are entitled to participate in the distribution of assets in proportion to their nominal shareholdings after payment of our creditors.

Indication of Takeover Bids

No takeover offers have been made by any third party in respect of our shares during the past or current financial year. Our articles of association do not contain provisions that are likely to have the effect of delaying, deferring or preventing a change in control of our company.

Provisions as to the Level of Equity Investments to be Notified to Us and the Danish Authorities

Shareholders in Danish companies with shares admitted to trading and official listing on Nasdaq Copenhagen are, pursuant to Section 38 of the Danish Capital Markets Act, required to give simultaneous notice to the company and the Danish Financial Supervisory Authority, or the FSA, of the shareholding in the company, when the shareholding reaches, exceeds or falls below thresholds of 5%, 10%, 15%, 20%, 25%, 50% or 90% and limits of one-third or two-thirds of the voting rights or nominal value of the total share capital.

A shareholder in a company means a natural or legal person who, directly or indirectly, holds: (i) shares in the company on behalf of itself and for its own account; (ii) shares in the company on behalf of itself, but for the account of another natural or legal person; or (iii) depository receipts, where such holder is considered a shareholder in relation to the underlying shares represented by the depository receipts.

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The duty to notify set forth above further applies to natural and legal persons who are entitled to acquire, sell or exercise voting rights which are:

- (i) held by a third party with whom that natural or legal person has concluded an agreement, which obliges them to adopt, by concerted exercise of the voting rights they hold, a lasting common policy towards the management of the issuer in question (common duty to inform for all parties to the agreement);
- (ii) held by a third party under an agreement concluded with that natural or legal person providing for the temporary transfer of the voting rights in question in return for consideration;
- (iii) attached to shares which are lodged as collateral for that natural or legal person, provided the person controls the voting rights and declares an intention of exercising them;
- (iv) attached to shares in which that natural or legal person has a lifelong right of disposal;
- (v) held, or may be exercised within the meaning of (i) to (iv), by an undertaking controlled by that person or entity;
- (vi) attached to shares deposited with that natural or legal person and which the person can exercise at its own discretion in the absence of specific instructions from the shareholders;
- (vii) held by a third party in its own name on behalf of that person; or
- (viii) exercisable by that person through a proxy where that person may exercise the voting rights at its discretion in the absence of specific instructions of the shareholder.

The duty to notify set forth above also applies to anyone, who directly or indirectly holds (a) financial instruments that afford the holder either an unconditional right to acquire or the discretion as to its right to acquire existing shares (e.g., share options); and/or (b) financial instruments based on existing shares and with an economic effect equal to that of the financial instruments mentioned in (a), regardless of them not affording the right to purchase existing shares (e.g., the ADSs or, under the circumstances, cash-settled derivatives linked to the value of our shares or ADSs representing our shares). Holding these kinds of financial instruments counts towards the thresholds mentioned above and may thus trigger a duty to notify by themselves or when accumulated with a holding of shares or ADSs. The FSA will in certain cases publish information concerning sanctions imposed, including, as a general rule, the name of the shareholder in question, as a consequence of non-compliance with the above rules.

The notification shall be made promptly but not later than four weekdays after the shareholder was aware or should have become aware of the completion of the transaction, and in accordance with the provisions of Danish Executive Order on Major Shareholders. The shareholder is deemed to have become aware of the completion of the transaction no later than two weekdays after the completion of the transaction. The shareholder shall disclose the change in voting rights and shares, including the number of voting rights (and the division of voting rights between share classes, if applicable) and shares held directly or indirectly by the shareholder following the transaction. The notification shall further state the transaction date on which the threshold was reached or no longer reached and the identity of the shareholder as well as the identity of any natural or legal person with the right to vote on behalf of the shareholder and in the case of a group structure, the chain of controlled undertakings through which voting rights are effectively held. The information shall be notified to the company and simultaneously submitted electronically to the FSA. Failure to comply with the notification requirements is punishable by fine or suspension of voting rights in instances of gross or repeated non-compliance.

When an obligation to notify rests on more than one natural or legal person, the notification may be made through a joint notification. However, use of a joint notification does not exempt the individual shareholders or natural or legal persons from their responsibilities in connection with the obligation to notify or the contents of the notification.

After receipt of the notification, but not later than three weekdays thereafter, the company shall publish the contents of the notification.

Furthermore, the general duty of notification under Section 55 of the DCA in respect of notification of significant holdings (similar to the thresholds set out in the Danish Capital Markets Act Section 38) applies, including when the limit of 100% of the share capital's voting rights or nominal value of the company is reached or are no longer reached.

EU Regulation No 596/2014 on Market Abuse and General Disclosure Requirements

EU Regulation No 596/2014 on market abuse, or the Market Abuse Regulation, applies to us and dealings concerning our shares and ADSs. In connection with our listing on The Nasdaq Global Select Market, we revised our internal code on possession and handling of inside information to cover trading in both our ordinary shares and ADSs and with respect to our board of directors', executive management's and employees' dealings in our shares or in financial instruments the value of which is determined by the value of our shares so that it also covers the ADSs. Furthermore, we have drawn up a list of those persons working for us who could have access to inside information on a regular or incidental basis and have informed such persons of the rules on insider trading and market manipulation, including the sanctions, which can be imposed in the event of a violation of those rules.

In addition, the company is obliged to disclose certain other information to the public pursuant to the Danish Capital Markets Act, the Danish Executive Order on an Issuers' Duty to Provide Information and the Issuer Rules of Nasdaq Copenhagen, regardless of whether this information amounts to inside information. Information which would have to be disclosed under these rules includes, for example: (i) changes to the board of directors, executive management and auditors; (ii) decisions to introduce incentive schemes; (iii) substantial changes in business activities; (iv) material acquisitions and divestments; (v) unexpected and significant deviations in the company's financial result or position; (vi) proposed changes in the capital structure; and (vii) annual and interim reports and accounts. Furthermore, the company is required to make sure that no unauthorized person gains access to inside information prior to its publication to the market.

The EU Short Selling Regulation (EU Regulation 236/2012) Includes Certain Notification Requirements in connection with Short Selling of Shares Admitted to Trading on a Trading Venue (including Nasdaq Copenhagen) and Securities or Derivatives that Relate to Such Shares (including the ADSs).

When a natural or legal person reaches, exceeds or falls below a net, short position of 0.2% of the issued share capital of a company that has shares admitted to trading on a trading venue (which includes the ADSs), such person shall make a private notification (i.e. such notification will not be made public) to the relevant competent authority, which in Denmark is the FSA. The obligation to notify the FSA, moreover, applies in each case where the short position reaches, exceeds or falls below 0.1% above the 0.2% threshold. In addition, when a natural or legal person reaches or falls below a net short position of 0.5% of the issued share capital of a company that has shares admitted to trading on a trading venue in the European Union and each 0.1% above that, such person shall make a public notification of its net short position via the FSA. The notification requirements apply to both physical and synthetic short positions. In addition uncovered short selling (naked short selling) of shares admitted to trading on a trading venue is prohibited. Furthermore, on March 16, 2020, the European Securities and Markets Authority, or ESMA, issued a decision which temporarily lowered the reporting threshold from 0.2% to 0.1% for net short position holders in shares traded on a trading venue in the European Union for three months due to COVID-19's impact on financial markets. On September 16, 2020, and December 17, 2020, ESMA issued a decision to renew the temporary requirement to temporarily lower the reporting threshold from 0.2% to 0.1% for net short position holders for an additional three months, which entails that the lowered threshold applied until March 19, 2021, to any natural or legal person, irrespective of their country of residence. ESMA has decided not to renew its decision on temporary lowered reporting threshold, hence the decision on temporarily lowered reporting threshold expired on March 19, 2021. On May 20, 2021, ESMA published an opinion wherein it recommended the European Commission to permanently lower the reporting threshold from 0.2% to 0.1%. On September 27, 2021, the European Commission adopted a delegated regulation reflecting this recommendation. After the European Commission has adopted the delegated regulation, the European Parliament and the Council have three months to formulate any objections. If they do not, the delegated regulation enters into force.

Mandatory Tender Offers

The Danish Capital Markets Act (Part 8) and the Danish Executive Order on Takeover include rules concerning public offers for the acquisition of shares admitted to trading on a regulated market (including Nasdaq Copenhagen).

If a shareholding is transferred, directly or indirectly, in a company with one or more share classes admitted to trading on a regulated market, to an acquirer or to persons acting in concert with such acquirer, the acquirer and the persons acting in concert with such acquirer, if applicable, shall give all shareholders of the company the option to dispose of their shares on identical terms, if the acquirer or the persons acting in concert with such acquirer gains control over the company as a result of the transfer.

Control as mentioned above exists if the acquirer or persons acting in concert with such acquirer, directly or indirectly, holds at least one-third of the voting rights in the company, unless it can be clearly proven in special cases that such ownership does not constitute control. An acquirer or persons acting in concert with such acquirer who does not hold at least one-third of the voting rights in a company, nevertheless has control when the acquirer has or persons acting in concert with such acquirer have:

- the right to control at least one-third of the voting rights in the company according to an agreement with other investors; or
- the right to appoint or dismiss a majority of the members of the central governing body.

Voting rights attached to treasury shares shall be included in the calculation of voting rights.

The Danish Capital Markets Act contains specific exemptions from the obligation to submit a mandatory takeover offer, including transfers of shares by inheritance or transfer within the same group and as a result of a creditor's debt enforcement proceedings. Exemptions from the mandatory tender offer rules may be granted under special circumstances by the FSA.

Limitation on Liability

Under Danish law, members of the board of directors or executive management may be held liable for damages in the event that loss is caused due to their negligence. They may be held jointly and severally liable for damages to the company and to third parties for acting in negligent violation of the articles of association and Danish law.

Comparison of Danish Corporate Law and Our Articles of Association and Delaware Corporate Law

The following comparison between Danish corporate law, which applies to us, and Delaware corporate law, the law under which many publicly listed companies in the United States are incorporated, discusses shareholder rights and obligations and certain additional matters. This summary is subject to Danish law, including the DCA, and Delaware corporate law, including the Delaware General Corporation Law. Further, please note that if you are a holder of the ADSs, then you are not treated as one of our shareholders under such laws and do not have any shareholder rights in Orphazyme A/S.

Shareholder Rights

Notice of Meeting

Denmark. According to the DCA and as implemented in our articles of association, general meetings in listed limited liability companies shall be convened by the board of directors with a minimum of three weeks'

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notice and a maximum of five weeks' notice. A convening notice shall also be forwarded to shareholders recorded in our shareholders' register who have requested such notification. There are specific requirements as to the information and documentation required to be disclosed in connection with the convening notice.

Delaware. Under Delaware law, unless otherwise provided in the certificate of incorporation or bylaws, written notice of any meeting of the stockholders must be given to each stockholder entitled to vote at the meeting not less than ten nor more than 60 days before the date of the meeting and shall specify the place, date, hour and purpose or purposes of the meeting.

Voting Rights

Denmark. Each share confers the right to cast one vote at the general meeting of shareholders, unless the articles of association provide otherwise. Our articles of association allow a person registered as a holder of our shares in VP Securities A/S and acting in a professional capacity on behalf of other natural or legal persons, including holders of ADS representing our ordinary shares to exercise voting rights attached to any such shares in a manner that is not identical to the exercise of the voting rights attached to our other shares held by such person. The right of a shareholder to vote is determined by the shares held by the shareholder at the record date. The record date is one week before the general meeting. Each holder of shares may cast as many votes as it holds shares. Voting instructions may be given only in respect of a number of ADSs representing an integral number of shares or other deposited securities. Shares that are held by us or our direct or indirect subsidiaries do not confer the right to vote.

Delaware. Under the Delaware General Corporation Law, each stockholder is entitled to one vote per share of stock, unless the certificate of incorporation provides otherwise. In addition, the certificate of incorporation may provide for cumulative voting at all elections of directors of the corporation, or at elections held under specified circumstances. Either the certificate of incorporation or the bylaws may specify the number of shares and/or the amount of other securities that must be represented at a meeting in order to constitute a quorum, but in no event can a quorum consist of less than one-third of the shares entitled to vote at a meeting.

Stockholders as of the record date for the meeting are entitled to vote at the meeting, and the board of directors may fix a record date that is no more than 60 nor less than 10 days before the date of the meeting, and if no record date is set then the record date is the close of business on the day next preceding the day on which notice is given, or if notice is waived then the record date is the close of business on the day next preceding the day on which the meeting is held. The determination of the stockholders of record entitled to notice or to vote at a meeting of stockholders shall apply to any adjournment of the meeting, but the board of directors may fix a new record date for the adjourned meeting.

Shareholder Proposals

Denmark. According to the DCA and our articles of association, extraordinary general meetings of shareholders will be held whenever our board of directors or our appointed auditor requires. In addition, one or more shareholders representing at least 5% of the registered share capital of the company may, in writing, require that a general meeting be convened. If such a demand is made, the board of directors shall convene the general meeting with three to five weeks' notice within 14 days thereafter.

All shareholders have the right to present proposals for adoption at the annual general meeting, provided that the proposals are submitted at least six weeks prior to the meeting. In the event that the request is made at a later date, the board of directors will determine whether the proposals were made in due time to be included on the agenda.

Delaware. Delaware law does not specifically grant stockholders the right to bring business before an annual or special meeting of stockholders. However, if a Delaware corporation is subject to the SEC's proxy rules, a stockholder who owns at least \$2,000 in market value, or 1% of the corporation's securities entitled to vote, may propose a matter for a vote at an annual or special meeting in accordance with those rules.

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Action by Written Consent

Denmark. Under Danish law, shareholders may take action and pass resolutions by written consent if such consent is unanimous. However, for a listed company, this method of adopting resolutions is generally not feasible.

Delaware. Although permitted by Delaware law, publicly listed companies do not typically permit stockholders of a corporation to take action by written consent.

Appraisal Rights

Denmark. The concept of appraisal rights does not exist under Danish law, except in connection with statutory redemption rights according to the DCA.

According to Section 73 of the DCA, a minority shareholder may require a majority shareholder that holds more than nine-tenths of the company's registered share capital and voting rights to redeem his or her shares. Similarly, shares in a company may be redeemed in whole or in part by a shareholder holding more than nine-tenths of the shares and the corresponding voting rights in the company, according to Section 70 of the DCA. In the event that the parties cannot agree to the redemption squeeze out price, this shall be determined by an independent evaluator appointed by the court. Additionally, there are specific regulations in Sections 249, 267, 285 and 305 of the DCA that require compensation in the event of national or cross-border mergers and demergers. Moreover, shareholders who vote against a cross-border merger or demerger are, according to Sections 286 and 306 of the DCA, entitled to have their shares redeemed.

Delaware. The Delaware General Corporation Law provides for stockholder appraisal rights, or the right to demand payment in cash of the judicially determined fair value of the stockholder's shares, in connection with certain mergers and consolidations.

Shareholder Suits

Denmark. Under Danish law, only a company itself can bring a civil action against a third party; an individual shareholder does not have the right to bring an action on behalf of a company. However, if shareholders representing at least one-tenth of the share capital have opposed at a general meeting a decision to grant discharge to a member of our board of directors or our executive management or refrain from bringing law suits against, among other persons, a member of our board of directors or executive management, a shareholder may bring a derivative action on behalf of our company against, among other persons, a member of our board of directors or executive management. An individual shareholder may, in its own name, have an individual right to take action against such third party in the event that the cause for the liability of that third party also constitutes a negligent act directly against such individual shareholder.

Delaware. Under the Delaware General Corporation Law, a stockholder may bring a derivative action on behalf of the corporation to enforce the rights of the corporation. An individual also may commence a class action suit on behalf of himself and other similarly situated stockholders where the requirements for maintaining a class action under Delaware law have been met. A person may institute and maintain such a suit only if that person was a stockholder at the time of the transaction which is the subject of the suit. In addition, under Delaware case law, the plaintiff normally must be a stockholder at the time of the transaction that is the subject of the suit and throughout the duration of the derivative suit. Delaware law also requires that the derivative plaintiff make a demand on the directors of the corporation to assert the corporate claim before the suit may be prosecuted by the derivative plaintiff in court, unless such a demand would be futile.

Repurchase of Shares

Denmark. Danish limited liability companies may not subscribe for newly issued shares in their own capital. Such companies may, however, according to the DCA Sections 196-201, acquire fully paid shares of themselves,

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provided that the board of directors has been authorized to do so by the shareholders at a general meeting. Such authorization can only be given for a maximum period of five years and the authorization shall fix (i) the maximum value of the shares and (ii) the minimum and the highest amount that the company may pay for the shares. Such purchase of shares may generally only be acquired using distributable reserves. In addition, the board of directors may, on behalf of the company, acquire the company's own shares, without authorization, in case it is necessary to avoid a considerable and imminent detrimental effect on the company and provided certain conditions are met. In case the company has acquired its own shares under such circumstances the board of directors is obligated to inform the shareholders of such acquisition at the next general meeting. See “—Authorizations to our Board of Directors.”

Delaware. Under the Delaware General Corporation Law, a corporation may purchase or redeem its own shares unless the capital of the corporation is impaired or the purchase or redemption would cause an impairment of the capital of the corporation. A Delaware corporation may, however, purchase or redeem out of capital any of its preferred shares or, if no preferred shares are outstanding, any of its own shares if such shares will be retired upon acquisition and the capital of the corporation will be reduced in accordance with specified limitations.

Anti-Takeover Provisions

Denmark. Under Danish law, it is possible to implement limited protective anti-takeover measures. Such provisions may include, among other things, (i) different share classes with different voting rights and (ii) notification requirements concerning participation in general meetings. We have currently not adopted any such provisions, except for the notification requirements concerning participation in general meetings. See description above under the caption “—Articles of Association and Danish Corporate Law—General Meetings and Voting Rights.”

Delaware. In addition to other aspects of Delaware law governing fiduciary duties of directors during a potential takeover, the Delaware General Corporation Law also contains a business combination statute that protects Delaware companies from hostile takeovers and from actions following the takeover by prohibiting some transactions once an acquirer has gained a significant holding in the corporation.

Section 203 of the Delaware General Corporation Law prohibits “business combinations,” including mergers, sales and leases of assets, issuances of securities and similar transactions by a corporation or a subsidiary with an interested stockholder that beneficially owns 15% or more of a corporation's voting stock, within three years after the person becomes an interested stockholder, unless:

- the transaction that will cause the person to become an interested stockholder is approved by the board of directors of the target prior to the transaction;
- after the completion of the transaction in which the person becomes an interested stockholder, the interested stockholder holds at least 85% of the voting stock of the corporation not including shares owned by persons who are directors and officers of interested stockholders and shares owned by specified employee benefit plans; or
- after the person becomes an interested stockholder, the business combination is approved by the board of directors of the corporation and holders of at least 66.67% of the outstanding voting stock, excluding shares held by the interested stockholder.

A Delaware corporation may elect not to be governed by Section 203 by a provision contained in the original certificate of incorporation of the corporation or an amendment to the original certificate of incorporation or to the bylaws of the company, which amendment must be approved by a majority of the shares entitled to vote and may not be further amended by the board of directors of the corporation. Such an amendment is not effective until 12 months following its adoption.

Inspection of Books and Records

Denmark. According to Section 150 of the DCA, a shareholder may, at the annual general meeting or at a general meeting whose agenda includes such item, request an inspection of the company's books regarding specific issues concerning the management of the company or specific annual reports. If approved by shareholders with a simple majority, one or more investigators are elected. If the proposal is not approved by a simple majority but 25% of the share capital votes in favor of the proposal, then any shareholder may, no later than four weeks after the general meeting, request the bankruptcy court for the district in which the company's registered office is situated to appoint investigators.

Delaware. Under the Delaware General Corporation Law, any stockholder may inspect certain of the corporation's books and records, for any proper purpose, during the corporation's usual hours of business.

Pre-Emptive Rights

Denmark. If our shareholders at a general meeting resolve to increase our share capital by a cash contribution, section 162 of the DCA will apply. Under that section, shareholders have a pre-emptive right to subscribe for new shares in proportion to their existing shareholdings. However, the pre-emptive right may be derogated from by a majority comprising at least two-thirds of the votes cast, as well as at least two-thirds of the share capital represented at the general meeting, provided the share capital increase takes place at market price or nine-tenths of the votes cast, as well as at least nine-tenths of the share capital represented at the general meeting if the share capital increase takes place below market price, unless (i) such capital increase is directed at certain but not all shareholders (in which case all shareholders must consent); or (ii) such capital increase is directed at our employees whereby a majority comprising at least two-thirds of the votes cast, as well as at least two-thirds of the share capital represented at the general meeting is required. Further, the pre-emptive rights may be derogated from by an exercise of the board of directors of a valid authorization in our articles of association, provided that the share capital increase takes place at or above market price. The board of directors may resolve to increase our share capital without pre-emptive subscription rights for existing shareholders pursuant to the authorizations described above under the caption "—Authorizations to our Board of Directors."

Unless future issuances of new shares are registered under the Securities Act or with any authority outside Denmark, U.S. shareholders and shareholders in jurisdictions outside Denmark may be unable to exercise their pre-emptive subscription rights under the law of their respective jurisdictions, including the U.S. securities law.

Delaware. Under the Delaware General Corporation Law, stockholders have no pre-emptive rights to subscribe for additional issues of stock or to any security convertible into such stock unless, and to the extent that, such rights are expressly provided for in the certificate of incorporation.

Dividends

Denmark. Under Danish law, the distribution of ordinary and interim dividends requires the approval of a company's shareholders at a company's general meeting. In addition the shareholders may authorize the board of directors to distribute interim dividends. We may only pay out dividends from our distributable reserves, which are defined as results from operations carried forward and reserves that are not bound by law after deduction of loss carried forward. It is possible under Danish law to pay out interim dividends. The decision to pay out interim dividends shall be accompanied by a balance sheet, and the board of directors determines whether it will be sufficient to use the statement of financial position from the annual report or if an interim statement of financial position for the period from the annual report period until the interim dividend payment shall be prepared. If the decision to distribute interim dividends is passed more than six months after the date of the statement of financial position as set out in our latest adopted annual report, an interim statement of financial position must be prepared and reviewed by our auditor. The statement of financial position or the interim statement of financial position, as applicable, must show that sufficient funds are available for distribution. Our general meeting of shareholders

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cannot resolve to distribute dividends at an amount exceeding the amount recommended or approved by our board of directors. Moreover, ordinary dividends and interim dividends may only be made out of distributable reserves and may not exceed what is considered sound and adequate with regard to our financial condition or be to the detriment of our creditors.

Delaware. Under the Delaware General Corporation Law, a Delaware corporation may pay dividends out of its surplus (the excess of net assets over capital), or in case there is no surplus, out of its net profits for the fiscal year in which the dividend is declared and/or the preceding fiscal year (provided that the amount of the capital of the corporation is not less than the aggregate amount of the capital represented by the issued and outstanding stock of all classes having a preference upon the distribution of assets). In determining the amount of surplus of a Delaware corporation, the assets of the corporation, including stock of subsidiaries owned by the corporation, must be valued at their fair market value as determined by the board of directors, without regard to their historical book value. Dividends may be paid in the form of shares, property or cash.

Shareholder Vote on Certain Reorganizations

Denmark. Under Danish law, all amendments to the articles of association shall be approved by the general meeting of shareholders with at least two-thirds of the votes cast and two-thirds of 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. The same applies to solvent liquidations, mergers with the company as the discontinuing entity, mergers with the company as the continuing entity if shares are issued in connection therewith and demergers. Under Danish law, it is debatable whether the shareholders must approve a decision to sell all or virtually all of the company's business/assets.

Delaware. Under the Delaware General Corporation Law, the vote of a majority of the outstanding shares of capital stock entitled to vote thereon generally is necessary to approve a merger or consolidation or the sale of all or substantially all of the assets of a corporation. The Delaware General Corporation Law permits a corporation to include in its certificate of incorporation a provision requiring for any corporate action the vote of a larger portion of the stock or of any class or series of stock than would otherwise be required. However, under the Delaware General Corporation Law, no vote of the stockholders of a surviving corporation to a merger is needed, unless required by the certificate of incorporation, if (1) the agreement of merger does not amend in any respect the certificate of incorporation of the surviving corporation, (2) the shares of stock of the surviving corporation are not changed in the merger and (3) the number of ordinary shares of the surviving corporation into which any other shares, securities or obligations to be issued in the merger may be converted does not exceed 20% of the surviving corporation's shares outstanding immediately prior to the effective date of the merger. In addition, stockholders may not be entitled to vote in certain mergers with other corporations that own 90% or more of the outstanding shares of each class of stock of such corporation, but the stockholders will be entitled to appraisal rights.

Mandatory Redemption of Shares

Denmark: Where a shareholder holds more than nine-tenths of the shares in a company and a corresponding proportion of the voting rights, such shareholder may, pursuant to the DCA, Section 70, demand that the other shareholders have their shares redeemed by that shareholder. In this case, the other shareholders must be requested, under the rules governing notices for general meeting, to transfer their shares to the shareholder within four weeks after the request to transfer their shares. In addition, the other shareholders shall through the Danish Business Authority's IT system be requested to transfer their shares within the same four-week period. Specific requirements apply to the contents of the notices to the other shareholders regarding the redemption. If the redemption price cannot be agreed upon, the redemption price must be determined by an independent expert appointed by the court in the jurisdiction of the company's registered office in accordance with the provisions of the DCA. However, the redemption price will be deemed fair under any circumstances, provided that (i) the redemption price is equal to the consideration paid by the bidder in connection with a voluntary tender offer by

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which the bidder obtained at least 90% of the voting rights or (ii) the redemption price is equal to the consideration paid by the bidder in connection with a mandatory tender offer. To the extent any minority shareholders have not transferred their shares to the acquiring shareholder before the expiry of the four-week period, the redeeming shareholder shall pay the redemption price to the remaining minority shareholders through the securities deposit. Upon such payment through the securities deposit, the minority shareholders will have been redeemed and the minority shareholders shall in such case through the Danish Business Authority's IT system be notified that the right to require determination of the redemption price by the independent expert expires at the end of a period, which cannot be less than three months pursuant to the DCA, Section 72.

Furthermore, where a shareholder holds more than nine-tenths of the shares in a company and a corresponding proportion of the voting rights, the other shareholders may require such shareholder to acquire their shares pursuant to Section 73 of the DCA. If the redemption price cannot be agreed upon, the redemption price must be determined by an independent expert appointed by the court in the jurisdiction of the company's registered office in accordance with the provisions of the DCA. Expenses relating to the determination of the redemption price must be paid by the shareholder requesting such determination. If the expert's valuation is higher than the price offered by the redeeming shareholder, the court may order the redeeming shareholder to pay the expenses relating to determination of the redemption price in full or in part.

Delaware: The Delaware General Corporation Law provides for stockholder appraisal rights, or the right to demand payment in cash of the judicially determined fair value of the stockholder's shares, in connection with certain mergers and consolidations.

Amendments to Governing Documents

Denmark. All resolutions made by the general meeting may be adopted by a simple majority of the votes, subject only to the mandatory provisions of the DCA and the articles of association. Resolutions concerning all amendments to the articles of association must be passed by two-thirds of the votes cast as well as two-thirds of 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.

Certain resolutions, which limit a shareholder's ownership or voting rights, are subject to approval by at least a nine-tenth majority of the votes cast and the share capital represented at the general meeting. Decisions to impose any or increase any obligations of the shareholders towards the company require unanimity.

Delaware. Under the Delaware General Corporation Law, a corporation's certificate of incorporation may be amended only if adopted and declared advisable by the board of directors and approved by a majority of the outstanding shares entitled to vote, and the bylaws may be amended with the approval of a majority of the outstanding shares entitled to vote and may, if so provided in the certificate of incorporation, also be amended by the board of directors.

Exchange Controls

There are no laws or regulations in Denmark that restrict the export or import of capital (except for certain investments in certain domains in accordance with applicable resolutions adopted by the United Nations or the European Union), including, but not limited to, foreign exchange controls, or which affect the remittance of dividends, interest or other payments to non-resident holders of our ordinary shares.

Transfer Agent and Registrar

The transfer agent and registrar for our shares is Computershare A/S, Lottenborgvej 26 D, 1., DK-2800 Kgs. Lyngby, Denmark. The Bank of New York Mellon serves as the depositary, registrar and transfer agent for the ADSs.

DESCRIPTION OF AMERICAN DEPOSITARY SHARES

American Depositary Shares

The Bank of New York Mellon, as depositary, registers and delivers ADSs. Each ADS represents one ordinary share (or a right to receive one ordinary share) deposited with Danske Bank A/S, as custodian for the depositary in the Kingdom of Denmark. Each ADS also represents any other securities, cash or other property that may be held by the depositary. The deposited shares together with any other securities, cash or other property held by the depositary are referred to as the deposited securities. A copy of our Deposit Agreement among us, the depositary, owners and holders of ADSs was filed with the SEC as an exhibit to our Annual Report on Form 20-F for the year ended December 31, 2020 filed on March 2, 2021 (File No. 001-39545).

Any ordinary shares that may be issued pursuant to this prospectus supplement and the accompanying prospectus, whether directly or upon exercise of warrants, can be deposited for delivery of ADSs. The ADSs may be uncertificated securities or certificated securities evidenced by American Depositary Receipts, or ADRs. The depositary's office at which the ADSs will be administered and its principal executive office are located at 240 Greenwich Street, New York, New York 10286.

You may hold ADSs either (A) directly (i) by having an American Depositary Receipt, also referred to as an ADR, which is a certificate evidencing a specific number of ADSs, registered in your name, or (ii) by having uncertificated ADSs registered in your name, or (B) indirectly by holding a security entitlement in ADSs through your broker or other financial institution that is a direct or indirect participant in DTC. If you hold ADSs directly, you are a registered ADS holder, also referred to as an ADS holder. This description assumes you are an ADS holder. If you hold the ADSs indirectly, you must rely on the procedures of your broker or other financial institution to assert the rights of ADS holders described in this section. You should consult with your broker or financial institution to find out what those procedures are.

Registered holders of uncertificated ADSs will receive statements from the depositary confirming their holdings.

As an ADS holder, we will not treat you as one of our shareholders and you will not have shareholder rights. Danish law governs shareholder rights. The depositary will be the holder of the shares underlying your ADSs. As a registered holder of ADSs, you will have ADS holder rights. A deposit agreement among us, the depositary, ADS holders and all other persons indirectly or beneficially holding ADSs sets out ADS holder rights as well as the rights and obligations of the depositary. New York law governs the deposit agreement and the ADSs.

The following is a summary of the material provisions of the deposit agreement. For more complete information, you should read the entire deposit agreement and the form of ADR. Directions on how to obtain copies of those documents are provided in the section titled "Where You Can Find Additional Information."

Dividends and Other Distributions

How Will You Receive Dividends and Other Distributions on the Shares?

The depositary has agreed to pay or distribute to ADS holders the cash dividends or other distributions it or the custodian receives on shares or other deposited securities, upon payment or deduction of its fees and expenses. You will receive these distributions in proportion to the number of shares your ADSs represent.

Cash

The depositary will convert any cash dividend or other cash distribution we pay on the shares into U.S. dollars, if it can do so on a reasonable basis and can transfer the U.S. dollars to the United States. If that is not possible or if any government approval is needed and cannot be obtained, the deposit agreement allows the

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depository to distribute the foreign currency only to those ADS holders to whom it is possible to do so. It will hold the foreign currency it cannot convert for the account of the ADS holders who have not been paid. It will not invest the foreign currency and it will not be liable for any interest.

Before making a distribution, any withholding taxes, or other governmental charges that must be paid will be deducted. See "Taxation." The depository will distribute only whole U.S. dollars and cents and will round fractional cents to the nearest whole cent. If the exchange rates fluctuate during a time when the depository cannot convert the foreign currency, you may lose some of the value of the distribution.

Shares

The depository may distribute additional ADSs representing any shares we distribute as a dividend or free distribution. The depository will only distribute whole ADSs. It will sell shares which would require it to deliver a fraction of an ADS (or ADSs representing those shares) and distribute the net proceeds in the same way as it does with cash. If the depository does not distribute additional ADSs, the outstanding ADSs will also represent the new shares. The depository may sell a portion of the distributed shares (or ADSs representing those shares) sufficient to pay its fees and expenses in connection with that distribution.

Rights to Purchase Additional Shares

If we offer holders of our securities any rights to subscribe for additional shares or any other rights, the depository may (i) exercise those rights on behalf of ADS holders, (ii) distribute those rights to ADS holders or (iii) sell those rights and distribute the net proceeds to ADS holders, in each case after deduction or upon payment of its fees and expenses. To the extent the depository does not do any of those things, it will allow the rights to lapse. In that case, you will receive no value for them. The depository will exercise or distribute rights only if we ask it to and provide satisfactory assurances to the depository that it is legal to do so. If the depository will exercise rights, it will purchase the securities to which the rights relate and distribute those securities or, in the case of shares, new ADSs representing the new shares, to subscribing ADS holders, but only if ADS holders have paid the exercise price to the depository. U.S. securities laws may restrict the ability of the depository to distribute rights or ADSs or other securities issued on exercise of rights to all or certain ADS holders, and the securities distributed may be subject to restrictions on transfer.

Other Distributions

The depository will send to ADS holders anything else we distribute on deposited securities by any means it thinks is legal, fair and practical. If it cannot make the distribution in that way, the depository has a choice. It may decide to sell what we distributed and distribute the net proceeds, in the same way as it does with cash. Or, it may decide to hold what we distributed, in which case ADSs will also represent the newly distributed property. However, the depository is not required to distribute any securities (other than ADSs) to ADS holders unless it receives satisfactory evidence from us that it is legal to make that distribution. The depository may sell a portion of the distributed securities or property sufficient to pay its fees and expenses in connection with that distribution. U.S. securities laws may restrict the ability of the depository to distribute securities to all or certain ADS holders, and the securities distributed may be subject to restrictions on transfer.

The depository is not responsible if it decides that it is unlawful or impractical to make a distribution available to any ADS holders. We have no obligation to register ADSs, shares, rights or other securities under the Securities Act. We also have no obligation to take any other action to permit the distribution of ADSs, shares, rights or anything else to ADS holders. This means that you may not receive the distributions we make on our shares or any value for them if it is illegal or impractical for us to make them available to you.

Deposit, Withdrawal and Cancellation

How are ADSs Issued?

The depositary will deliver ADSs if you or your broker deposits shares or evidence of rights to receive shares with the custodian. Upon payment of its fees and expenses and of any taxes or charges, such as stamp taxes or stock transfer taxes or fees, the depositary will register the appropriate number of ADSs in the names you request and will deliver the ADSs to or upon the order of the person or persons that made the deposit.

How Can ADS Holders Withdraw the Deposited Securities?

You may surrender your ADSs to the depositary for the purpose of withdrawal. Upon payment of its fees and expenses and of any taxes or charges, such as stamp taxes or stock transfer taxes or fees, the depositary will deliver the shares and any other deposited securities underlying the ADSs to the ADS holder or a person the ADS holder designates at the office of the custodian. Or, at your request, risk and expense, the depositary will deliver the deposited securities at its office, if feasible. However, the depositary is not required to accept surrender of ADSs to the extent it would require delivery of a fraction of a deposited share or other security. The depositary may charge you a fee and its expenses for instructing the custodian regarding delivery of deposited securities.

How do ADS Holders Interchange Between Certificated ADSs and Uncertificated ADSs?

You may surrender your ADR to the depositary for the purpose of exchanging your ADR for uncertificated ADSs. The depositary will cancel that ADR and will send to the ADS holder a statement confirming that the ADS holder is the registered holder of uncertificated ADSs. Upon receipt by the depositary of a proper instruction from a registered holder of uncertificated ADSs requesting the exchange of uncertificated ADSs for certificated ADSs, the depositary will execute and deliver to the ADS holder an ADR evidencing those ADSs.

Voting Rights

How do You Vote?

ADS holders may instruct the depositary how to vote the number of deposited shares their ADSs represent. If we request the depositary to solicit your voting instructions (and we are not required to do so), the depositary will notify you of a shareholders' meeting and send or make voting materials available to you. Those materials will describe the matters to be voted on and explain how ADS holders may instruct the depositary how to vote. For instructions to be valid, they must reach the depositary by a date set by the depositary. The depositary will try, as far as practical, subject to the laws of the Kingdom of Denmark and the provisions of our articles of association or similar documents, to vote or to have its agents vote the shares or other deposited securities as instructed by ADS holders. If we do not request the depositary to solicit your voting instructions, you can still send voting instructions, and, in that case, the depositary may try to vote as you instruct, but it is not required to do so.

Except by instructing the depositary as described above, you won't be able to exercise voting rights unless you surrender your ADSs and withdraw the shares. However, you may not know about the meeting enough in advance to withdraw the shares. In any event, the depositary will not exercise any discretion in voting deposited securities and it will only vote or attempt to vote as instructed.

We cannot assure you that you will receive the voting materials in time to ensure that you can instruct the depositary to vote your shares. In addition, the depositary and its agents are not responsible for failing to carry out voting instructions or for the manner of carrying out voting instructions. This means that you may not be able to exercise voting rights and there may be nothing you can do if your shares are not voted as you requested.

In order to give you a reasonable opportunity to instruct the depositary as to the exercise of voting rights relating to Deposited Securities, if we request the Depositary to act, we will provide the depositary with the proposed meeting date and details of the matters proposed to be voted on at least 30 days before the meeting date and the depositary will send voting materials to you approximately 21 days before the meeting date.

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Fees and Expenses

<i>Persons depositing or withdrawing shares or ADS holders must pay:</i>	<i>For:</i>
\$5.00 (or less) per 100 ADSs (or portion of 100 ADSs)	Issuance of ADSs, including issuances resulting from a distribution of shares or rights or other property Cancellation of ADSs for the purpose of withdrawal, including if the deposit agreement terminates
\$.05 (or less) per ADS	Any cash distribution to ADS holders
A fee equivalent to the fee that would be payable if securities distributed to you had been shares and the shares had been deposited for issuance of ADSs	Distribution of securities distributed to holders of deposited securities (including rights) that are distributed by the depository to ADS holders
\$.05 (or less) per ADS per calendar year	Depository services
Registration or transfer fees	Transfer and registration of shares on our share register to or from the name of the depository or its agent when you deposit or withdraw shares
Expenses of the depository	Cable (including SWIFT) and facsimile transmissions (when expressly provided in the deposit agreement) Converting foreign currency to U.S. dollars
Taxes and other governmental charges the depository or the custodian has to pay on any ADSs or shares underlying ADSs, such as stock transfer taxes, stamp duty or withholding taxes	As necessary
Any charges incurred by the depository or its agents for servicing the deposited securities	As necessary

The depository collects its fees for delivery and surrender of ADSs directly from investors depositing shares or surrendering ADSs for the purpose of withdrawal or from intermediaries acting for them. The depository collects fees for making distributions to investors by deducting those fees from the amounts distributed or by selling a portion of distributable property to pay the fees. The depository may collect its annual fee for depository services by deduction from cash distributions or by directly billing investors or by charging the book-entry system accounts of participants acting for them. The depository may collect any of its fees by deduction from any cash distribution payable (or by selling a portion of securities or other property distributable) to ADS holders that are obligated to pay those fees. The depository may generally refuse to provide fee-attracting services until its fees for those services are paid.

From time to time, the depository may make payments to us to reimburse us for costs and expenses generally arising out of establishment and maintenance of the ADS program, waive fees and expenses for services provided to us by the depository or share revenue from the fees collected from ADS holders. In performing its duties under the deposit agreement, the depository may use brokers, dealers, foreign currency dealers or other service providers that are owned by or affiliated with the depository and that may earn or share fees, spreads or commissions.

The depository may convert currency itself or through any of its affiliates, or the custodian or we may convert currency and pay U.S. dollars to the depository. Where the depository converts currency itself or through any of its affiliates, the depository acts as principal for its own account and not as agent, advisor, broker or fiduciary on behalf of any other person and earns revenue, including, without limitation, transaction spreads, that it will retain for its own account. The revenue is based on, among other things, the difference between the

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exchange rate assigned to the currency conversion made under the deposit agreement and the rate that the depository or its affiliate receives when buying or selling foreign currency for its own account. The depository makes no representation that the exchange rate used or obtained by it or its affiliate in any currency conversion under the deposit agreement will be the most favorable rate that could be obtained at the time or that the method by which that rate will be determined will be the most favorable to ADS holders, subject to the depository's obligation to act without negligence or bad faith. The methodology used to determine exchange rates used in currency conversions made by the depository is available upon request. Where the custodian converts currency, the custodian has no obligation to obtain the most favorable rate that could be obtained at the time or to ensure that the method by which that rate will be determined will be the most favorable to ADS holders, and the depository makes no representation that the rate is the most favorable rate and will not be liable for any direct or indirect losses associated with the rate. In certain instances, the depository may receive dividends or other distributions from us in U.S. dollars that represent the proceeds of a conversion of foreign currency or translation from foreign currency at a rate that was obtained or determined by us and, in such cases, the depository will not engage in, or be responsible for, any foreign currency transactions and neither it nor we make any representation that the rate obtained or determined by us is the most favorable rate and neither it nor we will be liable for any direct or indirect losses associated with the rate.

Payment of Taxes

You will be responsible for any taxes or other governmental charges payable on your ADSs or on the deposited securities represented by any of your ADSs. The depository may refuse to register any transfer of your ADSs or allow you to withdraw the deposited securities represented by your ADSs until those taxes or other charges are paid. It may apply payments owed to you or sell deposited securities represented by your ADSs to pay any taxes owed and you will remain liable for any deficiency. If the depository sells deposited securities, it will, if appropriate, reduce the number of ADSs to reflect the sale and pay to ADS holders any proceeds, or send to ADS holders any property, remaining after it has paid the taxes.

Tender and Exchange Offers; Redemption, Replacement or Cancellation of Deposited Securities

The depository will not tender deposited securities in any voluntary tender or exchange offer unless instructed to do so by an ADS holder surrendering ADSs and subject to any conditions or procedures the depository may establish.

If deposited securities are redeemed for cash in a transaction that is mandatory for the depository as a holder of deposited securities, the depository will call for surrender of a corresponding number of ADSs and distribute the net redemption money to the holders of called ADSs upon surrender of those ADSs.

If there is any change in the deposited securities such as a sub-division, combination or other reclassification, or any merger, consolidation, recapitalization or reorganization affecting the issuer of deposited securities in which the depository receives new securities in exchange for or in lieu of the old deposited securities, the depository will hold those replacement securities as deposited securities under the deposit agreement. However, if the depository decides it would not be lawful and practical to hold the replacement securities because those securities could not be distributed to ADS holders or for any other reason, the depository may instead sell the replacement securities and distribute the net proceeds upon surrender of the ADSs.

If there is a replacement of the deposited securities and the depository will continue to hold the replacement securities, the depository may distribute new ADSs representing the new deposited securities or ask you to surrender your outstanding ADRs in exchange for new ADRs identifying the new deposited securities.

If there are no deposited securities underlying ADSs, including if the deposited securities are cancelled, or if the deposited securities underlying ADSs have become apparently worthless, the depository may call for surrender of those ADSs or cancel those ADSs upon notice to the ADS holders.

Amendment and Termination

How May the Deposit Agreement be Amended?

We may agree with the depository to amend the deposit agreement and the ADRs without your consent for any reason. If an amendment adds or increases fees or charges, except for taxes and other governmental charges or expenses of the depository for registration fees, facsimile costs, delivery charges or similar items, or prejudices a substantial right of ADS holders, it will not become effective for outstanding ADSs until 30 days after the depository notifies ADS holders of the amendment. At the time an amendment becomes effective, you are considered, by continuing to hold your ADSs, to agree to the amendment and to be bound by the ADRs and the deposit agreement as amended.

How May the Deposit Agreement be Terminated?

The depository will initiate termination of the deposit agreement if we instruct it to do so. The depository may initiate termination of the deposit agreement if

- 60 days have passed since the depository told us it wants to resign but a successor depository has not been appointed and accepted its appointment;
- we delist the ADSs from an exchange in the United States on which they were listed and do not list the ADSs on another exchange in the United States or make arrangements for trading of ADSs on the U.S. over-the-counter market;
- we delist our shares from an exchange outside the United States on which they were listed and do not list the shares on another exchange outside the United States;
- the depository has reason to believe the ADSs have become, or will become, ineligible for registration on Form F-6 under the Securities Act;
- we appear to be insolvent or enter insolvency proceedings;
- all or substantially all the value of the deposited securities has been distributed either in cash or in the form of securities;
- there are no deposited securities underlying the ADSs or the underlying deposited securities have become apparently worthless; or
- there has been a replacement of deposited securities.

If the deposit agreement will terminate, the depository will notify ADS holders at least 90 days before the termination date. At any time after the termination date, the depository may sell the deposited securities. After that, the depository will hold the money it received on the sale, as well as any other cash it is holding under the deposit agreement, unsegregated and without liability for interest, for the pro rata benefit of the ADS holders that have not surrendered their ADSs. Normally, the depository will sell as soon as practicable after the termination date.

After the termination date and before the depository sells, ADS holders can still surrender their ADSs and receive delivery of deposited securities, except that the depository may refuse to accept a surrender for the purpose of withdrawing deposited securities or reverse previously accepted surrenders of that kind that have not settled if it would interfere with the selling process. The depository may refuse to accept a surrender for the purpose of withdrawing sale proceeds until all the deposited securities have been sold. The depository will continue to collect distributions on deposited securities, but, after the termination date, the depository is not required to register any transfer of ADSs or distribute any dividends or other distributions on deposited securities to the ADSs holder (until they surrender their ADSs) or give any notices or perform any other duties under the deposit agreement except as described in this paragraph.

Limitations on Obligations and Liability

Limits on our Obligations and the Obligations of the Depository; Limits on Liability to Holders of ADSs

The deposit agreement expressly limits our obligations and the obligations of the depository. It also limits our liability and the liability of the depository. We and the depository:

- are only obligated to take the actions specifically set forth in the deposit agreement without negligence or bad faith, and the depository will not be a fiduciary or have any fiduciary duty to holders of ADSs;
- are not liable if we are or it is prevented or delayed by law or by events or circumstances beyond our or its ability to prevent or counteract with reasonable care or effort from performing our or its obligations under the deposit agreement;
- are not liable if we or it exercises discretion permitted under the deposit agreement;
- are not liable for the inability of any holder of ADSs to benefit from any distribution on deposited securities that is not made available to holders of ADSs under the terms of the deposit agreement, or for any special, consequential or punitive damages for any breach of the terms of the deposit agreement;
- have no obligation to become involved in a lawsuit or other proceeding related to the ADSs or the deposit agreement on your behalf or on behalf of any other person;
- may rely upon any documents we believe or it believes in good faith to be genuine and to have been signed or presented by the proper person;
- are not liable for the acts or omissions of any securities depository, clearing agency or settlement system; and
- the depository has no duty to make any determination or provide any information as to our tax status, or any liability for any tax consequences that may be incurred by ADS holders as a result of owning or holding ADSs or be liable for the inability or failure of an ADS holder to obtain the benefit of a foreign tax credit, reduced rate of withholding or refund of amounts withheld in respect of tax or any other tax benefit.

In the deposit agreement, we and the depository agree to indemnify each other under certain circumstances.

Requirements for Depository Actions

Before the depository will deliver or register a transfer of ADSs, make a distribution on ADSs, or permit withdrawal of shares, the depository may require:

- payment of stock transfer or other taxes or other governmental charges and transfer or registration fees charged by third parties for the transfer of any shares or other deposited securities;
- satisfactory proof of the identity and genuineness of any signature or other information it deems necessary; and
- compliance with regulations it may establish, from time to time, consistent with the deposit agreement, including presentation of transfer documents.

The depository may refuse to deliver ADSs or register transfers of ADSs when the transfer books of the depository or our transfer books are closed or at any time if the depository or we think it advisable to do so.

Your Right to Receive the Shares Underlying your ADSs

ADS holders have the right to cancel their ADSs and withdraw the underlying shares at any time except:

- when temporary delays arise because: (i) the depository has closed its transfer books or we have closed our transfer books; (ii) the transfer of shares is blocked to permit voting at a shareholders' meeting; or (iii) we are paying a dividend on our shares;

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- when you owe money to pay fees, taxes and similar charges; or
- when it is necessary to prohibit withdrawals in order to comply with any laws or governmental regulations that apply to ADSs or to the withdrawal of shares or other deposited securities.

This right of withdrawal may not be limited by any other provision of the deposit agreement.

Direct Registration System

In the deposit agreement, all parties to the deposit agreement acknowledge that the Direct Registration System, also referred to as DRS, and Profile Modification System, also referred to as Profile, will apply to the ADSs. DRS is a system administered by DTC that facilitates interchange between registered holding of uncertificated ADSs and holding of security entitlements in ADSs through DTC and a DTC participant. Profile is a feature of DRS that allows a DTC participant, claiming to act on behalf of a registered holder of uncertificated ADSs, to direct the depository to register a transfer of those ADSs to DTC or its nominee and to deliver those ADSs to the DTC account of that DTC participant without receipt by the depository of prior authorization from the ADS holder to register that transfer.

In connection with and in accordance with the arrangements and procedures relating to DRS/Profile, the parties to the deposit agreement understand that the depository will not determine whether the DTC participant that is claiming to be acting on behalf of an ADS holder in requesting registration of transfer and delivery as described in the paragraph above has the actual authority to act on behalf of the ADS holder (notwithstanding any requirements under the Uniform Commercial Code). In the deposit agreement, the parties agree that the depository's reliance on and compliance with instructions received by the depository through the DRS/Profile system and in accordance with the deposit agreement will not constitute negligence or bad faith on the part of the depository.

Shareholder Communications; Inspection of Register of Holders of ADSs

The depository will make available for your inspection at its office all communications that it receives from us as a holder of deposited securities that we make generally available to holders of deposited securities. The depository will send you copies of those communications or otherwise make those communications available to you if we ask it to which we may choose not to. You have a right to inspect the register of holders of ADSs, but not for the purpose of contacting those holders about a matter unrelated to our business or the ADSs.

Jury Trial Waiver

The deposit agreement provides that, to the extent permitted by law, ADS holders waive the right to a jury trial of any claim they may have against us or the depository arising out of or relating to our shares, the ADSs or the deposit agreement, including any claim under the U.S. federal securities laws. If we or the depository opposed a jury trial demand based on the waiver, the court would determine whether the waiver was enforceable in the facts and circumstances of that case in accordance with applicable case law.

You will not, by agreeing to the terms of the deposit agreement, be deemed to have waived our or the depository's compliance with U.S. federal securities laws or the rules and regulations promulgated thereunder.

PLAN OF DISTRIBUTION

We have entered into a sales agreement with Cowen and Company, LLC, or Cowen, under which we may issue and sell from time to time up to \$50,000,000 of our ordinary shares represented by ADSs through Cowen as our sales agent. Sales of our ADSs, if any, will be made at market prices by any method that is deemed to be an “at the market offering” as defined in Rule 415(a)(4) under the Securities Act, including sales made directly on or through The Nasdaq Global Select Market or any other trading market for our ADSs. If authorized by us in writing, Cowen may purchase our ADSs as principal.

Cowen will offer our ADSs subject to the terms and conditions of the sales agreement on a daily basis or as otherwise agreed upon by us and Cowen. We will designate the maximum amount of ADSs to be sold through Cowen on a daily basis or otherwise determine such maximum amount together with Cowen. Subject to the terms and conditions of the sales agreement, Cowen will use its commercially reasonable efforts to sell on our behalf all of the ADSs requested to be sold by us. We may instruct Cowen not to sell ADSs if the sales cannot be effected at or above the price designated by us in any such instruction. Cowen or we may suspend the offering of our ADSs being made through Cowen under the sales agreement upon proper notice to the other party. Cowen and we each have the right, by giving written notice as specified in the sales agreement, to terminate the sales agreement in each party’s sole discretion at any time.

The aggregate compensation payable to Cowen as sales agent equals 3.0% of the gross sales price of the ADSs sold through it pursuant to the sales agreement. We have also agreed to reimburse Cowen for Cowen’s actual outside legal expenses, including fees and disbursements of counsel incurred by Cowen, up to a maximum aggregate amount of \$85,000 (inclusive of any amounts reimbursed for associated legal expenses of Cowen’s outside counsel for filings with FINRA). We estimate that the total expenses of the offering payable by us, excluding commissions payable to Cowen under the sales agreement, will be approximately \$300,000.

The remaining sales proceeds, after deducting any expenses payable by us and any transaction fees imposed by any governmental, regulatory, or self-regulatory organization in connection with the sales, will equal our net proceeds for the sale of such ADSs.

Cowen will provide written confirmation to us following the close of trading on The Nasdaq Global Select Market on each day in which ADSs are sold through it as sales agent under the sales agreement. Each confirmation will include the number of ADSs sold through it as sales agent on that day, the volume weighted average price of the ADSs sold, the percentage of the daily trading volume and the net proceeds to us.

The aggregate number of any such ADSs sold through Cowen under the sales agreement is expected to be disclosed on a weekly basis or such longer period as may be agreed with Nasdaq Copenhagen.

Settlement for sales of ADSs will occur, unless the parties agree otherwise, on the second business day that is also a trading day following the date on which any sales were made in return for payment of the net proceeds to us. There is no arrangement for funds to be received in an escrow, trust or similar arrangement.

In connection with the sales of our ADSs on our behalf, Cowen will be deemed to be an “underwriter” within the meaning of the Securities Act, and the compensation paid to Cowen will be deemed to be underwriting commissions or discounts. We have agreed in the sales agreement to provide indemnification and contribution to Cowen against certain liabilities, including liabilities under the Securities Act. As sales agent, Cowen will not engage in any transactions that stabilize our securities.

ADSs representing our ordinary shares are listed on The Nasdaq Global Select Market under the symbol “ORPH.” Our ordinary shares are listed on Nasdaq Copenhagen under the symbol “ORPHA.”

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Cowen and/or its affiliates have provided, and may in the future provide, various investment banking, commercial banking, financial advisory and other financial services to us and our affiliates for which services they have received, and may in the future receive, customary fees. In the course of their respective businesses, Cowen may actively trade our securities for their own respective accounts or for the accounts of customers, and, accordingly, Cowen may at any time hold long or short positions in such securities.

TAXATION

The following is a general summary of certain Danish and United States federal income tax consequences relevant to the decision to acquire ordinary shares or ADSs in this offering. The discussion is not intended to be, nor should it be construed as, legal or tax advice to any particular prospective purchaser. The discussion is based on laws and relevant interpretations thereof in effect as of the date of this prospectus supplement, all of which are subject to change or different interpretations, possibly with retroactive effect. The discussion does not address U.S. state or local tax laws, or tax laws of jurisdictions other than the Kingdom of Denmark and the United States. You should consult your tax advisors with respect to the consequences of the acquisition, ownership and disposition of the ordinary shares or ADSs.

Material U.S. Federal Income Tax Consequences for U.S. Holders

The following discussion describes the material U.S. federal income tax consequences relating to the acquisition, ownership and disposition of the ordinary shares or ADSs by U.S. Holders (as defined below). This discussion applies to U.S. Holders that purchase ordinary shares or ADSs pursuant to the offering and hold such ordinary shares or ADSs as capital assets (generally, property held for investment) within the meaning of Section 1221 of the U.S. Internal Revenue Code of 1986, as amended, or the Code. This discussion is based on the Code, U.S. Treasury regulations promulgated thereunder and administrative and judicial interpretations thereof, all as in effect on the date hereof and all of which are subject to change, possibly with retroactive effect. This discussion does not address all of the U.S. federal income tax consequences that may be relevant to specific U.S. Holders in light of their particular circumstances or to U.S. Holders subject to special treatment under U.S. federal income tax law (such as certain financial institutions, insurance companies, broker-dealers and traders in securities or other persons that generally mark their securities to market for U.S. federal income tax purposes, tax-exempt entities, retirement plans, regulated investment companies, real estate investment trusts, certain former citizens or residents of the United States, persons who hold ordinary shares or ADSs as part of a “straddle,” “hedge,” “conversion transaction,” “synthetic security” or other integrated investment, persons acquiring ordinary shares or ADSs in connection with a trade or business conducted outside of the United States, including a permanent establishment or a fixed base in Denmark, persons who received their ordinary shares or ADSs as compensatory payments, U.S. Holders that have a “functional currency” other than the U.S. dollar, persons that own directly, indirectly or through attribution 10% or more of our shares by vote or value, persons who are subject to special tax accounting under Section 451(b) of the Code, corporations that accumulate earnings to avoid U.S. federal income tax, partnerships and other pass-through entities or arrangements that are classified as partnerships for U.S. federal income tax purposes, and investors in such pass-through entities or arrangements). This discussion does not address any U.S. state or local or non-U.S. tax consequences or any U.S. federal estate, gift or alternative minimum tax consequences or the Medicare tax on net investment income.

As used in this discussion, the term “U.S. Holder” means a beneficial owner of ordinary shares or ADSs that is, for U.S. federal income tax purposes, (1) an individual who is a citizen or resident of the United States, (2) a corporation (or entity treated as a corporation for U.S. federal income tax purposes) created or organized in or under the laws of the United States, any state thereof, or the District of Columbia, (3) an estate the income of which is subject to U.S. federal income tax regardless of its source or (4) a trust (x) with respect to which a court within the United States is able to exercise primary supervision over its administration and one or more United States persons (as defined in the Code) have the authority to control all of its substantial decisions or (y) that has elected under applicable U.S. Treasury regulations to be treated as a domestic trust for U.S. federal income tax purposes.

If an entity or arrangement treated as a partnership for U.S. federal income tax purposes holds ordinary shares or ADSs, the U.S. federal income tax consequences relating to an investment in the ordinary shares or ADSs will depend in part upon the status and activities of such entity or arrangement and the particular partner. Any such partnership or partner therein should consult its own tax advisor regarding the U.S. federal income tax consequences applicable to it and its partners of the purchase, ownership and disposition of ordinary shares or ADSs.

Persons considering an investment in ordinary shares or ADSs should consult their own tax advisors as to the particular tax consequences applicable to them relating to the purchase, ownership and disposition of ordinary shares or ADSs, including the applicability of U.S. federal, state and local tax laws and non-U.S. tax laws.

Passive Foreign Investment Company Consequences

In general, a corporation organized outside the United States will be treated as a passive foreign investment company, or PFIC, for any taxable year in which either (1) at least 75% of its gross income is “passive income” or (2) on average at least 50% of its assets, determined on a quarterly weighted-average basis, are assets that produce passive income or are held for the production of passive income. Passive income for this purpose generally includes, among other things, dividends, interest, royalties, rents, and gains from the sale or exchange of property that gives rise to passive income. Assets that produce or are held for the production of passive income generally include cash, even if held as working capital or raised in a public offering, marketable securities, and other assets that may produce passive income. Generally, in determining whether a non-U.S. corporation is a PFIC, a proportionate share of the income and assets of each corporation in which it owns, directly or indirectly, at least a 25% interest (by value) is taken into account.

Our status as a PFIC will depend on the nature and composition of our income and the nature, composition and value of our assets (which may be determined based on the fair market value of each asset, with the value of goodwill and going concern value being determined in large part by reference to the market value of our ordinary shares or ADSs, which may be volatile). Our status may also depend, in part, on how quickly we utilize the cash proceeds from this offering. Based on our current estimates (and not final audited financials) of the composition of our income and valuation of our assets, including goodwill, we do not believe we were a PFIC for our taxable year ending June 30, 2021. There can be no assurance that the IRS will agree with our conclusion and that the IRS would not successfully challenge our position. The determination of whether we are a PFIC is a fact-intensive determination made on an annual basis and the applicable law is subject to varying interpretation. Furthermore, because there are uncertainties in the application of the relevant rules, it is possible that the IRS may challenge our classification of certain income and assets as non-passive or our valuation of our tangible and intangible assets, each of which may result in us being treated as a PFIC for our taxable year ending June 30, 2021 or us becoming a PFIC for the current taxable year or any future taxable years. Our PFIC status may change from year to year and we have not yet made any determination as to our expected PFIC status for the current year. Accordingly, there can be no assurance that we will not be considered a PFIC in the current year or for any future taxable year. Our U.S. counsel expresses no opinion with respect to our PFIC status for our taxable year ending June 30, 2021, and the current or any future taxable year.

If we are a PFIC in any taxable year during which a U.S. Holder owns ordinary shares or ADSs, the U.S. Holder could be liable for additional taxes and interest charges under the “PFIC excess distribution regime” upon (1) a distribution paid during a taxable year that is greater than 125% of the average annual distributions paid in the three preceding taxable years, or, if shorter, the U.S. Holder’s holding period for the ordinary shares or ADSs, and (2) any gain recognized on a sale, exchange or other disposition, including a pledge, of the ordinary shares or ADSs, whether or not we continue to be a PFIC. Under the PFIC excess distribution regime, the tax on such distribution or gain would be determined by allocating the distribution or gain ratably over the U.S. Holder’s holding period for ordinary shares or ADSs. The amount allocated to the current taxable year (i.e., the year in which the distribution occurs or the gain is recognized) and any year prior to the first taxable year in which we are a PFIC will be taxed as ordinary income earned in the current taxable year. The amount allocated to other taxable years will be taxed at the highest marginal rates in effect for individuals or corporations, as applicable, to ordinary income for each such taxable year, and an interest charge, generally applicable to underpayments of tax, will be added to the tax. In addition, any dividend distributions made to a U.S. Holder will not qualify for the lower rates of taxation applicable to qualified dividends discussed below under “Distributions.”

If we are a PFIC for any year during which a U.S. Holder holds ordinary shares or ADSs, we generally will continue to be treated as a PFIC by that holder for all succeeding years during which the U.S. Holder holds the

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ordinary shares or ADSs, unless we cease to meet the requirements for PFIC status and the U.S. Holder makes a “deemed sale” election with respect to the ordinary shares or ADSs. If the election is made, the U.S. Holder will be deemed to sell the ordinary shares or ADSs it holds at their fair market value on the last day of the last taxable year in which we qualified as a PFIC, and any gain recognized from such deemed sale would be taxed under the PFIC excess distribution regime. After the deemed sale election, the U.S. Holder’s ordinary shares or ADSs would not be treated as shares of a PFIC unless we subsequently become a PFIC.

If we are a PFIC for any taxable year during which a U.S. Holder holds ordinary shares or ADSs and one of our non-U.S. corporate subsidiaries is also a PFIC (i.e., a lower-tier PFIC), such U.S. Holder would be treated as owning a proportionate amount (by value) of the shares of the lower-tier PFIC and would be taxed under the PFIC excess distribution regime on distributions by the lower-tier PFIC and on gain from the disposition of shares of the lower-tier PFIC even though such U.S. Holder would not receive the proceeds of those distributions or dispositions. Each U.S. Holder is advised to consult its tax advisors regarding the application of the PFIC rules to our non-U.S. subsidiaries.

Certain elections may alleviate some of the adverse consequences of PFIC status and would result in an alternative treatment of our ordinary shares or ADSs. If we are a PFIC, a U.S. Holder will not be subject to tax under the PFIC excess distribution regime on distributions or gain recognized on ordinary shares or ADSs if such U.S. Holder makes a valid “mark-to-market” election for the ordinary shares or ADSs. The mark-to-market election is available only if we are a PFIC and our ordinary shares or ADSs are “regularly traded” on a “qualified exchange.” The ordinary shares or ADSs (respectively) will be treated as “regularly traded” in any calendar year in which more than a de minimis quantity of the ordinary shares or ADSs (respectively) are traded on a qualified exchange on at least 15 days during each calendar quarter (subject to the rule that trades that have as one of their principal purposes the meeting of the trading requirement are disregarded). It should be noted that only the ADSs and not our ordinary shares are listed on The Nasdaq Global Select Market. The Nasdaq Global Select Market is a qualified exchange for this purpose and, consequently, if the ADSs are regularly traded, the mark-to-market election will be available to a U.S. Holder. Consequently, our ordinary shares may not be marketable if Nasdaq Copenhagen (where our ordinary shares are currently listed) does not meet the applicable requirements. U.S. holders should consult their tax advisors regarding the availability of the mark-to-market election for ordinary shares that are not represented by ADSs.

If a mark-to-market election is in effect, a U.S. Holder generally would take into account, as ordinary income for each taxable year of the U.S. holder, the excess of the fair market value of ordinary shares or ADSs held at the end of such taxable year over the adjusted tax basis of such ordinary shares or ADSs. The U.S. Holder would also take into account, as an ordinary loss each year, the excess of the adjusted tax basis of such ordinary shares or ADSs over their fair market value at the end of the taxable year, but only to the extent of the excess of amounts previously included in income over ordinary losses deducted as a result of the mark-to-market election. The U.S. Holder’s tax basis in ordinary shares or ADSs would be adjusted to reflect any income or loss recognized as a result of the mark-to-market election. Any gain from a sale, exchange or other disposition of ordinary shares or ADSs in any taxable year in which we are a PFIC would be treated as ordinary income and any loss from such sale, exchange or other disposition would be treated first as ordinary loss (to the extent of any net mark-to-market gains previously included in income) and thereafter as capital loss.

A mark-to-market election will not apply to ordinary shares or ADSs for any taxable year during which we are not a PFIC, but will remain in effect with respect to any subsequent taxable year in which we become a PFIC. Such election will not apply to any non-U.S. subsidiaries that we may organize or acquire in the future. Accordingly, a U.S. Holder may continue to be subject to tax under the PFIC excess distribution regime with respect to any lower-tier PFICs that we may organize or acquire in the future notwithstanding the U.S. Holder’s mark-to-market election for the ordinary shares or ADSs.

The tax consequences that would apply if we are a PFIC would also be different from those described above if a U.S. Holder were able to make a valid qualified electing fund, or QEF, election. At this time, we do not

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expect to provide U.S. Holders with the information necessary for a U.S. Holder to make a QEF election. Prospective U.S. Holders should assume that a QEF election will not be available. U.S. Holders should consult their tax advisors to determine whether any of the other elections described above would be available and if so, what the consequences of the alternative treatments would be in their particular circumstances.

Each U.S. person that is an investor in a PFIC is generally required to file an annual information return on IRS Form 8621 containing such information as the U.S. Treasury Department may require. The failure to file IRS Form 8621 could result in the imposition of penalties and the extension of the statute of limitations with respect to U.S. federal income tax.

The U.S. federal income tax rules relating to PFICs are very complex. Prospective U.S. Holders are strongly urged to consult their own tax advisors with respect to the impact of PFIC status on the purchase, ownership and disposition of ordinary shares or ADSs, the consequences to them of an investment in a PFIC, any elections available with respect to the ordinary shares or ADSs and the IRS information reporting obligations with respect to the purchase, ownership and disposition of ordinary shares or ADSs of a PFIC.

Distributions

As described in the section titled “Dividend Policy,” we do not anticipate declaring or paying dividends to holders of the ordinary shares or ADSs in the foreseeable future. However, if we make a distribution contrary to this expectation, subject to the discussion above under “—Passive Foreign Investment Company Consequences,” a U.S. Holder that receives a distribution with respect to ordinary shares or ADSs generally will be required to include the gross amount (including any amounts withheld in respect of foreign taxes) of such distribution in gross income as a dividend when actually or constructively received to the extent of the U.S. Holder’s pro rata share of our current and/or accumulated earnings and profits (as determined under U.S. federal income tax principles). To the extent a distribution received by a U.S. Holder is not a dividend because it exceeds the U.S. Holder’s pro rata share of our current and accumulated earnings and profits, it will be treated first as a tax-free return of capital and reduce (but not below zero) the adjusted tax basis of the U.S. Holder’s ordinary shares or ADSs. To the extent the distribution exceeds the adjusted tax basis of the U.S. Holder’s ordinary shares or ADSs, the remainder will be taxed as capital gain. Because we may not calculate our earnings and profits in accordance with U.S. federal income tax principles, U.S. Holders can expect all distributions to be reported to them as dividends.

Distributions on ordinary shares or ADSs that are treated as dividends generally will not be eligible for the “dividends received” deduction generally allowed to corporate shareholders with respect to dividends received from U.S. corporations. Dividends paid by a “qualified foreign corporation” are eligible for taxation to non-corporate U.S. Holders at a reduced capital gains rate rather than the marginal tax rates generally applicable to ordinary income provided that certain requirements are met. A non-United States corporation (other than a corporation that is classified as a PFIC for the taxable year in which the dividend is paid or the preceding taxable year) generally will be considered to be a qualified foreign corporation (a) if it is eligible for the benefits of a comprehensive tax treaty with the United States which the Secretary of Treasury of the United States determines is satisfactory for purposes of this provision and which includes an exchange of information provision, or (b) with respect to any dividend it pays on shares that are readily tradable on an established securities market in the United States. Only the ADSs and not our ordinary shares are listed on The Nasdaq Global Select Market, which is an established securities market in the United States, and we expect the ADSs to be readily tradable on The Nasdaq Global Select Market. There can be no assurance that the ADSs will be considered to be readily tradable on an established securities market in the United States in later years. However, the Company, which is incorporated under the laws of the Kingdom of Denmark, believes that it qualifies as a resident of the Kingdom of Denmark for purposes of, and is eligible for the benefits of, the Convention between the Government of the United States of America and the Government of the Kingdom of Denmark for the Avoidance of Double Taxation and the Prevention of Fiscal Evasion with Respect to Taxes on Income, signed on August 19, 1999, as

amended and currently in force, or the U.S.-Denmark Tax Treaty, although there can be no assurance in this regard. Further, the IRS has determined that the U.S.-Denmark Tax Treaty is satisfactory for purposes of the qualified dividend rules and that it includes an exchange-of-information program. Therefore, subject to the discussion under “—Passive Foreign Investment Company Consequences” above and provided we are not a PFIC for the taxable year in which the dividend is paid or the preceding taxable year, dividends paid on the ADSs and the ordinary shares will generally be “qualified dividend income” in the hands of individual U.S. holders, provided that a holding period requirement (more than 60 days of ownership, without protection from the risk of loss, during the 121-day period beginning 60 days before the ex-dividend date) and certain other requirements are met. Each U.S. Holder is advised to consult its tax advisors regarding the availability of the reduced tax rate on dividends with regard to its particular circumstances.

Distributions on ordinary shares or ADSs that are treated as dividends generally will constitute income from sources outside the United States for foreign tax credit purposes and generally will constitute passive category income. Subject to certain complex conditions and limitations, Danish taxes withheld on any distributions on ordinary shares or ADSs may be eligible as a credit or deduction against a U.S. Holder’s federal income tax liability. The rules relating to the determination of the U.S. foreign tax credit are complex, and U.S. Holders should consult their tax advisors regarding the availability of a foreign tax credit in their particular circumstances and the possibility of claiming an itemized deduction (in lieu of the foreign tax credit) for any foreign taxes paid or withheld. In addition, the creditability of foreign taxes could be affected by actions taken by intermediaries in the chain of ownership between the holders of ADSs and our company if, as a result of such actions, the holders of ADSs are not properly treated as beneficial owners of the underlying ordinary shares. Each U.S. Holder should consult its own tax advisors regarding the foreign tax credit rules.

In general, the amount of a distribution paid to a U.S. Holder in a foreign currency will be the U.S. dollar value of the foreign currency calculated by reference to the spot exchange rate on the day the depository receives the distribution, in the case of the ADSs, or on the day the distribution is received by the U.S. Holder, in the case of ordinary shares, regardless of whether the foreign currency is converted into U.S. dollars at that time. Any foreign currency gain or loss a U.S. Holder realizes on a subsequent conversion of foreign currency into U.S. dollars will be U.S. source ordinary income or loss. If dividends received in a foreign currency are converted into U.S. dollars on the day they are received, a U.S. Holder generally will not be required to recognize foreign currency gain or loss in respect of the dividend.

Sale, Exchange or Other Disposition of Ordinary Shares or ADSs

Subject to the discussion above under “—Passive Foreign Investment Company Consequences,” a U.S. Holder generally will recognize capital gain or loss for U.S. federal income tax purposes upon the sale, exchange or other disposition of ordinary shares or ADSs in an amount equal to the difference, if any, between the U.S. dollar value of the amount realized (i.e., the amount of cash plus the fair market value of any property received) on the sale, exchange or other disposition and such U.S. Holder’s adjusted tax basis in the ordinary shares or ADSs. A U.S. Holder’s adjusted tax basis in its ordinary shares or ADSs generally will be equal to the cost of such ordinary shares or ADSs. Such capital gain or loss generally will be long-term capital gain taxable at a reduced rate for non-corporate U.S. Holders or long-term capital loss if, on the date of sale, exchange or other disposition, the ordinary shares or ADSs were held by the U.S. Holder for more than one year. Any capital gain of a non-corporate U.S. Holder that is not long-term capital gain is taxed at ordinary income rates. The deductibility of capital losses is subject to limitations. Any gain or loss recognized from the sale or other disposition of ordinary shares or ADSs will generally be gain or loss from sources within the United States for U.S. foreign tax credit purposes.

For a cash basis taxpayer, units of foreign currency paid or received are translated into U.S. dollars at the spot rate on the settlement date of the purchase or sale. In that case, no foreign currency exchange gain or loss will result from currency fluctuations between the trade date and the settlement date of such a purchase or sale.

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An accrual basis taxpayer, however, may elect the same treatment required of cash basis taxpayers with respect to purchases and sales of ordinary shares or ADSs that are traded on an established securities market, provided the election is applied consistently from year to year. Such election may not be changed without the consent of the IRS. For an accrual basis taxpayer who does not make such election, units of foreign currency paid or received are translated into U.S. dollars at the spot rate on the trade date of the purchase or sale. Such an accrual basis taxpayer may recognize exchange gain or loss based on currency fluctuations between the trade date and the settlement date. Any foreign currency gain or loss a U.S. Holder realizes will be U.S. source ordinary income or loss.

Information Reporting and Backup Withholding

U.S. Holders may be required to file certain U.S. information reporting returns with the IRS with respect to an investment in ordinary shares or ADSs, including, among others, IRS Form 8938 (Statement of Specified Foreign Financial Assets). As described above under “—Passive Foreign Investment Company Consequences,” each U.S. Holder who is a shareholder of a PFIC must file an annual report containing certain information. U.S. Holders paying more than US\$100,000 for ordinary shares or ADSs may be required to file IRS Form 926 (Return by a U.S. Transferor of Property to a Foreign Corporation) reporting this payment. Substantial penalties may be imposed upon a U.S. Holder that fails to comply with the required information reporting. U.S. Holders are urged to consult their tax advisors regarding their information reporting obligations, if any, with respect to their ownership and disposition of ordinary shares or ADSs.

Dividends on and proceeds from the sale or other disposition of ordinary shares or ADSs may be reported to the IRS unless the U.S. Holder establishes an adequate basis for exemption. In addition, U.S. holders may be subject to backup withholding on such payments, unless the U.S. Holder provides a taxpayer identification number and a duly executed IRS Form W-9 or otherwise establishes an exemption. Backup withholding is not an additional tax. Any amounts withheld under the backup withholding rules generally will be allowed as a refund or a credit against a U.S. Holder's U.S. federal income tax liability if the required information is furnished by the U.S. Holder on a timely basis to the IRS.

U.S. Holders should consult their own tax advisors regarding the backup withholding tax and information reporting rules.

EACH PROSPECTIVE INVESTOR IS URGED TO CONSULT ITS OWN TAX ADVISOR ABOUT THE TAX CONSEQUENCES TO IT OF AN INVESTMENT IN ORDINARY SHARES OR ADSS IN LIGHT OF THE INVESTOR'S OWN CIRCUMSTANCES.

Material Danish Income Tax Consequences

The following is a summary of material Danish tax considerations relating to the ownership and disposition of ordinary shares or ADSs. The summary only sets out the tax position of the direct owners of the shares or ADSs and assumes that the holders of the shares or ADSs are the beneficial owners of the shares or ADSs and income deriving from the shares or ADSs. The summary is for general information purposes only and does not constitute exhaustive tax or legal advice.

It is noted specifically that the summary does not address all possible Danish tax consequences relating to the ownership and disposition of ordinary shares or ADSs. The summary does accordingly not apply to investors to whom special tax rules apply, and, therefore, may not be relevant, for example, to investors subject to the Danish Tax on Pension Yields Act (i.e., pension savings), professional investors, certain institutional investors, insurance companies, pension companies, banks, stockbrokers and investors with tax liability on return on pension investments. The summary does further not apply to non-Danish tax resident investors that carry on business activities in Denmark through a permanent establishment to which the ordinary shares or ADSs are allocated.

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In the context of the following section, “companies” mean entities that are treated as separate taxable entities under domestic tax laws of their jurisdiction of incorporation.

The summary is based solely on the tax laws of Denmark in effect on the date of this prospectus supplement. Danish tax laws may be subject to change, potentially with retroactive effect.

Potential investors in the ordinary shares or ADSs are advised to consult their tax advisors regarding the applicable tax consequences of ownership and disposition of the ordinary shares or ADSs based on their particular circumstances.

Tax Treatment of ADSs under Danish Tax Law

It is currently not clear under Danish tax legislation or case law how ADSs are to be treated for Danish tax purposes as this, inter alia, depends on the agreement between the domestic custodian bank (the ADS issuer) and the ADS holders.

This summary assumes that the ADS holder in respect of the ADSs is treated as the direct owner of the shares underlying the ADSs and, accordingly, as the shareholder for Danish domestic tax law purposes, and that the ADS holder is deemed the beneficial owner of any dividend distributed on the underlying shares for Danish domestic tax law purposes as well as under any applicable tax treaty. Based on this assumption, the ADSs listed in the U.S. should, for Danish tax purposes, be treated as listed shares since the company’s ordinary shares are admitted to trading on a regulated market.

Danish Tax Resident Individuals

Sale of Ordinary Shares or ADSs

Capital gains from the sale of shares realized by Danish tax resident individuals are taxed as share income at a rate of 27% on the first DKK 56,500 (approximately \$8,800 based on current exchange rates) (for cohabiting spouses, a total of DKK 113,000 (approximately \$17,600 based on current exchange rates)) and at a rate of 42% on share income exceeding DKK 56,500 (approximately \$8,800 based on current exchange rates) (for cohabiting spouses over DKK 113,000 (approximately \$17,600 based on current exchange rates)) (all 2021 amounts and thresholds). The threshold is subject to annual adjustments and include all share income (i.e., all capital gains on shares and dividends derived by the individual or cohabiting spouses, respectively).

Gains and losses on the sale of shares are calculated as the difference between the purchase price and the sales price. The purchase price is generally determined using the average method as a proportionate part of the aggregate purchase price for all the shareholder’s shares in the company (i.e., not the purchase price paid for each share).

Losses on the sale of listed shares can only be offset against other share income deriving from listed shares (i.e., dividends and capital gains on the sale of listed shares) and subject to the Danish tax authorities having received certain information concerning the ownership of the shares in due time. Unused losses will automatically be offset against a cohabiting spouse’s share income deriving from listed shares and any additional losses can be carried forward and offset against future share income deriving from listed shares.

Dividends

Dividends paid to Danish tax resident individuals are included in the individual’s share income and taxed as such, as outlined above. All share income must be included when calculating whether the threshold amounts mentioned above are exceeded. Dividends paid to Danish tax resident individuals are generally subject to withholding tax at the rate of 27% and taxed as share income.

Non-Danish Tax Resident Individuals

Sale of Ordinary Shares or ADSs

Non-Danish tax resident individuals, including individuals tax resident in the United States, are normally not subject to Danish taxation on any gains realized on the sale of shares, irrespective of the ownership period, subject to certain anti-avoidance rules seeking to prevent that taxable dividend payments are converted to tax exempt capital gains (see below).

Dividends

Dividends paid to non-Danish tax resident individuals, including individuals tax resident in the United States, are generally subject to withholding tax at the rate of 27%. No additional Danish tax will be imposed.

In the event that the shareholder is tax resident in a state with which Denmark has entered into a tax treaty and is entitled to benefits under such tax treaty, the shareholder may seek a refund from the Danish Tax Agency of the tax withheld in excess of the applicable treaty rate (Danish tax treaties typically provide for a 15% tax rate). Denmark has entered into tax treaties with approximately 80 countries, including the United States and almost all EU member states. The treaty between Denmark and the United States generally provides for a 15% tax rate.

Similarly, Danish domestic tax law provides for a 15% tax rate, if the shareholder holds less than 10% of the nominal share capital in the company and is tax resident in a state that is obligated to exchange information with Denmark under a tax treaty or an international agreement, convention or other administrative agreement on assistance in tax matters. If the shareholder is tax resident outside the EU, it is an additional requirement for application of the 15% tax rate that the shareholder together with related shareholders holds less than 10% of the share capital of the company.

Any reduced tax rate according to an applicable tax treaty and/or Danish domestic tax law will not affect the withholding rate (27%). In order to receive a refund (from 27% to e.g., 15%), the shareholder must make a claim for such refund through certain certification procedures.

The Danish Tax Agency has published guidance on the documentation necessary for processing refund claims. The guidance is available in English from the Danish tax authorities' website, <https://skat.dk/skat.aspx?oId=2244931&vId=0&lang=US>. The information on, or information that can be accessed through, such website is not part of and should not be incorporated by reference into this prospectus supplement or the accompanying prospectus. We have included such website address as an inactive textual reference only.

Dividends paid to non-Danish individuals tax resident in a country which is "blacklisted" by EU (i.e. at present American Samoa, the Republic of Fiji, Guam, Republic of Palau, Panama, the Independent State of Samoa, Republic of Trinidad and Tobago, U.S. Virgin Islands and the Republic of Vanuatu); and who is holding or within the last five years has been holding i) minimum 25% of the share capital of the Company; or ii) more than 50% of the voting rights on the share capital of the Company are subject to a Danish tax rate of 44% (which is paid as a withholding taxation). The blacklist is updated as per October 2021 and is to be updated again in February 2022.

Danish Tax Resident Companies

Sale of Ordinary Shares or ADSs

For the purpose of taxation of sales of shares by corporate shareholders (and dividends received by corporate shareholders, see below), a distinction is made between Subsidiary Shares, Group Shares, Tax Exempt Portfolio Shares and Taxable Portfolio Shares (note that the ownership threshold described below is applied on

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the basis of the number of all shares issued by the company, and not on the basis of the number of the ordinary shares or ADSs issued):

“Subsidiary Shares,” which are generally defined as shares owned by a shareholder holding at least 10% of the share capital of the issuing company;

“Group Shares,” which are generally defined as shares in a company in which the shareholder of the company and the issuing company are subject to Danish joint taxation or satisfy the requirements for international joint taxation under Danish law;

“Tax-Exempt Portfolio Shares,” which are generally defined as unlisted shares owned by a shareholder holding less than 10% of the share capital of the issuing company; and

“Taxable Portfolio Shares,” which are defined as shares that do not qualify as Subsidiary Shares, Group Shares or Tax-Exempt Portfolio Shares, e.g. shares admitted to trading on a regulated market (such as the ordinary shares and the ADSs) owned by a shareholder holding less than 10% of the nominal share capital of the issuing company.

Gains and losses on disposal of Subsidiary Shares, Group Shares and Tax-Exempt Portfolio Shares realized by Danish tax resident companies are generally not included in the taxable income of the shareholder, subject to certain anti-avoidance rules (see below).

Capital gains on listed Taxable Portfolio Shares are taxable at the general corporate tax rate of currently 22% and losses on such shares are generally deductible.

Gains and losses on listed Taxable Portfolio Shares are taxed under the mark-to-market principle irrespective of realization.

According to the mark to market principle, each year’s taxable gain or loss on Taxable Portfolio Shares is calculated as the difference between the market value of the shares at the beginning of the tax year and the market value of the shares at the end of the tax year. Thus, taxation will take place on an accrual basis even if no shares have been disposed of and no gains or losses have been realized.

Dividends

Dividends received on Subsidiary Shares and Group Shares are generally tax-exempt, subject to certain anti-avoidance rules (see below).

Dividends received on Taxable Portfolio Shares are taxable at the general corporate tax rate of 22% and tax is generally withheld similarly at 22%.

Non-Danish Tax Resident Companies

Sale of Ordinary Shares or ADSs

Non-Danish tax resident companies, including companies tax resident in the United States, are generally not taxed in Denmark on gains realized on the sale of shares, subject to certain anti-avoidance rules (see below).

Dividends

Dividends received on Subsidiary Shares are exempt from Danish withholding tax provided that taxation shall be waived or reduced under the Parent-Subsidiary Directive (2011/96/EU) or under an applicable tax treaty.

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Similarly, dividends received on Group Shares, which are not Subsidiary Shares, are exempt from Danish withholding tax if the shareholder is resident in the European Union or the EEA and provided that taxation shall be waived or reduced under the Parent-Subsidiary Directive (2011/96/EU) or under an applicable tax treaty had the shares been Subsidiary Shares.

In other cases, dividends will generally be subject to tax at a rate of 22%. However, the withholding rate is 27%, meaning that all foreign corporate shareholders receiving taxable dividends distributed from Danish companies will be able to ask the Danish Tax Agency for a refund of minimum 5% of the total dividend.

Further, in the event that the shareholder is tax resident in a state with which Denmark has entered into a tax treaty and is entitled to the benefits under such tax treaty, the shareholder may seek a refund from the Danish Tax Agency of the tax withheld in excess of the applicable treaty rate (Danish tax treaties typically provide for a 15% tax rate). Denmark has entered into tax treaties with approximately 80 countries, including the United States and almost all EU member states. The treaty between Denmark and the United States generally provides for a 15% tax rate.

Similarly, Danish domestic tax law provides for an applicable 15% tax rate, if the shareholder holds less than 10% of the share capital in the company and is tax resident in a state that is obligated to exchange information with Denmark under a tax treaty or an international agreement, convention or other administrative agreement on assistance in tax matters. If the shareholder is tax resident outside the EU, it is an additional requirement for eligibility for the 15% tax rate that the shareholder together with related shareholders holds less than 10% of the nominal share capital of the company.

Any reduced tax rate according to an applicable tax treaty (and/or the 15% tax rate provided for under Danish domestic tax law) will not affect the withholding rate (27%). In order to receive a refund (from 27% to e.g., 15%), the shareholder must make a claim against the Danish Tax Agency for such refund through certain certification procedures.

The Danish Tax Agency has published guidance on the documentation necessary for processing refund claims. The guidance is available in English from the Danish tax authorities' website, <https://skat.dk/skat.aspx?oId=2244931&vId=0&lang=US>. The information on, or information that can be accessed through, such website is not part of and should not be incorporated by reference into this prospectus supplement or the accompanying prospectus. We have included such website address as an inactive textual reference only.

Dividends received on Subsidiary Shares or Group Shares held by non-Danish companies tax resident in a country which is "blacklisted" by EU (i.e. at present American Samoa, the Republic of Fiji, Guam, Republic of Palau, Panama, the Independent State of Samoa, Republic of Trinidad and Tobago, U.S. Virgin Islands and the Republic of Vanuatu) are subject to a Danish tax rate of 44% (which is paid as a withholding taxation). The blacklist is updated as per October 2021 and is to be updated again in February 2022.

Danish Anti-avoidance Rules

Payments may be subject to Danish withholding tax irrespective of the above, if the holder of ADSs or ordinary shares is not the beneficial owner of the shares and dividend (e.g. if the holder of ADSs or ordinary shares reassigns the payments to a person or entity not itself entitled to the above exemptions).

Further, Danish law has certain general anti-avoidance rules, which focus on substance over form. Under these rules the Danish tax authorities can set aside a setup, which constitutes a fictitious arrangement, which is carried out for the main purposes (or with one of the main purposes) of tax avoidance and resulting in no taxes being paid. This is the case where the relevant scheme presents a number of unusual features which suggest that it had not been entered into for commercial business reasons but to unduly obtain tax benefits. Subject to the conditions of the specific GAAR an investor might be denied the benefits of the Parent-Subsidiary Directive (2011/96/EU) or a tax treaty, and Danish withholding tax of 27% (or 44%) will in such cases be levied.

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Finally, it should be noted that it is the shareholder who owns the share, i.e. the ordinary share or the ADS, at the time of the general meeting where the decision to distribute dividend is passed, who is subject to Danish taxation on the dividend, and thereby entitled to make a tax reclaim, if any.

LEGAL MATTERS

The validity of the securities in respect of which this prospectus supplement is being delivered and certain legal matters with respect to Danish law will be passed upon by Gorrissen Federspiel Advokatpartnerselskab, Copenhagen, Denmark. We are being represented by Cooley LLP, New York, New York with respect to certain legal matters as to United States federal securities and New York State law. Shearman & Sterling LLP, New York, New York is acting as counsel for Cowen in connection with this offering with respect to United States federal securities and New York State law and Plesner Advokatpartnerselskab, Copenhagen, Denmark, is acting as counsel for Cowen in connection with this offering with respect to Danish law.

EXPERTS

The consolidated financial statements of Orphazyme A/S appearing in Orphazyme A/S's Annual Report (Form 20-F) for the year ended December 31, 2020, have been audited by EY Godkendt Revisionspartnerselskab, independent registered public accounting firm, as set forth in their report thereon, included therein, and incorporated herein by reference. Such consolidated financial statements are incorporated herein by reference in reliance upon such report given on the authority of such firm as experts in accounting and auditing.

The registered business address of EY Godkendt Revisionspartnerselskab is Dirch Passers Allé 36, 2000 Frederiksberg, Denmark.

SERVICE OF PROCESS AND ENFORCEMENT OF LIABILITIES

We are organized under the laws of Denmark, with a domicile in the municipality of Copenhagen, Denmark.

Some of the members of the board of directors and the executive board named herein are residents of Denmark or other jurisdictions outside the United States. A substantial portion of ours and such persons' assets are located in Denmark or other jurisdictions outside the United States. As a result, it may not be possible for investors to effect service of process upon such persons or us with respect to litigation that may arise under U.S. law or to enforce against them or our company judgments obtained in U.S. courts, whether or not such judgments were made pursuant to civil liability provisions of the federal or state securities laws of the United States or any other laws of the United States.

There is not currently a treaty between the United States and Denmark providing for reciprocal recognition and enforceability of judgments rendered in connection with civil and commercial disputes and, accordingly, that a final judgment (other than arbitration awards) rendered by a U.S. court based on civil liability would not be enforceable in Denmark. It is uncertain whether Danish courts would allow actions to be predicated on the securities laws of the United States or other jurisdictions outside Denmark. Danish courts are likely to deny claims for punitive damages and may grant a reduced amount of damages compared to U.S. courts.

WHERE YOU CAN FIND ADDITIONAL INFORMATION

This prospectus supplement and the accompanying prospectus are part of the registration statement on Form F-3 we filed with the SEC under the Securities Act. This prospectus supplement and the accompanying prospectus do not contain all of the information set forth in the registration statement and the exhibits to the registration statement. For further information with respect to us and the securities we are offering under this prospectus supplement, we refer you to the registration statement and the exhibits and schedules filed as a part of the registration statement, which may be obtained from the SEC or us, as provided below. Statements in this prospectus supplement are summaries and each statement is qualified in all respects by reference to the document to which it refers. You should refer to the actual documents for a more complete description of the relevant matters.

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You should rely only on the information contained in this prospectus supplement and the accompanying prospectus or incorporated by reference herein or therein. We have not authorized anyone else to provide you with different information. We are not making an offer of these securities in any state where the offer is not permitted. You should not assume that the information in this prospectus supplement and the accompanying prospectus is accurate as of any date other than the date on the front page of the applicable document, regardless of the time of delivery of this prospectus supplement or any sale of the securities offered by this prospectus supplement.

We are subject to the periodic reporting and other informational requirements of the Exchange Act. Under the Exchange Act, we file annual reports and other information with the SEC. As a foreign private issuer, we are exempt from, among other things, the rules under the Exchange Act prescribing the furnishing and content of proxy statements and our officers, directors and principal shareholders are exempt from the reporting and short-swing profit recovery provisions contained in Section 16 of the Exchange Act.

The SEC maintains a website that contains reports, proxy and information statements and other information regarding issuers that file electronically with the SEC, including us. The address of the SEC website is www.sec.gov.

We also maintain a website at www.orphazyme.com through which you can access our SEC filings. The information contained on, or accessible through, our website is not incorporated by reference into this prospectus supplement or the accompanying prospectus, and you should not consider any information contained in, or that can be accessed through, our website as part of this prospectus supplement or the accompanying prospectus or in deciding whether to purchase ordinary shares, including ordinary shares in the form of ADSs. We have included our website address as an inactive textual reference only.

INCORPORATION BY REFERENCE

The SEC allows us to “incorporate by reference” information into this prospectus supplement and the accompanying prospectus, which means that we can disclose important information to you by referring you to other documents filed separately with the SEC. The information incorporated by reference is deemed to be part of this prospectus supplement and the accompanying prospectus, and subsequent information that we file with the SEC will automatically update and supersede that information. Any statement contained in a previously filed document incorporated by reference will be deemed to be modified or superseded for purposes of this prospectus supplement and the accompanying prospectus to the extent that a statement contained in this prospectus supplement or the accompanying prospectus modifies or replaces that statement.

This prospectus supplement incorporates by reference the documents set forth below that have previously been filed with the SEC:

- our Annual Report on [Form 20-F](#) for the year ended December 31, 2020, filed with the SEC on March 2, 2021;
- our reports on Form 6-K furnished to the SEC on [February 26, 2021](#), [March 1, 2021](#), [March 2, 2021](#), [March 3, 2021](#), [March 26, 2021](#), [March 29, 2021](#), [April 22, 2021](#), [May 7, 2021](#), [June 11, 2021](#), [June 21, 2021](#), [June 28, 2021](#), [August 31, 2021](#), [October 5, 2021](#), [October 7, 2021](#), [November 1, 2021](#) and [November 4, 2021](#); and
- the description of our ordinary shares and American Depositary Shares contained in our registration statement on [Form 8-A](#) (File No. 001-39545), filed with the SEC on September 22, 2020, including any amendments or reports filed for the purpose of updating such description.

We are also incorporating by reference all subsequent annual reports on Form 20-F that we file with the SEC and certain reports on Form 6-K that we furnish to the SEC after the date of this prospectus supplement (if

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such reports on Form 6-K expressly state that they are incorporated in whole or in part by reference into the registration statement of which this prospectus supplement and the accompanying prospectus forms a part) prior to the termination of this offering. In all cases, you should rely on the later information over different information included in this prospectus supplement.

Unless expressly incorporated by reference, nothing in this prospectus supplement or the accompanying prospectus shall be deemed to incorporate by reference information furnished to, but not filed with, the SEC. Copies of all documents incorporated by reference in this prospectus supplement and the accompanying prospectus, other than exhibits to those documents unless such exhibits are specifically incorporated by reference in this prospectus supplement and the accompanying prospectus, will be provided at no cost to each person, including any beneficial owner, who receives a copy of this prospectus supplement on the written or oral request of that person made to:

Orphazyme A/S
Ole Maaløes Vej 3, DK-2200
Copenhagen N
Denmark
Tel: (+45) 28 98 90 55
Attention: Investor Relations

SEC Position on Indemnification for Securities Act Liabilities

Insofar as indemnification for liabilities arising under the Securities Act may be permitted to our directors, officers and controlling persons pursuant to the foregoing provisions, or otherwise, we have been informed that in the opinion of the SEC such indemnification is against public policy as expressed in the Securities Act and is, therefore, unenforceable.

PROSPECTUS

\$75,000,000



Orphazyme A/S

Ordinary Shares

(including Ordinary Shares represented by American Depositary Shares)

We may offer and sell up to \$75,000,000 of our ordinary shares, including ordinary shares represented by ADSs, from time to time in one more offerings. This prospectus describes the general manner in which these securities will be offered. We will provide the specific terms of these offerings in one or more supplements to this prospectus. We may also authorize one or more free writing prospectuses to be provided to you in connection with these offerings. The prospectus supplement and any related free writing prospectus may also add, update or change information contained in this prospectus. You should carefully read this prospectus, the applicable prospectus supplement and any related free writing prospectus, as well as any documents incorporated by reference, before investing in any of the securities being offered.

This prospectus may not be used to consummate a sale of any securities unless accompanied by a prospectus supplement.

Our ordinary shares may be represented by American Depositary Shares, or ADSs. Each ADS represents the right to receive one ordinary share.

ADSs representing our ordinary shares are listed on The Nasdaq Global Select Market, under the symbol "ORPH." Our ordinary shares are listed on Nasdaq Copenhagen A/S, or Nasdaq Copenhagen, under the symbol "ORPHA." On October 13, 2021, the last reported sale price of ADSs, as reported on The Nasdaq Global Select Market, was \$4.91 per ADS. The applicable prospectus supplement will contain information, where applicable, as to other listings, if any, on The Nasdaq Global Select Market or other securities exchange of the securities covered by the prospectus supplement.

Securities may be sold to or through one or more underwriters or dealers, directly to purchasers or through agents designated from time to time. For additional information on the methods of sale, you should refer to the section titled "Plan of Distribution" in this prospectus and in the applicable prospectus supplement. If any underwriters or agents are involved in the sale of any securities with respect to which this prospectus is being delivered, the names of such underwriters or agents and any applicable discounts or commissions and options to purchase additional shares will be set forth in a prospectus supplement. The price to the public of such securities and the net proceeds that we expect to receive from such sale will also be set forth in a prospectus supplement. No securities may be sold without delivery of this prospectus and the applicable prospectus supplement describing the method and terms of the offering of such securities.

We are an "emerging growth company" as that term is used in the Jumpstart Our Business Startups Act of 2012 and, as such, have elected to comply with certain reduced public company reporting requirements for this prospectus and future filings. See "Prospectus Summary—Implications of Being an Emerging Growth Company" and "Prospectus Summary—Implications of Being a Foreign Private Issuer" for additional information.

Investing in these securities involves a high degree of risk. You should review carefully the risks and uncertainties described under the heading "[Risk Factors](#)" on page 3 of this prospectus and any similar section contained in the applicable prospectus supplement and in any free writing prospectuses we have authorized for use in connection with a specific offering, and under similar headings in the other documents that are incorporated by reference into this prospectus.

None of the Securities and Exchange Commission, any state securities commission, the Danish Financial Supervisory Authority, nor any other foreign securities commission has approved or disapproved of these securities or determined if this prospectus is truthful or complete. Any representation to the contrary is a criminal offense.

The date of this prospectus is October 22, 2021.

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ABOUT THIS PROSPECTUS

This prospectus is part of a registration statement on Form F-3 that we filed with the U.S. Securities and Exchange Commission, or the SEC, using a “shelf” registration process. Under this shelf registration process, we may offer and sell, from time to time in one or more offerings, up to a total dollar amount of \$75,000,000 of ordinary shares, including ordinary shares represented by ADSs.

This prospectus provides you with a general description of the securities we may offer. Each time we offer securities under this prospectus, we will provide a prospectus supplement that will contain more specific information about the terms of that offering. We may also authorize one or more free writing prospectuses to be provided to you that may contain material information relating to these offerings. The prospectus supplement and any related free writing prospectus that we may authorize to be provided to you may also add, update or change any of the information contained in this prospectus or in the documents that we have incorporated by reference into this prospectus. We urge you to read carefully this prospectus, any applicable prospectus supplement and any related free writing prospectuses we have authorized for use in connection with a specific offering, together with the information incorporated herein by reference as described under the heading “Incorporation By Reference,” before investing in any of the securities being offered.

THIS PROSPECTUS MAY NOT BE USED TO CONSUMMATE A SALE OF SECURITIES UNLESS IT IS ACCOMPANIED BY A PROSPECTUS SUPPLEMENT. Neither we, nor any agent, underwriter or dealer has authorized any person to give any information or to make any representation other than

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those contained or incorporated by reference in this prospectus, any applicable prospectus supplement or any related free writing prospectus prepared by or on behalf of us or to which we have referred you. This prospectus, any applicable supplement to this prospectus or any related free writing prospectus do not constitute an offer to sell or the solicitation of an offer to buy any securities other than the registered securities to which they relate, nor do this prospectus, any applicable supplement to this prospectus or any related free writing prospectus constitute an offer to sell or the solicitation of an offer to buy securities in any jurisdiction to any person to whom it is unlawful to make such offer or solicitation in such jurisdiction.

You should not assume that the information contained in this prospectus, any applicable prospectus supplement or any related free writing prospectus is accurate on any date subsequent to the date set forth on the front of the document or that any information we have incorporated by reference is correct on any date subsequent to the date of the document incorporated by reference, even though this prospectus, any applicable prospectus supplement or any related free writing prospectus is delivered, or securities are sold, on a later date.

This prospectus and the information incorporated herein by reference contains summaries of certain provisions contained in some of the documents described herein, but reference is made to the actual documents for complete information. All of the summaries are qualified in their entirety by the actual documents. Copies of some of the documents referred to herein have been filed, will be filed or will be incorporated by reference as exhibits to the registration statement of which this prospectus is a part, and you may obtain copies of those documents as described below under the heading “Where You Can Find Additional Information.”

Unless otherwise indicated or the context otherwise requires, all references in this prospectus to the terms “Orphazyme,” “the Company,” “we,” “us” and “our” refer to Orphazyme A/S and its wholly owned subsidiaries. In this prospectus, any reference to any provision of any legislation shall include any amendment, modification, re-enactment or extension thereof. Words importing the singular shall include the plural and vice versa, and words importing the masculine gender shall include the feminine or neutral gender. All references to “shares” in this prospectus refer to ordinary shares of Orphazyme A/S with a nominal value of DKK 1 per share.

For investors outside the United States: We have not done anything that would permit the offering or possession or distribution of this prospectus in any jurisdiction where action for that purpose is required, other than in the United States. Persons outside the United States who come into possession of this prospectus must inform themselves about, and observe any restrictions relating to, the offering of the securities described herein and the distribution of this prospectus outside the United States.

We are incorporated in Denmark, and many of our outstanding securities are owned by non-U.S. residents. Under the rules of the SEC, we are currently eligible for treatment as a “foreign private issuer.” As a foreign private issuer, we are not required to file periodic reports and financial statements with the SEC as frequently or as promptly as domestic registrants whose securities are registered under the U.S. Securities Exchange Act of 1934, as amended, or the Exchange Act.

TRADEMARKS

This prospectus includes trademarks, tradenames and service marks, certain of which belong to us and others that are the property of other organizations. Solely for convenience, trademarks and tradenames referred to in this prospectus appear without the ® and ™ symbols, but the absence of those references is not intended to indicate, in any way, that we will not assert our rights or that the applicable owner will not assert its rights to these trademarks and tradenames to the fullest extent under applicable law. We do not intend our use or display of other parties’ trademarks, trade names or service marks to imply, and such use or display should not be construed to imply, a relationship with, or endorsement or sponsorship of us by, these other parties.

MARKET AND INDUSTRY DATA

This prospectus includes statistical and other industry and market data that we obtained from industry publications and research, surveys and studies conducted by third parties, as well estimates by our management based on such data. Management estimates are derived from publicly available information, our knowledge of our industry and assumptions based on such information and knowledge, which we believe to be reasonable. The market data and estimates used in this prospectus involve a number of assumptions and limitations, and you are cautioned not to give undue weight to such data and estimates. We believe that the information from these industry publications, surveys and studies is reliable. The industry in which we operate is subject to a high degree of uncertainty and risk due to a variety of important factors, including those described in the sections titled “Risk Factors” and “Special Note Regarding Forward-Looking Statements” in this prospectus, any applicable prospectus supplement and any related free writing prospectus and under similar sections contained in other documents that are incorporated by reference herein. These and other factors could cause results to differ materially from those expressed in the estimates made by the independent parties and by us.

PRESENTATION OF FINANCIAL INFORMATION

We maintain our books and records in Danish kroner and we prepare our audited consolidated financial statements in accordance with International Financial Reporting Standards, or IFRS, as issued by the International Accounting Standards Board, or IASB. None of the consolidated financial statements incorporated by reference into this prospectus were prepared in accordance with accounting principles generally accepted in the United States, or U.S. GAAP. All references in this prospectus to “\$” are to U.S. dollars, to “DKK” are to Danish kroner and to “€” are to the Euro.

PROSPECTUS SUMMARY

This summary does not contain all of the information that may be important to you in making your investment decision. In addition to this summary, you should carefully read the entire prospectus, the applicable prospectus supplement and any related free writing prospectus, including the risks of investing in our ordinary shares and the ADSs discussed under the heading “Risk Factors” contained herein and in the applicable prospectus supplement and any related free writing prospectus, and under similar headings in the other documents that are incorporated by reference into this prospectus before deciding whether to invest in the ordinary shares and ADSs. You should also carefully read the information incorporated by reference into this prospectus, including our financial statements, and the exhibits to the registration statement of which this prospectus is a part.

Overview

We are a late-stage biopharmaceutical company committed to developing a new therapeutic option for people living with Niemann-Pick disease type C, or NPC, a rare, genetic, progressively debilitating and often fatal neurodegenerative disease. In November 2020, we submitted a marketing authorization application, or MAA, for our investigational product candidate, arimoclomol, in NPC to the European Medicines Agency, or EMA, and expect an opinion from the Committee for Human Medicinal Products, or CHMP, in the first quarter of 2022. We had also previously submitted a new drug application, or NDA, to the U.S Food and Drug Administration, or FDA, for arimoclomol in NPC. In June 2021, we received a complete response letter from the FDA following its review of the NDA, and in October 2021, we held a Type-A meeting with the FDA to assess a path forward for arimoclomol for NPC in the U.S. Arimoclomol is an orally- or naso/gastrically-administered small molecule that crosses the blood-brain barrier and is designed to selectively amplify the natural role of endogenous HSPs, which protect against cellular toxicity caused by protein misfolding, aggregation and lysosomal dysfunction. In our Phase 2/3 clinical trial of arimoclomol in NPC, we have observed evidence of slowing of disease progression. We also believe that arimoclomol has been well tolerated in clinical trials including more than 500 human subjects for various indications.

Implications of Being an Emerging Growth Company

As a company with less than \$1.07 billion in revenue during our last fiscal year, we qualify as an “emerging growth company,” as defined in the U.S. Jumpstart Our Business Startups Act of 2012, or JOBS Act. An emerging growth company may take advantage of specified reduced reporting and other burdens that are otherwise applicable generally to public companies. These provisions include an exemption from the auditor attestation requirement in the assessment of our internal control over financial reporting pursuant to the Sarbanes-Oxley Act of 2002, or Sarbanes-Oxley Act.

We may choose to take advantage of some but not all of these provisions, and therefore the information that we provide holders of ordinary shares and ADSs may be different than the information you might receive from other public companies in which you hold equity. In addition, Section 107 of the JOBS Act also provides that an emerging growth company can take advantage of an extended transition period for complying with new or revised accounting standards applicable to public companies in the United States. As a public company in Denmark, we are unable to take advantage of the extended transition period.

We may take advantage of these provisions for up to five years from the initial public offering of our ADSs or such earlier time that we are no longer an emerging growth company. We will cease to be an emerging growth company upon the earliest of the following:

- the last day of the first fiscal year in which our annual revenues were at least \$1.07 billion;

- the last day of the fiscal year following the fifth anniversary of the initial public offering of ADSs;
- the date on which we have issued more than \$1 billion of non-convertible debt securities over a three-year period; and
- the last day of the fiscal year during which we meet the following conditions: (i) the worldwide market value of our common equity securities held by non-affiliates as of our most recently completed second fiscal quarter is at least \$700 million, (ii) we have been subject to U.S. public company reporting requirements for at least 12 months and (iii) we have filed at least one annual report as a U.S. public company.

Implications of Being a Foreign Private Issuer

We are also considered a “foreign private issuer” under U.S. securities laws. In our capacity as a foreign private issuer, we are exempt from certain rules under the U.S. Securities Exchange Act of 1934, as amended, or the Exchange Act, that impose certain disclosure obligations and procedural requirements for proxy solicitations under Section 14 of the Exchange Act. In addition, our officers, directors and principal shareholders are exempt from the reporting and “short-swing” profit recovery provisions of Section 16 of the Exchange Act and the rules under the Exchange Act with respect to their purchases and sales of our securities. Moreover, we are not required to file periodic reports and financial statements with the SEC as frequently or as promptly as U.S. companies whose securities are registered under the Exchange Act. In addition, we are not required to comply with Regulation FD, which restricts the selective disclosure of material information.

Both foreign private issuers and emerging growth companies are also exempt from certain more stringent executive compensation disclosure rules for public companies in the United States under the Dodd-Frank Wall Street Reform and Consumer Protection Act of 2010. Even if we no longer qualify as an emerging growth company, so long as we remain a foreign private issuer, we will continue to be exempt from such compensation disclosures.

We may take advantage of these exemptions until such time as we are no longer a foreign private issuer. We will remain a foreign private issuer until such time that more than 50% of our outstanding voting securities are held by U.S. residents and any of the following three circumstances applies: (1) the majority of our executive officers or directors are U.S. citizens or residents; (2) more than 50% of our assets are located in the United States; or (3) our business is administered principally in the United States.

Corporate History and Information

We were incorporated on June 19, 2009 as a private limited liability company under Danish law and later converted into a Danish public limited liability company on October 20, 2017. We are registered with the Danish Business Authority (Erhvervsstyrelsen) in Copenhagen, Denmark under company registration number (CVR) no. 32266355. We were publicly listed on Nasdaq Copenhagen, Denmark, in November 2017 under the symbol “ORPHA” and on The Nasdaq Global Select Market, United States, in September 2020 under the symbol “ORPH.”

Our headquarters and principal executive offices are located at Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark, and our telephone number is +45 39 17 82 72. Our website address is www.orphazyme.com. The information contained on, or accessible through, our website is not incorporated by reference into this prospectus, and you should not consider any information contained in, or that can be accessed through, our website as part of this prospectus or in deciding whether to purchase ordinary shares, including ordinary shares in the form of ADSs. We have included our website address as an inactive textual reference only.

RISK FACTORS

Investing in our securities involves a high degree of risk. You should carefully review the risks and uncertainties described under the heading “Risk Factors” contained in the applicable prospectus supplement and any related free writing prospectus, and in our Annual Report on Form 20-F for the year ended December 31, 2020 and reports on Form 6-K as updated by our subsequent filings, which are incorporated by reference into this prospectus, before deciding whether to purchase any of the securities being registered pursuant to the registration statement of which this prospectus is a part. Each of the risk factors could adversely affect our business, results of operations, financial condition and cash flows, as well as adversely affect the value of an investment in our securities, and the occurrence of any of these risks might cause you to lose all or part of your investment. Additional risks not presently known to us or that we currently believe are immaterial may also significantly impair our business operations.

SPECIAL NOTE REGARDING FORWARD-LOOKING STATEMENTS

This prospectus and the documents incorporated by reference herein contain forward-looking statements within the meaning of Section 27A of the Securities Act of 1933, as amended, or the Securities Act, and Section 21E of the Securities Exchange Act of 1934, as amended, or the Exchange Act, that reflect our current expectations and views of future events. Discussions containing these forward-looking statements may be found, among other places, in the sections titled “Information on the Company,” “Risk Factors” and “Operating and Financial Review and Prospects” incorporated by reference from our most recent Annual Report on Form 20-F, as well as any amendments thereto, filed with the SEC. Known and unknown risks, uncertainties and other factors, including those listed under “Risk Factors” in this prospectus, the applicable prospectus supplement and in any related free writing prospectuses and under similar headings in documents that are incorporated by reference into this prospectus, including our most recent Annual Report on Form 20-F and reports on Form 6-K as updated by our subsequent filings, may cause our actual results, performance or achievements to be materially different from those expressed or implied by the forward-looking statements.

You can identify some of these forward-looking statements by words or phrases, such as “may,” “will,” “expect,” “anticipate,” “aim,” “estimate,” “intend,” “plan,” “believe,” “is/are likely to,” “potential,” “continue” or other similar expressions. We have based these forward-looking statements largely on our current expectations and projections about future events that we believe may affect our financial condition, results of operations, business strategy and financial needs. These forward-looking statements include statements relating to:

- the ability of our clinical trials to demonstrate acceptable safety and efficacy of our product candidate, and other positive results;
- the timing, progress and results of clinical trials for our product candidate, including statements regarding the timing of initiation and completion of studies or trials and related preparatory work, the period during which the results of the trials will become available, and our research and development programs;
- the timing, scope and likelihood of regulatory filings, submissions and approvals, including the MAA and NDA process for arimoclomol for the treatment of NPC and the potential regulatory approval of arimoclomol;
- our ability to obtain marketing approvals of our product candidate and to meet existing or future regulatory standards or comply with post-approval requirements;
- our expectations regarding our ability to fund our operating expenses and capital expenditure requirements;
- our payments of future milestone payments to our licensing partners, and the expected timing of such payments;
- our expectations regarding the potential market size and the size of the patient populations for our product candidate, if approved for commercial use;
- our expectations regarding the potential advantages of our product candidate over existing therapies;
- the impact of the ongoing COVID-19 pandemic on our business and operations;
- our expectations regarding the outcome and impact of class action lawsuits and any other litigation on our business and operations;
- our potential to enter into new collaborations;
- our expectations with regard to our ability to identify and develop additional product candidates or product candidates for other indications or technologies with significant commercial potential that are consistent with our commercial objectives;

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- our expectations with regard to the willingness and ability of our current and future licensing and collaboration partners to pursue the development of our product candidate;
- the commercialization and market acceptance of our product candidate;
- our marketing and manufacturing capabilities;
- the pricing of and reimbursement for our product candidate;
- the implementation of our business model and strategic plans for our business and product candidate;
- our ability to operate our businesses without infringing the intellectual property rights and proprietary technology of third parties;
- the scope of protection we are able to establish and maintain for intellectual property rights covering our product candidate;
- our analysis of our actual or potential patent infringement claims and the rights of our collaboration partners with respect to such claims;
- estimates of our expenses, future revenue, capital requirements, our needs for additional financing and our ability to obtain additional capital;
- regulatory development in the United States, Europe and other jurisdictions;
- our exposure to scrutiny as a public company in the United States;
- our ability to effectively manage our anticipated growth;
- our ability to attract and retain qualified employees and key personnel;
- our financial performance;
- our expectations regarding the time during which we will be an emerging growth company under the JOBS Act and qualify as a foreign private issuer; and
- developments and projections relating to our competitors and our industry, including competing therapies.

You should refer to the “Risk Factors” section contained in this prospectus, the applicable prospectus supplement and any related free writing prospectus, and under similar headings in the other documents that are incorporated by reference into this prospectus, for a discussion of important factors that may cause our actual results to differ materially from those expressed or implied by our forward-looking statements. As a result of these factors, we cannot assure you that the forward-looking statements in this prospectus will prove to be accurate. Furthermore, if our forward-looking statements prove to be inaccurate, the inaccuracy may be material. In light of the significant uncertainties in these forward-looking statements, you should not regard these statements as a representation or warranty by us or any other person that we will achieve our objectives and plans in any specified time frame or at all.

The forward-looking statements made in this prospectus and the documents incorporated by reference herein relate only to events or information as of the date on which the statements are made in this prospectus and the documents incorporated by reference herein. Except as required by law, we undertake no obligation to update or revise publicly any forward-looking statements, whether as a result of new information, future events or otherwise, after the date on which the statements are made or to reflect the occurrence of unanticipated events. You should read this prospectus, the documents incorporated by reference herein and the documents that we refer to in this prospectus and have filed as exhibits to this prospectus completely and with the understanding that our actual future results may be materially different from what we expect. For all forward-looking statements, we claim the protection of the safe harbor for forward-looking statements contained in the Private Securities Litigation Reform Act of 1995.

OFFER STATISTICS AND EXPECTED TIMETABLE

We may offer and sell an indeterminate number of ordinary shares, including ordinary shares represented by ADSs, from time to time in one or more offerings pursuant to this prospectus (as may be detailed in a prospectus supplement) up to a total dollar amount of \$75,000,000. The actual price per share of the securities that we will offer pursuant hereto will depend on a number of factors that may be relevant as of the time of offer. See “Plan of Distribution.”

REASONS FOR THE OFFERING AND USE OF PROCEEDS

Unless otherwise set forth in a prospectus supplement, we currently intend to use the net proceeds of any offering of securities for working capital and other general corporate purposes. Accordingly, we will have significant discretion in the use of any net proceeds. We may provide additional information on the use of the net proceeds from the sale of the offered securities in an applicable prospectus supplement relating to the offered securities.

OFFER AND LISTING DETAILS

Our ordinary shares have been listed on Nasdaq Copenhagen under the symbol “ORPHA” since November 2017. ADSs representing our ordinary shares have been listed on The Nasdaq Global Select Market under the symbol “ORPH” since September 2020. Prior to those dates, there was no public trading market for our ordinary shares or for ADSs.

DESCRIPTION OF SHARE CAPITAL AND ARTICLES OF ASSOCIATION

The following describes our issued share capital, summarizes the material provisions of our articles of association and highlights certain differences in corporate law in the Kingdom of Denmark and Delaware corporate law, the law under which many publicly listed companies in the United States are incorporated. Please note that this summary is not intended to be exhaustive. For further information, please refer to the full version of our articles of association, which are incorporated by reference into the registration statement of which this prospectus forms a part.

General

We were incorporated on June 19, 2009 as a private limited liability company under Danish law and later converted into a Danish public limited liability company on October 20, 2017. We are registered with the Danish Business Authority (Erhvervsstyrelsen) in Copenhagen, Denmark under company registration number (CVR) no. 32266355. Our ADSs have been listed on The Nasdaq Global Select Market under the symbol “ORPH” since September 2020. Our ordinary shares have been listed in Denmark on Nasdaq Copenhagen under the symbol “ORPHA” since November 2017. Our company has been established with the objectives of engaging in medical research, production and sale of such products and related business.

Our headquarters and principal executive offices are located at Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark, and our telephone number is +45 39 17 82 72. Our website address is www.orphazyme.com. The information contained on, or accessible through, our website is not incorporated by reference into this prospectus, and you should not consider any information contained in, or that can be accessed through, our website as part of this prospectus or in deciding whether to purchase ordinary shares, including ordinary shares in the form of ADSs. We have included our website address as an inactive textual reference only.

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Development of Share Capital

Since November 2017, we had one class of shares (prior to this date we had multiple classes of shares).

As of June 30, 2021, our registered, issued and fully paid outstanding share capital was DKK 34,952,241 distributed into 34,952,241 shares of nominal value DKK 1 each, including 21,067,199 ADSs, each representing one ordinary share.

The development of our share capital since December 31, 2016 and up to and including the date of this prospectus is set forth in the table below.

Date of approval	Capital Increase, No. of Shares	Gross Proceeds, DKK000s	Share Capital, No. of Shares after change	Issued Share DKK Capital after change
Share capital at December 31, 2016			A-shares: 125,000 B-shares: 2,050,208 C-shares: 1,185,333	3,360,541
2017				
Capital increase by cash contribution, January 26, 2017	534,007	48,060	A-shares: 125,000 B-shares: 2,050,208 C-shares: 1,719,340	3,894,548
Capital increase by cash contribution, January 26, 2017	772,022	69,482	A-shares: 125,000 B-shares: 2,050,208 C-shares: 2,491,362	4,666,570
Capital increase by cash contribution, June 29, 2017	435,640	39,208	A-shares: 125,000 B-shares: 2,050,208 C-shares: 2,927,002	5,102,210
Conversion into a public limited liability company, October 20, 2017	—	—	A-shares: 125,000 B-shares: 2,050,208 C-shares: 2,927,002	5,102,210
Consolidation of share classes, November 2, 2017	—	—	5,102,210	5,102,210
Issuance of bonus shares, November 2, 2017	6,487,882	—	11,590,092	11,590,092
Initial public offering, November 6, 2017	7,500,000	600,000	19,090,092	19,090,092
Exercise of warrants, November 20, 2017	838,092	1,161	19,928,184	19,928,184
2018				
Issuance of bonus shares, January 29, 2018	11,380	—	19,939,564	19,939,564

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Date of approval	Capital Increase, No. of Shares	Gross Proceeds, DKK000s	Share Capital, No. of Shares after change	Issued Share DKK Capital after change
2019				
Issuance of bonus shares, January 31, 2019	26,060	—	19,965,624	19,965,624
Issuance of matching shares, March 4, 2019	19,175	19	19,984,799	19,984,799
2020				
Issuance of bonus shares, January 31, 2020	20,650	—	20,005,449	20,005,449
Capital increase by cash contribution, February 6, 2020	7,032,937	745,491	27,038,386	27,038,386
Exercise of restricted share units, March 27, 2020(1)	4,616	282	27,043,002	27,043,002
Exercise of restricted share units, March 27, 2020(1)	1,927	118	27,044,929	27,044,929
Exercise of restricted share units, March 27, 2020(1)	3,451	211	27,048,380	27,048,380
Vesting and exercise of matching shares, July 29, 2020	31,250	31	27,079,630	27,079,630
Exercise of restricted share units, March 27, 2020(1)	1,927	118	27,081,557	27,081,557
U.S. initial public offering, September 28, 2020	7,616,146	534,534	34,697,703	34,697,703
2021				
Vesting and exercise of matching shares, including US IPO bonus grants to certain participants, February 1, 2021	170,131	170	34,867,834	34,867,834
Issuance of bonus shares, February 25, 2021	22,553	23	34,890,387	34,890,387
Directed issuance of new shares, February 25, 2021	58,000	58	34,948,387	34,948,387
Exercise of restricted share units, March 24, 2021	3,854	236	34,952,241	34,952,241

- (1) Share issue was approved by our board of directors on March 27, 2020 and subsequently registered with the Danish Business Authority following exercise by the respective directors.

Authorizations to Our Board of Directors

Our board of directors is authorized to increase our share capital as follows:

- In accordance with article 3.1 of our articles of association, our board of directors is, until March 25, 2026, authorized 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.
- In accordance with article 3.2 of our articles of association, our board of directors is, until November 2, 2022, authorized to increase our share capital in one or more issues without pre-emption rights for our existing shareholders by up to a nominal amount of DKK 1,300,000 in connection with the issue of new shares to members of our board of directors, our executives and/or our employees. 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.
- In accordance with article 3.3 of our articles of association, our board of directors is, until November 2, 2022, authorized to increase our share capital in one or more issues of new shares without preemption

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rights for our existing shareholders by up to a nominal amount of DKK 15,750,000 in connection with 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, which will be below market price. The value of such new shares to be issued can in any case not exceed a maximum of \$2.5 million with a fixed exchange rate of DKK 6.30 per 1 USD based on the average closing price of the our ordinary shares on Nasdaq Copenhagen for the 30 days immediately prior to the date of issuance.

- In accordance with article 3.4 of our articles of association, our board of directors is, until January 25, 2025, authorized to increase our share capital in one or more issues of new shares with preemption rights for our 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.
- In accordance with article 3.5 of our articles of association, our board of directors is, until March 25, 2026, authorized to increase our share capital in one or more issues of new shares without pre-emption rights for our existing shareholders by up to a nominal amount of DKK 1,300,000 in connection with the issue of new shares to members of our board of directors, our executives and/or our employees. The new shares shall be issued against cash payment at a subscription price to be determined by our board of directors, which may be below the market price.
- In accordance with article 3.6 of our articles of association, the authorizations granted to our board of directors pursuant to articles 3.2 and 3.5 can in the aggregate only be exercised to increase our share capital by a maximum nominal amount of DKK 2,000,000.

As of the date of this prospectus, our board of directors partially exercised the authorization in article 3.2 of our articles of association to increase our share capital following which a nominal value of DKK 294,331 of the authorization has been issued. In addition, our board of directors has partly exercised the authorization in article 3.3 to increase our share capital following which a nominal value of DKK 80,643 of the authorization has been issued.

Further, our board of directors is authorized on behalf of the company until March 25, 2026 to acquire our own shares for a total nominal value of up to 10% of our share capital for the time being, so long as the company's holding of treasury shares after such acquisition does not exceed 20% of the company's share capital. The price paid for such shares may not deviate by more than 10% from the share price quoted on Nasdaq Copenhagen at the time of acquisition.

As of June 30, 2021, the total number of additional shares our board of directors is authorized to issue was 49,364,793.

Our ADSs are listed on The Nasdaq Global Select Market under the symbol "ORPH" and our ordinary shares are listed in Denmark on Nasdaq Copenhagen under the symbol "ORPHA."

Pre-emptive Rights

If our shareholders at a general meeting resolve to increase our share capital by a cash contribution, section 162 of the DCA will apply. Under that section, shareholders have a pre-emptive right to subscribe for new shares in proportion to their existing shareholdings. However, the pre-emptive right may be derogated from by a majority comprising at least two-thirds of the votes cast, as well as at least two-thirds of the share capital represented at the general meeting, provided the share capital increase takes place at market price or nine-tenths of the votes cast, as well as at least nine-tenths of the share capital represented at the general meeting if the share capital increase takes place below market price, unless (i) such capital increase is directed at certain but not all shareholders (in which case all shareholders must consent); or (ii) such capital increase is directed at our

employees whereby a majority comprising at least two-thirds of the votes cast, as well as at least two-thirds of the share capital represented at the general meeting is required. Further, the pre-emptive rights may be derogated from by an exercise of the board of directors of a valid authorization in our articles of association, provided that the share capital increase takes place at or above market price.

Shareholders' Register

We are obliged to maintain a shareholders' register (Ejerbog). The shareholders' register is maintained by Computershare A/S, Lottenborgvej 26 D, 1., DK-2800 Kgs. Lyngby, Denmark, our Danish share registrar and transfer agent. It is mandatory that the shareholders' register is maintained within the European Union and that it is available to public authorities.

Pursuant to the DCA, public and private limited liability companies are required to register with the Danish Business Authority information regarding shareholders who own at least 5% of the share capital or the voting rights. Pursuant to this provision, we file registrations with the Danish Public Shareholders' Register of the Danish Business Authority. Shareholders that exceed or fall below the ownership threshold must notify us, and we will subsequently file the information with the Danish Business Authority. Reporting is further required upon passing or falling below thresholds of 5%, 10%, 15%, 20%, 25%, 50%, 90%, and 100% as well as one-third and two-thirds of the votes or the share capital. This also applies to beneficial holders of our shares, such as holders of the ADSs.

Articles of Association and Danish Corporate Law

General Meetings and Voting Rights

Our general meetings shall be held in the Capital Region of Denmark. Our annual general meeting 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. Not later than eight weeks before the contemplated date of the annual general meeting, we shall publish the date of the general meeting and the deadline for submitting requests for specific proposals to be included in the agenda.

Extraordinary general meetings shall be held when determined by our board of directors or requested by our auditor. Furthermore, our board of directors shall convene an extraordinary general meeting within two weeks of receipt of a written request from shareholders representing no less than 5% of the share capital containing specific proposals for the business to be transacted at such extraordinary general meeting.

General meetings shall be convened by our board of directors at least three weeks' and not more than five weeks' notice. The notice shall be published on our website. Furthermore, a notice of the general meeting shall be sent electronically to all shareholders recorded in our register of shareholders who have requested such notice.

In accordance with Danish law, the notice shall specify the time and place of the general meeting and the agenda containing the business to be transacted at the general meeting. If a proposal to amend our articles of association is to be considered at the general meeting, the main contents of the proposal shall be specified in the notice. Our general meetings shall be held in English. Our 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 our board of directors, in Danish.

Every shareholder is entitled to have specific business transacted at the general meeting, provided that the shareholder submits a written request to that effect to our board of directors not later than six weeks before the date of the general meeting.

The right of a shareholder to attend a general meeting and to vote is determined by the shares held by the shareholder at the record date. The record date is one week before the general meeting. The shares held by each

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shareholder are determined at the record date based on the number of shares held by that shareholder as registered in our register of shareholders and any notification of ownership received by us for the purpose of registration in our register of shareholders, but which have not yet been registered.

At the general meeting each share of the nominal value of DKK 1 shall carry one vote. Our articles of association permit a person registered as a holder of our shares in VP Securities A/S and acting in a professional capacity to exercise on behalf of other natural or legal persons, including holders of ADSs representing our ordinary shares, voting rights attached to any such shares in a manner that is not identical to the exercise of the voting rights attached to our other shares held by such person.

A shareholder who is entitled to attend the general meeting pursuant to our articles of association and who wants to attend the general meeting shall notify us of his/her attendance no later than three days prior to the date of the general meeting. A shareholder may, subject to having notified us of his/her attendance in accordance with our articles of association, attend in person or by proxy, and the shareholder or the proxy may attend together with an adviser.

The right to vote may be exercised by a written and dated instrument of proxy in accordance with applicable laws. Our board of directors may be appointed as proxy. A shareholder who is entitled to participate in the general meeting according to our articles of association may vote by postal vote in accordance with the DCA. Such postal votes shall be received by us no later than the business day before the general meeting. Postal votes cannot be withdrawn. In accordance with Danish law, the notice shall specify the time and place of the general meeting and the agenda containing the business to be transacted at the general meeting. If a proposal to amend our articles of association is to be considered at the general meeting, the main contents of the proposal shall be specified in the notice.

Our articles of association permit our board to decide to hold general meetings partially or fully by electronic means in accordance with our articles of association and applicable Danish law.

Resolutions by the General Meetings and Amendments to the Articles of Association

Resolutions at general meetings shall be passed by a simple majority of votes cast, unless otherwise prescribed by law or by our articles of association. Adoption of changes to our articles of association, our dissolution, merger or demerger requires that the resolution is adopted by 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 our board of directors or other bodies. The provisions in our articles of association relating to a change of the rights of shareholders or a change to the capital are not more stringent than required by the DCA.

Redemption and Conversion Provisions

Except as provided for in the DCA, no shareholder is under an obligation to have its shares redeemed in whole or in part by us or by any third party, and none of the shares carry any redemption or conversion rights or any other special rights.

Dissolution and Liquidation

In the event of dissolution and liquidation, our shareholders are entitled to participate in the distribution of assets in proportion to their nominal shareholdings after payment of our creditors.

Indication of Takeover Bids

No takeover offers have been made by any third party in respect of our shares during the past or current financial year. Our articles of association do not contain provisions that are likely to have the effect of delaying, deferring or preventing a change in control of our company.

Provisions as to the Level of Equity Investments to be Notified to Us and the Danish Authorities

Shareholders in Danish companies with shares admitted to trading and official listing on Nasdaq Copenhagen are, pursuant to Section 38 of the Danish Capital Markets Act, required to give simultaneous notice to the company and the Danish Financial Supervisory Authority, or the FSA, of the shareholding in the company, when the shareholding reaches, exceeds or falls below thresholds of 5%, 10%, 15%, 20%, 25%, 50% or 90% and limits of one-third or two-thirds of the voting rights or nominal value of the total share capital.

A shareholder in a company means a natural or legal person who, directly or indirectly, holds: (i) shares in the company on behalf of itself and for its own account; (ii) shares in the company on behalf of itself, but for the account of another natural or legal person; or (iii) depository receipts, where such holder is considered a shareholder in relation to the underlying shares represented by the depository receipts.

The duty to notify set forth above further applies to natural and legal persons who are entitled to acquire, sell or exercise voting rights which are:

- (i) held by a third party with whom that natural or legal person has concluded an agreement, which obliges them to adopt, by concerted exercise of the voting rights they hold, a lasting common policy towards the management of the issuer in question (common duty to inform for all parties to the agreement);
- (ii) held by a third party under an agreement concluded with that natural or legal person providing for the temporary transfer of the voting rights in question in return for consideration;
- (iii) attached to shares which are lodged as collateral for that natural or legal person, provided the person controls the voting rights and declares an intention of exercising them;
- (iv) attached to shares in which that natural or legal person has a lifelong right of disposal;
- (v) held, or may be exercised within the meaning of (i) to (iv), by an undertaking controlled by that person or entity;
- (vi) attached to shares deposited with that natural or legal person and which the person can exercise at its own discretion in the absence of specific instructions from the shareholders;
- (vii) held by a third party in its own name on behalf of that person; or
- (viii) exercisable by that person through a proxy where that person may exercise the voting rights at its discretion in the absence of specific instructions of the shareholder.

The duty to notify set forth above also applies to anyone, who directly or indirectly holds (a) financial instruments that afford the holder either an unconditional right to acquire or the discretion as to its right to acquire existing shares (e.g., share options); and/or (b) financial instruments based on existing shares and with an economic effect equal to that of the financial instruments mentioned in (a), regardless of them not affording the right to purchase existing shares (e.g., the ADSs or, under the circumstances, cash-settled derivatives linked to the value of our shares or ADSs representing our shares). Holding these kinds of financial instruments counts towards the thresholds mentioned above and may thus trigger a duty to notify by themselves or when accumulated with a holding of shares or ADSs. The FSA will in certain cases publish information concerning sanctions imposed, including, as a general rule, the name of the shareholder in question, as a consequence of non-compliance with the above rules.

The notification shall be made promptly but not later than four weekdays after the shareholder was aware or should have become aware of the completion of the transaction, and in accordance with the provisions of Danish Executive Order on Major Shareholders. The shareholder is deemed to have become aware of the completion of the transaction no later than two weekdays after the completion of the transaction. The shareholder shall disclose the change in voting rights and shares, including the number of voting rights (and the division of voting rights between share classes, if applicable) and shares held directly or indirectly by the shareholder following the

transaction. The notification shall further state the transaction date on which the threshold was reached or no longer reached and the identity of the shareholder as well as the identity of any natural or legal person with the right to vote on behalf of the shareholder and in the case of a group structure, the chain of controlled undertakings through which voting rights are effectively held. The information shall be notified to the company and simultaneously submitted electronically to the FSA. Failure to comply with the notification requirements is punishable by fine or suspension of voting rights in instances of gross or repeated non-compliance.

When an obligation to notify rests on more than one natural or legal person, the notification may be made through a joint notification. However, use of a joint notification does not exempt the individual shareholders or natural or legal persons from their responsibilities in connection with the obligation to notify or the contents of the notification.

After receipt of the notification, but not later than three weekdays thereafter, the company shall publish the contents of the notification.

Furthermore, the general duty of notification under Section 55 of the DCA in respect of notification of significant holdings (similar to the thresholds set out in the Danish Capital Markets Act Section 38) applies, including when the limit of 100% of the share capital's voting rights or nominal value of the company is reached or are no longer reached.

EU Regulation No 596/2014 on Market Abuse and General Disclosure Requirements

EU Regulation No 596/2014 on market abuse, or the Market Abuse Regulation, applies to us and dealings concerning our shares and ADSs. In connection with our listing on The Nasdaq Global Select Market, we revised our internal code on possession and handling of inside information to cover trading in both our ordinary shares and ADSs and with respect to our board of directors', executive management's and employees' dealings in our shares or in financial instruments the value of which is determined by the value of our shares so that it also covers the ADSs. Furthermore, we have drawn up a list of those persons working for us who could have access to inside information on a regular or incidental basis and have informed such persons of the rules on insider trading and market manipulation, including the sanctions, which can be imposed in the event of a violation of those rules.

In addition, the company is obliged to disclose certain other information to the public pursuant to the Danish Capital Markets Act, the Danish Executive Order on an Issuers' Duty to Provide Information and the Issuer Rules of Nasdaq Copenhagen, regardless of whether this information amounts to inside information. Information which would have to be disclosed under these rules includes, for example: (i) changes to the board of directors, executive management and auditors; (ii) decisions to introduce incentive schemes; (iii) substantial changes in business activities; (iv) material acquisitions and divestments; (v) unexpected and significant deviations in the company's financial result or position; (vi) proposed changes in the capital structure; and (vii) annual and interim reports and accounts. Furthermore, the company is required to make sure that no unauthorized person gains access to inside information prior to its publication to the market.

The EU Short Selling Regulation (EU Regulation 236/2012) Includes Certain Notification Requirements in connection with Short Selling of Shares Admitted to Trading on a Trading Venue (including Nasdaq Copenhagen) and Securities or Derivatives that Relate to Such Shares (including the ADSs).

When a natural or legal person reaches, exceeds or falls below a net, short position of 0.2% of the issued share capital of a company that has shares admitted to trading on a trading venue (which includes the ADSs), such person shall make a private notification (i.e. such notification will not be made public) to the relevant competent authority, which in Denmark is the FSA. The obligation to notify the FSA, moreover, applies in each case where the short position reaches, exceeds or falls below 0.1% above the 0.2% threshold. In addition, when a natural or legal person reaches or falls below a net short position of 0.5% of the issued share capital of a

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company that has shares admitted to trading on a trading venue in the European Union and each 0.1% above that, such person shall make a public notification of its net short position via the FSA. The notification requirements apply to both physical and synthetic short positions. In addition uncovered short selling (naked short selling) of shares admitted to trading on a trading venue is prohibited. Furthermore, on March 16, 2020, the European Securities and Markets Authority, or ESMA, issued a decision which temporarily lowered the reporting threshold from 0.2% to 0.1% for net short position holders in shares traded on a trading venue in the European Union for three months due to COVID-19's impact on financial markets. On September 16, 2020, and December 17, 2020, ESMA issued a decision to renew the temporary requirement to temporarily lower the reporting threshold from 0.2% to 0.1% for net short position holders for an additional three months, which entails that the lowered threshold applied until March 19, 2021, to any natural or legal person, irrespective of their country of residence. ESMA has decided not to renew its decision on temporary lowered reporting threshold, hence the decision on temporarily lowered reporting threshold expired on March 19, 2021. On May 20, 2021, ESMA published an opinion wherein it recommended the European Commission to permanently lower the reporting threshold from 0.2% to 0.1%. On September 27, 2021, the European Commission adopted a delegated regulation reflecting this recommendation. After the European Commission has adopted the delegated regulation, the European Parliament and the Council have three months to formulate any objections. If they do not, the delegated regulation enters into force.

Mandatory Tender Offers

The Danish Capital Markets Act (Part 8) and the Danish Executive Order on Takeover include rules concerning public offers for the acquisition of shares admitted to trading on a regulated market (including Nasdaq Copenhagen).

If a shareholding is transferred, directly or indirectly, in a company with one or more share classes admitted to trading on a regulated market, to an acquirer or to persons acting in concert with such acquirer, the acquirer and the persons acting in concert with such acquirer, if applicable, shall give all shareholders of the company the option to dispose of their shares on identical terms, if the acquirer or the persons acting in concert with such acquirer gains control over the company as a result of the transfer.

Control as mentioned above exists if the acquirer or persons acting in concert with such acquirer, directly or indirectly, holds at least one-third of the voting rights in the company, unless it can be clearly proven in special cases that such ownership does not constitute control. An acquirer or persons acting in concert with such acquirer who does not hold at least one-third of the voting rights in a company, nevertheless has control when the acquirer has or persons acting in concert with such acquirer have:

- the right to control at least one-third of the voting rights in the company according to an agreement with other investors; or
- the right to appoint or dismiss a majority of the members of the central governing body.

Voting rights attached to treasury shares shall be included in the calculation of voting rights.

The Danish Capital Markets Act contains specific exemptions from the obligation to submit a mandatory takeover offer, including transfers of shares by inheritance or transfer within the same group and as a result of a creditor's debt enforcement proceedings. Exemptions from the mandatory tender offer rules may be granted under special circumstances by the FSA.

Limitation on Liability

Under Danish law, members of the board of directors or executive management may be held liable for damages in the event that loss is caused due to their negligence. They may be held jointly and severally liable for damages to the company and to third parties for acting in negligent violation of the articles of association and Danish law.

Comparison of Danish Corporate Law and Our Articles of Association and Delaware Corporate Law

The following comparison between Danish corporate law, which applies to us, and Delaware corporate law, the law under which many publicly listed companies in the United States are incorporated, discusses shareholder rights and obligations and certain additional matters. This summary is subject to Danish law, including the DCA, and Delaware corporate law, including the Delaware General Corporation Law. Further, please note that if you are a holder of the ADSs, then you are not treated as one of our shareholders under such laws and do not have any shareholder rights in Orphazyme A/S.

Shareholder Rights

Notice of Meeting

Denmark. According to the DCA and as implemented in our articles of association, general meetings in listed limited liability companies shall be convened by the board of directors with a minimum of three weeks' notice and a maximum of five weeks' notice. A convening notice shall also be forwarded to shareholders recorded in our shareholders' register who have requested such notification. There are specific requirements as to the information and documentation required to be disclosed in connection with the convening notice.

Delaware. Under Delaware law, unless otherwise provided in the certificate of incorporation or bylaws, written notice of any meeting of the stockholders must be given to each stockholder entitled to vote at the meeting not less than ten nor more than 60 days before the date of the meeting and shall specify the place, date, hour and purpose or purposes of the meeting.

Voting Rights

Denmark. Each share confers the right to cast one vote at the general meeting of shareholders, unless the articles of association provide otherwise. Our articles of association allow a person registered as a holder of our shares in VP Securities A/S and acting in a professional capacity on behalf of other natural or legal persons, including holders of ADS representing our ordinary shares to exercise voting rights attached to any such shares in a manner that is not identical to the exercise of the voting rights attached to our other shares held by such person. The right of a shareholder to vote is determined by the shares held by the shareholder at the record date. The record date is one week before the general meeting. Each holder of shares may cast as many votes as it holds shares. Voting instructions may be given only in respect of a number of ADSs representing an integral number of shares or other deposited securities. Shares that are held by us or our direct or indirect subsidiaries do not confer the right to vote.

Delaware. Under the Delaware General Corporation Law, each stockholder is entitled to one vote per share of stock, unless the certificate of incorporation provides otherwise. In addition, the certificate of incorporation may provide for cumulative voting at all elections of directors of the corporation, or at elections held under specified circumstances. Either the certificate of incorporation or the bylaws may specify the number of shares and/or the amount of other securities that must be represented at a meeting in order to constitute a quorum, but in no event can a quorum consist of less than one-third of the shares entitled to vote at a meeting.

Stockholders as of the record date for the meeting are entitled to vote at the meeting, and the board of directors may fix a record date that is no more than 60 nor less than 10 days before the date of the meeting, and if no record date is set then the record date is the close of business on the day next preceding the day on which notice is given, or if notice is waived then the record date is the close of business on the day next preceding the day on which the meeting is held. The determination of the stockholders of record entitled to notice or to vote at a meeting of stockholders shall apply to any adjournment of the meeting, but the board of directors may fix a new record date for the adjourned meeting.

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Shareholder Proposals

Denmark. According to the DCA and our articles of association, extraordinary general meetings of shareholders will be held whenever our board of directors or our appointed auditor requires. In addition, one or more shareholders representing at least 5% of the registered share capital of the company may, in writing, require that a general meeting be convened. If such a demand is made, the board of directors shall convene the general meeting with three to five weeks' notice within 14 days thereafter.

All shareholders have the right to present proposals for adoption at the annual general meeting, provided that the proposals are submitted at least six weeks prior to the meeting. In the event that the request is made at a later date, the board of directors will determine whether the proposals were made in due time to be included on the agenda.

Delaware. Delaware law does not specifically grant stockholders the right to bring business before an annual or special meeting of stockholders. However, if a Delaware corporation is subject to the SEC's proxy rules, a stockholder who owns at least \$2,000 in market value, or 1% of the corporation's securities entitled to vote, may propose a matter for a vote at an annual or special meeting in accordance with those rules.

Action by Written Consent

Denmark. Under Danish law, shareholders may take action and pass resolutions by written consent if such consent is unanimous. However, for a listed company, this method of adopting resolutions is generally not feasible.

Delaware. Although permitted by Delaware law, publicly listed companies do not typically permit stockholders of a corporation to take action by written consent.

Appraisal Rights

Denmark. The concept of appraisal rights does not exist under Danish law, except in connection with statutory redemption rights according to the DCA.

According to Section 73 of the DCA, a minority shareholder may require a majority shareholder that holds more than nine-tenths of the company's registered share capital and voting rights to redeem his or her shares. Similarly, shares in a company may be redeemed in whole or in part by a shareholder holding more than nine-tenths of the shares and the corresponding voting rights in the company, according to Section 70 of the DCA. In the event that the parties cannot agree to the redemption squeeze out price, this shall be determined by an independent evaluator appointed by the court. Additionally, there are specific regulations in Sections 249, 267, 285 and 305 of the DCA that require compensation in the event of national or cross-border mergers and demergers. Moreover, shareholders who vote against a cross-border merger or demerger are, according to Sections 286 and 306 of the DCA, entitled to have their shares redeemed.

Delaware. The Delaware General Corporation Law provides for stockholder appraisal rights, or the right to demand payment in cash of the judicially determined fair value of the stockholder's shares, in connection with certain mergers and consolidations.

Shareholder Suits

Denmark. Under Danish law, only a company itself can bring a civil action against a third party; an individual shareholder does not have the right to bring an action on behalf of a company. However, if shareholders representing at least one-tenth of the share capital have opposed at a general meeting a decision to grant discharge to a member of our board of directors or our executive management or refrain from bringing law

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suits against, among other persons, a member of our board of directors or executive management, a shareholder may bring a derivative action on behalf of our company against, among other persons, a member of our board of directors or executive management. An individual shareholder may, in its own name, have an individual right to take action against such third party in the event that the cause for the liability of that third party also constitutes a negligent act directly against such individual shareholder.

Delaware. Under the Delaware General Corporation Law, a stockholder may bring a derivative action on behalf of the corporation to enforce the rights of the corporation. An individual also may commence a class action suit on behalf of himself and other similarly situated stockholders where the requirements for maintaining a class action under Delaware law have been met. A person may institute and maintain such a suit only if that person was a stockholder at the time of the transaction which is the subject of the suit. In addition, under Delaware case law, the plaintiff normally must be a stockholder at the time of the transaction that is the subject of the suit and throughout the duration of the derivative suit. Delaware law also requires that the derivative plaintiff make a demand on the directors of the corporation to assert the corporate claim before the suit may be prosecuted by the derivative plaintiff in court, unless such a demand would be futile.

Repurchase of Shares

Denmark. Danish limited liability companies may not subscribe for newly issued shares in their own capital. Such companies may, however, according to the DCA Sections 196-201, acquire fully paid shares of themselves, provided that the board of directors has been authorized to do so by the shareholders at a general meeting. Such authorization can only be given for a maximum period of five years and the authorization shall fix (i) the maximum value of the shares and (ii) the minimum and the highest amount that the company may pay for the shares. Such purchase of shares may generally only be acquired using distributable reserves. In addition, the board of directors may, on behalf of the company, acquire the company's own shares, without authorization, in case it is necessary to avoid a considerable and imminent detrimental effect on the company and provided certain conditions are met. In case the company has acquired its own shares under such circumstances the board of directors is obligated to inform the shareholders of such acquisition at the next general meeting. See “—Authorizations to our Board of Directors.”

Delaware. Under the Delaware General Corporation Law, a corporation may purchase or redeem its own shares unless the capital of the corporation is impaired or the purchase or redemption would cause an impairment of the capital of the corporation. A Delaware corporation may, however, purchase or redeem out of capital any of its preferred shares or, if no preferred shares are outstanding, any of its own shares if such shares will be retired upon acquisition and the capital of the corporation will be reduced in accordance with specified limitations.

Anti-Takeover Provisions

Denmark. Under Danish law, it is possible to implement limited protective anti-takeover measures. Such provisions may include, among other things, (i) different share classes with different voting rights and (ii) notification requirements concerning participation in general meetings. We have currently not adopted any such provisions, except for the notification requirements concerning participation in general meetings. See description above under the caption “—Articles of Association and Danish Corporate Law—General Meetings and Voting Rights.”

Delaware. In addition to other aspects of Delaware law governing fiduciary duties of directors during a potential takeover, the Delaware General Corporation Law also contains a business combination statute that protects Delaware companies from hostile takeovers and from actions following the takeover by prohibiting some transactions once an acquirer has gained a significant holding in the corporation.

Section 203 of the Delaware General Corporation Law prohibits “business combinations,” including mergers, sales and leases of assets, issuances of securities and similar transactions by a corporation or a

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subsidiary with an interested stockholder that beneficially owns 15% or more of a corporation's voting stock, within three years after the person becomes an interested stockholder, unless:

- the transaction that will cause the person to become an interested stockholder is approved by the board of directors of the target prior to the transaction;
- after the completion of the transaction in which the person becomes an interested stockholder, the interested stockholder holds at least 85% of the voting stock of the corporation not including shares owned by persons who are directors and officers of interested stockholders and shares owned by specified employee benefit plans; or
- after the person becomes an interested stockholder, the business combination is approved by the board of directors of the corporation and holders of at least 66.67% of the outstanding voting stock, excluding shares held by the interested stockholder.

A Delaware corporation may elect not to be governed by Section 203 by a provision contained in the original certificate of incorporation of the corporation or an amendment to the original certificate of incorporation or to the bylaws of the company, which amendment must be approved by a majority of the shares entitled to vote and may not be further amended by the board of directors of the corporation. Such an amendment is not effective until 12 months following its adoption.

Inspection of Books and Records

Denmark. According to Section 150 of the DCA, a shareholder may, at the annual general meeting or at a general meeting whose agenda includes such item, request an inspection of the company's books regarding specific issues concerning the management of the company or specific annual reports. If approved by shareholders with a simple majority, one or more investigators are elected. If the proposal is not approved by a simple majority but 25% of the share capital votes in favor of the proposal, then any shareholder may, no later than four weeks after the general meeting, request the bankruptcy court for the district in which the company's registered office is situated to appoint investigators.

Delaware. Under the Delaware General Corporation Law, any stockholder may inspect certain of the corporation's books and records, for any proper purpose, during the corporation's usual hours of business.

Pre-Emptive Rights

Denmark. If our shareholders at a general meeting resolve to increase our share capital by a cash contribution, section 162 of the DCA will apply. Under that section, shareholders have a pre-emptive right to subscribe for new shares in proportion to their existing shareholdings. However, the pre-emptive right may be derogated from by a majority comprising at least two-thirds of the votes cast, as well as at least two-thirds of the share capital represented at the general meeting, provided the share capital increase takes place at market price or nine-tenths of the votes cast, as well as at least nine-tenths of the share capital represented at the general meeting if the share capital increase takes place below market price, unless (i) such capital increase is directed at certain but not all shareholders (in which case all shareholders must consent); or (ii) such capital increase is directed at our employees whereby a majority comprising at least two-thirds of the votes cast, as well as at least two-thirds of the share capital represented at the general meeting is required. Further, the pre-emptive rights may be derogated from by an exercise of the board of directors of a valid authorization in our articles of association, provided that the share capital increase takes place at or above market price. The board of directors may resolve to increase our share capital without pre-emptive subscription rights for existing shareholders pursuant to the authorizations described above under the caption "—Authorizations to our Board of Directors."

Unless future issuances of new shares are registered under the Securities Act or with any authority outside Denmark, U.S. shareholders and shareholders in jurisdictions outside Denmark may be unable to exercise their pre-emptive subscription rights under the law of their respective jurisdictions, including the U.S. securities law.

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Delaware. Under the Delaware General Corporation Law, stockholders have no pre-emptive rights to subscribe for additional issues of stock or to any security convertible into such stock unless, and to the extent that, such rights are expressly provided for in the certificate of incorporation.

Dividends

Denmark. Under Danish law, the distribution of ordinary and interim dividends requires the approval of a company's shareholders at a company's general meeting. In addition the shareholders may authorize the board of directors to distribute interim dividends. We may only pay out dividends from our distributable reserves, which are defined as results from operations carried forward and reserves that are not bound by law after deduction of loss carried forward. It is possible under Danish law to pay out interim dividends. The decision to pay out interim dividends shall be accompanied by a balance sheet, and the board of directors determines whether it will be sufficient to use the statement of financial position from the annual report or if an interim statement of financial position for the period from the annual report period until the interim dividend payment shall be prepared. If the decision to distribute interim dividends is passed more than six months after the date of the statement of financial position as set out in our latest adopted annual report, an interim statement of financial position must be prepared and reviewed by our auditor. The statement of financial position or the interim statement of financial position, as applicable, must show that sufficient funds are available for distribution. Our general meeting of shareholders cannot resolve to distribute dividends at an amount exceeding the amount recommended or approved by our board of directors. Moreover, ordinary dividends and interim dividends may only be made out of distributable reserves and may not exceed what is considered sound and adequate with regard to our financial condition or be to the detriment of our creditors.

Delaware. Under the Delaware General Corporation Law, a Delaware corporation may pay dividends out of its surplus (the excess of net assets over capital), or in case there is no surplus, out of its net profits for the fiscal year in which the dividend is declared and/or the preceding fiscal year (provided that the amount of the capital of the corporation is not less than the aggregate amount of the capital represented by the issued and outstanding stock of all classes having a preference upon the distribution of assets). In determining the amount of surplus of a Delaware corporation, the assets of the corporation, including stock of subsidiaries owned by the corporation, must be valued at their fair market value as determined by the board of directors, without regard to their historical book value. Dividends may be paid in the form of shares, property or cash.

Shareholder Vote on Certain Reorganizations

Denmark. Under Danish law, all amendments to the articles of association shall be approved by the general meeting of shareholders with at least two-thirds of the votes cast and two-thirds of 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. The same applies to solvent liquidations, mergers with the company as the discontinuing entity, mergers with the company as the continuing entity if shares are issued in connection therewith and demergers. Under Danish law, it is debatable whether the shareholders must approve a decision to sell all or virtually all of the company's business/assets.

Delaware. Under the Delaware General Corporation Law, the vote of a majority of the outstanding shares of capital stock entitled to vote thereon generally is necessary to approve a merger or consolidation or the sale of all or substantially all of the assets of a corporation. The Delaware General Corporation Law permits a corporation to include in its certificate of incorporation a provision requiring for any corporate action the vote of a larger portion of the stock or of any class or series of stock than would otherwise be required. However, under the Delaware General Corporation Law, no vote of the stockholders of a surviving corporation to a merger is needed, unless required by the certificate of incorporation, if (1) the agreement of merger does not amend in any respect the certificate of incorporation of the surviving corporation, (2) the shares of stock of the surviving corporation are not changed in the merger and (3) the number of ordinary shares of the surviving corporation into which any other shares, securities or obligations to be issued in the merger may be converted does not exceed 20% of the

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surviving corporation's shares outstanding immediately prior to the effective date of the merger. In addition, stockholders may not be entitled to vote in certain mergers with other corporations that own 90% or more of the outstanding shares of each class of stock of such corporation, but the stockholders will be entitled to appraisal rights.

Mandatory Redemption of Shares

Denmark: Where a shareholder holds more than nine-tenths of the shares in a company and a corresponding proportion of the voting rights, such shareholder may, pursuant to the DCA, Section 70, demand that the other shareholders have their shares redeemed by that shareholder. In this case, the other shareholders must be requested, under the rules governing notices for general meeting, to transfer their shares to the shareholder within four weeks after the request to transfer their shares. In addition, the other shareholders shall through the Danish Business Authority's IT system be requested to transfer their shares within the same four-week period. Specific requirements apply to the contents of the notices to the other shareholders regarding the redemption. If the redemption price cannot be agreed upon, the redemption price must be determined by an independent expert appointed by the court in the jurisdiction of the company's registered office in accordance with the provisions of the DCA. However, the redemption price will be deemed fair under any circumstances, provided that (i) the redemption price is equal to the consideration paid by the bidder in connection with a voluntary tender offer by which the bidder obtained at least 90% of the voting rights or (ii) the redemption price is equal to the consideration paid by the bidder in connection with a mandatory tender offer. To the extent any minority shareholders have not transferred their shares to the acquiring shareholder before the expiry of the four-week period, the redeeming shareholder shall pay the redemption price to the remaining minority shareholders through the securities deposit. Upon such payment through the securities deposit, the minority shareholders will have been redeemed and the minority shareholders shall in such case through the Danish Business Authority's IT system be notified that the right to require determination of the redemption price by the independent expert expires at the end of a period, which cannot be less than three months pursuant to the DCA, Section 72.

Furthermore, where a shareholder holds more than nine-tenths of the shares in a company and a corresponding proportion of the voting rights, the other shareholders may require such shareholder to acquire their shares pursuant to Section 73 of the DCA. If the redemption price cannot be agreed upon, the redemption price must be determined by an independent expert appointed by the court in the jurisdiction of the company's registered office in accordance with the provisions of the DCA. Expenses relating to the determination of the redemption price must be paid by the shareholder requesting such determination. If the expert's valuation is higher than the price offered by the redeeming shareholder, the court may order the redeeming shareholder to pay the expenses relating to determination of the redemption price in full or in part.

Delaware: The Delaware General Corporation Law provides for stockholder appraisal rights, or the right to demand payment in cash of the judicially determined fair value of the stockholder's shares, in connection with certain mergers and consolidations.

Amendments to Governing Documents

Denmark. All resolutions made by the general meeting may be adopted by a simple majority of the votes, subject only to the mandatory provisions of the DCA and the articles of association. Resolutions concerning all amendments to the articles of association must be passed by two-thirds of the votes cast as well as two-thirds of 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.

Certain resolutions, which limit a shareholder's ownership or voting rights, are subject to approval by at least a nine-tenth majority of the votes cast and the share capital represented at the general meeting. Decisions to impose any or increase any obligations of the shareholders towards the company require unanimity.

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Delaware. Under the Delaware General Corporation Law, a corporation's certificate of incorporation may be amended only if adopted and declared advisable by the board of directors and approved by a majority of the outstanding shares entitled to vote, and the bylaws may be amended with the approval of a majority of the outstanding shares entitled to vote and may, if so provided in the certificate of incorporation, also be amended by the board of directors.

Exchange Controls

There are no laws or regulations in Denmark that restrict the export or import of capital (except for certain investments in certain domains in accordance with applicable resolutions adopted by the United Nations or the European Union), including, but not limited to, foreign exchange controls, or which affect the remittance of dividends, interest or other payments to non-resident holders of our ordinary shares.

Transfer Agent and Registrar

The transfer agent and registrar for our shares is Computershare A/S, Lottenborgvej 26 D, 1., DK-2800 Kgs. Lyngby, Denmark. The Bank of New York Mellon serves as the depositary, registrar and transfer agent for the ADSs.

DESCRIPTION OF AMERICAN DEPOSITARY SHARES

American Depositary Shares

The Bank of New York Mellon, as depositary, registers and delivers ADSs. Each ADS represents one ordinary share (or a right to receive one ordinary share) deposited with Danske Bank A/S, as custodian for the depositary in the Kingdom of Denmark. Each ADS also represents any other securities, cash or other property that may be held by the depositary. The deposited shares together with any other securities, cash or other property held by the depositary are referred to as the deposited securities. A copy of our Deposit Agreement among us, the depositary, owners and holders of ADSs was filed with the SEC as an exhibit to our Annual Report on Form 20-F for the year ended December 31, 2020 filed on March 2, 2021 (File No. 001-39545).

Any ordinary shares that may be issued pursuant to this prospectus and the applicable prospectus supplement, whether directly or upon exercise of warrants, can be deposited for delivery of ADSs. The ADSs may be uncertificated securities or certificated securities evidenced by American Depositary Receipts, or ADRs. The depositary's office at which the ADSs will be administered and its principal executive office are located at 240 Greenwich Street, New York, New York 10286.

You may hold ADSs either (A) directly (i) by having an American Depositary Receipt, also referred to as an ADR, which is a certificate evidencing a specific number of ADSs, registered in your name, or (ii) by having uncertificated ADSs registered in your name, or (B) indirectly by holding a security entitlement in ADSs through your broker or other financial institution that is a direct or indirect participant in DTC. If you hold ADSs directly, you are a registered ADS holder, also referred to as an ADS holder. This description assumes you are an ADS holder. If you hold the ADSs indirectly, you must rely on the procedures of your broker or other financial institution to assert the rights of ADS holders described in this section. You should consult with your broker or financial institution to find out what those procedures are.

Registered holders of uncertificated ADSs will receive statements from the depositary confirming their holdings.

As an ADS holder, we will not treat you as one of our shareholders and you will not have shareholder rights. Danish law governs shareholder rights. The depositary will be the holder of the shares underlying your ADSs. As a registered holder of ADSs, you will have ADS holder rights. A deposit agreement among us, the depositary, ADS holders and all other persons indirectly or beneficially holding ADSs sets out ADS holder rights as well as the rights and obligations of the depositary. New York law governs the deposit agreement and the ADSs.

The following is a summary of the material provisions of the deposit agreement. For more complete information, you should read the entire deposit agreement and the form of ADR. Directions on how to obtain copies of those documents are provided in the section titled "Where You Can Find Additional Information."

Dividends and Other Distributions

How Will You Receive Dividends and Other Distributions on the Shares?

The depositary has agreed to pay or distribute to ADS holders the cash dividends or other distributions it or the custodian receives on shares or other deposited securities, upon payment or deduction of its fees and expenses. You will receive these distributions in proportion to the number of shares your ADSs represent.

Cash

The depositary will convert any cash dividend or other cash distribution we pay on the shares into U.S. dollars, if it can do so on a reasonable basis and can transfer the U.S. dollars to the United States. If that is not possible or if any government approval is needed and cannot be obtained, the deposit agreement allows the depositary to distribute the foreign currency only to those ADS holders to whom it is possible to do so. It will hold the foreign currency it cannot convert for the account of the ADS holders who have not been paid. It will not invest the foreign currency and it will not be liable for any interest.

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Before making a distribution, any withholding taxes, or other governmental charges that must be paid will be deducted. See “Taxation.” The depositary will distribute only whole U.S. dollars and cents and will round fractional cents to the nearest whole cent. If the exchange rates fluctuate during a time when the depositary cannot convert the foreign currency, you may lose some of the value of the distribution.

Shares

The depositary may distribute additional ADSs representing any shares we distribute as a dividend or free distribution. The depositary will only distribute whole ADSs. It will sell shares which would require it to deliver a fraction of an ADS (or ADSs representing those shares) and distribute the net proceeds in the same way as it does with cash. If the depositary does not distribute additional ADSs, the outstanding ADSs will also represent the new shares. The depositary may sell a portion of the distributed shares (or ADSs representing those shares) sufficient to pay its fees and expenses in connection with that distribution.

Rights to Purchase Additional Shares

If we offer holders of our securities any rights to subscribe for additional shares or any other rights, the depositary may (i) exercise those rights on behalf of ADS holders, (ii) distribute those rights to ADS holders or (iii) sell those rights and distribute the net proceeds to ADS holders, in each case after deduction or upon payment of its fees and expenses. To the extent the depositary does not do any of those things, it will allow the rights to lapse. In that case, you will receive no value for them. The depositary will exercise or distribute rights only if we ask it to and provide satisfactory assurances to the depositary that it is legal to do so. If the depositary will exercise rights, it will purchase the securities to which the rights relate and distribute those securities or, in the case of shares, new ADSs representing the new shares, to subscribing ADS holders, but only if ADS holders have paid the exercise price to the depositary. U.S. securities laws may restrict the ability of the depositary to distribute rights or ADSs or other securities issued on exercise of rights to all or certain ADS holders, and the securities distributed may be subject to restrictions on transfer.

Other Distributions

The depositary will send to ADS holders anything else we distribute on deposited securities by any means it thinks is legal, fair and practical. If it cannot make the distribution in that way, the depositary has a choice. It may decide to sell what we distributed and distribute the net proceeds, in the same way as it does with cash. Or, it may decide to hold what we distributed, in which case ADSs will also represent the newly distributed property. However, the depositary is not required to distribute any securities (other than ADSs) to ADS holders unless it receives satisfactory evidence from us that it is legal to make that distribution. The depositary may sell a portion of the distributed securities or property sufficient to pay its fees and expenses in connection with that distribution. U.S. securities laws may restrict the ability of the depositary to distribute securities to all or certain ADS holders, and the securities distributed may be subject to restrictions on transfer.

The depositary is not responsible if it decides that it is unlawful or impractical to make a distribution available to any ADS holders. We have no obligation to register ADSs, shares, rights or other securities under the Securities Act. We also have no obligation to take any other action to permit the distribution of ADSs, shares, rights or anything else to ADS holders. This means that you may not receive the distributions we make on our shares or any value for them if it is illegal or impractical for us to make them available to you.

Deposit, Withdrawal and Cancellation

How are ADSs Issued?

The depositary will deliver ADSs if you or your broker deposits shares or evidence of rights to receive shares with the custodian. Upon payment of its fees and expenses and of any taxes or charges, such as stamp taxes or stock transfer taxes or fees, the depositary will register the appropriate number of ADSs in the names you request and will deliver the ADSs to or upon the order of the person or persons that made the deposit.

How Can ADS Holders Withdraw the Deposited Securities?

You may surrender your ADSs to the depository for the purpose of withdrawal. Upon payment of its fees and expenses and of any taxes or charges, such as stamp taxes or stock transfer taxes or fees, the depository will deliver the shares and any other deposited securities underlying the ADSs to the ADS holder or a person the ADS holder designates at the office of the custodian. Or, at your request, risk and expense, the depository will deliver the deposited securities at its office, if feasible. However, the depository is not required to accept surrender of ADSs to the extent it would require delivery of a fraction of a deposited share or other security. The depository may charge you a fee and its expenses for instructing the custodian regarding delivery of deposited securities.

How do ADS Holders Interchange Between Certificated ADSs and Uncertificated ADSs?

You may surrender your ADR to the depository for the purpose of exchanging your ADR for uncertificated ADSs. The depository will cancel that ADR and will send to the ADS holder a statement confirming that the ADS holder is the registered holder of uncertificated ADSs. Upon receipt by the depository of a proper instruction from a registered holder of uncertificated ADSs requesting the exchange of uncertificated ADSs for certificated ADSs, the depository will execute and deliver to the ADS holder an ADR evidencing those ADSs.

Voting Rights

How do You Vote?

ADS holders may instruct the depository how to vote the number of deposited shares their ADSs represent. If we request the depository to solicit your voting instructions (and we are not required to do so), the depository will notify you of a shareholders' meeting and send or make voting materials available to you. Those materials will describe the matters to be voted on and explain how ADS holders may instruct the depository how to vote. For instructions to be valid, they must reach the depository by a date set by the depository. The depository will try, as far as practical, subject to the laws of the Kingdom of Denmark and the provisions of our articles of association or similar documents, to vote or to have its agents vote the shares or other deposited securities as instructed by ADS holders. If we do not request the depository to solicit your voting instructions, you can still send voting instructions, and, in that case, the depository may try to vote as you instruct, but it is not required to do so.

Except by instructing the depository as described above, you won't be able to exercise voting rights unless you surrender your ADSs and withdraw the shares. However, you may not know about the meeting enough in advance to withdraw the shares. In any event, the depository will not exercise any discretion in voting deposited securities and it will only vote or attempt to vote as instructed.

We cannot assure you that you will receive the voting materials in time to ensure that you can instruct the depository to vote your shares. In addition, the depository and its agents are not responsible for failing to carry out voting instructions or for the manner of carrying out voting instructions. This means that you may not be able to exercise voting rights and there may be nothing you can do if your shares are not voted as you requested.

In order to give you a reasonable opportunity to instruct the depository as to the exercise of voting rights relating to Deposited Securities, if we request the Depository to act, we will provide the depository with the proposed meeting date and details of the matters proposed to be voted on at least 30 days before the meeting date and the depository will send voting materials to you approximately 21 days before the meeting date.

Fees and Expenses

Persons depositing or withdrawing shares or ADS holders must pay:
\$5.00 (or less) per 100 ADSs (or portion of 100 ADSs)

For:
Issuance of ADSs, including issuances resulting from a distribution of shares or rights or other property
Cancellation of ADSs for the purpose of withdrawal, including if the deposit agreement terminates

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\$.05 (or less) per ADS	Any cash distribution to ADS holders
A fee equivalent to the fee that would be payable if securities distributed to you had been shares and the shares had been deposited for issuance of ADSs	Distribution of securities distributed to holders of deposited securities (including rights) that are distributed by the depositary to ADS holders
\$.05 (or less) per ADS per calendar year	Depository services
Registration or transfer fees	Transfer and registration of shares on our share register to or from the name of the depositary or its agent when you deposit or withdraw shares
Expenses of the depositary	Cable (including SWIFT) and facsimile transmissions (when expressly provided in the deposit agreement) Converting foreign currency to U.S. dollars
Taxes and other governmental charges the depositary or the custodian has to pay on any ADSs or shares underlying ADSs, such as stock transfer taxes, stamp duty or withholding taxes	As necessary
Any charges incurred by the depositary or its agents for servicing the deposited securities	As necessary

The depositary collects its fees for delivery and surrender of ADSs directly from investors depositing shares or surrendering ADSs for the purpose of withdrawal or from intermediaries acting for them. The depositary collects fees for making distributions to investors by deducting those fees from the amounts distributed or by selling a portion of distributable property to pay the fees. The depositary may collect its annual fee for depository services by deduction from cash distributions or by directly billing investors or by charging the book-entry system accounts of participants acting for them. The depositary may collect any of its fees by deduction from any cash distribution payable (or by selling a portion of securities or other property distributable) to ADS holders that are obligated to pay those fees. The depositary may generally refuse to provide fee-attracting services until its fees for those services are paid.

From time to time, the depositary may make payments to us to reimburse us for costs and expenses generally arising out of establishment and maintenance of the ADS program, waive fees and expenses for services provided to us by the depositary or share revenue from the fees collected from ADS holders. In performing its duties under the deposit agreement, the depositary may use brokers, dealers, foreign currency dealers or other service providers that are owned by or affiliated with the depositary and that may earn or share fees, spreads or commissions.

The depositary may convert currency itself or through any of its affiliates, or the custodian or we may convert currency and pay U.S. dollars to the depositary. Where the depositary converts currency itself or through any of its affiliates, the depositary acts as principal for its own account and not as agent, advisor, broker or fiduciary on behalf of any other person and earns revenue, including, without limitation, transaction spreads, that it will retain for its own account. The revenue is based on, among other things, the difference between the exchange rate assigned to the currency conversion made under the deposit agreement and the rate that the depositary or its affiliate receives when buying or selling foreign currency for its own account. The depositary makes no representation that the exchange rate used or obtained by it or its affiliate in any currency conversion under the deposit agreement will be the most favorable rate that could be obtained at the time or that the method

by which that rate will be determined will be the most favorable to ADS holders, subject to the depositary's obligation to act without negligence or bad faith. The methodology used to determine exchange rates used in currency conversions made by the depositary is available upon request. Where the custodian converts currency, the custodian has no obligation to obtain the most favorable rate that could be obtained at the time or to ensure that the method by which that rate will be determined will be the most favorable to ADS holders, and the depositary makes no representation that the rate is the most favorable rate and will not be liable for any direct or indirect losses associated with the rate. In certain instances, the depositary may receive dividends or other distributions from us in U.S. dollars that represent the proceeds of a conversion of foreign currency or translation from foreign currency at a rate that was obtained or determined by us and, in such cases, the depositary will not engage in, or be responsible for, any foreign currency transactions and neither it nor we make any representation that the rate obtained or determined by us is the most favorable rate and neither it nor we will be liable for any direct or indirect losses associated with the rate.

Payment of Taxes

You will be responsible for any taxes or other governmental charges payable on your ADSs or on the deposited securities represented by any of your ADSs. The depositary may refuse to register any transfer of your ADSs or allow you to withdraw the deposited securities represented by your ADSs until those taxes or other charges are paid. It may apply payments owed to you or sell deposited securities represented by your ADSs to pay any taxes owed and you will remain liable for any deficiency. If the depositary sells deposited securities, it will, if appropriate, reduce the number of ADSs to reflect the sale and pay to ADS holders any proceeds, or send to ADS holders any property, remaining after it has paid the taxes.

Tender and Exchange Offers; Redemption, Replacement or Cancellation of Deposited Securities

The depositary will not tender deposited securities in any voluntary tender or exchange offer unless instructed to do so by an ADS holder surrendering ADSs and subject to any conditions or procedures the depositary may establish.

If deposited securities are redeemed for cash in a transaction that is mandatory for the depositary as a holder of deposited securities, the depositary will call for surrender of a corresponding number of ADSs and distribute the net redemption money to the holders of called ADSs upon surrender of those ADSs.

If there is any change in the deposited securities such as a sub-division, combination or other reclassification, or any merger, consolidation, recapitalization or reorganization affecting the issuer of deposited securities in which the depositary receives new securities in exchange for or in lieu of the old deposited securities, the depositary will hold those replacement securities as deposited securities under the deposit agreement. However, if the depositary decides it would not be lawful and practical to hold the replacement securities because those securities could not be distributed to ADS holders or for any other reason, the depositary may instead sell the replacement securities and distribute the net proceeds upon surrender of the ADSs.

If there is a replacement of the deposited securities and the depositary will continue to hold the replacement securities, the depositary may distribute new ADSs representing the new deposited securities or ask you to surrender your outstanding ADRs in exchange for new ADRs identifying the new deposited securities.

If there are no deposited securities underlying ADSs, including if the deposited securities are cancelled, or if the deposited securities underlying ADSs have become apparently worthless, the depositary may call for surrender of those ADSs or cancel those ADSs upon notice to the ADS holders.

Amendment and Termination

How May the Deposit Agreement be Amended?

We may agree with the depository to amend the deposit agreement and the ADRs without your consent for any reason. If an amendment adds or increases fees or charges, except for taxes and other governmental charges or expenses of the depository for registration fees, facsimile costs, delivery charges or similar items, or prejudices a substantial right of ADS holders, it will not become effective for outstanding ADSs until 30 days after the depository notifies ADS holders of the amendment. At the time an amendment becomes effective, you are considered, by continuing to hold your ADSs, to agree to the amendment and to be bound by the ADRs and the deposit agreement as amended.

How May the Deposit Agreement be Terminated?

The depository will initiate termination of the deposit agreement if we instruct it to do so. The depository may initiate termination of the deposit agreement if

- 60 days have passed since the depository told us it wants to resign but a successor depository has not been appointed and accepted its appointment;
- we delist the ADSs from an exchange in the United States on which they were listed and do not list the ADSs on another exchange in the United States or make arrangements for trading of ADSs on the U.S. over-the-counter market;
- we delist our shares from an exchange outside the United States on which they were listed and do not list the shares on another exchange outside the United States;
- the depository has reason to believe the ADSs have become, or will become, ineligible for registration on Form F-6 under the Securities Act;
- we appear to be insolvent or enter insolvency proceedings;
- all or substantially all the value of the deposited securities has been distributed either in cash or in the form of securities;
- there are no deposited securities underlying the ADSs or the underlying deposited securities have become apparently worthless; or
- there has been a replacement of deposited securities.

If the deposit agreement will terminate, the depository will notify ADS holders at least 90 days before the termination date. At any time after the termination date, the depository may sell the deposited securities. After that, the depository will hold the money it received on the sale, as well as any other cash it is holding under the deposit agreement, unsegregated and without liability for interest, for the pro rata benefit of the ADS holders that have not surrendered their ADSs. Normally, the depository will sell as soon as practicable after the termination date.

After the termination date and before the depository sells, ADS holders can still surrender their ADSs and receive delivery of deposited securities, except that the depository may refuse to accept a surrender for the purpose of withdrawing deposited securities or reverse previously accepted surrenders of that kind that have not settled if it would interfere with the selling process. The depository may refuse to accept a surrender for the purpose of withdrawing sale proceeds until all the deposited securities have been sold. The depository will continue to collect distributions on deposited securities, but, after the termination date, the depository is not required to register any transfer of ADSs or distribute any dividends or other distributions on deposited securities to the ADSs holder (until they surrender their ADSs) or give any notices or perform any other duties under the deposit agreement except as described in this paragraph.

Limitations on Obligations and Liability

Limits on our Obligations and the Obligations of the Depository; Limits on Liability to Holders of ADSs

The deposit agreement expressly limits our obligations and the obligations of the depository. It also limits our liability and the liability of the depository. We and the depository:

- are only obligated to take the actions specifically set forth in the deposit agreement without negligence or bad faith, and the depository will not be a fiduciary or have any fiduciary duty to holders of ADSs;
- are not liable if we are or it is prevented or delayed by law or by events or circumstances beyond our or its ability to prevent or counteract with reasonable care or effort from performing our or its obligations under the deposit agreement;
- are not liable if we or it exercises discretion permitted under the deposit agreement;
- are not liable for the inability of any holder of ADSs to benefit from any distribution on deposited securities that is not made available to holders of ADSs under the terms of the deposit agreement, or for any special, consequential or punitive damages for any breach of the terms of the deposit agreement;
- have no obligation to become involved in a lawsuit or other proceeding related to the ADSs or the deposit agreement on your behalf or on behalf of any other person;
- may rely upon any documents we believe or it believes in good faith to be genuine and to have been signed or presented by the proper person;
- are not liable for the acts or omissions of any securities depository, clearing agency or settlement system; and
- the depository has no duty to make any determination or provide any information as to our tax status, or any liability for any tax consequences that may be incurred by ADS holders as a result of owning or holding ADSs or be liable for the inability or failure of an ADS holder to obtain the benefit of a foreign tax credit, reduced rate of withholding or refund of amounts withheld in respect of tax or any other tax benefit.

In the deposit agreement, we and the depository agree to indemnify each other under certain circumstances.

Requirements for Depository Actions

Before the depository will deliver or register a transfer of ADSs, make a distribution on ADSs, or permit withdrawal of shares, the depository may require:

- payment of stock transfer or other taxes or other governmental charges and transfer or registration fees charged by third parties for the transfer of any shares or other deposited securities;
- satisfactory proof of the identity and genuineness of any signature or other information it deems necessary; and
- compliance with regulations it may establish, from time to time, consistent with the deposit agreement, including presentation of transfer documents.

The depository may refuse to deliver ADSs or register transfers of ADSs when the transfer books of the depository or our transfer books are closed or at any time if the depository or we think it advisable to do so.

Your Right to Receive the Shares Underlying your ADSs

ADS holders have the right to cancel their ADSs and withdraw the underlying shares at any time except:

- when temporary delays arise because: (i) the depository has closed its transfer books or we have closed our transfer books; (ii) the transfer of shares is blocked to permit voting at a shareholders' meeting; or (iii) we are paying a dividend on our shares;
- when you owe money to pay fees, taxes and similar charges; or
- when it is necessary to prohibit withdrawals in order to comply with any laws or governmental regulations that apply to ADSs or to the withdrawal of shares or other deposited securities.

This right of withdrawal may not be limited by any other provision of the deposit agreement.

Direct Registration System

In the deposit agreement, all parties to the deposit agreement acknowledge that the Direct Registration System, also referred to as DRS, and Profile Modification System, also referred to as Profile, will apply to the ADSs. DRS is a system administered by DTC that facilitates interchange between registered holding of uncertificated ADSs and holding of security entitlements in ADSs through DTC and a DTC participant. Profile is a feature of DRS that allows a DTC participant, claiming to act on behalf of a registered holder of uncertificated ADSs, to direct the depository to register a transfer of those ADSs to DTC or its nominee and to deliver those ADSs to the DTC account of that DTC participant without receipt by the depository of prior authorization from the ADS holder to register that transfer.

In connection with and in accordance with the arrangements and procedures relating to DRS/Profile, the parties to the deposit agreement understand that the depository will not determine whether the DTC participant that is claiming to be acting on behalf of an ADS holder in requesting registration of transfer and delivery as described in the paragraph above has the actual authority to act on behalf of the ADS holder (notwithstanding any requirements under the Uniform Commercial Code). In the deposit agreement, the parties agree that the depository's reliance on and compliance with instructions received by the depository through the DRS/Profile system and in accordance with the deposit agreement will not constitute negligence or bad faith on the part of the depository.

Shareholder Communications; Inspection of Register of Holders of ADSs

The depository will make available for your inspection at its office all communications that it receives from us as a holder of deposited securities that we make generally available to holders of deposited securities. The depository will send you copies of those communications or otherwise make those communications available to you if we ask it to which we may choose not to. You have a right to inspect the register of holders of ADSs, but not for the purpose of contacting those holders about a matter unrelated to our business or the ADSs.

Jury Trial Waiver

The deposit agreement provides that, to the extent permitted by law, ADS holders waive the right to a jury trial of any claim they may have against us or the depository arising out of or relating to our shares, the ADSs or the deposit agreement, including any claim under the U.S. federal securities laws. If we or the depository opposed a jury trial demand based on the waiver, the court would determine whether the waiver was enforceable in the facts and circumstances of that case in accordance with applicable case law.

You will not, by agreeing to the terms of the deposit agreement, be deemed to have waived our or the depository's compliance with U.S. federal securities laws or the rules and regulations promulgated thereunder.

PLAN OF DISTRIBUTION

We may offer and sell the securities described in this prospectus in one or more of the following ways (or in any combination) from time to time:

- through underwriters or dealers;
- directly to a limited number of purchasers or to a single purchaser;
- through agents; or
- through any other method permitted by applicable law and described in the applicable prospectus supplement.

The distribution of securities may be carried out, from time to time, in one or more transactions, including:

- block transactions and transactions on The Nasdaq Global Select Market or any other securities exchange or quotation or trading service on which such securities may be listed, quoted or traded at the time of sale;
- purchases by a broker-dealer as principal and resale by the broker-dealer for its own account pursuant to a prospectus supplement;
- ordinary brokerage transactions and transactions in which a broker-dealer solicits purchasers;
- sales “at the market” to or through a market maker or into an existing trading market, on an exchange or otherwise; or
- sales in other ways not involving market makers or established trading markets, including direct sales to purchasers.

A prospectus supplement or supplements (and any related free writing prospectus that we may authorize to be provided to you) will describe the terms of the offering of the securities, including, to the extent applicable:

- the name or names of any underwriters, dealers or agents and the amounts of securities underwritten or purchased by each of them, if any;
- the method of distribution;
- the public offering price or purchase price and the proceeds to us from that sale;
- the expenses of the offering;
- any discounts or commissions to be allowed or paid to the underwriters, dealers or agents;
- all other items constituting underwriting compensation and the discounts and commissions to be allowed or paid to dealers, if any; and
- any other information regarding the distribution of the securities that we believe to be material.

Any ordinary shares will be listed on Nasdaq Copenhagen and any ADSs will be listed on The Nasdaq Global Select Market. Underwriters may offer and sell the securities at a fixed price or prices, which may be changed, or from time to time at market prices prevailing at the time of sale, at prices related to prevailing market prices or at negotiated prices. We may, from time to time, authorize agents acting on a best or reasonable efforts basis as our agents to solicit or receive offers to purchase the securities upon the terms and conditions as are set forth in the applicable prospectus supplement. Underwriters, dealers and agents participating in the distribution of the securities may be deemed to be underwriters within the meaning of the Securities Act, and may be deemed to have received compensation from us in the form of underwriting discounts or commissions and may also receive commissions from purchasers of securities for whom they may act as agent. Underwriters may sell securities to or through dealers, and dealers may receive compensation in the form of discounts, concessions or commissions from the underwriters and/or commissions from the purchasers for whom they may act as agent.

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Underwriters, dealers and agents who participate in the distribution of securities and their controlling persons may be entitled, under agreements that may be entered into with us to indemnification by us against certain liabilities, including liabilities under the Securities Act, or to contribution with respect to payments that the underwriters, dealers or agents and their controlling persons may be required to make in respect of those liabilities.

We may also make direct sales through subscription rights distributed to our existing shareholders on a pro rata basis, which may or may not be transferable. In any distribution of subscription rights to our shareholders, if all of the underlying securities are not subscribed for, we may then sell the unsubscribed securities directly to third parties or may engage the services of one or more underwriters, dealers or agents, including standby underwriters, to sell the unsubscribed securities to third parties.

Certain persons participating in an offering may engage in over-allotment, stabilizing transactions, short-covering transactions and penalty bids in accordance with applicable laws and regulations, including Regulation M under the Exchange Act, that stabilize, maintain or otherwise affect the price of the offered securities at a level above that which might otherwise prevail in the open market and affect the market abuse regulation (Commission Regulation (EU) no. 596/2014 of 16 April 2014), including delegated acts. Over-allotments or short sales of the securities involve the sale by persons participating in the offering of more securities than were sold to them. In these circumstances, these persons would cover such over-allotments or short positions by making purchases in the open market or by exercising their over-allotment option, if any. In addition, these persons may stabilize or maintain the price of the securities by bidding for or purchasing securities in the open market or by imposing penalty bids, whereby selling concessions allowed to dealers participating in the offering may be reclaimed if securities sold by them are repurchased in connection with stabilization transactions. If any such activities will occur, they will be described in the applicable prospectus supplement. These transactions may be discontinued at any time.

The specific terms of any lock-up provisions in respect of any given offering will be described in the applicable prospectus supplement.

The underwriters, dealers and agents may engage in transactions with us, or perform services for us, in the ordinary course of business for which they receive compensation.

TAXATION

Taxation in Denmark

A general summary of certain Danish tax considerations relating to the purchase, ownership and disposition of any of the securities offered by this prospectus will be set forth or incorporated by reference in a prospectus supplement relating to the offering of those securities.

Taxation in the United States

A general summary of the material U.S. federal income tax consequences relating to the purchase, ownership and disposition of any of the securities offered by this prospectus will be set forth or incorporated by reference in a prospectus supplement relating to the offering of those securities.

EXPENSES

The following table sets forth the expenses, other than any underwriting discounts and commissions or agency fees and other items constituting underwriters' or agents' compensation, expected to be incurred by us in connection with a possible offering of securities registered under the registration statement of which this prospectus is a part. All amounts are estimated other than the SEC registration fee.

SEC registration fee	\$ 6,952.50
FINRA filing fees	\$ 11,750.00
Legal fees and expenses	*
Accounting fees and expenses	*
Printing expenses	*
Miscellaneous expenses	*
Total	\$ *

* To be provided in a prospectus supplement or in a report on Form 6-K subsequently incorporated by reference into this prospectus.

LEGAL MATTERS

Unless the applicable prospectus supplement indicates otherwise, the validity of the securities in respect of which this prospectus is being delivered and certain legal matters with respect to Danish law will be passed upon by Gorrissen Federspiel Advokatpartnerselskab, Copenhagen, Denmark. Certain matters in respect of U.S. securities laws may be opined upon by Cooley LLP. Additional legal matters may be passed upon for any underwriters, dealers or agents by counsel that we will name in the applicable prospectus supplement.

EXPERTS

The consolidated financial statements of Orphazyme A/S appearing in Orphazyme A/S's Annual Report (Form 20-F) for the year ended December 31, 2020, have been audited by EY Godkendt Revisionspartnerselskab, independent registered public accounting firm, as set forth in their report thereon, included therein, and incorporated herein by reference. Such consolidated financial statements are incorporated herein by reference in reliance upon such report given on the authority of such firm as experts in accounting and auditing.

The registered business address of EY Godkendt Revisionspartnerselskab is Dirch Passers Allé 36, 2000 Frederiksberg, Denmark.

SERVICE OF PROCESS AND ENFORCEMENT OF LIABILITIES

We are organized under the laws of Denmark, with a domicile in the municipality of Copenhagen, Denmark.

Some of the members of the board of directors and the executive board named herein are residents of Denmark or other jurisdictions outside the United States. A substantial portion of ours and such persons' assets are located in Denmark or other jurisdictions outside the United States. As a result, it may not be possible for investors to effect service of process upon such persons or us with respect to litigation that may arise under U.S. law or to enforce against them or our company judgments obtained in U.S. courts, whether or not such judgments were made pursuant to civil liability provisions of the federal or state securities laws of the United States or any other laws of the United States.

There is not currently a treaty between the United States and Denmark providing for reciprocal recognition and enforceability of judgments rendered in connection with civil and commercial disputes and, accordingly, that a final judgment (other than arbitration awards) rendered by a U.S. court based on civil liability would not be enforceable in Denmark. It is uncertain whether Danish courts would allow actions to be predicated on the securities laws of the United States or other jurisdictions outside Denmark. Danish courts are likely to deny claims for punitive damages and may grant a reduced amount of damages compared to U.S. courts.

WHERE YOU CAN FIND ADDITIONAL INFORMATION

This prospectus is part of the registration statement on Form F-3 we filed with the SEC under the Securities Act. This prospectus does not contain all of the information set forth in the registration statement and the exhibits to the registration statement. For further information with respect to us and the securities we are offering under this prospectus, we refer you to the registration statement and the exhibits and schedules filed as a part of the registration statement, which may be obtained from the SEC or us, as provided below. Statements in this prospectus or any prospectus supplement are summaries and each statement is qualified in all respects by reference to the document to which it refers. You should refer to the actual documents for a more complete description of the relevant matters.

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You should rely only on the information contained in this prospectus or incorporated by reference. We have not authorized anyone else to provide you with different information. We are not making an offer of these securities in any state where the offer is not permitted. You should not assume that the information in this prospectus is accurate as of any date other than the date on the front page of this prospectus, regardless of the time of delivery of this prospectus or any sale of the securities offered by this prospectus.

We are subject to the periodic reporting and other informational requirements of the Exchange Act. Under the Exchange Act, we file annual reports and other information with the SEC. As a foreign private issuer, we are exempt from, among other things, the rules under the Exchange Act prescribing the furnishing and content of proxy statements and our officers, directors and principal shareholders are exempt from the reporting and short-swing profit recovery provisions contained in Section 16 of the Exchange Act.

The SEC maintains a website that contains reports, proxy and information statements and other information regarding issuers that file electronically with the SEC, including us. The address of the SEC website is www.sec.gov.

We also maintain a website at www.orphazyme.com through which you can access our SEC filings. The information contained on, or accessible through, our website is not incorporated by reference into this prospectus, and you should not consider any information contained in, or that can be accessed through, our website as part of this prospectus or in deciding whether to purchase ordinary shares, including ordinary shares in the form of ADSs. We have included our website address as an inactive textual reference only.

INCORPORATION BY REFERENCE

The SEC allows us to “incorporate by reference” information into this prospectus, which means that we can disclose important information to you by referring you to other documents filed separately with the SEC. The information incorporated by reference is deemed to be part of this prospectus, and subsequent information that we file with the SEC will automatically update and supersede that information. Any statement contained in a previously filed document incorporated by reference will be deemed to be modified or superseded for purposes of this prospectus to the extent that a statement contained in this prospectus modifies or replaces that statement.

This prospectus and any accompanying prospectus supplement incorporate by reference the documents set forth below that have previously been filed with the SEC:

- our Annual Report on [Form 20-F](#) for the year ended December 31, 2020, filed with the SEC on March 2, 2021;
- our reports on Form 6-K furnished to the SEC on [February 26, 2021](#), [March 1, 2021](#), [March 2, 2021](#), [March 3, 2021](#), [March 26, 2021](#), [March 29, 2021](#), [April 22, 2021](#), [May 7, 2021](#), [June 11, 2021](#), [June 21, 2021](#), [June 28, 2021](#), [August 31, 2021](#), [October 5, 2021](#) and [October 7, 2021](#); and
- the description of our ordinary shares and American Depositary Shares contained in our registration statement on [Form 8-A](#) (File No. 001-39545), filed with the SEC on September 22, 2020, including any amendments or reports filed for the purpose of updating such description.

We are also incorporating by reference all subsequent annual reports on Form 20-F that we file with the SEC and certain reports on Form 6-K that we furnish to the SEC after the date of this prospectus (if such reports on Form 6-K expressly state that they are incorporated in whole or in part by reference into the registration statement of which this prospectus forms a part) prior to the termination of this offering. In all cases, you should rely on the later information over different information included in this prospectus or any accompanying prospectus supplement.

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Unless expressly incorporated by reference, nothing in this prospectus shall be deemed to incorporate by reference information furnished to, but not filed with, the SEC. Copies of all documents incorporated by reference in this prospectus, other than exhibits to those documents unless such exhibits are specifically incorporated by reference in this prospectus, will be provided at no cost to each person, including any beneficial owner, who receives a copy of this prospectus on the written or oral request of that person made to:

Orphazyme A/S
Ole Maaløes Vej 3, DK-2200
Copenhagen N
Denmark
Tel: (+45) 28 98 90 55
Attention: Investor Relations

SEC Position on Indemnification for Securities Act Liabilities

Insofar as indemnification for liabilities arising under the Securities Act may be permitted to our directors, officers and controlling persons pursuant to the foregoing provisions, or otherwise, we have been informed that in the opinion of the SEC such indemnification is against public policy as expressed in the Securities Act and is, therefore, unenforceable.

Up to \$50,000,000



Orphazyme A/S

Ordinary Shares

(including Ordinary Shares represented by American Depositary Shares)

PROSPECTUS SUPPLEMENT

Cowen

November 4, 2021
