

**UNITED STATES
SECURITIES AND EXCHANGE COMMISSION
Washington, D.C. 20549**

FORM 20-F

(Mark One)

REGISTRATION STATEMENT PURSUANT TO SECTION 12(b) OR (g) OF THE SECURITIES EXCHANGE ACT OF 1934

OR

ANNUAL REPORT PURSUANT TO SECTION 13 OR 15(d) OF THE SECURITIES EXCHANGE ACT OF 1934

For the fiscal year ended December 31, 2021

OR

TRANSITION REPORT PURSUANT TO SECTION 13 OR 15(d) OF THE SECURITIES EXCHANGE ACT OF 1934

For the transition period from _____ to _____

OR

SHELL COMPANY REPORT PURSUANT TO SECTION 13 OR 15(d) OF THE SECURITIES EXCHANGE ACT OF 1934

Date of event requiring this shell company report:

Commission file number: 001-39545

Orphazyme A/S

(Exact name of Registrant as specified in its charter)

Not applicable

(Translation of Registrant's name into English)

The Kingdom of Denmark

(Jurisdiction of incorporation or organization)

Ole Maaløes Vej 3, DK-2200

Copenhagen N

Denmark

(Address of principal executive offices)

Anders Fink Vadsholt

Chief Executive Officer and Chief Financial Officer

Orphazyme A/S

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Tel: (+45) 28 98 90 55

(Name, Telephone, E-mail and/or Facsimile number and Address of Company Contact Person)

Securities registered or to be registered, pursuant to Section 12(b) of the Act. **None**

Securities registered or to be registered pursuant to Section 12(g) of the Act. **None**

Securities for which there is a reporting obligation pursuant to Section 15(d) of the Act. **Ordinary shares, nominal value DKK 1 per share**

Indicate the number of outstanding shares of each of the issuer's classes of capital stock or common stock as of the close of business covered by the annual report.

34,952,241 Ordinary Shares (including shares underlying American Depositary Shares)

19,886,184 American Depositary Shares

Indicate by check mark if the registrant is a well-known seasoned issuer, as defined in Rule 405 of the Securities Act. Yes No

If this report is an annual or transition report, indicate by check mark if the registrant is not required to file reports pursuant to Section 13 or 15(d) of the Securities Exchange Act of 1934. Yes No

Note – Checking the box above will not relieve any registrant required to file reports pursuant to Section 13 or 15(d) of the Securities Exchange Act of 1934 from their obligations under those Sections.

Indicate by check mark whether the registrant (1) has filed all reports required to be filed by Section 13 or 15(d) of the Securities Exchange Act of 1934 during the preceding 12 months (or for such shorter period that the registrant was required to file such reports), and (2) has been subject to such filing requirements for the past 90 days. Yes No

Indicate by check mark whether the registrant has submitted electronically every Interactive Data File required to be submitted pursuant to Rule 405 of Regulation S-T (§232.405 of this chapter) during the preceding 12 months (or for such shorter period that the registrant was required to submit such files). Yes No

Indicate by check mark whether the registrant is a large accelerated filer, an accelerated filer, a non-accelerated filer, or an emerging growth company. See the definitions of "large accelerated filer," "accelerated filer," and "emerging growth company" in Rule 12b-2 of the Exchange Act.

Large accelerated filer Accelerated filer Non-accelerated filer
Emerging growth company

If an emerging growth company that prepares its financial statements in accordance with U.S. GAAP, indicate by check mark if the registrant has elected not to use the extended transition period for complying with any new or revised financial accounting standards † provided pursuant to Section 13(a) of the Exchange Act.

† The term "new or revised financial accounting standard" refers to any update issued by the Financial Accounting Standards Board to its Accounting Standards Codification after April 5, 2012.

Indicate by check mark whether the registrant has filed a report on and attestation to its management's assessment of the effectiveness of its internal control over financial reporting under Section 404(b) of the Sarbanes-Oxley Act (15 U.S.C. 7262(b)) by the registered public accounting firm that prepared or issued its audit report.

Indicate by check mark which basis of accounting the registrant has used to prepare the financial statements included in this filing:

U.S. GAAP International Financial Reporting Standards as issued by the International Accounting Standards Board Other

If "Other" has been checked in response to the previous question indicate by check mark which financial statement item the registrant has elected to follow. Item 17 Item 18

If this is an annual report, indicate by check mark whether the registrant is a shell company (as defined in Rule 12b-2 of the Exchange Act). Yes No

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INTRODUCTION

In this Annual Report on Form 20-F, the terms “Orphazyme,” “the Company,” “we,” “us” and “our” refer to the parent company Orphazyme A/S together with its wholly owned subsidiaries. The term “Orphazyme A/S” is used when addressing issues specifically related to the parent entity. 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.

Pursuant to Rule 12b-23(a) of the Securities Exchange Act of 1934, as amended, certain information for the 2021 Form 20-F of Orphazyme A/S set out herein is being incorporated by reference from the Company’s statutory Annual Report 2021 (hereafter “Annual Report 2021”). Therefore, the information in this Form 20-F should be read in conjunction with the Annual Report 2021 (see Exhibit 15.1), which was furnished to the SEC on Form 6-K on June 9, 2022.

PRESENTATION OF FINANCIAL AND OTHER 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 in this Annual Report on Form 20-F were prepared in accordance with accounting principles generally accepted in the United States, or U.S. GAAP.

All references in this Annual Report on Form 20-F to “\$” are to U.S. dollars (USD), to “DKK” are to Danish kroner and to “€” are to the Euro. Except with respect to U.S. dollar amounts presented as contractual terms, amounts denominated in U.S. dollars when received or paid and unless otherwise indicated, certain Danish kroner amounts contained in this Annual Report on Form 20-F have been translated into U.S. dollars at the rate of \$1.00 to DKK 6.5704, which was the noon buying rate of the Federal Reserve Bank of New York on December 31, 2021. Except with respect to Euro amounts presented as contractual terms, amounts denominated in Euros when received or paid and unless otherwise indicated, Danish kroner amounts have been translated into Euros at the rate of €1.00 to DKK 7.4364, which was the closing rate of the European Central Bank on December 31, 2021. These translations should not be considered representations that any such amounts have been, could have been or could be converted into U.S. dollars or Euros at that or any other exchange rate as of that or any other rate. We have made rounding adjustments to some of the figures included in this Annual Report on Form 20-F. Accordingly, numerical figures shown as totals in some tables may not be an arithmetic aggregation of the figures that precede them.

All references to “shares” in this Annual Report on Form 20-F refer to ordinary shares of Orphazyme A/S with a nominal value of DKK 1 per share.

This Annual Report on Form 20-F 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 Annual Report on Form 20-F 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.

This Annual Report on Form 20-F 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 Annual Report on Form 20-F involve a number of assumptions and limitations, and the reader is 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 “Item 3.D-Risk Factors” and in the next section entitled “Cautionary Statement Regarding Forward-Looking Statements.” These and other factors could cause results to differ materially from those expressed in the estimates made by the independent parties and by us.

Enforceability of Civil Liabilities

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

As of the date of this Annual Report on Form 20-F, two-thirds of the members of the board of directors reside in other jurisdictions outside of Denmark and the United States and the one member of our executive management named herein is a resident of Denmark. 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.

CAUTIONARY STATEMENT REGARDING FORWARD-LOOKING STATEMENTS

This Annual Report on Form 20-F contains forward-looking statements that reflect our current expectations and views of future events. Known and unknown risks, uncertainties and other factors, including those listed under “Risk Factors,” 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:

- our estimates regarding future financial results and condition;
- our ability to retain qualified employees, consultants and key personnel;
- the outcome of the class action lawsuit filed against Orphazyme A/S;
- the impact of the COVID-19 pandemic on our business and operations;
- our exposure to scrutiny as a U.S. public company; and
- 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.

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. Important risks and factors that could cause our actual results to be materially different from our expectations are generally set forth in “Item 3.D-Risk Factors”, “Item 4.B-Business Overview”, “Item 5. Operating and Financial Review and Prospects”, and other sections in this Annual Report on Form 20-F.

The forward-looking statements made in this Annual Report on Form 20-F relate only to events or information as of the date on which the statements are made 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 Annual Report on Form 20-F and the documents that we refer to in this Annual Report on Form 20-F and have filed with the SEC completely and with the understanding that our actual future results may be materially different from what we expect.

IMPORTANT NOTICE REGARDING DELISTING AND TERMINATION OF AMERICAN DEPOSITARY SHARES

In March 2022, we voluntary delisted ADSs representing our ordinary shares from the Nasdaq Global Select Market in the United States, which became effective March 31, 2022.

The Bank of New York Mellon, serves as depositary, or the Depositary, for our American Depositary Share, or ADS, facility. Each ADS represents one of our ordinary shares. The Depositary has announced that it will terminate the Deposit Agreement, dated September 28, 2020 among us, the Depositary, and owners and holders of ADSs effective at 5:00 PM (Eastern Time) on July 6, 2022. Under the terms of the Deposit Agreement, ADS holders have until at least July 11, 2022 to surrender ADSs for delivery of the underlying ordinary shares. ADS holders who surrender ADSs for delivery of the underlying shares must pay a cable fee of \$17.50, a cancellation fee of up to \$0.05 per ADS surrendered and any applicable U.S. or local taxes or governmental charges. Subsequent to July 11, 2022, the Depositary may attempt to sell the underlying shares. If the Depositary has sold such shares, holders of ADSs must surrender such securities in order to obtain payment of the sale proceeds of the underlying ordinary shares, net of the expenses of sale, any applicable U.S. or local taxes or government charges and a cancellation fee of up to \$0.05 per ADS. To surrender ADSs, the address of the Depositary is: The Bank of New York Mellon, 240 Greenwich Street, Depositary Receipts Division – 8th Floor, Attention: Cancellation Desk, New York, NY 10286. Registered or overnight mail is the suggested method of delivering ADSs to the Depositary. For Settlement specific inquiries, please contact DRsettlements@bnymellon.com.

PART I

ITEM 1. IDENTITY OF DIRECTORS, SENIOR MANAGEMENT AND ADVISERS

Not applicable

ITEM 2. OFFER STATISTICS AND EXPECTED TIMETABLE

Not applicable

ITEM 3. KEY INFORMATION

A. [Reserved]

B. Capitalization and indebtedness

Not applicable

C. Reasons for the offer and use of proceeds

Not applicable.

D. Risk factors.

Investing in our ordinary shares and ADSs involves a high degree of risk. You should carefully consider the risks described below, and all other information contained in or incorporated by reference in this Annual Report on Form 20-F, including our Audited Consolidated Financial Statements before making an investment decision regarding our securities. The occurrence of any of the events or developments described below could harm our business and financial condition. If any of the following risks actually occurs, our business, cash flows, financial condition and/or prospects could suffer materially. In such event, the price of the ordinary shares and ADSs could decline, which would cause you to lose all or part of your investment.

Risk Factors Summary

Recent Developments

In March 2022, we voluntarily delisted ADSs representing our ordinary shares from the Nasdaq Global Select Market in the United States, which became effective March 31, 2022. On May 31, 2022, we completed the sale of substantially all of the Company's assets and business activities to KemPharm Denmark A/S (KemPharm), a wholly owned subsidiary of KemPharm Inc., a specialty pharmaceutical company focused on the discovery and development of novel treatments for rare central nervous system diseases, and KemPharm retained all of Orphazyme's remaining Danish employees. As of the date of publication of this Annual Report on Form 20-F, we have limited ongoing operational business activities and only two employees. The Bank of New York Mellon, which serves as depository for our ADS facility has announced that it will terminate the Deposit Agreement governing the ADSs effective at 5:00 PM (Eastern Time) on July 6, 2022.

Our business is subject to a number of risks and uncertainties, including those risks discussed at-length below this summary. These risks include, among others, the following:

Risks Related to our Financial Position and Limited Business Activities

- We have limited operating business activities and only two employees. As such our future business prospects are uncertain.
- We may become subject to claims in connection with past asset disposals.
- We may choose to wind down our business, which may result in additional costs.

Risks Related to Employees, Key Personnel and Third-Party vendors

- We rely on our CEO/CFO to operate our business.

Risks Related to Our Securities

- The company has voluntarily requested to delisted its ADSs from Nasdaq Global Select Market in the U.S., and the ADSs are not listed for trading on any other national securities exchange in the United States, which could cause the ADSs to lose some or all of their value.
- The ADS Facility will be terminated on July 6, 2022, which will present costs and risks to ADS holders.
- We intend to deregister the ordinary shares under the Exchange Act in March 2023, following which we would no longer be required to file periodic reports with the SEC thereunder.
- The trading price of our equity securities may be volatile due to factors beyond our control.
- Investors should be aware that the rights provided to our shareholders and holders of ADSs under Danish corporate law and our articles of association differ in certain respects from the rights that you would typically enjoy as a shareholder of a U.S. company under applicable U.S. federal and state laws.
- As a result of material weaknesses, management concluded that our internal control over financial reporting was not effective at the reasonable assurance level as of December 31, 2021. Therefore, we may not be able to accurately report our financial results in a timely manner or prevent fraud, which may adversely affect our business, investor confidence in our company and the market price of our shares and ADSs.

General Risk Factors

- We are subject to securities litigation, which is expensive and could divert management’s attention.

Risks Related to our Financial Position and Limited Business Activities

On May 31, 2022 we completed the sale of substantially all of the Company’s assets and business activities to KemPharm Denmark A/S, a wholly owned subsidiary of KemPharm Inc. We now have limited ongoing operational business activities and only two employees.

Until recently, Orphazyme was a biopharmaceutical company involved in the research and development of novel therapeutics for the treatment of neurodegenerative rare diseases. In May 2022, we announced that we had signed an agreement to sell substantially all of the Company’s assets and business activities, including those relating to the development and approval of arimoclomol and the full claw back liability related to the French early access program, to KemPharm Denmark A/S (“KemPharm”), a wholly owned subsidiary of KemPharm Inc. for a total of USD 12.8 million in cash and assumed liabilities estimated to equal approximately USD 5.2 million (the “Sale of Assets”). KemPharm is a specialty pharmaceutical company focused on the discovery and development of novel treatments for rare central nervous system diseases. As we have limited ongoing operational business activities and only two employees our future business prospects are uncertain.

We may become subject to claims in connection with past asset disposals.

In May 2022, we sold substantially all of our assets and business activities to KemPharm on an “as-is” basis and we have not given any substantial representations or warranties in favor of KemPharm. While we are not aware of any outstanding matters that would reasonably form a basis for a claim related to the Sale of Assets, if we become subject to liability based upon our contractual obligations to KemPharm or otherwise, it could have a material adverse effect on our financial position.

We may choose to wind down our business, which may result in additional costs.

We may liquidate and distribute remaining cash to shareholders, after satisfaction of any obligations. We would incur third party costs associated with any distribution which would further limit funds to shareholders. There would be significant costs associated with winding down, such as separation of employees, and termination of contracts, and there could be taxes payable, all of which will further reduce the cash resources available.

Our business could be adversely affected by the effects of health epidemics, including those related to the ongoing COVID-19 pandemic.

While a limited risk given that substantially all of our assets and business activities have been sold and we have limited ongoing operational business activities, our business could be adversely affected by health epidemics. In particular, ongoing constraints associated with the COVID-19 pandemic have had, and are expected to continue to have, a significant impact on businesses and commerce worldwide, including as a result of future worker shortages; supply chain disruptions; suspensions among facilities and production across a variety of industries; and fluctuating demand for certain goods and services. While vaccines have been approved and are being deployed, the timing and likelihood of achieving widespread global vaccination remains uncertain, and the vaccines may be less effective against new variants, potentially leading to the reimpositions of restrictions in an effort to mitigate risks to public health, especially as more infectious variants of the virus emerge for a prolonged period of time, further delaying the return of the global economy to pre-pandemic levels.

At this time, there is no material impact directly related to the COVID-19 pandemic on our consolidated financial statements, including the judgements and estimates applied. The ultimate impact of the COVID-19 pandemic or impacts from any other health epidemic are highly uncertain. The current pandemic and its resulting impacts on our business or the global economy as a whole remain subject to change and could have a material impact on our limited operations. To the extent the pandemic adversely affects our business, cash flows, financial condition and/or prospects, it may also have the effect of heightening many of the other risks described in this “Risk Factors” section.

Risks Related to Our Employees, Key Personnel and our Dependence on Third Parties

We rely on our CEO/CFO to operate our business.

We rely on the efforts of our Chief Executive Officer and Chief Financial Officer, Anders Vadsholt, to operate our business. The loss of Mr. Vadsholt could require us to incur additional costs to recruit replacements, which could have a material adverse effect on our business. While we have entered into an employment agreement with Mr. Vadsholt, we can make no assurances that he will continue to be employed by us.

There are measures in our employment contracts designed to give us time and flexibility to seek alternative solutions in the event of an executive departure. Mr. Vadsholt has an employment agreement which includes a six month notice period if Mr. Vadsholt wishes to end his employment with us, and we can terminate his employment by giving 12 months’ notice.

We engage a number of third-party suppliers and service providers to supply critical services, such as IT services. Disruptions to the business, financial stability or operations of these suppliers and service providers, including due to strikes, labor disputes or other disruptions to the workforce, for instance, if, as a result of COVID-19, employees are not able to come to work, or to their willingness and ability to produce or deliver such products or provide such services in a manner that satisfies the requirements put forth by the authorities, or in a manner that satisfies our own requirements, this could adversely affect our business, cash flows, financial condition and/or prospects. If these suppliers and service providers were unable or unwilling to continue to provide their products or services in the manner expected, or at all, we could encounter difficulty finding alternative suppliers. Even if we are able to secure appropriate alternative suppliers in a timely manner, costs for such products or services could increase significantly. Any of these events could adversely affect our business, cash flows, financial condition and/or prospects.

Risks Related to Legal and Regulatory Compliance Matters

We are subject to risks related to data privacy concerns, cyber security breaches and failure to comply with laws, regulations, standards, and contracts relating to data privacy and security.

We are subject to evolving data protection laws, privacy and security requirements and other regulatory restrictions in the various jurisdictions in which we operate. These laws are subject to differing interpretations and may be inconsistent among jurisdictions, and guidance on implementation and compliance practices are often updated or otherwise revised, which adds to the complexity of processing personal information. During the course of our business, we come in to the possession of sensitive personal data and may store or process such information outside the country in which it was collected. This information needs to be handled by us in compliance with such obligations. These and other obligations could require us or our partners to incur additional costs to achieve compliance, limit our competitiveness, necessitate the acceptance of more onerous obligations in our contracts, restrict our ability to use, store, transfer, and process data, impact our or our

partners' ability to process or use data in order to support the provision of our products or services, affect our or our partners' ability to offer our products and services or operate in certain locations, result in increased expenses, reduce overall demand for our products and services and make it more difficult to meet expectations of or commitments to customers or collaborators.

Furthermore, our failure to keep apprised of, and comply with, privacy, data use and security laws, standards and regulations, including, for instance, unauthorized disclosure of, or access to, data, could result in the suspension or revocation of our approvals or registrations, the limitation, suspension or termination of services or the imposition of administrative, civil or criminal penalties, including fines. For example, under the EU General Data Protection Regulation that entered into force in May 25, 2018, fines may be as high as 20 million Euros or 4% of the annual worldwide revenue, whichever is higher, for certain infringements. Laws such as the GDPR and EU member state laws may also apply to health-related and other personal information that we process. These laws impose strict obligations on the ability to process health-related and other personal information of data subjects in the European Union and the United Kingdom, including, among other things, standards relating to the privacy and security of personal data, which require the adoption of administrative, physical and technical safeguards designed to protect such information. These laws may affect our use, collection, analysis, and transfer (including cross-border transfer) of such personal information. These laws include several requirements relating to transparency requirements related to communications with data subjects regarding the processing of their personal data, obtaining the consent of the individuals to whom the personal data relates, limitations on data processing, establishing a legal basis for processing, notification of data processing obligations or security incidents to appropriate data protection authorities or data subjects, the security and confidentiality of the personal data and various rights that data subjects may exercise. The GDPR prohibits the transfer, without an appropriate legal basis, of personal data to countries outside of the European Economic Area, or EEA, such as the United States, which are not considered by the European Commission to provide an adequate level of data protection. Switzerland and the United Kingdom has adopted similar restrictions. Although there are legal mechanisms to allow for the transfer of personal data from the EEA and Switzerland to the United States, uncertainty about compliance with EU data protection laws remains and such mechanisms may not be available or applicable with respect to the personal data processing activities necessary to research, develop, and market our products and services. For example, ongoing legal challenges in Europe to the mechanisms allowing companies to transfer personal data from the EEA to the United States could result in further limitations on the ability to transfer personal data across borders, particularly if governments are unable or unwilling to reach new or maintain existing agreements that support cross-border data transfers, such as the invalidated EU-U.S. and Swiss-U.S. Privacy Shield Frameworks. Additionally, other countries have passed or are considering passing laws requiring local data residency and/or restricting the international transfer of data. Further, the UK's decision to leave the EU, often referred to as Brexit, has created uncertainty with regard to data protection regulation in the United Kingdom. The Data Protection Act 2018, that "implements" and complements the GDPR achieved Royal Assent on May 23, 2018 and is now effective in the United Kingdom. The EU Commission issued an adequacy decision approving the United Kingdom as an adequate country in accordance with GDPR art. 45, provided significant changes are not made to UK law on data protection. We may, however, incur liabilities, expenses, costs, and other operational losses under GDPR and applicable EU Member States and the United Kingdom privacy laws in connection with any measures we take to comply with them.

Additionally, California has enacted legislation that has been dubbed the first "GDPR-like" law in the United States. Known as the California Consumer Privacy Act, or CCPA, it creates new individual privacy rights for consumers (as that word is broadly defined in the law) and places increased privacy and security obligations on entities handling personal data of consumers or households. The CCPA went into effect on January 1, 2020 and requires covered companies to provide new disclosures to California consumers, provides consumers with new data privacy rights, imposes new operational requirements for covered businesses, creates a statutory damages framework, and allows for a new cause of action for data breaches. Although there are limited exemptions for clinical trial data, the CCPA could impact our business activities depending on how it is interpreted and exemplifies the vulnerability of our business to not only cyber threats but also the evolving regulatory environment related to personal data and protected health information.

Any failure by our vendors to comply with applicable law, regulations or contractual obligations related to data privacy and security could result in proceedings against us by governmental entities or others.

We publish privacy policies, self-certifications, and other documentation regarding our collection, processing, use and disclosure of personal information and/or other confidential information. Although we endeavor to comply with our published policies, certifications, and documentation, we may at times fail to do so

or may be perceived to have failed to do so. Moreover, despite our efforts, we may not be successful in achieving compliance if our employees or vendors to comply with our published policies, certifications, and documentation. Such failures can subject us to potential international, local, state and federal action if they are found to be deceptive, unfair, or misrepresentative of our actual practices. Moreover, patients or subjects about whom we or our partners obtain information, as well as the providers who share this information with us, may contractually limit our ability to use and disclose the information. Claims that we have violated individuals' privacy rights or failed to comply with data protection laws or applicable privacy notices even if we are not found liable, could be expensive and time-consuming to defend and could result in adverse publicity that could harm our business.

Because of the breadth of these laws, it is possible that some of our current or future business activities could be subject to challenge under one or more of such privacy and data security laws. The heightening compliance environment and the need to build and maintain robust and secure systems to comply with different privacy compliance and/or reporting requirements in multiple jurisdictions could increase the possibility that a healthcare company may fail to comply fully with one or more of these requirements. If our operations are alleged to be or are found to be in violation of any of the privacy or data security laws or regulations described above that are applicable to us, or any other laws that apply to us, we may be subject to government investigations and enforcement actions, private litigation, penalties, including potentially significant criminal, civil, and administrative penalties, damages, fines, imprisonment, contractual damages, reputational harm, diminished profits and future earnings, additional reporting requirements, and/or oversight if we become subject to a consent decree or similar agreement to resolve allegations of non-compliance with these laws, and the curtailment or restructuring of our operations, any of which could adversely affect our ability to operate our business and our results of operations. Accordingly, our failure to keep apprised of, and comply with, privacy, data use and security laws, standards and regulations could have a material adverse effect on our reputation and negatively affect our business, results of operations, cash flows, financial condition and/or prospects.

Cyber security attacks on our servers, information systems and databases, or the third-party servers, information systems and databases on which our information is stored or processed, could compromise the security, availability, or integrity of our data or could cause interruptions in the operations of our business. We cannot guarantee that our security measures will be sufficient to protect against unauthorized access to or other compromise of the personal or confidential information we process. The techniques used to sabotage or to obtain unauthorized access to our systems, networks and/or physical facilities in which data is stored or through which data is transmitted change frequently, and we may be unable to implement adequate preventative measures or stop security breaches. The recovery systems, security protocols, network protection mechanisms and other security measures that we have integrated into our platform, systems, networks and physical facilities, which are designed to protect against, detect and minimize security breaches, may not be adequate to prevent or detect service interruption, system failure or data loss. Notwithstanding safeguards, cyber security breaches, internal security breaches, physical security breaches or other unauthorized or accidental access to our servers, other information systems or databases could result in tampering with, or the theft or publication of, sensitive information or the deletion or modification of data, or could otherwise cause interruptions in our operations, which could have a material adverse effect on our business, results of operations, cash flows, financial condition and/or prospects.

The tampering with, disruption to, or the theft or publication of, sensitive information or the deletion or modification of records held either in our systems or the systems of others to which we have access, could subject us to increased costs and exposure to litigation. We have contractual and legal obligations to notify relevant stakeholders of security breaches. Most jurisdictions have enacted laws requiring companies to notify individuals, regulatory authorities, and others of security breaches involving certain types of data. In addition, our agreements with certain partners may require us to notify them in the event of a security breach. The loss of confidential information could result in the payment of damages and reputational harm and could have a material adverse effect on our business, results of operations, cash flows, financial condition and/or prospects.

Despite the implementation of security measures, our internal computer systems and those of any third-party contractors and consultants that we may employ are vulnerable to damage from computer viruses, natural disasters, terrorism, war and telecommunication and electrical failures. If such an event were to occur and cause interruptions in our operations, it could result in a material disruption of our product candidate development programs. For example, the loss of pre-clinical studies or clinical trial data from completed, ongoing or planned trials could result in delays in our regulatory approval efforts and significantly increase our costs to recover or reproduce the data. To the extent that any such disruption results in a loss of or damage to our data or applications or other data or applications relating to our technology or product candidate, or inappropriate

disclosure or theft of confidential or proprietary information, we could incur liabilities, our competitive position could be harmed and the further development of our product candidates could be delayed.

The financial exposure from the items referenced above could either not be insured against or not fully covered through any insurance that we maintain and could have a material adverse effect on our business, results of operations, cash flows, financial condition and/or prospects. In addition, we cannot be sure that our existing insurance coverage and coverage for errors and omissions will continue to be available on acceptable terms or that our insurers will not deny coverage as to any future claim. There can be no assurance that the limitations of liability in our contracts would be enforceable or adequate or would otherwise protect us from liabilities or damages.

Our insurance policies protect us only from some business risks, which leaves us exposed to significant uninsured liabilities.

We do not carry insurance for all categories of risk that our business may encounter. Although we have product liability and clinical study liability insurance that we believe is appropriate, this insurance is subject to deductibles and coverage limitations. Our current product liability insurance may not continue to be available to us on acceptable terms, if at all, and, if available, coverage may not be adequate to protect us against any future product liability claims. If we are unable to obtain insurance at an acceptable cost or on acceptable terms or otherwise protect against potential product liability claims, we could be exposed to significant liabilities. A product liability claim or other claim with respect to uninsured liabilities or for amounts in excess of insured liabilities could have a material adverse effect on our business, results of operations, cash flows, financial condition and/or prospects. As a result of the Sale of Assets to KemPharm, certain of our clinical study and product related liability insurances will be transferred to KemPharm. Further, we have terminated our cyber security insurance due to our limited operating activities and employees.

Risks Related to Our Securities and our Foreign Private Issuer Status

The company has voluntarily delisted its ADSs from Nasdaq Global Select Market in the U.S., and the ADSs are not listed for trading on any other national securities exchange in the United States, which could cause the ADSs to lose some or all of their value.

The company has voluntarily delisted its ADSs from Nasdaq and the ADSs are not listed for trading on any other national securities exchange in the United States, which could cause the ADSs to lose some or all of their value. In the absence of an active trading market for the ADSs, investors may not be able to sell their ADSs at the time that they would like to sell such securities or to sell them at all.

The ADS Facility will be terminated on July 6, 2022, which will present costs and risks to ADS holders.

The Bank of New York Mellon, which serves as Depositary for our ADS program, has announced that it will terminate the Deposit Agreement, dated September 28, 2020 among us, the Depositary, and owners and holders of ADSs effective at 5:00 PM (Eastern Time) on July 6, 2022. Under the terms of the Deposit Agreement, ADS holders have until at least July 11, 2022 to surrender ADSs for delivery of the underlying ordinary shares. ADS holders who surrender ADSs for delivery of the underlying shares must pay a cable fee of \$17.50, a cancellation fee of up to \$0.05 per ADS surrendered and any applicable U.S. or local taxes or governmental charges. Subsequent to July 11, 2022, the Depositary may attempt to sell the underlying shares. If the Depositary has sold such shares, holders of ADSs must surrender such securities in order to obtain payment of the sale proceeds of the underlying ordinary shares, net of the expenses of sale, any applicable U.S. or local taxes or government charges and a cancellation fee of up to \$0.05 per ADS. The termination of the ADS program presents costs and risks to ADS holders.

We intend to deregister the ordinary shares under the Exchange Act in March 2023, upon which our SEC reporting obligations will be suspended.

Although we have voluntarily delisted the ADSs representing our ordinary shares from the Nasdaq Global Select Market in the United States., which became effective March 31, 2022, we are required to continue our SEC reporting obligations until at least March 31, 2023, when we anticipate filing a Form 15F with the SEC to suspend our reporting obligations under the Exchange Act. At such time, we would cease to file periodic or current reports with the Commission, which would limit the information available to investors in the United States in order to evaluate an investment in our company.

The trading price of our equity securities may be volatile due to factors beyond our control.

The market prices of the ordinary shares or ADSs and shares may be volatile, particularly as the ADSs have been delisted from the Nasdaq Global Select Market and have not been listed on any other U.S. national securities exchange. The ADSs may trade over-the-counter in the United States, if they trade at all, although no assurance can be provided that an active market will develop therefor prior to the time that the ADS program will be terminated on July 6, 2022.

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;
- 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;
- commentary by investors and/or media on the prospects for our business, the ordinary shares or ADSs on the internet and/or social media and resulting in trading of our ordinary shares or ADSs;
- unusual trading in our ordinary shares or ADSs or securities derivative thereof, including pursuant to naked, or uncovered, short positions;
- fluctuations in the valuation of companies perceived by investors to be comparable to us;
- currency fluctuations;
- additions or departures of key management, including our CEO/CFO, or members of the Board of Directors;
- disputes or other developments related to proprietary rights, including patents, litigation matters, asset disposals, prior clinical development;
- announcement or expectation of additional debt or equity financing efforts;
- continuing uncertainty related to 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, which may limit or prevent investors from readily selling their shares or ordinary shares or ADSs and may otherwise negatively affect the liquidity of the trading market for ordinary shares or ADSs.

We intend to retain all available funds and any future earnings and, consequently, your ability to achieve a return on your investment will depend on appreciation in the price of the ordinary shares or ADSs.

We have never declared or paid any cash dividends on our shares, and we intend to retain all available funds and any future earnings to fund the development of our business. Therefore, you are not likely to receive any dividends on your ordinary shares or ADSs for the foreseeable future and the success of an investment in ordinary shares or ADSs will depend upon any future appreciation in their value. Consequently, investors may need to sell all or part of their holdings of ordinary shares or ADSs after price appreciation, which may never occur, as the only way to realize any future gains on their investment. There is no guarantee that the ordinary shares or ADSs will appreciate in value or even maintain the price at which our investors have purchased them. Investors seeking cash dividends should not purchase the ordinary shares or ADSs.

In addition, if we choose to pay dividends in the future, exchange rate fluctuations may affect the amount of Danish kroner that we are able to distribute, and the amount in U.S. dollars that our shareholders receive upon the payment of cash dividends or other distributions we declare and pay in Danish kroner, if any. Any dividends will generally be subject to Danish withholding tax. See "Item 10.E-Taxation - Material Danish

Income Tax Consequences” for a more detailed description of Danish taxes on dividends. These factors could harm the value of the ordinary shares or ADSs.

Investors should be aware that the rights provided to our shareholders and holders of ADSs under Danish corporate law and our articles of association differ in certain respects from the rights that you would typically enjoy as a shareholder of a U.S. company under applicable U.S. federal and state laws.

We are a Danish company with limited liability. Our corporate affairs are governed by our Articles of Association and by the laws governing companies incorporated in Denmark. The rights of shareholders and the responsibilities of members of our board of directors may be different from the rights and obligations of shareholders and boards of directors in companies governed by the laws of U.S. jurisdictions. In the performance of its duties, our board is required by Danish law to consider the interests of our company, its shareholders, its employees and other stakeholders. It is possible that some of these parties will have interests that are different from, or in addition to, the interests of our shareholders. See “Description of Share Capital and Articles of Association—Articles of Association and Danish Corporate Law.”

Under Danish corporate law, except in certain limited circumstances, which require at a minimum that a proposal for inspection has been supported by shareholders representing a minimum of 25% of the voting rights and the share capital being present at a general meeting, our shareholders may not ask for an inspection of our corporate records, while under Delaware corporate law any shareholder, irrespective of the size of such shareholder’s shareholdings, may do so. Shareholders of a Danish limited liability company are also unable to initiate a derivative action, a remedy typically available to shareholders of U.S. companies, in order to enforce a right of our company, in case we fail to enforce such right ourselves, other than in certain cases of board member/management liability under limited circumstances. In addition, a majority of our shareholders may release a member of our board of directors or our executive management from any claim of liability we may have, including if such board member or member of our executive management has acted in bad faith, negligently or fraudulently. However, a shareholder may bring a derivative action on behalf of our company against, among other persons, a member of our board of directors or our executive management, provided that the circumstances of the act or omission giving rise to the claim of liability was not known to the shareholder at the time of such shareholder resolution, or if shareholders representing at least 10% of the share capital represented at the relevant general meeting have opposed such shareholder resolution. In contrast, most U.S. federal and state laws prohibit a company or its shareholders from releasing a board member from liability altogether if such board member has acted in bad faith or has breached such board member’s duty of loyalty. Additionally, distribution of dividends from Danish companies to foreign companies and individuals can be subject to non-refundable withholding tax, and not all receiving countries allow for deduction. See “Material Danish Income Tax Consequences” for a more detailed description of the withholding tax. Also, the rights of a creditor of the company may not be as strong under Danish insolvency law as under U.S. or other insolvency law, and consequently creditors may recover less in the event our company is subject to insolvency compared to a similar case involving a U.S. debtor. In addition, the use of the tax asset consisting of the accumulated tax losses requires that we are able to generate positive taxable income, and the use of tax losses carried forward to offset against future income is subject to certain restrictions and can be restricted further by future amendments to Danish tax law. Finally, Danish corporate law may not provide appraisal rights in the case of a business combination equivalent to those generally afforded a shareholder of a U.S. company under applicable U.S. laws. For additional information on these and other aspects of Danish corporate law and our articles of association, see Exhibit 2.1 “Description of Share Capital and Articles of Association.” As a result of these differences between Danish corporate law and our articles of association, on the one hand, and U.S. federal and state laws, on the other hand, in certain instances, you could receive less protection as an equity holder of our company than you would as a shareholder of a U.S. company.

You may not be able to exercise your right to vote the ordinary shares underlying your ADSs even for so long as the ADS program remains in existence.

The ADSs program will be terminated on July 6, 2022. Prior to such date, a holder of our ADSs is not treated as one of our shareholders and does not have shareholder rights. The depositary is the holder of the ordinary shares underlying ADSs. A holder of ADSs has contractual ADS holder rights. The deposit agreement among us, the depositary and you, as an ADS holder, and all other persons directly and indirectly holding ADSs sets out ADS holder rights as well as the rights and obligations of the depositary.

ADS holders may only exercise voting rights with respect to the shares underlying their respective ADSs in accordance with the provisions of the deposit agreement, which provides that a holder may vote the ordinary shares underlying any ADSs for any particular matter to be voted on by our shareholders either by

withdrawing the ordinary shares underlying the ADSs or by instructing the depositary how to vote those ordinary shares. However, even if you are able to instruct the depositary to vote the ordinary shares underlying your ADSs, we cannot guarantee you that the depositary will vote in accordance with your instructions and you may not know about the meeting far enough in advance to withdraw those ordinary shares.

Our articles of association permit differentiated voting, allowing the depositary to vote the ordinary 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 ordinary 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, as far as practical, to vote the ordinary shares underlying the ADSs as instructed by the ADS holders. In such an instance, if we ask for your instructions, the depositary, upon timely notice from us, will notify you of the upcoming vote and arrange to deliver our voting materials to you. We cannot guarantee that you will receive the voting materials in time to ensure that you can instruct the depositary to vote your ordinary shares or to withdraw your ordinary shares so that you can vote them yourself. Voting instructions may be given only in respect of a number of ADSs representing an integral number of ordinary shares or other deposited securities. 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 any right to vote that you may have with respect to the underlying ordinary shares, and there may be nothing you can do if the ordinary shares underlying your ADSs are not voted as you requested. In addition, the depositary is only required to notify you of any particular vote if it receives notice from us in advance of the scheduled meeting.

ADS investors may also not realize all of the benefits of being a shareholder in our company. For instance, the votes of ADS holders will not be represented directly on our books, but only through a vote by the depositary of the underlying ordinary shares, which vote will reflect the ADS majority's election on the vote of all such ordinary shares. Separately, we may elect to offer subscription rights to our shareholders without offering such rights to ADS holders as such subscription rights will be offered to the depositary as shareholder. The depositary has substantial discretion as to what will happen with any offered subscription rights and may determine that it is not legal or reasonably practicable to make such rights available to ADS holders, in which case the depositary will endeavor to sell such rights and distribute the proceeds to ADS holders, which it may not be able to do at the then-current market price or at all. If the depositary is unable to distribute or sell such rights, they will lapse, and ADS holders will receive no value. See Exhibit 2.4 under "Dividends and Other Distributions."

Holders of ADSs may be subject to limitations on the transfer of ADSs and the withdrawal of the underlying ordinary shares for so long as the ADS program remains in existence and upon its termination.

ADSs are transferable on the books of the depositary. However, the depositary may close its books at any time or from time to time when it deems expedient in connection with the performance of its duties. The depositary may refuse to deliver, transfer or register transfers of ADSs generally when our books or the books of the depositary are closed, or at any time if we or the depositary think it is advisable to do so because of any requirement of law, government or governmental body, or under any provision of the deposit agreement, or for any other reason subject to your right to cancel ADSs and withdraw the underlying shares. Temporary delays in the cancellation of ADSs and withdrawal of the underlying shares may arise because the depositary has closed its transfer books or we have closed our transfer books, the transfer of ordinary shares is blocked to permit voting at a shareholders' meeting or we are paying a dividend on our ordinary shares. In addition, you may not be able to cancel ADSs and withdraw the underlying ordinary shares when you owe money for fees, taxes and similar charges and 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 ordinary shares or other deposited securities. See Exhibit 2.4.

The ADSs program will be terminated on July 6, 2022. Under the terms of the Deposit Agreement, ADS holders have until at least July 11, 2022 to surrender ADSs for delivery of the underlying ordinary shares. ADS holders who surrender ADSs for delivery of the underlying shares must pay a cable fee of \$17.50, a cancellation fee of up to \$0.05 per ADS surrendered and any applicable U.S. or local taxes or governmental charges. Subsequent to July 11, 2022, the Depositary may attempt to sell the underlying shares. If the Depositary has sold such shares, holders of ADSs must surrender such securities in order to obtain payment of the sale proceeds of the underlying ordinary shares, net of the expenses of sale, any applicable U.S. or local taxes or government charges and a cancellation fee of up to \$0.05 per ADS. To surrender ADSs, the address of the Depositary is: The Bank of New York Mellon, 240 Greenwich Street, Depositary Receipts Division – 8th Floor, Attention: Cancellation Desk, New York, NY 10286. Registered or overnight mail is the suggested

method of delivering ADSs to the Depository. For Settlement specific inquiries, please contact DRsettlements@bnymellon.com.

You may not receive distributions on our ordinary shares represented by the ADSs or any value for them if it is illegal or impractical to make them available to holders of ADSs.

Although we do not have any present plans to declare or pay any dividends, in the event we declare and pay any dividends, the depository for the ADSs has agreed to pay to you the cash dividends or other distributions it or the custodian receives on our ordinary shares after deducting its fees and expenses. You will receive these distributions in proportion to the number of our ordinary shares your ADSs represent. However, in accordance with the limitations set forth in the deposit agreement, it may be unlawful or impractical to make a distribution available to holders of ADSs. We have no obligation to register under U.S. securities laws any offering of ADSs, ordinary shares or other securities received through such distributions. We also have no obligation to take any other action to permit distribution of ADSs, ordinary shares, rights or anything else to holders of the ADSs. This means that you may not receive the distributions we make on our ordinary shares or any value from them if it is unlawful or impractical to make them available to you. These restrictions may have an adverse effect on the value of your ADSs. See Exhibit 2.4.

ADSs holders may not be entitled to a jury trial with respect to claims arising under the deposit agreement, which could cause less favorable results to the plaintiff(s) in any such action.

The deposit agreement governing the ADSs representing our ordinary shares provides that holders and beneficial owners of ADSs irrevocably waive the right to a trial by jury in any legal proceeding arising out of or relating to the deposit agreement or the ADSs, including claims under federal securities laws, against us or the depository to the fullest extent permitted by applicable law. If this jury trial waiver provision is prohibited by applicable law, an action could nevertheless proceed under the terms of the deposit agreement with a jury trial. To our knowledge, the enforceability of a jury trial waiver under the federal securities laws has not been finally adjudicated by a federal court. However, we believe that a jury trial waiver provision is generally enforceable under the laws of the State of New York, which govern the deposit agreement, by a court of the State of New York or a federal court, which have non-exclusive jurisdiction over matters arising under the deposit agreement, applying such law. In determining whether to enforce a jury trial waiver provision, New York courts and federal courts will consider whether the visibility of the jury trial waiver provision within the agreement is sufficiently prominent such that a party has knowingly waived any right to trial by jury. We believe that this is the case with respect to the deposit agreement and the ADSs. In addition, New York courts will not enforce a jury trial waiver provision in order to bar a viable setoff or counterclaim sounding in fraud or one which is based upon a creditor's negligence in failing to liquidate collateral upon a guarantor's demand, or in the case of an intentional tort claim (as opposed to a contract dispute), none of which we believe are applicable in the case of the deposit agreement or the ADSs. No condition, stipulation or provision of the deposit agreement or ADSs serves as a waiver by any holder or beneficial owner of ADSs or by us or the depository of compliance with any provision of the federal securities laws. If you or any other holder or beneficial owner of ADSs brings a claim against us or the depository in connection with matters arising under the deposit agreement or the ADSs, you or such other holder or beneficial owner may not be entitled to a jury trial with respect to such claims, which may have the effect of limiting and discouraging lawsuits against us and / or the depository. If a lawsuit is brought against us and/or the depository under the deposit agreement, it may be heard only by a judge or justice of the applicable trial court, which would be conducted according to different civil procedures and may cause different results than a trial by jury would have had, including results that could be less favorable to the plaintiff(s) in any such action, depending on, among other things, the nature of the claims, the judge or justice hearing such claims, and the venue of the hearing.

Holders of the ADSs will not be able to exercise the pre-emptive subscription rights related to the ordinary shares that they represent, and may suffer dilution of their equity holding in the event of future issuances of our ordinary shares.

Under the Danish Companies Act, or DCA, our shareholders benefit from a pre-emptive subscription right on the issuance of ordinary shares for cash consideration only and not in the event of issuance of shares against non-cash contribution or debt conversion. Shareholders' pre-emptive subscription rights, in the event of issuances of ordinary shares against cash payment, may be disappplied by a resolution of the shareholders at a general meeting of our shareholders and/or the ordinary shares may be issued on the basis of an authorization granted to the board of directors pursuant to which the board may disapply the shareholders' pre-emptive subscription rights. Our shareholders have authorized our board of directors to issue securities, including in connection with issues of new ordinary shares without pre-emptive rights for our existing shareholders at or

above market price against cash payment, issues of new ordinary shares without pre-emptive rights to members of our board of directors, our executives and/or our employees and to certain specific third-parties which may be below the market price against cash payment or, for certain third-parties, by issuance of bonus shares as well as issues of new ordinary shares with pre-emptive rights for our existing shareholders against cash payment or conversion of debt which may be below the market price. Ordinary shares may be issued at or above the market price or below the market price, as such term is construed under Danish law, in the case of rights issues or pursuant to a resolution of the shareholders. The absence of pre-emptive rights for existing equity holders may cause dilution to such holders.

Furthermore, ADS holders would not be entitled, even if such rights accrued to our shareholders in any given instance, to receive such pre-emptive subscription rights related to the ordinary shares that they represent. Rather, the depositary is required to endeavor to sell any such subscription rights that may accrue to the ordinary shares underlying the ADSs and to remit the net proceeds therefrom to the ADS holders pro rata. In addition, if the depositary is unable to sell rights, the depositary will allow the rights to lapse, in which case you will receive no value for these rights.

Claims of U.S. civil liabilities may not be enforceable against us.

We are incorporated under the laws of Denmark. Substantially all of our assets are located outside the United States. The majority of our board members and employees reside outside the United States. As a result, it may not be possible for investors to effect service of process within the United States upon such persons or to enforce against them or us in U.S. courts, including judgments predicated upon the civil liability provisions of the U.S. securities laws.

The United States and Denmark currently do not have a treaty providing for the reciprocal recognition and enforcement of judgments (other than arbitration awards) in civil and commercial matters. Consequently, a final judgment for payment given by a U.S. court, whether or not predicated solely upon U.S. securities laws, would not be enforceable in Denmark.

In order to obtain a judgment that is enforceable in Denmark, the party in whose favor a final and conclusive judgment of the U.S. court has been rendered will be required to file its claim again with a court of competent jurisdiction in Denmark. The Danish court will not be bound by the judgment by the U.S. court, but the judgment may be submitted as evidence. It is up to the Danish court to assess the judgment by the U.S. court and decide if and to what extent the judgment should be followed. Danish courts are likely to deny claims for punitive damages and may grant a reduced amount of damages compared to U.S. courts.

Based on the lack of a treaty as described above, U.S. investors may not be able to enforce against us or members of our board of directors or our executive management, or certain experts named herein who are residents of Denmark or countries other than the United States any judgments obtained in U.S. courts in civil and commercial matters, including judgments under the U.S. federal securities laws.

We are an “emerging growth company” and as a result of the reduced disclosure and governance requirements applicable to emerging growth companies, the ordinary shares or ADSs may be less attractive to investors.

We are an “emerging growth company” as defined in the Jumpstart Our Business Startups Act of 2012. For as long as we continue to be an emerging growth company, we may take advantage of exemptions from various reporting requirements that are applicable to other public companies that are not emerging growth companies, including not being required to comply with the auditor attestation requirements of Section 404(b) of the Sarbanes-Oxley Act of 2002, or Section 404(b), exemptions from the requirements of holding a nonbinding advisory vote on executive compensation and shareholder approval of any golden parachute payments not previously approved. We may take advantage of these exemptions until we are no longer an emerging growth company. We could be an emerging growth company for up to five years, although circumstances could cause us to lose that status earlier, including if the aggregate market value of our ordinary shares, including ordinary shares represented by ADSs, held by non-affiliates exceeds \$700 million as of the end of our second fiscal quarter before that time, in which case we would no longer be an emerging growth company as of the following December 31st (the last day of our fiscal year). We cannot predict if investors will find the ordinary shares or ADSs less attractive because we may rely on these exemptions. If some investors find the ordinary shares or ADSs less attractive as a result, there may be a less active trading market for the ordinary shares or ADSs and the price of the ordinary shares or ADSs may be more volatile.

As a foreign private issuer, we rely on certain home country corporate governance practices.

We currently qualify as a foreign private issuer. As a result, we rely on home country governance requirements and certain exemptions thereunder. For example, we are exempt from certain rules under the Exchange Act that regulate disclosure obligations and procedural requirements related to the solicitation of proxies, consents or authorizations applicable to a security registered under the Exchange Act, including the U.S. proxy rules under Section 14 of the Exchange Act. In addition, our officers and directors are exempt from the reporting and “short-swing” profit recovery provisions of Section 16 of the Exchange Act and related rules with respect to their purchases and sales of our securities. Moreover, while we currently publish annual and semi-annual reports on our website pursuant to the rules of Nasdaq Copenhagen and expect to file such financial reports with the SEC, we are not required to file periodic reports with the SEC as frequently or as promptly as U.S. public companies. Specifically, we are not required to file quarterly reports on Form 10-Q or current reports on Form 8-K that a domestic company would be required to file under the Exchange Act. Accordingly, there may be less publicly available information concerning our company than there would be if we were not a foreign private issuer.

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

Based on the composition of our income and valuation of our assets, including goodwill, and certain assumptions with respect to the characterization of our income as active or passive, 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, generally 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 Consequences for U.S. Holders”) 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 Consequences for U.S. Holders.” 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.

If a U.S. Holder is treated as owning at least 10% of our ordinary shares or ADSs, such holder may be subject to adverse U.S. federal income tax consequences.

If a U.S. Holder is treated as owning, directly, indirectly or constructively, at least 10% of the value or voting power of our ordinary shares or ADSs, such U.S. Holder may be treated as a “United States shareholder” with respect to each “controlled foreign corporation” in our group, if any. Because our group currently includes at least one U.S. subsidiary, under current law, any of our current non-U.S. subsidiaries and any future newly formed or acquired non-U.S. subsidiaries will be treated as controlled foreign corporations, regardless of whether we are treated as a controlled foreign corporation. A United States shareholder of a controlled foreign corporation may be required to annually report and include in its U.S. taxable income its pro rata share of “Subpart F income,” “global intangible low-taxed income” and investments in U.S. property by controlled foreign corporations, regardless of whether we make any distributions. An individual that is a United States shareholder with respect to a controlled foreign corporation generally would not be allowed certain tax deductions or foreign tax credits that would be allowed to a United States shareholder that is a U.S. corporation. Failure to comply with controlled foreign corporation reporting obligations may subject a United States shareholder to significant monetary penalties. We cannot provide any assurances that we will furnish to any United States shareholder information that may be necessary to comply with the reporting and tax paying obligations applicable under the controlled foreign corporation rules of the Code. U.S. Holders should consult their tax advisors regarding the potential application of these rules to their investment in ordinary shares or ADSs.

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 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.

We are exposed to changes in foreign currency exchange rates and interest rates.

Substantially all of our income is expected to be in U.S. dollars and Euros, while part of our operating costs are currently denominated in Danish kroner, although in the future such Danish kroner denominated operating costs are likely to constitute a smaller percentage of our total operating costs. We do not currently have in place hedging contracts to cover our currency risks and, accordingly, fluctuations in Danish kroner against, in particular U.S. dollars, could have an adverse effect on our business, results of operations, cash flows, financial condition and/or prospects.

Our interest rate risk mainly derives from the fact that we hold a large cash position. Significant negative changes in interest rates could affect the value of our funds and any placement thereof and may thereby adversely affect our business, results of operations, cash flows, financial condition and/or prospects.

Shareholders outside Denmark may be subject to exchange rate risk.

The ordinary shares underlying the ADSs are denominated in Danish kroner. Accordingly, an investment in the ordinary shares or ADSs by an investor whose principal currency is not Danish kroner may expose such investor to foreign currency exchange rate risk. Any depreciation of Danish kroner against such foreign currency would reduce the value of the investment in the ordinary shares or ADSs, as applicable, in terms of such foreign currency.

We incur costs as a result of operating as a public company in the United States and Denmark, and our management is required to devote substantial time to new compliance initiatives.

As a U.S. public company, we incur significant legal, accounting, and other expenses. We are subject to the reporting requirements of the Securities Exchange Act of 1934, or the Exchange Act, the Sarbanes-Oxley Act, the Dodd-Frank Wall Street Reform and Consumer Protection Act, and other applicable securities rules and regulations. Compliance with these rules and regulations requires increased investment in legal and financial compliance, makes some activities more difficult, time consuming or costly and increases demand on our systems and resources, and this will continue to be the case particularly after we are no longer an “emerging growth company” and/or a foreign private issuer. The Exchange Act requires that, as a public company, we file annual and current reports with respect to our business, financial condition and result of operations. However, as a foreign private issuer, we are not required to file quarterly and certain other current reports with respect to our business and results. We currently make annual and semi-annual reporting with respect to our listing on Nasdaq Copenhagen.

Moreover, these rules and regulations may make it more difficult and more expensive for us to obtain director and officer liability insurance, which in turn could make it more difficult for us to attract and retain qualified senior management personnel or members for our board of directors.

However, these rules and regulations are often subject to varying interpretations, in many cases due to their lack of specificity, and, as a result, their application in practice may evolve over time as new guidance is provided by regulatory and governing bodies. This could result in continuing uncertainty regarding compliance matters and higher costs necessitated by ongoing revisions to disclosure and governance practices.

Further, being a U.S. listed company and a Danish public company with ordinary shares admitted to trading on Nasdaq Copenhagen impacts the disclosure of information and requires compliance with two sets of applicable rules. From time to time, this may result in uncertainty regarding compliance matters and result in higher costs necessitated by legal analysis of dual legal regimes, ongoing revisions to disclosure and adherence to heightened governance practices. As a result of the enhanced disclosure requirements of the U.S. securities laws, business and financial information that we report is broadly disseminated and highly visible to investors, which we believe may increase the likelihood of threatened or actual litigation, including by competitors and other third parties, which could, even if unsuccessful, divert financial resources and the attention of our management from our operations.

As a result of material weaknesses, we have concluded that our internal control over financial reporting was not effective at the reasonable assurance level as of December 31, 2021. Therefore, we may not be able to accurately report our financial results in a timely manner or prevent fraud, which may adversely affect our business, investor confidence in our company and the market price of our shares and ADSs.

We have identified material weaknesses in the design and operating effectiveness of our internal control over financial reporting such that we have concluded that our internal control over financial reporting was not effective at the reasonable assurance level as of December 31, 2021. A material weakness is a deficiency, or a combination of deficiencies, in internal control over financial reporting, such that there is a reasonable possibility that a material misstatement of annual or interim financial statements will not be prevented or detected on a timely basis. In connection with the preparation of this annual report on Form 20-F, we have identified material weaknesses, as follows: (i) ineffective control environment driven by the lack of a sufficient number of trained professionals with an appropriate level of internal control knowledge, training and experience; (ii) insufficient control activities, as we did not have adequate formal documentation of certain policies and procedures, implementation of all required business process controls, including effective review

process of key financial information, and sufficient documentation to evidence the design and operating effectiveness of the control activities; (iii) insufficient information & communication as we did not implement effective controls over the segregation of duties and certain information technology general controls for information systems that are relevant to the preparation of our financial statements; and (iv) insufficient monitoring activities, as we did not have the evidence to support evaluation of the effectiveness of monitoring controls to ascertain whether the components of internal control are present and functioning. Thus, we are unable to evaluate whether other deficiencies, individually or in combination, result in a reasonable possibility that a material misstatement of our annual financial statements will not be prevented or detected on a timely basis.

On May 31, 2022 we completed the sale of substantially all of the Company's assets and business activities, including those relating to the development and approval of arimoclomol and the full claw back liability related to the French early access program, to KemPharm Denmark A/S (KemPharm), a wholly owned subsidiary of KemPharm Inc. Following completion of the Sale of Assets, we have limited ongoing operational business activities and two employees.

Although we have voluntarily delisted the ADSs representing our ordinary shares from the Nasdaq Global Select Market in the United States, which became effective March 31, 2022, we are required to continue our SEC reporting obligations until at least March 31, 2023, when we anticipate filing a Form 15F with the SEC to suspend our reporting obligations under the Exchange Act.

In light of our limited operational business activities and resources, we are evaluating remediation activities that are commensurate with the size and operations of the company going forward. There can be no assurance that we will enter into such activities prior to such date, if ever, or comply with the requirements of Section 404 of the Sarbanes-Oxley Act before within such time frame or at all.

If we fail to remediate the material weaknesses to rectify our ineffective internal controls over financial reporting, identify new material weaknesses in our internal controls over financial reporting, are unable to comply with the requirements of Section 404 of the Sarbanes-Oxley Act in a timely manner, are unable to conclude that our internal controls over financial reporting are effective, or if our independent registered public accounting firm is unable to express an opinion as to the effectiveness of our internal controls over financial reporting when we are no longer an emerging growth company, investors may lose confidence in the accuracy and completeness of our financial reports and the market price of our ADSs could be negatively affected. As a result of such failures, we could also become subject to investigations by the stock exchanges on which our securities are listed, the SEC, or other regulatory authorities, and become subject to litigation from investors and shareholders, which could harm our reputation and financial condition or divert financial and management resources from our regular business activities.

We are a Danish company with limited liability. The rights of our shareholders may be different from the rights of shareholders in companies governed by the laws of U.S. jurisdictions.

We are a Danish company with limited liability. Our corporate affairs are governed by our Articles of Association and by the laws governing companies incorporated in Denmark. The rights of shareholders and the responsibilities of members of our board of directors may be different from the rights and obligations of shareholders and boards of directors in companies governed by the laws of U.S. jurisdictions. In the performance of its duties, our board is required by Danish law to consider the interests of our company, its shareholders, its employees and other stakeholders, in all cases with due observation of the principles of reasonableness and fairness. It is possible that some of these parties will have interests that are different from, or in addition to, the interests of our shareholders. See Exhibit 2.1 under "Articles of Association and Danish Corporate Law."

General Risk Factors

We are subject to securities litigation, which is expensive and could divert management's attention.

The market price of the ordinary shares or ADSs may be volatile and, in the past, companies that have experienced volatility in the market price of their stock have been subject to securities class action litigation. On July 9, 2021, a putative class action lawsuit, captioned *Busic v. Orphazyme A/S, et al.*, No. 21- cv-03640 (N.D. Ill.), was filed against us and certain of our current and former directors and officers in the U.S. District Court for the Northern District of Illinois. On November 19, 2021, the lead plaintiff filed an amended complaint asserting claims under the Securities Act and the Exchange for alleged misrepresentations and/or omissions in

our registration statement for our IPO of ADS in the United States and in subsequent public statements, which seeks an unspecified amount of damages on behalf of a putative class comprised of all persons and entities other than the defendants that purchased or otherwise acquired the ADS in connection with the IPO in September 2020 and/or between September 29, 2020 and November 4, 2021. See “Business – Legal Proceedings.” The litigation remains ongoing. However, regardless of outcome, *Busic* or other potential securities litigation against us could result in substantial costs and divert our management’s attention from other business concerns, which could seriously harm our business.

Our operations as a global company subject us to various risks, and our failure to manage these risks could adversely affect our business, results of operations, cash flows, financial condition and/or prospects.

We face significant operational risks as a result of doing business globally, such as:

- fluctuations in currency exchange rates (in particular, U.S. dollars, Euros and Danish kroner);
- potentially adverse tax consequences, including the complexities of multinational value-added tax systems, tax inefficiencies related to our corporate structure, and restrictions on the repatriation of earnings;
- export restrictions, trade regulations and foreign tax laws;
- political, social, and economic instability, global health epidemics or other contagious diseases, terrorist attacks and security concerns in general;
- the burdens of complying with a wide variety of multinational laws and different legal standards; and
- increased financial accounting and reporting burdens and complexities.

If one or more of these risks are realized, it could have a material adverse effect on our business, cash flows, financial condition and/or prospects.

Global economic uncertainty and other global economic or political and regulatory developments could have a material adverse effect on our business, cash flows, financial condition and/or prospects.

Growth in the global pharmaceutical market has become increasingly tied to (i) global economic growth as an economic downturn may, for example as the result of COVID-19 paralyzing economic activities, reduce the amount of funding for the pharmaceutical sector as a whole or certain diseases targeted by us and (ii) political conditions, tension and uncertainty which could, for instance, impact the regulations applicable to us.

Uncertain political and geopolitical conditions currently exist in various parts of the world. At the end of 2021 and into 2022, tensions between the United States and Russia escalated when Russia amassed large numbers of military ground forces and support personnel on the Ukraine-Russia border and in February 2022, Russia launched a wide-ranging attack on Ukraine. In response, NATO has deployed additional military forces to Eastern Europe, and the United Kingdom, the European Union and the United States announced certain sanctions against Russia. The conflict in Ukraine and any retaliatory measures taken by the United States and NATO could threaten global security and result in further regional conflict and otherwise have a lasting impact on regional and global economies, any or all of which could adversely affect our business. In addition, the full effects of the United Kingdom’s exit from the EU in January 2020 are impossible to predict but may result in significant market volatility and dislocation, and adversely affect the United Kingdom, European and global economy.

Future legal or regulatory changes in jurisdictions where we currently operate, or in such jurisdictions in which we may choose to operate in the future, could materially and adversely affect our business, results of operations, cash flows, financial condition and/or prospects, including by imposing regulatory and operational restrictions and compliance obligations on our business, reducing our revenue or increasing our expenses.

The above circumstances, individually or in the aggregate, could have a material adverse effect on our business, cash flows, financial condition and/or prospects.

ITEM 4. INFORMATION ON THE COMPANY

A. History and development of the company.

Orphazyme A/S was 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 in Denmark on Nasdaq Copenhagen in November 2017, under the ticker “ORPHA.” In September 2020, following a U.S. public offering, American Depositary Shares (“ADSs”) representing ordinary shares of Orphazyme began trading on Nasdaq Global Select Market, under the ticker “ORPH.” In March 2022, we voluntarily delisted the ADSs from the Nasdaq Global Select Market, which became effective March 31, 2022.

Our headquarters and principal executive office is located at Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark, and our telephone number is +45 39 17 82 72. The mailing address of our wholly owned subsidiary, Orphazyme US, Inc., is P.O. Box 583, New York, NY 10116, United States of America. Our wholly owned subsidiary, Orphazyme Schweiz GmbH, has offices at Grafenauweg 10, Zug, 6301, Switzerland. We have discontinued operations at our U.S. and Swiss entities and are considering winding down these subsidiaries at an appropriate time.

On March 11, 2022 the Board of Directors of Orphazyme initiated in-court restructuring proceedings whereby we pursued potential opportunities for a sale of our assets and operations, a sale of the Company and/or a refinancing (potentially in combination with a compulsory composition).

In May 2022, substantially all of the Company’s assets and business activities, including those relating to the development and approval of arimoclomol and the full claw back liability related to the French early access program, were sold to KemPharm Denmark A/S (“KemPharm”), a wholly owned subsidiary of KemPharm Inc. for a total of USD 12.8 million in cash and assumed liabilities estimated to equal approximately USD 5.2 million (the “Sale of Assets”).

Following the signing of the agreement with KemPharm, we submitted a restructuring proposal to the Danish Maritime and Commercial High Court and Orphazyme’s known creditors, which amongst other things, included a proposal to complete the Sale of Assets to KemPharm. The restructuring proposal was approved on May 30, 2022 by Orphazyme’s known creditors and affirmed by the Danish Maritime and Commercial High Court. The in-court restructuring proceedings were discontinued with immediate effect on May 30, 2022 and the Sale of Assets was completed on May 31, 2022.

Completion of the Sale of Assets is expected to provide full coverage to creditors with undisputed claims based on the claims filed during the restructuring. All (undisputed and unconditional) debts related to the time prior to restructuring have been or will be paid in full, including all obligations outstanding under the Company’s debt facility with Kreos Capital.

As substantially all of our assets and business activities have been sold to KemPharm, we have limited ongoing operational business activities and, as of June 24, 2022, we had only two full-time employees (FTEs).

Our capital expenditures primarily relate to leases. For further information please see Note 3.2 ‘Leases’ in our Audited Consolidated Financial Statements included under “Item 18 – Financial Statements” of this Annual Report on Form 20-F. The lease on our Danish Headquarters was assigned to KemPharm, June 1 2022, and KemPharm is allowing us to maintain an executive office at the premises. We have terminated the lease on our U.S. office in Chicago, effective June 22, 2022.

We have not had any public takeover offers in respect of Orphazyme A/S’ shares to date. Pursuant to the in-court restructuring proceedings, we received certain non-binding offers to purchase all of Orphazyme’s assets and operations as outlined above.

Our website address is www.orphazyme.com. The information contained on, or accessible through, our website is not incorporated by reference into this annual report, and you should not consider any information contained in, or that can be accessed through, our website as part of this annual report or in deciding whether to purchase or sell our ordinary shares (including ordinary shares in the form of ADSs). We have included our website address as an inactive textual reference only.

The United States Securities and Exchange Commission (the “SEC”) maintains a website at www.sec.gov which contains in electronic form each of the reports, proxy and information statements, and other information that we have filed electronically with the SEC.

B. Business overview

In 2021, and until recently, the primary focus of Orphazyme was on seeking marketing authorization in the United States and Europe for its investigational product candidate arimoclomol for the treatment of Niemann-Pick disease type-C (NPC), preparing to commercialize arimoclomol in NPC, if approved, and completing two late-stage clinical studies – a Phase 2/3 trial evaluating arimoclomol in Amyotrophic Lateral Sclerosis (ALS) and a Phase 2/3 trial evaluating arimoclomol in inclusion body myositis (IBM).

A number of unfavorable events occurred during 2021 and early 2022, which significantly impacted our business.

In March 2021, we announced that our Phase 2/3 trial in IBM did not meet its primary and secondary endpoints, and in May 2021 we announced that our Phase 3 trial in ALS did not meet its primary and secondary endpoints. As a result, we ceased development in these indications. These clinical trials were some of the largest and longest performed in these indications and we believe the data will contribute meaningfully to the scientific dialogue in these diseases.

In June 2021, we received a Complete Response Letter (CRL) from the U.S. Food and Drug Administration (FDA) for our New Drug Application (NDA) for arimoclomol in NPC. The CRL outlined the need for 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 that additional data would be needed to supplement confirmatory evidence beyond the single Phase 2/3 trial for NPC.

Following the CRL, we took swift and decisive action and in June 2021 we announced a restructuring plan. We re-focused our activities and obligations on gaining approval for arimoclomol in NPC in Europe and evaluating the regulatory path forward in the U.S. We significantly scaled back our global organization, including teams based in the U.S. and Europe, with the purpose of reducing the number of employees to those supporting essential activities including the regulatory processes in U.S. and Europe and our Early Access Program (EAP).

We continued to build the profile of arimoclomol with the scientific community during 2021. In June 2021 we announced presentation of 24-month interim results from the open-label extension (OLE) trial, providing efficacy and safety data for arimoclomol in NPC for up to 36 months, at the Parseghian Scientific Conference for NPC Research and in August 2021 we announced publication of data from the 12-month double-blind portion of the Phase 2/3 NPC-002 trial in the *Journal of Inherited Metabolic Disease* (JIMD). Further, we held a Type A meeting with the FDA in October 2021, where the FDA recommended we submit additional data, information and analyses and engage in further interactions to identify a pathway to resubmission of the NDA. In addition, we continued to engage with European regulators as they evaluated our Marketing Authorisation Application (MAA).

We ended 2021 with DKK 102 million in cash and cash equivalents.

In January 2022, Christophe Bourdon, our Chief Executive Officer (CEO), resigned from his position as CEO of Orphazyme to take on the role of CEO at another company. Anders Vadsholt was appointed CEO of Orphazyme, effective March 1, 2022, in addition to his position as Chief Financial Officer (CFO).

In February 2022, we were notified of a negative trend vote by the European Medicine’s Agency (EMA) Committee for Medicinal Products for Human Use (CHMP) relating to our MAA for arimoclomol in NPC in Europe. We withdrew our MAA in March 2022, ahead of the CHMP’s final vote.

In light of our financial situation at the time and the negative trend vote from the CHMP, the Board of Directors initiated in-court restructuring proceedings of Orphazyme under the Danish Insolvency Act, which commenced on March 11, 2022. In accordance with the in-court restructuring proceedings, we published a statutory restructuring plan on March 31, 2022, which was adopted by Orphazyme’s creditors on April 7, 2022 (the Statutory Restructuring Plan). The aim of the in-court restructuring proceedings was to explore whether a basis could be established for all or part of our operations to continue, including a basis for injecting further capital, and/or a basis for a sale of all or parts of our assets. Following initiation of the in-court restructuring proceedings, we voluntarily delisted our American Depositary Shares (ADSs) representing Orphazyme’s

ordinary shares from the Nasdaq Global Select Market in the United States and filed a Form 25 with the Securities and Exchange Commission (SEC) on March 21, 2022.

In accordance with the Statutory Restructuring Plan, in April 2022 we announced that we had received certain non-binding offers to purchase all of our assets and operations. We subsequently entered into exclusive negotiations with a selected potential buyer.

In May 2022, we announced that we had signed an agreement to sell substantially all of the Company's assets and business activities, including those relating to the development and approval of arimoclomol and the full claw back liability related to the French early access program, to KemPharm Denmark A/S ("KemPharm"), a wholly owned subsidiary of KemPharm Inc. for a total of USD 12.8 million in cash and assumed liabilities estimated to equal approximately USD 5.2 million (the "Sale of Assets"). KemPharm is a specialty pharmaceutical company focused on the discovery and development of novel treatments for rare central nervous system diseases. Under the terms of the agreement, KemPharm agreed to acquire substantially all of Orphazyme's assets and business activities, including those relating to the development and approval of arimoclomol, retain a majority of Orphazyme's remaining Danish employees, continue the early access programs with arimoclomol and pursue the potential approval of arimoclomol as a treatment option for NPC.

Following the signing of the agreement with KemPharm, we submitted a restructuring proposal to the Danish Maritime and Commercial High Court and Orphazyme's known creditors, which amongst other things, included a proposal to complete the Sale of Assets to KemPharm. The restructuring proposal was approved on May 30, 2022 by Orphazyme's known creditors and affirmed by the Danish Maritime and Commercial High Court. Following the approval of the restructuring proposal, the in-court restructuring proceedings were discontinued with immediate effect on May 30, 2022 and the Sale of Assets was completed on May 31, 2022. Completion of the Sale of Assets is expected to provide full coverage to creditors with undisputed claims based on the claims filed during the restructuring. All (undisputed and unconditional) debts related to the time prior to restructuring have been or will be paid in full, including all obligations outstanding under the Company's debt facility with Kreos Capital.

As substantially all of our assets and business activities have been sold to KemPharm, we have limited ongoing operational business activities and, as of June 24, 2022, we had only two employees. We announced that Stephanie Okey, Carolee Barlow and Martin Bonde stepped down from their positions as members of the Board of Directors as of May 23, 2022.

Intellectual Property

As of June 1, 2022, our trademark portfolio contained approximately eight trademark registrations since other trademark registrations and trademark applications have been assigned to KemPharm. We have registered "ORPHAZYME" as a word mark in relevant trademark classes and jurisdictions (in the European Union, United Kingdom, and International Madrid Protocol designating the United States, China, Israel, India, Japan and Russia).

Material Agreements

KemPharm Asset Purchase Agreement

On May 15, 2022, we entered into an asset purchase agreement with KemPharm Denmark A/S ("KemPharm"), a wholly owned subsidiary of KemPharm Inc. Pursuant to this agreement KemPharm (i) acquired substantially all of our employees and most of our assets, except for certain exempted assets; (ii) took over certain liabilities, including employee liabilities, liabilities relating to an early access patient program and certain milestone payments linked to approvals of medicinal products; and (iii) took over certain of our continued agreements relevant to the continued operations of the business operated by us. KemPharm paid a total of USD 12.8 million in cash and assumed liabilities estimated to equal approximately USD 5.2 million as consideration. This transaction was completed on May 31, 2022.

C. Organizational structure.

Orphazyme A/S is the parent company of two wholly-owned subsidiaries for which it holds 100% of the voting power: Orphazyme US, Inc., incorporated in the United States in April 2018, and Orphazyme Schweiz GmbH, incorporated in Switzerland in March 2020. Orphazyme A/S has discontinued operations at its U.S. and Swiss entities and is considering winding down these subsidiaries at an appropriate time.

D. Property, plant and equipment.

At December 31, 2021, we still leased office and laboratory space from COBIS A/S on Ole Maaløes Vej 3, 2200 Copenhagen N, Denmark. Since then, we have vacated our laboratory space and assigned the remaining office facility to KemPharm Denmark ApS as of June 1, 2022. We also leased an office in Chicago, Illinois for our U.S. subsidiary, for which the lease was terminated as of June 22, 2022.

For further information on our leases, please see Note 3.2 ‘Leases’ to our Audited Consolidated Financial Statements included under Item 18 of this Annual Report on Form 20-F.

For further information our property, plant and equipment, please see Note 3.3 ‘Property, Plant, and Equipment’ to our Audited Consolidated Financial Statements included under Item 18 of this Annual Report on Form 20-F.

ITEM 4A. UNRESOLVED STAFF COMMENTS

Not applicable.

ITEM 5. OPERATING AND FINANCIAL REVIEW AND PROSPECTS

You should read the following discussion and analysis of our financial condition and results of operations together with our Audited Consolidated Financial Statements included in Item 18 of this Annual Report on Form 20-F and other financial information included in this Annual Report on Form 20-F and our statutory Annual Report 2021. The following discussion is based on our financial information prepared in accordance with IFRS, as issued by the IASB, which might differ in material respects from accounting principles generally accepted in other jurisdictions, including U.S. GAAP. Some of the information contained in this discussion and analysis or set forth elsewhere in this Annual Report on Form 20-F, including information with respect to our plans and strategy for our business, includes forward-looking statements that involve risks and uncertainties. You should review the “Risk Factors” section of this Annual Report on Form 20-F for a discussion of important factors that could cause actual results to differ materially from the results described in or implied by the forward-looking statements contained in the following discussion and analysis.

Critical Accounting Policies

Our consolidated financial statements are prepared in accordance with International Financial Reporting Standards (IFRS) as issued by the International Accounting Standards Board (IASB) and in conformity with IFRS as adopted by the European Union (EU). For a description of our significant accounting policies, including significant accounting estimates and judgements, reference is made to Notes 1.3 ‘Significant Accounting Policies’ and 1.4 ‘Significant Accounting Estimates and Judgements’ to our Audited Consolidated Financial Statements included under Item 18 of this Annual Report on Form 20-F.

New IFRS Standards Applicable to the Company

For a description of recent accounting pronouncements applicable to us reference is made to Note 1.5 ‘New IFRS Standards Applicable to the Group’ to our Audited Consolidated Financial Statements included under Item 18 of this Annual Report on Form 20-F.

A. Operating results

Comparison of the Results of Operations for the Years Ended December 31, 2021 and 2020

For a discussion on the financial development and condition of the Company for the fiscal year ending December 31, 2021, and comparison with the year ended December 31, 2020, reference is made to the section ‘Financial Review’ contained on pages 7 – 8 in our Annual Report 2021, and to the Audited Consolidated Financial Statements included under “Item 18 – Financial Statements”, “Item 4 – Information on the Company – B. Business Overview” and “Item 3 – Key Information – D. Risk Factors” of this Annual Report 2021 on Form 20-F.

The analysis and discussion included in our Annual Report 2021 is primarily based on the Audited Consolidated Financial Statements, which are prepared in accordance with IFRS as issued by the IASB.

Comparison of the Results of Operations for the Years Ended December 31, 2020 and 2019

For a discussion on the financial development and condition of the Company for the fiscal year ending December 31, 2020, and comparison with the year ended December 31, 2019, reference is made to “Item 5A –

Operating Results”, “Item 18 – Financial Results” and “Item 3 – Key Information – D. Risk Factors” in our Annual Report 2020 on Form 20-F, filed with the SEC on March 2, 2021.

Notes 1.3 through 1.5 of our Audited Consolidated Financial Statements in this Annual Report on Form 20-F include a description of significant accounting policies and critical estimates and judgements, including implementation of new and revised IFRS standards and interpretations.

Segment information

Although Orphazyme established a US subsidiary in 2018 and a Swiss subsidiary in 2020, the Group is managed and operated as one business unit that is reflected in the internal reporting. No separate lines of business or separate business entities have been identified with respect to any product candidate or geographical market and no segment information is currently disclosed in the Group’s internal reporting. For the years ended December 31, 2020 and 2019, the Group generated no revenue and for the year ended December 31, 2021 the Danish entity generated revenue which is disclosed in a separate note. For the years ended December 31, 2021, 2020 and 2019 all material non-current assets are located in Denmark.

Inflation

Inflation for the years ended December 31, 2021, 2020 and 2019 has not had a material impact on our financial results.

Foreign currencies

For the impact of foreign currency fluctuations on the Company, reference is made to Note 4.4 ‘Financial Risks’ to our Audited Consolidated Financial Statements included under “Item 18 – Financial Statements” of this Annual Report on Form 20-F.

B. Liquidity and capital resources

To date, we have raised aggregate gross proceeds of DKK 2,271 million (approximately \$321 million) through sales of equity securities. This includes gross proceeds of DKK 600 million raised in our initial public offering in Denmark in November 2017, gross proceeds of DKK 745 million raised in our directed issue and private placement in February 2020, gross proceeds of DKK 534 million raised in our global offering in September 2020 and gross proceeds of USD 835,668 from the issue of new shares as a result of the utilization of the Company’s U.S. At-the-Market Offering Program in February 2022.

Since our inception, we have incurred significant operating losses relating to the advancement of our product candidate arimoclomol.

As of December 31, 2021, we held DKK 102 million (\$15.5 million) in cash.

In May 2022, we sold substantially all of the Company’s assets and business activities, including those relating to the development and approval of arimoclomol and the full claw back liability related to the French early access program, to KemPharm Denmark A/S (KemPharm), a wholly owned subsidiary of KemPharm Inc. for a total of USD 12.8 million in cash and assumed liabilities estimated to equal approximately USD 5.2 million (the “Sale of Assets”). Following completion of the sale of substantially all the Company’s assets and business activities to KemPharm in May 2022, Orphazyme has limited ongoing operational business activities and only two employees.

Financial resources

Reference is made to our Audited Consolidated Financial Statements included under “Item 18 – Financial Statements” in this Annual Report on Form 20-F including our ‘Consolidated Statements of Financial Position’ and ‘Consolidated Statement of Cash flows’. Further reference is made to “Item 3 – Key Information – D. Risk Factors” and “Item 4 – Information on the Company – B. Business Overview”.

Comparison of Cash Flow for the Years Ended December 31, 2021 and 2020

Reference is made to our ‘Consolidated Statement of Cash flows’ in our Audited Consolidated Financial Statements included under “Item 18 – Financial Statements” in this Annual Report on Form 20-F and to ‘Financial Review – Cash Flows’ on page 8 of our Annual Report 2021.

Comparison of Cash Flow for the Years Ended December 31, 2020 and 2019

Reference is made to our Audited Consolidated Financial Statements included under “Item 18 – Financial Results” in our Annual Report 2020 on Form 20-F, including our ‘Consolidated Statements of Financial Position’ and ‘Consolidated Statement of Cash flows’, and to “Item 5B – Liquidity and Financial Resources” in our Annual Report 2020 on Form 20-F, filed with the SEC on March 2, 2021.

Debt financing

Reference is made to the Audited Consolidated Financial Statements included under “Item 18 – Financial Statements” of this Annual Report on Form 20-F, including ‘Consolidated Statements of Financial Position’, Note 3.7 ‘Financial Assets and Liabilities’ and Note 3.9 ‘Commitments and Contingencies’.

Financial Instruments

Reference is made to Note 4.4 ‘Financial Risks’ to the Audited Consolidated Financial Statements included under “Item 18 – Financial Statements” of this Annual Report on Form 20-F.

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 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 a requirement to include only two years of audited financial statements and only two years of related Results of Operations disclosure; and an exemption from the auditor attestation requirement in the assessment of our internal control over financial reporting pursuant to the Sarbanes-Oxley Act.

We may choose to take advantage of some but not all of these reduced burdens, and therefore the information that we provide holders of 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. We currently prepare our consolidated financial statements in accordance with IFRS as issued by the IASB, so we are unable to make use of the extended transition period.

We may take advantage of these provisions for up to five years 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 this offering;
- 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.

C. Research and Development, Patents and Licenses, etc.

In May 2022, we sold substantially all of the Company’s assets and business activities, including those relating to the development and approval of arimoclomol and the full claw back liability related to the French early access program, to KemPharm Denmark A/S (KemPharm), a wholly owned subsidiary of KemPharm Inc.

The Company retained certain trademarks and licenses which are outlined in "Item 4 - Information on the Company - B. Business Overview"

D. Trend Information

In May 2022, we sold substantially all of the Company's assets and business activities, including those relating to the development and approval of arimoclomol and the full claw back liability related to the French early access program, to KemPharm Denmark A/S (KemPharm), a wholly owned subsidiary of KemPharm Inc. for a total of USD 12.8 million in cash and assumed liabilities estimated to equal approximately USD 5.2 million (the Sale of Assets). Following completion of the sale of substantially all the Company's assets and business activities to KemPharm in May 2022, Orphazyme has limited ongoing operational business activities and, as of June 24, 2022, we had only two employees.

In June 2022, Orphazyme A/S repaid the full remaining loan to Kreos Capital VI (UK) Ltd. At the repayment all pledges and securities withdrew. All (undisputed and unconditional) debts related to the time prior to restructuring have been or will be paid in full in the coming months.

Orphazyme's current share-based compensation programs are expected to have a limited value, due to the decreasing share price and the disclosed Sale of Assets.

For the full-year 2022 we anticipate an operating profit in the range DKK 10 – 30 million. We expect to end 2022 with more than DKK 30 million in cash and equivalents. There are inherent risks and uncertainties in our Outlook for 2022 given the recent closing of the KemPharm transaction, transfer of operating activities from Orphazyme to KemPharm and our future prospects.

E. Critical Accounting Estimates

Our consolidated financial statements are prepared in accordance with IFRS, which requires management to make certain estimates and assumptions, either at the balance sheet date or during the year, which affect the reported amounts of revenues, expenses, assets, liabilities and contingent amounts.

Our significant accounting policies are set out in "Item 18. Financial Statements—Note 1.3 Significant Accounting Policies". Given the uncertainties inherent in our business activities, we must make certain estimates and assumptions that require difficult, subjective and complex judgments. These are set out in "Item 18. Financial Statements—Note 1.4 Significant Accounting Estimates and Judgements" and the specific referenced notes.

ITEM 6. DIRECTORS, SENIOR MANAGEMENT AND EMPLOYEES

A. Directors and senior management

The below sets forth certain information relating to our executive management and directors as of the date of this Annual Report on Form 20-F.

Executive Management

Anders Vadsholt, 53, has served as our CEO and CFO since March 2022, succeeding Christophe Bourdon, former CEO, who resigned in January 2022 to take a position as CEO of LEO Pharma A/S. Mr. Vadsholt has also served as our Interim CEO, December 2020 to March 2021, and as our Chief Financial Officer since May 2016 and is registered as part of executive management with the Danish Business Authority. Mr. Vadsholt is the owner and a member of the executive management of Alpha Healthcare Investments ApS since 2015, as well as a partner at Obton Solenergi Sinope Komplementaranpartsselskab since 2015. In the past five years, Mr. Vadsholt has been a member of the board of directors of Oxthera AB from 2019 to 2021, a member of the executive management of Lakeside Invest ApS from 2015 to 2018 and Copenhagen Innovation Capital Management ApS from 2015 to 2018. Mr. Vadsholt holds an MBA in Finance and Strategy from the University of Melbourne, a Master of Science in Corporate Law and Economics from Copenhagen Business School.

Directors

Georges Gemayel, 62, has served as a member of our board of directors since November 2012 and as Chairman of our board of directors since September 2014. Mr. Gemayel is currently Interim CEO and chairman

of the Board of Gemini Therapeutics, chairman of the board of directors of Dynacure SAS, Enterome SA , and Orphazyme, and member of the Board of Directors of Supernus Pharmaceuticals Inc. Mr. Gemayel has been a partner in Gemayel Investment LLC since 2012, as well as a director of the non-governmental organization, St. Andrew's School in Ngong Inc. and a trustee of the Gemayel Family Foundation. In the past five years, Mr. Gemayel was a member of the board of directors of Momenta Pharmaceuticals Inc, chairman of the board of directors of Dimension Therapeutics Inc., Epitherapeutics ApS, OxThera AB and Vascular Magnetics Inc., a member of the board of directors of NPS Pharmaceuticals Inc. and Raptor Pharmaceuticals Corp., as well as a director of the nongovernmental organization, International Institute of New England. Mr. Gemayel holds a Master's degree and a PhD degree in Pharmacology from Paris-Sud University and a Docteur d' Exercice en Pharmacie from St. Joseph University.

Bo Jesper Hansen, 63, has served as a member of our board of directors since December 2010 and as Deputy Chairman since October 2017. Dr. Hansen is currently Chairman of the Board of Laborie Inc., Deputy Chairman of Swedish Orphan Biovitrum (SOBI) AB, member of the Board of directors Innoventa Medica ApS, and Reapplix A/S. Venture Partner at Wellington Partners Life Science Fund LP; Advisory Consultant for Aescap 2.0, Nordic Capital, EQT AB and Broad Street Principal Investments Europe Ltd. & senior business advisor for HBM Ventures Ltd. In the past five years, Dr. Hansen was chairman of the board of directors and a member of the executive management of chairman of the board of directors of Karo Pharma AB, Ablynx NV (which was acquired by Sanofi in 2019), Karolinska Development AB and a member of the board of directors of Ascelia Pharma AB, Newron Pharmaceuticals SpA, Hyperion Therapeutics Inc.(which was acquired by Horizon Pharma in 2015), Azanta A/S (which was acquired by Norgine B.V. in 2020) and Inspyr Inc. Dr. Hansen holds an M.D. and Ph.D. degree in Medicine from the University of Copenhagen.

Andrew Mercieca, 56, has served as a member of our board of directors since 2022. He is currently Director of Octant Limited, a company providing Financial Consultancy services. Mr. Mercieca has extensive experience in International Finance and across Life Science and Technology sectors in corporate, private and non-profit organisations. In the past five years, Mr. Mercieca was Chief Financial Officer and member of the Board of Trustees of LifeArc, a UK medical research charity focused on the translation of scientific discoveries and innovations for the benefit of patients and held the position of Trustee at Plan Zheroes, an innovative charitable organization focused on poverty alleviation in London. Mr. Mercieca holds a BA in Accounting and Finance from the University of Portsmouth and is a fellow of the Institute of Chartered Accountants in England & Wales.

Recent Developments

As announced in May 2022, it is the intention of the Board of Directors to propose re-election of the current Deputy Chairman of the Board of Directors, Bo Jesper Hansen, election of the current CEO and CFO, Anders Fink Vadsholt, as a new member of the Board of Directors, and election of the Company's former restructuring administrator, John Sommer Schmidt, as a new member of the Board of Directors at the Annual General Meeting of Orphazyme expected to be held on June 29, 2022. The current Chairman of the Board of Directors, Georges Gemayel, and member of the Board of Directors, Andrew Mercieca, will not be seeking re-election at the Annual General Meeting.

Family Relationships

There are no family relationships among any of our executive officers or directors.

B. Compensation

Compensation of Executive Management and Directors

Please see Note 4.5 'Remuneration of Board of Directors and Executive Management', which discusses compensation of our executive management, including short-term incentives, Note 2.5 'Employee Costs', which discusses the long-term incentive programs in which our executive management participates and Notes 2.6d and 2.6e to our Audited Consolidated Financial Statements included under "Item 18 – Financial Statements" of this Annual Report on Form 20-F which relate to the Sign-on bonus shares granted to former CEO Kim Stratton and Sign-on bonus shares granted to former CEO Christophe Bourdon, respectively.

Executive Management Agreements

Our executive management consists of our Chief Executive Officer and Chief Financial Officer, Anders Vadsholt. The below section describes the service agreements in place during 2021 with our former CEO, Christophe Bourdon; our CEO and CFO, Anders Vadsholt; and the service agreement in place in 2020 with our former CEO, Kim Stratton. Note 4.5 in our Audited Consolidated Financial Statements in this Annual Report on Form 20-F discusses compensation of our executive management, including short-term incentives, and Note 2.5 discusses the long-term incentive programs in which our executive management participates.

Anders Vadsholt, CEO and CFO

We entered into a service agreement for the position of CFO with Anders Vadsholt in October 2017 with an effective date of November 1, 2017, which was later amended to adjust for, among other things, annual salary increases. Pursuant to the service agreement, as amended, Mr. Vadsholt is entitled to (i) an annual base salary of approximately DKK 2.3 million, (ii) participate in our incentive schemes and (iii) standard benefits (such as a mileage allowance, insurance coverage and a company-paid computer). Following the resignation of Ms. Stratton as CEO in December 2020, Mr. Vadsholt acted as interim CEO with no additional compensation. Following the resignation of Mr. Bourdon, Mr. Vadsholt is acting as joint CEO and CFO, and received a salary increase to DKK 3.0 million and a one-time sign-on bonus of gross DKK 1.266 million.

Mr. Vadsholt may terminate his employment with us by giving six months' notice and we can terminate his employment with us by giving 12 months' notice.

The service agreement with Mr. Vadsholt provides for the payment of a takeover retention bonus equaling 12 months' base salary if Mr. Vadsholt is still employed by us and not under notice on the first anniversary of completion of certain transactions resulting in a change of control. Mr. Vadsholt will still be entitled to this bonus if he is under notice on the first anniversary if we terminate his employment without a reasonably justifiable cause or if he gives the notice due to a gross breach by us. The service agreement with Mr. Vadsholt also provides for the payment of post-employment compensation to his dependents in the event of his death.

Mr. Vadsholt is subject to a non-competition clause and a non-solicitation of customers clause applicable during his employment and for a period of 12 months following expiry of his employment. Mr. Vadsholt is entitled to separate compensation under his non-competition and non-solicitation clauses. Pursuant to mandatory Danish law, Mr. Vadsholt's non-competition clause lapses if his employment is terminated by the Company for a reason that is not attributable to him.

Christophe Bourdon, former CEO (April 2021 to February 2022)

We entered into a service agreement with Christophe Bourdon with an effective date of April 1, 2021.

Pursuant to the service agreement, Mr. Bourdon was entitled to (i) an annual base salary of approximately DKK 3.9 million, (ii) a sign-on bonus consisting of RSU's equal to 60% of the fixed annual gross salary and (iii) compensation for any documented loss of bonus at his previous employer up to a net amount of USD 350,000 (payable in 50% cash and 50% RSUs) and (iv) standard benefits (such as company car, insurance coverage and a company-paid computer). Mr. Bourdon was also eligible to benefits related to relocation and to participate in our incentive schemes, including an annual maximum cash bonus of 50% of his annual gross salary. In addition, under the long-term incentive program (LTIP), Mr. Bourdon was eligible to receive RSU's and PSU's for a total amount of up to 95% of Mr. Bourdon's annual gross salary. For further details on the terms and conditions of the LTIP, including definitions of capitalized terminology, see Note 2.6 of our Audited Consolidated Financial Statements in the Annual Report on Form 20-F.

In April 2021, Mr. Bourdon was granted sign-on RSUs equal to 60% of the fixed annual gross salary. Pursuant to this grant, Mr. Bourdon was eligible to receive 34,941 RSUs. In connection with the modified 2021 LTIP, the sign-on RSUs granted to Mr. Bourdon in April 2021 were reset after the same principles as the modified 2021 LTIP but with immediate vesting upon grant. Pursuant to this grant, Mr. Bourdon was eligible to receive 73,262 RSUs.

Christophe Bourdon, CEO, left the Company on February 28, 2022. In the period March 1, to March 31, 2022, Mr. Bourdon assisted the Company and its management as a full time consultant.

In accordance with the Termination Agreement, Mr. Bourdon was entitled to receive remuneration as usual until February 28, 2022 and was not entitled to receive any separate compensation in respect of the consultancy services provided in the period March 1 to March 31, 2022.

To the extent that Mr. Bourdon is entitled to a short-term incentive program (STIP) bonus for 2021 and 2022 in accordance with the terms and conditions of the applicable STIP, including that applicable targets are met, any bonus will be calculated and paid out on a pro rata basis in accordance with the terms of the STIP and Mr. Bourdon's service agreement. Mr. Bourdon is not entitled to additional bonus, any further grants or payments under the LTIP and STIP and is not entitled to receive the Extraordinary Retention Bonus (part 2) or the Takeover Bonus. Any granted Restricted Share Units and Performance Share Units as well as any sign-on Restricted Share Units not exercised before February 28, 2022 will lapse without further notice or compensation.

Mr. Bourdon's non-competition clause shall remain in force until and including November 30, 2022. The non-solicitation clause shall remain in force until and including February 28, 2023. Mr. Bourdon is not entitled to any compensation in this respect.

Shareholding Requirements

Our executive management are at all times required to hold a minimum amount of our shares with a value equal to each of their respective annual fixed salaries at the time of acquisition of the shares. The required shareholding may be built up over a specified period.

No member of the board of directors is entitled to any kind of remuneration upon retirement from his or her position as a member of the board of directors. We have not allocated funds for any pension benefits, severance schemes or similar measures, or undertaken any other obligations to do so on behalf of the board of directors, and we have no obligation to do so.

We previously adopted a Remuneration Policy for the board of directors and executive management of the company in accordance with Sections 139 and 139(a) of the Danish Companies Act, or DCA.

Equity Incentive Plans

Please see Note 2.6a 'Long-term incentive program (equity-settled)', Note 2.6b 'Phantom share-based incentive program (cash-settled)', and to Note 2.6c 'Restricted Share Units (cash-settled)' to our Audited Consolidated Financial Statements included under "Item 18 – Financial Statements" of this Annual Report on Form 20-F.

Insurance and Indemnification

According to the Danish Companies Act, the general meeting is permitted to discharge our board members and members of our executive management from liability for any particular financial year based on a resolution relating to the period covered by the financial statements for the previous financial year. This discharge means that the general meeting will relieve such board members and members of our executive management from liability to us. However, the general meeting cannot discharge any claims by individual shareholders or other third parties. In addition, the discharge can be set aside in case the general meeting prior to its decision to discharge was not presented with all reasonable information necessary for the general meeting to assess the matter at hand.

Additionally, we have indemnified our board members and members of our executive management and employees, in relation to certain claims. We have not, however, indemnified our board members, executive management and employees, in respect of: (i) claims against a person pursuant to Danish law raised before the Danish Courts, except claims arising from the offer, sale and listing of the our securities in the United States and/or our subsequent status as a listed company in the United States, including in respect of our reports filed with or furnished to the U.S. Securities and Exchange Commission; (ii) claims against a person for damages and legal costs related to criminal and/or grossly negligent or willful acts or omissions committed by the indemnified person; (iii) claims against an indemnified person, which is attributable to the gaining or purported gaining of any profit or advantage to which the indemnified person or any related natural or legal person was not legally entitled; (iv) claims covered by insurance; (v) claims brought against the indemnified person by the Company or any subsidiary of the Company; and (vi) any sum payable to a regulatory authority by way of a penalty in respect of the indemnified person's personal non-compliance with any requirement of a regulatory nature howsoever arising. The indemnification is limited to a maximum amount per claim per person equivalent to the gross proceeds obtained by us in connection with our initial public offering of ADSs in the United States.

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

There is a risk that such indemnification will be deemed void under Danish law, either because the indemnification is deemed contrary to the rules on discharge of liability in the DCA (*Selskabsloven*) as set forth above, because the indemnification is deemed contrary to sections 19 and 23 of the Danish Liability and Compensation Act (*Erstatningsloven*), which contain mandatory provisions on recourse claims between an employee (including members of our executive management) and the company, or because the indemnification is deemed contrary to the general provisions of the Danish Contracts Act (*Aftaleloven*).

In addition, we provide our board members and executive management with directors' and officers' liability insurance.

C. Board Practices

We have a two-tier governance structure consisting of our board of directors and our executive officers which include executive management registered with the Danish Business Authority. The two bodies are separate and have no overlapping members. The business address of our board of directors and executive management is Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark.

Our board of directors is responsible for the overall and strategic management and proper organization of our business and operations and supervises our activities, management and organization. Our board of directors appoints and dismisses the members of our executive management, who are responsible for the day-to-day management of our business. In accordance with our articles of association, at our general meeting it is contemplated to reduce the number of directors to the statutory minimum of three members. The members of our board of directors elected at the general meeting are elected for a term of one year and may be re-elected.

As of the date of this Annual Report on Form 20-F, our board of directors is comprised of three members elected at the general meeting, and consists of the Chairman, the Deputy Chairman and one additional board member. Based upon information requested from and provided by each director concerning such director's background, employment and affiliations, including family relationships, our board of directors determined that all of our directors are "independent directors" as defined under current rules and regulations of the SEC. In making such determination, our board of directors considered whether any director has a material relationship with us that could compromise their ability to exercise independent judgment in carrying out their responsibilities.

Reference is made to the section 'Corporate Governance' on pages 11 – 14 of our Annual Report 2021 for information on board practices. The year of election of each member of the Board of Directors is included in the table on page 12 and is also included in the Board of Directors and Executive Management section on Pages 22 – 23 of the Annual Report 2021, along with details on the individual Board Members.

Committees of our Board of Directors

Our board of directors has three standing committees: an audit committee, a remuneration committee and a nomination committee. The board of directors decided to discontinue the Science Committee with effect from May 2022. Reference is made to section 'Corporate Governance' on pages 11 – 14 of our Annual Report 2021 for further details and discussion of our board committees.

D. Employees

As of December 31, 2021 we had 62 full time equivalent employees (FTEs). Most employees were in the areas of finance, legal, IT and administration (25%), clinical (22%) and commercial/pre-launch (20%). In 2020 we had 141 FTEs as at December 31, 2020. The number of employees was significantly reduced in 2021 by the restructuring of the Company in June 2021 following receipt of the CRL from FDA. Further reductions in employees occurred in early 2022 following the initiation of in-court restructuring proceedings. In March 2022, following the in-court restructuring proceedings, there were approximately 20 employees at Orphazyme. Following completion of the Sale of Assets in May 2022, all remaining Danish employees were transferred to KemPharm. As of the date of this Annual Report on Form 20-F, Orphazyme has two full-time employees.

E. Share ownership.

For information on our board of directors' and executive management's individual holdings of shares and restricted share units as of December 31, 2021 please see Note 4.5 'Remuneration of Board of Directors and Executive Management' and Note 4.5 'Related Parties' to our Audited Consolidated Financial Statements included under "Item 18 – Financial Statements" of this Annual Report on Form 20-F.

In the period from January 1, 2022 until the date of this Annual Report on Form 20F, no shares were sold or purchased by the members of our board of directors or executive management. The internal rules on trading in Orphazyme securities by members of the Board of Directors and Executive Management prohibits trading of Orphazyme shares for a period of one week before the end of a quarter and until one trading day after the day of publication of an annual or interim report.

ITEM 7. MAJOR SHAREHOLDERS AND RELATED PARTY TRANSACTIONS

A. Major shareholders

Except as specifically noted, the following information contains information with respect to the beneficial ownership of our ordinary shares as of May 31, 2022 by:

- each of our directors and executive officers;
- all of our directors and executive officers as a group; and
- each person known to us to beneficially own more than 5% of our ordinary shares.

Beneficial ownership is determined in accordance with the rules and regulations of the SEC. In computing the number of ordinary shares beneficially owned by a person and the percentage ownership of that person, we have included ordinary shares that the person has the right to acquire within 60 days of May 31, 2021, including through the exercise of any option, warrant or other right or the conversion of any other security. These ordinary shares, however, are not included in the computation of the percentage ownership of any other person.

The calculations of the percentage of beneficial ownership are based on 35,312,241 ordinary shares (including ordinary shares in the form of ADSs) issued and outstanding as of May 31, 2022.

Except as otherwise indicated, addresses of the directors, executive officers and named beneficial owners are in care of Orphazyme A/S, Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark.

Based on the latest major shareholder filings submitted to Orphazyme as required under EU Transparency Directive, LSP V Cooperatieve U.A., of Johannes Vermeer, Plein 9, 1071 DV Amsterdam, Netherlands holds directly and indirectly 2,710,829 shares of a nominal value of DKK 1 each in the Company.

Name of beneficial owner	Number of ordinary shares beneficially owned	Percentage of ordinary shares beneficially owned
5% or Greater Shareholders:		
LSP V Cooperatieve U.A.	2,710,829	7.7%
Orpha Pooling B.V. (1)	2,431,672	6.9%
LSP (directly) (2)	279,157	0.8%
Executive Officers and Directors:		
Anders Vadsholt (3)	159,517	*
Georges Gemayel (4)	100,809	*
Bo Jesper Hansen	143,234	*
Andrew Mercieca	0	*
All current directors and executive officer as a group (4 persons)	403,560	1.1%

* Represents beneficial ownership of less than one percent.

(1) Orpha Pooling B.V. is an investment vehicle 88.2% owned by LSP V Coöperatieve U.A. and 11.8% owned by ALS Invest 2 B.V. LSP Management B.V. is the director of LSP V Coöperatieve U.A. and the director of Orpha Pooling B.V. and exercises voting rights on behalf of Orpha Pooling B.V.

- (2) LSP holds directly 279,157 shares of a nominal value of DKK 1 each in the Company and LSP holds indirectly through Orpha Pooling B.V. 2,431,672 shares of a nominal value of DKK 1 each in the Company.
- (3) Consists of (i) 20,400 ordinary shares held directly by Anders Vadsholt and close family members, (ii) 121,556 ordinary shares held by Alpha Healthcare Investments ApS, and (iii) 17,561 ordinary shares from the exercise of Matching Shares after December 21, 2020. Alpha Healthcare Investments ApS is an investment company wholly-owned by Anders Vadsholt.
- (4) Consists of (i) 3,451 ordinary shares held directly by Georges Gemayel and (ii) 97,358 ordinary shares held by GFD Investments LLC. Georges Gemayel is the manager and the Gemayel Family 2016 Irrevocable Trust is the sole member of GFD Investments LLC.

None of our major shareholders have voting rights different to other shareholders.

There have been several notable changes in our major shareholders in the past year, including the following former major shareholders who reduced their positions to below 5% or exited the shares entirely: Sunstone Life Science Ventures A/S, Aescap Venture Management B.V. and Kurma Partners.

As of May 31, 2022, we estimate that approximately 1.08% of our ordinary shares are held by 16 record holders in the United States. The actual number of holders is greater than these numbers of record holders and includes beneficial owners whose ordinary shares or ADSs are held in street name by brokers and other nominees. This number of holders of record also does not include holders whose shares may be held in trust by other entities.

B. Related party transactions

For a discussion of related party transactions, please see Note 4.6 'Related Parties' to our Audited Consolidated Financial Statements included under "Item 18 – Financial Statements" of this Annual Report on Form 20-F.

C. Interests of experts and counsel.

Not applicable.

ITEM 8. FINANCIAL INFORMATION

A. Consolidated Statements and Other Financial Information

Financial Statements

See "Item 18—Financial Statements" which contains our Audited Consolidated Financial Statements prepared in accordance with IFRS.

Legal Proceedings

From time to time, we may be party to governmental, litigation, administrative or arbitration proceedings arising in the ordinary course of our business. Regardless of the outcome, any future governmental, litigation, administrative or arbitration proceedings could have an adverse impact on us because of defense and settlement costs, diversion of management resources and other factors, which may be material.

Busic v. Orphazyme A/S, et.al.

On July 9, 2021, a putative class action lawsuit, captioned *Busic v. Orphazyme A/S, et al.*, No. 21- cv-03640 (N.D. Ill.), was filed against us and certain of our current and former directors and officers in the U.S. District Court for the Northern District of Illinois. On September 13, 2021, the Court appointed a lead plaintiff for the putative class. On November 19, 2021, the lead plaintiff filed an amended complaint asserting claims under the Securities Act and the Exchange for alleged misrepresentations and/or omissions in our registration statement for our IPO of ADS in the United States and in subsequent public statements, which seeks an unspecified amount of damages on behalf of a putative class comprised of all persons and entities other than the defendants that purchased or otherwise acquired the ADS in connection with the IPO in September 2020 and/or between September 29, 2020 and November 4, 2021.

On January 21, 2022, the defendants filed a motion to dismiss the amended complaint. On March 11, 2022, the lead plaintiff filed his opposition to the motion to dismiss, as well as a motion to strike certain exhibits that the defendants submitted in connection with their motion to dismiss. On April 4, 2022, the defendants filed their opposition to the motion to strike. On April 11, 2022, the defendants filed their reply in support of the motion to dismiss. On April 14, 2022, the lead plaintiff filed his reply in support of the motion to strike. On April 15, 2022, the lead plaintiff filed a notice of supplemental authority in further opposition to the motion to dismiss. On April 18, 2022, the Court held a telephonic hearing on the motion to dismiss and motion to strike. We anticipate that the Court will issue rulings on the motions in July or August 2022. We believe that the claims raised in the suit are without merit and we intend to continue to vigorously defend the company.

Dividends

We have never declared or made any dividend payments on our ordinary shares for the last two financial years. The Company has limited ongoing operational business activities and, as of the date of this Annual Report on Form 20-F, does not expect to make dividend payments within the foreseeable future.

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.

B. Significant Changes.

Please refer to Note 1.6 ‘Significant Events after the Reporting Period’ to our Audited Consolidated Financial Statements included under “Item 18 – Financial Statements” of this Annual Report on Form 20-F.

ITEM 9. THE OFFER AND LISTING.

A. Offer and listing details.

Not applicable.

B. Plan of distribution.

Not applicable.

C. Markets.

Our ordinary shares have been trading on Nasdaq Copenhagen under the symbol “ORPHA” since November 6, 2017.

Our ADSs were listed on the Nasdaq Global Select Market under the symbol “ORPH” from September 28, 2020 through March 31, 2022, when we voluntarily delisted the ADSs.

D. Selling shareholders

Not applicable.

E. Dilution.

Not applicable.

F. Expenses of the issue.

Not applicable.

ITEM 10. ADDITIONAL INFORMATION.

A. Share capital.

Not applicable

B. Memorandum and articles of association.

Please refer to Exhibit 2.1 to this Annual Report on Form 20-F for a Description of Share Capital and Articles of Association.

C. Material contracts.

For information on our material contracts, please see “Item 4.B – Business Overview,” “Item 6. Directors, Senior Management and Employees,” and “Item 7.B - Related Party Transactions” of this Annual Report on Form 20-F.

D. Exchange controls.

There are no governmental laws, decrees, or regulations in Denmark (including, but not limited to, foreign exchange controls) that restrict the export or import of capital, or that affect the remittance of dividends,

interest or other payments to non-resident holders of the shares or the ADSs (please see below Taxation section for a discussion of Danish withholding tax on dividends). There are no limitations on the right of non-resident or foreign owners to hold or vote the shares or the ADSs imposed by the laws of Denmark or our Articles of Association solely due to the fact that such holders are non-residents or foreign owners.

E. Taxation.

The following is a general summary of certain Danish and United States federal income tax consequences relevant to holders of ordinary shares or ADSs in Orphazyme. The discussion is not intended to be, nor should it be construed as, legal or tax advice. The discussion is based on laws and relevant interpretations thereof in effect as of the date of this Annual Report on Form 20-F, 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 hold 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) 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, 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 our cash. Based on the composition of our income and valuation of our assets, including goodwill, and certain assumptions with respect to the characterization of our income as active or passive, 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 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). The Nasdaq Global Select Market is a qualified exchange for this purpose and, consequently, if the ADSs were regularly traded, the mark-to-market election would be available to a U.S. Holder. However, our ADSs are no longer listed on the Nasdaq Global Select Market or any other national securities exchange in the United States. 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 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

Distributions

As described in the section Item 8A Financial Information- Consolidated Statements and Other Financial Information - Dividends 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. The ADSs are no longer listed on the Nasdaq Global Select Market, which is an established securities market in the United States, or any other national securities exchange in the United States. 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.

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 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.

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 Annual Report on Form 20-F. 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.

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 57,200 (approximately \$9,300) and for cohabiting spouses, a total of DKK 114,400 (approximately \$18,500); and at a rate of 42% on share income exceeding DKK 57,200 (approximately \$9,300) and for cohabiting spouses over DKK 114,400 approximately (\$18,500) (all 2022 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 amounts mentioned above are exceeded. Dividends paid to Danish tax resident individuals are generally subject to withholding tax at the rate of 27%.

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 Annual Report on Form 20-F. We have included such website address as an inactive textual reference only.

The Danish Tax Ministry has in May 2020 announced that the Ministry will issue a proposal of a new structure, implying that the current refund system should be replaced by a specific upfront withholding taxation rate depending on the respective shareholder.

A 44% Danish withholding taxation applies to dividends paid on shares to a shareholder who 1) is resident in a country which is "blacklisted" by EU (i.e. at present American Samoa, Anguilla, Barbados, U.S. Virgin Islands, the Republic of Fiji, Guam, Republic of Palau, Panama, the Independent State of Samoa, Republic of Seychelles, Republic of Trinidad and Tobago and the Republic of Vanuatu); and 2) is holding or within the last five years has hold i) minimum 25% of the share capital of the issuing company; or ii) more than 50% of the voting rights on the share capital of the issuing company.

Danish Tax Resident Companies

Sale of Ordinary Shares or ADSs

For the purpose of taxation of sales of shares made 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 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 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. 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 almost all foreign corporate shareholders receiving taxable dividends distributed from Danish companies will be able to ask for a refund of minimum 5 percentage point 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 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?old=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 Annual Report on Form 20-F. We have included such website address as an inactive textual reference only.

The Danish Tax Ministry has in May 2020 announced that the Ministry will issue a proposal of a new structure, implying that the current refund system should be replaced by a specific upfront withholding taxation rate depending on the respective shareholder.

A 44% Danish withholding taxation applies to dividends paid to a shareholder which is holding Subsidiary Shares or Group Shares and is resident in a country which is "blacklisted" by EU (i.e. at present American Samoa, Anguilla, Barbados, U.S. Virgin Islands, the Republic of Fiji, Guam, Republic of Palau, Panama, the Independent State of Samoa, Republic of Seychelles, Republic of Trinidad and Tobago and the Republic of Vanuatu).

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.

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.

F. Dividends and paying agents.

Not applicable.

G. Statement by experts.

Not applicable.

H. Documents on display.

We are subject to the information reporting requirements of the Exchange Act applicable to foreign private issuers and under those requirements will file reports with the SEC. Those reports may be inspected without charge at the locations described below. As a foreign private issuer, we are exempt from the rules under the Exchange Act related to 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. In addition, we are not required under the Exchange Act to file periodic reports and financial statements with the SEC as frequently or as promptly as United States companies whose securities are registered under the Exchange Act. Nevertheless, we will file with the SEC an Annual Report on Form 20-F containing financial statements that have been examined and reported on, with an opinion expressed by an independent registered public accounting firm.

We maintain a corporate website at www.orphazyme.com. Information contained on, or that can be accessed through, our website does not constitute a part of this Annual Report on Form 20-F and our website address is included herein as an inactive textual reference only.

The SEC maintains a website (www.sec.gov) that contains reports, proxy and information statements and other information regarding registrants, such as us, that file electronically with the SEC. With respect to references made in this Annual Report on Form 20-F to any contract or other document of our company, such references are not necessarily complete and you should refer to the exhibits attached or incorporated by reference to this Annual Report on Form 20-F for copies of the actual contract or document.

I. Subsidiary information.

Not applicable.

ITEM 11. QUANTITATIVE AND QUALITATIVE DISCLOSURES ABOUT MARKET RISK.

Not applicable.

ITEM 12. DESCRIPTION OF SECURITIES OTHER THAN EQUITY SECURITIES.

A. Debt securities.

Not applicable.

B. Warrants and rights.

Not applicable.

C. Other securities.

Not applicable.

D. American depositary shares.

The Bank of New York Mellon, as depositary, registers and delivers our 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 will also represent 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. The depositary's office at which the ADSs are 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 Depositary Trust Company, or 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.

Fees and Expenses

The following table shows the fees and charges that a holder of our ADSs may have to pay, either directly or indirectly. The majority of these costs are set by the depository bank and are subject to change:

Fee	Service
\$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 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 the 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

ADS holders are 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.

See Exhibit 2.4 to this Annual Report on Form 20-F for additional information on the ADSs.

Recent Developments

In March 2022, we voluntary delisted ADSs representing our ordinary shares from the Nasdaq Global Select Market in the United States, which became effective March 31, 2022.

The Depository has announced that it will terminate the Deposit Agreement on July 6, 2022. Under the terms of the Deposit Agreement, ADS holders have until at least July 11, 2022 to surrender ADSs for delivery of the underlying ordinary shares. ADS holders who surrender ADSs for delivery of the underlying shares must pay a cable fee of \$17.50, a cancellation fee of up to \$0.05 per ADS surrendered and any applicable U.S. or local taxes or governmental charges. Subsequent to July 11, 2022, the Depository may attempt to sell the underlying shares. If the Depository has sold such shares, holders of ADSs must surrender such securities in order to obtain payment of the sale proceeds of the underlying ordinary shares, net of the expenses of sale, any applicable U.S. or local taxes or government charges and a cancellation fee of up to \$0.05 per ADS. To surrender ADSs, the address of the Depository is: The Bank of New York Mellon, 240 Greenwich Street, Depository Receipts Division – 8th Floor, Attention: Cancellation Desk, New York, NY 10286. Registered or overnight mail is the suggested method of delivering ADSs to the Depository. For Settlement specific inquiries, please contact DRsettlements@bnymellon.com.

PART II

ITEM 13. DEFAULTS, DIVIDEND ARREARAGES AND DELINQUENCIES.

None

ITEM 14. MATERIAL MODIFICATIONS TO THE RIGHTS OF SECURITY HOLDERS AND USE OF PROCEEDS.

None.

ITEM 15. CONTROLS AND PROCEDURES.

A. Disclosure Controls and Procedures.

Our management, with the participation of our Chief Executive Officer and Chief Financial Officer, has evaluated the effectiveness of our disclosure controls and procedures (as defined in Rules 13a- 15(e) and 15d- 15(e) under the Securities Exchange Act of 1934, as amended), as of the end of the period covered by this Annual Report on Form 20-F. Based on such evaluation, our principal executive officer and principal financial officer has concluded that as of such date, our disclosure controls and procedures were not effective as of December 31, 2021, because of the material weaknesses in our internal control over financial reporting which are detailed in “Item 15.B-Management’s Annual Report on Internal Control over Financial Reporting”.

B. Management’s Annual Report on Internal Control over Financial Reporting.

Our management is responsible for establishing and maintaining adequate internal controls over financial reporting (as defined in Rules 13a-15(f) and 15d-15(f) under the Exchange Act) and for the assessment of the effectiveness of our internal control over financial reporting. In making this assessment, it used the criteria set forth by the Committee of Sponsoring Organizations of the Treadway Commission (COSO 2013) in the Internal Control-Integrated Framework. Based on its assessment and those criteria, our management identified the following weakness in our internal control over financial reporting and therefore determined that our internal controls over financial reporting were not effective at the reasonable assurance level as of December 31, 2021.

As defined in the standards established by the U.S. Public Company Accounting Oversight Board, a “material weakness” is a deficiency, or combination of deficiencies, in internal control over financial reporting, such that there is a reasonable possibility that a material misstatement of our company’s annual or interim consolidated financial statements will not be prevented or detected on a timely basis.

Material weaknesses related to the lack of sufficiently designed and implemented internal controls.

In connection with our financial statement preparation process for the years ended December 31, 2021, under the supervision and with the participation of our Chief Executive Officer and Chief Financial Officer, we identified material weaknesses. A project plan to design and implement SOX compliant internal controls was developed and approved by the Audit Committee in Spring 2021. However, in June 2021, following the receipt of the Complete Response Letter from the U.S. Food and Drug Administration (FDA), we restructured the business, reprioritizing our resources. The implementation of the internal controls project was impacted and ultimately paused.

Therefore, the company did not appropriately implement components of the COSO framework and adequate internal controls over financial reporting had not been effectively designed and implemented as of December 31, 2021. The material weaknesses identified by us relate to (i) ineffective control environment driven by the lack of a sufficient number of trained professionals with an appropriate level of IFRS, SEC reporting and internal control knowledge, training and experience; (ii) insufficient control activities, as we did not have adequate formal documentation of certain policies and procedures, implementation of all required business process controls, including effective review process of key financial information, and sufficient documentation to evidence the design and operating effectiveness of the control activities; (iii) insufficient information and communication as we did not implement effective controls over the segregation of duties and certain information technology general controls for information systems that are relevant to the preparation of our financial statements; and (iv) insufficient monitoring activities, as we did not have the evidence to support

evaluation of the effectiveness of monitoring controls to ascertain whether the components of internal control are present and functioning. Thus, we are unable to evaluate whether other deficiencies, individually or in combination, result in a reasonable possibility that a material misstatement of our annual financial statements will not be prevented or detected on a timely basis.

Therefore, we have assessed that we lacked sufficient internal controls to support effective financial reporting as of December 31, 2021, which constitutes a material weakness.

As a result of the material weaknesses described above, management concluded our internal control over financial reporting was not effective at the reasonable assurance level as of December 31, 2021.

Management’s Plan for Remediation of Material Weaknesses

On May 31, 2022 we completed the sale of substantially all of the Company’s assets and business activities, including those relating to the development and approval of arimoclomol and the full claw back liability related to the French early access program, to KemPharm Denmark A/S (KemPharm), a wholly owned subsidiary of KemPharm Inc. Following completion of the Sale of Assets, we have limited ongoing operational business activities and, as of June 24, 2022, we had two employees.

Considering our limited operational business activities and resources the company will evaluate remediation activities that are commensurate with the size and operations of the company going forward.

C. Attestation Report of the Registered Public Accounting Firm.

This Annual Report on form 20-F does not include an attestation report of the company’s registered public accounting firm because we qualify as an emerging growth company under the JOBS Act.

D. Changes in Internal Control Over Financial Reporting.

Other than as discussed above, there has been no change in our internal control over financial reporting (as defined in Rule 13a-15(f) under the Exchange Act) during the period covered by this annual report that has materially affected, or is reasonably likely to materially affect, internal control over financial reporting.

Item 16. [Reserved]

ITEM 16A. AUDIT COMMITTEE FINANCIAL EXPERT

The audit committee is comprised of three non-executive directors, Andrew Mercieca (Chair), Bo Jesper Hansen and George Gemayel. Each member of the audit committee is an “independent director” as set forth in Rule 10A-3 under the Exchange Act. Our board of directors has determined that Andrew Mercieca is an “audit committee financial expert” as defined by applicable SEC rules.

ITEM 16B. CODE OF BUSINESS CONDUCT AND ETHICS

We have adopted a code of business conduct and ethics (see Exhibit 14.1) that is applicable to all our board members and employees. A copy of the code of conduct is also available on our website at www.orphazyme.com. The board of directors or committees thereof must approve any waivers from the provisions of the code of conduct for the benefit of a director or an executive officer. The board of directors, committees thereof or our compliance officer must approve any waivers from the provisions of the code of conduct for other employees. Any waivers from the provisions of the code of conduct will be disclosed in accordance with applicable securities laws.

ITEM 16C. PRINCIPAL ACCOUNTANT FEES AND SERVICES.

Our financial statements have been prepared in accordance with IFRS and are audited by EY Godkendt Revisionspartnerselskab (“EY”), of Copenhagen, Denmark, our independent registered public accounting firm registered with the Public Company Accounting Oversight Board in the United States (PCAOB Auditor ID 1757).

EY has served as our independent registered public accountant for each of the years ended December 31, 2019, December 31, 2020 and December 31, 2021.

The following table shows the aggregate fees billed to us, including some of our subsidiaries, for services rendered by EY Godkendt Revisionspartnerselskab.

DKK 000	2021	2020
Audit Fees	2,650	2,646
Audit-related Fees	783	3,820
Tax	—	—
All Other Fees	—	—
Total fees to principal accountants	3,433	6,466

Audit Fees

Audit Fees consist of fees billed for professional services rendered by EY for the audit of our annual consolidated financial statements or services that are normally provided by the accountant in connection with statutory and regulatory filings or engagements for those fiscal years.

Audit-Related Fees

Audit-related Fees consist of assurance and related services performed by EY that are reasonably related to the performance of the audit of our consolidated financial statements and are not reported under "Audit Fees".

In 2021, Audit-Related Fees consists of services provided by EY for other permitted services, including fees for work performed by EY in connection with the U.S. At -the-Market offering program in November 2021.

In 2020, this also includes fees for work performed by EY in connection with the U.S. listing in September 2020.

Tax Fees

Tax fees consist of aggregate fees for professional services rendered by the principal accountant for tax compliance, tax advice, and tax planning. No such fees were incurred for the financial periods ending December 31, 2020 and December 31, 2021.

All Other Fees

All other fees consist of aggregate fees for products and services provided by the principal accountant, other than the services outlined above. No such fees were incurred for the financial periods ending December 31, 2020 and December 31, 2021.

Pre-approval policies

The Audit Committee assesses and pre-approves all services provided by the statutory auditors. The pre-approval includes the type of service and a fee budget.

ITEM 16D. EXEMPTIONS FROM THE LISTING STANDARDS FOR AUDIT COMMITTEES.

Not applicable.

ITEM 16E. PURCHASES OF EQUITY SECURITIES BY THE ISSUER AND AFFILIATED PURCHASERS.

Not applicable.

ITEM 16F. CHANGE IN REGISTRANT’S CERTIFYING ACCOUNTANT.

Not applicable.

ITEM 16G. CORPORATE GOVERNANCE

As a foreign private issuer, we take advantage of the following exemptions from SEC reporting obligations:

- Exemption from filing quarterly reports on Form 10-Q or provide current reports on Form 8-K disclosing significant events within four days of their occurrence.
- Exemption from Section 16 rules regarding sales of our securities by insiders, which will provide less data in this regard than shareholders of U.S. companies that are subject to the Exchange Act.

We intend to take all actions necessary for us to maintain compliance as a foreign private issuer under the applicable corporate governance requirements of the Sarbanes-Oxley Act, the rules adopted by the SEC. Accordingly, our shareholders and holders of ADSs will not have the same protections afforded to shareholders of companies that are subject to the domestic reporting requirements of the SEC. We may utilize these exemptions for as long as we continue to qualify as a foreign private issuer. For an overview of our corporate governance principles, please visit the “Governance” section of our website (www.orphazyme.com) under the “Investor & Media” section.

ITEM 16H. MINE SAFETY DISCLOSURE

Not applicable.

ITEM 16I. DISCLOSURES REGARDING FOREIGN JURISDICTIONS THAT PREVENT INSPECTIONS

Not applicable.

PART III

ITEM 17. FINANCIAL STATEMENTS.

See “Item 18 – Financial Statements.”

ITEM 18. FINANCIAL STATEMENTS.

The financial statements filed as part of this Annual Report on Form 20-F begin on page F-1.

ITEM 19. EXHIBITS.

Exhibit Number	Description
1.1*	Articles of Association of Orphazyme A/S, as amended February 15, 2022
2.1*	Description of Share Capital and Articles of Association
2.2**	Deposit Agreement, dated September 28, 2020 among Orphazyme A/S, The Bank of New York Mellon, as depositary, and all owners and holders of American Depositary Shares issued thereunder (incorporated by reference to Exhibit 2.2 to the Annual Report on Form 20-F (File No. 001-39545) filed with the Commission on March 2, 2021).
2.3**	Form of American Depositary Receipt evidencing American Depositary Shares (included in Exhibit 2.2)
2.4**	Description of American Depositary Shares (incorporated by reference to the section entitled “Description of American Depositary Shares” in the Form 424B4 (File number 333-248607) filed with the Commission on September 28, 2020).
4.1*+	Asset Purchase Agreement by and among KemPharm, Inc., KemPharm Denmark A/S and Orphazyme in restructuring, dated May 15, 2022.
4.2**#	Orphazyme A/S - Long-Term Incentive Program (incorporated by reference to Exhibit 10.1 to the Form S-8 Registration Statement (File No. 333-249407) filed with the Commission on October 9, 2020)
4.3*#	Orphazyme A/S - Long-Term Incentive Program 2021 – U.S. Participants.
4.4*#	Orphazyme A/S –Long-Term Incentive Program 2021 – Danish and Swiss Participants.
4.5**#	Orphazyme A/S – Modifications to Long-Term Incentive Program 2021 (incorporated by reference to Exhibit 99.1 to the Form 6-K (File No. 001-39545) filed with the Commission on October 7, 2021).
4.6*#	Orphazyme A/S – Board Incentive Program 2021.
8.1**	Subsidiaries of the registrant (incorporated by reference to Exhibit 21.1 to the Form F-1 Registration Statement (File No. 333-248607) filed with the Commission on September 4, 2020)
12.1*	Certification of Principal Executive Officer Pursuant to Rules 13a-14(a) and 15d-14(a) under the Securities Exchange Act of 1934, as Adopted Pursuant to Section 302 of the Sarbanes-Oxley Act of 2002.
12.2*	Certification of Principal Financial Officer Pursuant to Rules 13a-14(a) and 15d-14(a) under the Securities Exchange Act of 1934, as Adopted Pursuant to Section 302 of the Sarbanes-Oxley Act of 2002.
13.1*	Certification of Principal Executive Officer Pursuant to 18 U.S.C. Section 1350, as Adopted Pursuant to Section 906 of the Sarbanes-Oxley Act of 2002.
13.2*	Certification of Principal Financial Officer Pursuant to 18 U.S.C. Section 1350, as Adopted Pursuant to Section 906 of the Sarbanes-Oxley Act of 2002.
14.1*	Orphazyme A/S – Code of Business Conduct and Ethics.

- 15.1**^ [Registrant's Annual Report 2021 \(incorporated by reference to Exhibit 99.2 to the Form 6-K \(File No. 001-39545\) filed with the Commission on June 9, 2022\).](#)
- 101.INS Inline XBRL Instance Document- the instance document does not appear in the Interactive Data File because its XBRL tags are embedded within the Inline XBRL document.
- 101.SCH Inline XBRL Taxonomy Extension Schema Document
- 101.CAL Inline XBRL Taxonomy Extension Calculation Linkbase Document
- 101.DEF Inline XBRL Taxonomy Extension Definition Linkbase Document
- 101.LAB Inline XBRL Taxonomy Extension Label Linkbase Document
- 101.PRE Inline XBRL Taxonomy Extension Presentation Linkbase Document
- 104 The cover page from the Company's Annual Report on Form 20-F for the year ended December 31, 2021, has been formatted in Inline XBRL.

* Filed herewith

** Previously filed

Indicates a management contract or any compensatory plan, contract or arrangement.

+ Schedules have been omitted pursuant to Item 601(a)(5) of Regulation S-K. A copy of any omitted schedule will be furnished to the Securities and Exchange Commission upon request; provided, however, that the parties may request confidential treatment pursuant to Rule 24b-2 of the Exchange Act for any document so furnished.

^ Certain of the information included within Exhibit 15.1, which is provided pursuant to Rule 12b-23(a) of the Securities Exchange Act of 1934, as amended, is incorporated by reference in this Form 20-F, as specified elsewhere in this Form 20-F. With the exception of the items and pages so specified, the Annual Report is not deemed to be filed as part of this Annual Report on Form 20-F.

SIGNATURES

The registrant hereby certifies that it meets all of the requirements for filing on Form 20-F and that it has duly caused and authorized the undersigned to sign this annual report on its behalf.

Orphazyme A/S

Date: June 28, 2022

By: _____
/s/ Anders Fink Vadsholt
Anders Fink Vadsholt
Chief Executive Officer and Chief Financial Officer

INDEX TO THE CONSOLIDATED FINANCIAL STATEMENTS

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Report of Independent Registered Public Accounting Firm

To the Shareholders and the Board of Directors of Orphazyme A/S

Opinion on the Financial Statements

We have audited the accompanying consolidated statements of financial position of Orphazyme A/S (the Company) as of December 31, 2021 and 2020, the related consolidated statements of profit or loss and other comprehensive income, changes in shareholders' equity and cash flows for each of the three years in the period ended December 31, 2021, and the related notes (collectively referred to as the "consolidated financial statements"). In our opinion, the consolidated financial statements present fairly, in all material respects, the financial position of the Company at December 31, 2021 and 2020, and the results of its operations and its cash flows for each of the three years in the period ended December 31, 2021, in conformity with International Financial Reporting Standards (IFRS) as issued by the International Accounting Standards Board (IASB).

Basis for Opinion

These financial statements are the responsibility of the Company's management. Our responsibility is to express an opinion on the Company's financial statements based on our audits. We are a public accounting firm registered with the Public Company Accounting Oversight Board (United States) (PCAOB) and are required to be independent with respect to the Company in accordance with the U.S. federal securities laws and the applicable rules and regulations of the Securities and Exchange Commission and the PCAOB.

We conducted our audits in accordance with the standards of the PCAOB. Those standards require that we plan and perform the audit to obtain reasonable assurance about whether the financial statements are free of material misstatement, whether due to error or fraud. The Company is not required to have, nor were we engaged to perform, an audit of its internal control over financial reporting. As part of our audits we are required to obtain an understanding of internal control over financial reporting but not for the purpose of expressing an opinion on the effectiveness of the Company's internal control over financial reporting. Accordingly, we express no such opinion.

Our audits included performing procedures to assess the risks of material misstatement of the financial statements, whether due to error or fraud, and performing procedures that respond to those risks. Such procedures included examining, on a test basis, evidence regarding the amounts and disclosures in the financial statements. Our audits also included evaluating the accounting principles used and significant estimates made by management, as well as evaluating the overall presentation of the financial statements. We believe that our audits provide a reasonable basis for our opinion.

/s/ EY Godkendt Revisionspartnerselskab

We have served as the Company's auditor since 2015.

Copenhagen, Denmark

June 28, 2022

2021 Consolidated Financial Statements

CONSOLIDATED STATEMENTS OF PROFIT OR LOSS AND OTHER COMPREHENSIVE INCOME

For the years ended December 31,

'DKK 000, except per share and share data	Note	2021	2020	2019
Net revenue	2.1	36,193	—	—
Research and development expenses	2.2, 2.3	(329,980)	(361,284)	(285,413)
General and administrative expenses	2.4	(339,516)	(247,250)	(50,541)
Operating loss		(633,303)	(608,534)	(335,954)
Financial income	2.7	12,432	2,444	316
Financial expenses	2.7	(10,609)	(29,071)	(7,359)
Loss before tax		(631,480)	(635,161)	(342,997)
Income tax benefit	2.8	4,941	1,915	5,500
Net loss for the year		(626,539)	(633,246)	(337,497)
Items that will be reclassified subsequently to the Statement of Profit or Loss:				
Exchange difference from translation of foreign operations		(302)	605	67
Total comprehensive loss		(626,841)	(632,641)	(337,430)
Weighted-average shares outstanding		34,924,702	28,366,469	20,024,692
Loss per share, basic and diluted (DKK)	4.3	(17.94)	(22.32)	(16.85)

The accompanying notes form an integral part of these consolidated financial statements.

CONSOLIDATED STATEMENTS OF FINANCIAL POSITION

As of December 31,

DKK 000			
ASSETS	Note	2021	2020
Non-current assets			
Intangible assets	3.1	2,152	12,454
Right-of-use assets	3.2	5,434	14,859
Property, plant, and equipment	3.3	2,985	4,687
Corporation tax receivable	2.8	2,750	2,750
Deferred tax assets	2.8	—	2,065
Prepayments and deposits	3.4	964	2,014
Total non-currents assets		14,285	38,829
Current assets			
Corporation tax receivable	2.8	7,229	5,500
Trade receivables	3.5	29,268	—
Prepayments and other receivables	3.4	20,192	51,235
Inventory	3.6	—	—
Cash	3.8	102,255	726,929
Total current assets		158,944	783,664
Total assets		173,229	822,493
EQUITY AND LIABILITIES			
	Note	2021	2020
Equity			
Share capital	4.2	34,952	34,698
Share premium		2,082,486	2,082,254
Other reserves		2,899	6,494
Accumulated deficit		(2,110,998)	(1,502,921)
Total equity		9,339	620,525
Non-current liabilities			
Borrowings	3.7	2,482	23,830
Lease liabilities	3.2	3,925	9,877
Discount and rebate liabilities	3.7	28,293	—
Other non-current liabilities	3.7	98	1,634
Total non-current liabilities		34,798	35,341
Current liabilities			
Provisions		—	—
Borrowings	3.7	30,983	33,349
Lease liabilities	3.2	2,578	3,657
Trade payables and accruals	3.7	57,524	72,135
Tax payables		584	4,159
Discount and rebate liabilities	3.7	7,900	—
Other liabilities	3.7	29,523	53,327
Total current liabilities		129,092	166,627
Total equity and liabilities		173,229	822,493

The accompanying notes form an integral part of these consolidated financial statements.

CONSOLIDATED STATEMENTS OF CHANGES IN SHAREHOLDERS' EQUITY

DKK 000

	Notes	Share capital	Share premium	Other reserves		Accumulated deficit	Total
				Foreign currency translation reserve	Share-based compensation – acquisition of intangible assets		
Balance as of December 31, 2018		<u>19,939</u>	<u>924,021</u>	<u>42</u>	<u>9,070</u>	<u>(564,823)</u>	<u>388,249</u>
Net loss for the year						(337,497)	(337,497)
Other comprehensive income (loss)				67		—	67
Total other comprehensive income (loss)		—	—	67	—	(337,497)	(337,430)
Transactions with owners:							
Capital increase in connection with issuance of bonus shares	3.1	26			(1,197)	1,171	—
Issuance of Matching Shares, net of costs	2.6	19					19
Share-based compensation expense	2.6					2,131	2,131
Total transactions with owners		45	—	—	(1,197)	3,302	2,150
Balance as of December 31, 2019		<u>19,984</u>	<u>924,021</u>	<u>109</u>	<u>7,873</u>	<u>(899,018)</u>	<u>52,969</u>
Net loss for the year						(633,246)	(633,246)
Other comprehensive income				605			605
Total other comprehensive income (loss)		—	—	605	—	(633,246)	(632,641)
Transactions with owners:							
Capital increase in connection with issuance of bonus shares	3.1	21			(2,094)	2,073	—
Capital increase in connection with exercise of RSUs	4.2	13	717				730
Capital increase related to directed issue and private placement	4.2	7,033	738,458				745,491
Transaction costs related to directed issue and private placement			(51,243)				(51,243)
Capital increase in connection with US listing	4.2	7,616	526,918				534,534
Transaction costs related to the US listing			(56,616)				(56,616)
Issuance of Matching Shares, net of costs	2.6	31					31
Share-based compensation expense	2.6					27,270	27,270
Total transactions with owners		14,714	1,158,233	—	(2,094)	29,343	1,200,196
Balance as of December 31, 2020		<u>34,698</u>	<u>2,082,254</u>	<u>714</u>	<u>5,780</u>	<u>(1,502,921)</u>	<u>620,525</u>
Net loss for the year						(626,539)	(626,539)
Other comprehensive income				(302)			(302)
Total other comprehensive income (loss)		—	—	(302)	—	(626,539)	(626,841)
Transactions with owners:							
Capital increase, issuance of Matching Shares, net of costs	4.2	170					170
Capital increase, Bonus Shares	3.1	22			(1,645)	1,623	—
Cash settlement of Bonus Shares	3.1				(1,648)		(1,648)
Capital increase, issuance of sign-on bonus shares to former CEO	4.2	58					58
Capital increase, exercise of RSUs	4.2	4	232				236
Share-based compensation expense	2.6					16,838	16,838
Total transactions with owners		254	232	—	(3,293)	18,461	15,654
Balance as of December 31, 2021		<u>34,952</u>	<u>2,082,486</u>	<u>412</u>	<u>2,487</u>	<u>(2,110,998)</u>	<u>9,339</u>

The accompanying notes form an integral part of these consolidated financial statements.

CONSOLIDATED STATEMENTS OF CASH FLOWS

For the years ended December 31,

DKK 000	Note	2021	2020	2019
Net loss		(626,539)	(633,246)	(337,497)
Reversal of non-cash items:				
Equity-settled share-based compensation expense	2.6	16,019	28,105	2,549
Depreciation and amortization	2.2, 2.4	18,111	5,200	3,803
Financial income		(12,432)	(2,444)	(316)
Financial expenses		10,608	29,071	7,359
Income tax benefit		(4,941)	(1,915)	(5,500)
Exchange rate adjustments		—	—	—
Change in working capital:				
Change in prepayments, deposits, and other receivables	3.4, 3.5	2,826	(33,662)	4,920
Change in trade payables, accruals, and other liabilities	3.7	(3,758)	76,424	(2,844)
Corporation taxes received		5,500	5,500	5,500
Corporation taxes paid		(1,738)	(1,431)	—
Interest received		37	45	388
Interest paid		(6,263)	(10,723)	(5,181)
Net cash used in operating activities		(602,571)	(539,076)	(326,818)
Investing activities				
Purchase of intangible assets	3.1	(902)	(2,736)	(508)
Purchase of property, plant, and equipment	3.3	(92)	(2,365)	(2,777)
Proceeds from sale of property, plant and equipment		1,040	—	—
Net cash used in investing activities		46	(5,101)	(3,285)
Financing activities				
Proceeds from borrowings	3.7	—	—	62,758
Repayment of borrowings		(25,657)	(10,535)	—
Repayment of lease obligations	3.2	(3,503)	(2,970)	(3,838)
Proceeds from issuance of shares		464	1,280,786	19
Cash settlement of Bonus Shares		(1,648)	—	—
Transaction costs related to issuance of shares		—	(107,859)	—
Net cash provided by financing activities		(30,344)	1,159,422	58,939
Net change in cash		(632,869)	615,245	(271,164)
Effects of changes in exchange rates		8,195	(11,904)	46
Cash at the beginning of the year		726,929	123,588	394,706
Cash at the end of the year		102,255	726,929	123,588

The accompanying notes form an integral part of these consolidated financial statements.

SECTION 1 Basis of preparation and significant accounting policies

1.1 CORPORATE INFORMATION

Orphazyme A/S (the “Company”) was, as of December 31, 2021 and until recently, involved in the research and development of novel therapeutics for the treatment of neurodegenerative rare diseases, including Niemann-Pick disease type C, or NPC.

The Company is headquartered in Copenhagen, Denmark and is publicly traded on Nasdaq Copenhagen. In September 2020, the Company listed American Depositary Shares (ADSs) on the Nasdaq Global Select Market. In March 2022, Orphazyme A/S voluntarily delisted the ADSs from the Nasdaq Global Select Market in the US.

In April 2018, a wholly-owned subsidiary, Orphazyme U.S., Inc., was incorporated in Delaware, USA and in March 2020, a wholly-owned subsidiary, Orphazyme Schweiz GmbH, was incorporated in Zug, Switzerland (together with Orphazyme A/S, “Orphazyme” or “the Group”).

In February 2022, Orphazyme announced that it had been notified by the Committee for Medicinal Products for Human Use (CHMP) of the European Medicines Agency (EMA) of a negative trend vote as part of the ongoing review of the Marketing Authorisation Application (MAA) for its investigational product candidate, arimoclomol, for the treatment of Niemann-Pick disease type C (NPC) following an oral explanation.

In light of the negative trend vote and considering Orphazyme’s financial situation at the time, the Board of Directors of Orphazyme filed a petition for an in-court restructuring of Orphazyme, which commenced in March 2022.

In May 2022, as part of the in-court restructuring proceedings, Orphazyme sold substantially all of its assets and business activities to KemPharm Denmark A/S (KemPharm), a wholly owned subsidiary of KemPharm Inc., a specialty pharmaceutical company focused on the discovery and development of novel treatments for rare central nervous system diseases, and KemPharm retained all of Orphazyme’s remaining Danish employees. The in-court restructuring proceedings were discontinued on May 30, 2022.

As of the date of publication of this annual report, Orphazyme has limited ongoing operational business activities and only two employees.

These consolidated financial statements were approved and authorized for issuance by the Board of Directors on June 7, 2022.

Information on COVID-19

As of 31 December 2021 and as of the date of publication of the annual report, there is no material impact directly related to COVID-19 on the Group’s consolidated financial statements, including the judgements and estimates applied. Specifically, following the sale of substantially all of its assets and business activities to KemPharm, Orphazyme is no longer involved in the conduct of clinical trials and has limited operational business activities and employees. Other parts of the business and operations may be adversely impacted by the effects of COVID-19, for example: the productivity of our staff; our relationships with vendors and other parties and significant disruption of global financial markets. We will continue to monitor the COVID-19 pandemic and its potential impact on our business and financials.

1.2 BASIS OF PREPARATION

The consolidated financial statements of the Group have been prepared in accordance with International Financial Reporting Standards (IFRS) as issued by the International Accounting Standards Board (IASB). All entities in the Orphazyme Group follow the same Group accounting policies.

The consolidated financial statements have been prepared on a going concern basis and are presented in Danish Kroner, or DKK, which is both the functional and presentation currency of the Company. The functional currency of Orphazyme US, Inc. is the US dollar (USD) and the functional currency of Orphazyme Schweiz GmbH is the Swiss Franc (CHF). Where indicated, amounts are rounded to the nearest thousand.

Materiality

The consolidated financial statements and parent company financial statements are prepared based on the concept of materiality, which considers both quantitative and qualitative factors. Items that are considered individually significant or are required under the minimum presentation requirements of IFRS are presented separately. If items are individually immaterial, they are aggregated with other items of similar nature in the financial statements or in the notes.

1.3 SIGNIFICANT ACCOUNTING POLICIES

A detailed description of accounting policies and significant accounting estimates and judgements related to specific financial statement line items is presented in each note to the relevant line item. The consolidated financial statements and parent company financial statements have been prepared on a historical cost basis except for share-based compensation and the embedded derivative in our borrowings, which is measured at fair value.

Principles of consolidation

The consolidated financial statements of the Group include the financial statements of the parent company, Orphazyme A/S (the “Parent Company”), Orphazyme US, Inc. and Orphazyme Schweiz GmbH, fully-owned subsidiaries over which the Parent Company has control. A company controls an entity when the company (i) is exposed to, or has rights to, variable returns from its involvement with the entity, (ii) has power over the entity (i.e. existing rights that give it the current ability to direct the activities of the entity), and (iii) has the ability to use its power to affect the returns of the entity. The Parent Company reassesses whether it controls an entity if facts and circumstances indicate that there are changes to one or more of the three elements of control. Consolidation of an entity begins when the Parent Company obtains control and ceases when the Parent Company has lost control of the entity. On consolidation, intercompany income and expenses, intercompany receivables, and payables, and unrealized gains and losses on transactions between the consolidated companies are eliminated.

Translation of foreign currencies

Items included in the financial statements of each of the Orphazyme entities are measured using the currency of the primary economic environment in which the entity operates, or functional currency. On initial recognition, transactions denominated in foreign currencies are recorded using the foreign exchange spot rate at the transaction date. For monetary assets and liabilities, differences arising between the foreign exchange spot rates at the transaction date and the date of settlement or period-end exchange rates are recognized in the Statement of Profit or Loss as financial income or financial expenses. On consolidation, the assets and liabilities of Orphazyme US, Inc. and Orphazyme Schweiz GmbH are translated from the subsidiary’s functional currency to DKK at the exchange rate in effect at the balance sheet date and the Statement of Profit or Loss and Other Comprehensive Income is translated from the subsidiary’s functional currency to DKK at the date of the underlying transaction or average exchange rate of the period if there are no significant fluctuations in exchange rate throughout the period. The exchange rate differences arising on translation for consolidation are recognized in other comprehensive income (loss).

Statement of cash flows

The statement of cash flows is presented using the indirect method and shows cash flows resulting from operating activities, investing activities, financing activities, and the Group’s cash at the beginning and end of the year, including any effects of exchange rate changes.

Cash flows used in operating activities converts items in the Statement of Profit or Loss from the accrual basis of accounting to the cash basis of accounting. Non-cash items such as foreign exchange gains and losses, depreciation, amortization, and changes in working capital are reversed from the net loss for the year and actual cash receipts and payments are included.

Cash flows from investing activities shows payments related primarily to the purchase of licenses and property, plant, and equipment.

Cash flows from financing activities shows proceeds from share issuance, borrowings, net of transaction costs, repayment of debt, and lease payments.

Segment information

Although Orphazyme established a US subsidiary in 2018 and a Swiss subsidiary in 2020, the Group is managed and operated as one business unit that is reflected in the internal reporting. No separate lines of business or separate business entities have been identified with respect to any product candidate or geographical market and no segment information is currently disclosed in the Group's internal reporting. For the years ended December 31, 2020 and 2019, the Group generated no revenue and for the year ended December 31, 2021 the Danish entity generated revenue which is disclosed in a separate note. For the years ended December 31, 2021, 2020 and 2019 all material non-current assets are located in Denmark.

1.4 SIGNIFICANT ACCOUNTING ESTIMATES AND JUDGEMENTS

The use of reasonable estimates and judgements is an essential part of the preparation of the consolidated financial statements and parent company financial statements. Given the uncertainties inherent in the Group's business activities, Management must make certain significant accounting estimates and judgements, which affect the application of accounting policies and therefore the reported amounts of assets, liabilities, expenses, and disclosures in the consolidated financial statements and parent company financial statements. The significant accounting estimates and judgements identified are those that have a significant risk of resulting in a material adjustment to the consolidated financial statements. Management bases its estimates on historical experience, assumptions, and information currently available and deemed to be reasonable at the time the consolidated financial statements are prepared. However, actual amounts may differ from the estimated amounts as more detailed information becomes available. Estimates and assumptions are reviewed on an ongoing basis and, if necessary, changes are recognized in the period in which the estimate is revised. Management has made significant accounting estimates and judgements in the following areas, which are further presented in each note to the relevant financial statement line items:

- Estimate of net revenue and clawback liability recognized using the 'expected value' method (Note 2.1)
- Estimate of research and development expenses associated with clinical trials (Note 2.3) and related prepayments (Note 3.4) and accruals (Note 3.6)
- Estimate of inputs and assumptions used in share-based compensation valuation models (Note 2.6)
- Estimate of the fair value of licenses (Note 3.1)
- Estimate relating to the incremental borrowing rate to measure lease liabilities (Note 3.2)
- Judgement and estimate relating to pre-launch drug product inventory (Note 3.5)
- Judgement regarding the recognition of deferred tax assets related to taxable losses to be carried forward (Note 2.8)
- Judgement regarding management's assessment of the company's ability to continue as a going concern (Note 4.1)

Please refer to the specific referenced notes for further information on the significant accounting estimates and judgements as well as assumptions applied.

1.5 NEW IFRS STANDARDS APPLICABLE TO THE GROUP

The Group applied for the first-time certain standards and amendments, which are effective for annual periods beginning on or after 1 January 2021 (unless otherwise stated). The Group has not early adopted any other standard, interpretation or amendment that has been issued but is not yet effective.

Interest Rate Benchmark Reform – Phase 2: Amendments to IFRS 9, IAS 39, IFRS 7, IFRS 4 and IFRS 16

The amendments provide temporary reliefs which address the financial reporting effects when an interbank offered rate (IBOR) is replaced with an alternative nearly risk-free interest rate (RFR). The amendments include the following practical expedients:

- A practical expedient to require contractual changes, or changes to cash flows that are directly required by the reform, to be treated as changes to a floating interest rate, equivalent to a movement in a market rate of interest
- Permit changes required by IBOR reform to be made to hedge designations and hedge documentation without the hedging relationship being discontinued
- Provide temporary relief to entities from having to meet the separately identifiable requirement when an RFR instrument is designated as a hedge of a risk component

These amendments had no impact on the consolidated financial statements of the Group. The Group intends to use the practical expedients in future periods if they become applicable.

Covid-19-Related Rent Concessions beyond 30 June 2021 Amendments to IFRS 16

On 28 May 2020, the IASB issued Covid-19-Related Rent Concessions - amendment to IFRS 16 Leases. The amendments provide relief to lessees from applying IFRS 16 guidance on lease modification accounting for rent concessions arising as a direct consequence of the Covid-19 pandemic. As a practical expedient, a lessee may elect not to assess whether a Covid-19 related rent concession from a lessor is a lease modification. A lessee that makes this election accounts for any change in lease payments resulting from the Covid-19 related rent concession the same way it would account for the change under IFRS 16 if the change were not a lease modification.

The amendment was intended to apply until 30 June 2021, but as the impact of the Covid-19 pandemic is continuing, on 31 March 2021, the IASB extended the period of application of the practical expedient to 30 June 2022. The amendment applies to annual reporting periods beginning on or after 1 April 2021. However, the Group has not received Covid-19-related rent concessions, but plans to apply the practical expedient if it becomes applicable within the allowed period of application.

New IFRS Standards not yet effective

The IASB has issued a number of new standards and updated some existing standards, which are effective for accounting periods beginning January 1, 2022 or later. Therefore, they are not incorporated in these consolidated financial statements and parent company financial statements. There are no standards presently known that are not yet effective and that would be expected to have a material impact on our current or future reporting periods.

1.6 SIGNIFICANT EVENTS AFTER THE REPORTING PERIOD

In January 2022, Christophe Bourdon resigned his position Chief Executive Officer of Orphazyme, to take on the role of CEO at another company. Anders Vadsholt was appointed CEO of Orphazyme, effective March 1, 2022 in addition to his position as Chief Financial Officer.

In February 2022, the Company issued new shares as a result of the utilization of the Company's U.S. At-the-Market Offering Program with Cowen and Company, LLC ("Cowen"). On February 11, 2022, a total of 360,000 ordinary shares of nominally DKK 1 each, represented by American Depositary Shares ("ADSs"), were issued by the Company and sold in the market by Cowen as the sales agent at market price as determined by the Company's Board of Directors in accordance with the authorization in article 3.1 of the Company's Articles of Association. Gross proceeds from the issue of new shares was USD835,668.00.

In February 2022, Andrew Mercieca was elected as new member of the Board of Directors at the Extraordinary General Meeting.

In February 2022, the Company was notified of a negative trend vote by the European Medicines Agency's (EMA) Committee for Medicinal Products for Human Use (CHMP), for its Marketing Authorisation Application (MAA) for arimoclomol for NPC, causing the Company to initiate in-court restructuring proceedings in March 2022.

During the restructuring proceedings, mass redundancies took place and employees were released from their duties to reduce payroll costs. The MAA filed with EMA was withdrawn in March 2022. In addition, the Company voluntarily delisted the ADSs from the Nasdaq Global Select Market in the US, which became effective in March 2022.

In May 2022, the Company announced that it had signed an agreement to sell substantially all of the Company's assets and business activities, including those relating to the development and approval of arimoclomol and the full claw back liability related to the French early access program, to KemPharm Denmark A/S (KemPharm), a wholly owned subsidiary of KemPharm Inc., for a total of USD 12.8 million in cash and assumed liabilities estimated to equal approximately USD 5.2 million (the Sale of Assets). KemPharm is a specialty pharmaceutical company focused on the discovery and development of novel treatments for rare central nervous system diseases. Under the terms of the agreement, KemPharm agreed to acquire substantially all of Orphazyme's assets and business activities, including those relating to the development and approval of arimoclomol, retain a majority of Orphazyme's remaining Danish employees, continue the early access programs with arimoclomol and pursue the potential approval of arimoclomol as a treatment option for NPC. The transaction has been considered a non-adjusting event and did not indicate any impairment of assets for the year ending December 31, 2021.

The Restructuring Proposal was adopted by the creditors and affirmed by the Danish Maritime and Commercial High Court May 30, 2022. The restructuring proceedings were discontinued on May 30, 2022 and by completing the Sale of Assets to KemPharm Denmark A/S on May 31, 2022. Orphazyme will pay its debts to the creditors in accordance with the Restructuring Proposal.

In May 2022, Stephanie Okey, Carolee Barlow and Martin Bonde stepped down from their positions as members of the Board of Directors, with the view to reduce the number of members of the Board of Directors to three members.

In June 2022, Orphazyme A/S repaid the full remaining loan to Kreos Capital VI (UK) Ltd. At the repayment all pledges and securities withdrew. All (undisputed and unconditional) debts related to the time prior to restructuring have been or will be paid in full in the coming months.

Orphazyme's current share-based compensation programs are expected to have a limited value, due to the decreasing share price and the disclosed Sale of Assets.

SECTION 2 Result of the Year

2.1 NET REVENUE

§ ACCOUNTING POLICIES

Orphazyme recognizes revenue when fulfilling its performance obligation by transferring control of promised goods or services to its customer, in an amount that reflects the consideration that the entity expects to receive in exchange for those goods or services. Revenue is recognized net of sales deductions, including discounts and rebates and revenue based taxes. Orphazyme recognizes revenue in accordance with IFRS 15 Revenue from Contracts with Customers and, as a result, follows the five-step model when recognizing revenue: 1) identifying a contract; 2) identifying the performance obligations; 3) determining the transaction price; 4) allocating the price to the performance obligations; and 5) recognizing revenue when the performance obligations have been fulfilled.

Net revenue comprises revenue from the sale of arimoclomol for the treatment of NPC under the remunerated early access compassionate use program ("nATU") in France. An early access compassionate use program is a program giving specific patients access to a drug, which is not yet approved for commercial sale.

Only drugs targeting serious or rare indications and for which there is currently no appropriate treatment are considered for early access compassionate use programs. Further, to be considered for the early access compassionate use program, the drug must have proven efficacy and safety and must either be undergoing price negotiations or seeking marketing approval.

Revenue is recognized when the drug products are sold to the customer, i.e., at the time when control over the drug product is transferred to the third-party customer. Under the French nATU, the manufacturer can set its own price for the drug products until a price agreement with the authorities is in place. Any excess in the price charged by the manufacturer compared to the price agreed with the health authorities once the drug product is approved in France must be repaid. The repayment is considered in the clawback liability. The liability is disclosed in note 3.7, which also explains the net to gross revenue.

All sales and distributions of arimoclomol are included in the service agreement with Clinigen Health Limited, who keep Orphazyme goods on a consignment stock until it is transferred to third-party customer.

The following table presents net revenue for the years ended December 31:

DKK 000	2021	2020	2019
Revenue by type			
Revenue from sale of goods	36,193	—	—
Revenue by partner			
Clinigen Health Limited	36,193	—	—
Geographical areas			
France	36,193	—	—

Estimate of net revenue and clawback liability recognized using the ‘expected value’ method

Accounting for net revenue and clawback liability requires determination of the most appropriate method for the expected final transaction price, which depends on the terms and conditions in the contracts with the French Health Authorities, and is subject to price negotiations with the French Health Authorities, following a market approval. This estimate also requires assumptions in respect inputs to the method, including current pricing of comparable marketed products within the rare disease area in France. Management has considered the expected final sales price as well as the price of similar drug products.

Management has based their initial sales prices on comparable drug products for arimoclomol, and the estimate of the clawback liability on the basis of the average cost of treatment which the Authorities are expected to cover.

In the estimate for clawback liability, Management have applied relevant available market data. Management’s assumptions are based on available relevant market information regarding average treatment cost of the most comparable drugs possible in the rare disease area in Europe. The Company is operating within rare disease therapeutic area where there is unmet treatment need and hence a limited number of comparable commercialized drugs products. The limited available relevant market information for directly comparable commercialized drugs within rare diseases increases the uncertainty in managements estimate. However, as mentioned in note 1.6 and 3.7, the clawback liability has been transferred to Kempharm at the carrying amount subsequently to the balance sheet date and on the basis of the amounts estimated by management.

2.2 RESEARCH AND DEVELOPMENT EXPENSES

§ ACCOUNTING POLICIES

Research expenses comprise of costs incurred during the very early stages of the drug development cycle from initial drug discovery until the drug is ready for administration to humans. The activities initially focus on identifying a single drug candidate with a profile that will support a decision to initiate development activities. Before selection of the final drug candidate, it is tested in animals to gather efficacy, toxicity and pharmacokinetic information.

Development expenses comprise costs incurred during the different phases of clinical drug development starting in phase 1, when the drug is administered to humans for the first time, through phases 2

and 3, and subsequent activities to obtain marketing authorizations, which will permit Orphazyme to eventually market and sell the drug products.

In line with industry practice, Orphazyme expenses all research costs. Development costs that do not meet the definition of an asset are also expensed as incurred. Due to regulatory and other uncertainties inherent in the development of new products, development costs do not qualify for capitalization as intangible assets until marketing approval by a regulatory authority is obtained or highly probable. In addition, pre-launch inventory costs are recognized under Research and Development (see Note 3.6). Cost of goods sold relating to products sold are included in the cost recognized under Research and Development.

Clinical trial costs are a significant component of research and development expenses. The Groups's clinical trials are performed by third-party Contract Research Organizations (CROs) and in order to estimate the amount of costs to charge to expense Management has developed expense models for each clinical trial based on estimates and assumptions.

The clinical trials generally have three distinctive stages.

- Start-up stage: initial setting up of the trial
- Treatment stage: site and trial management during the dosing period
- Wrap-up stage: close down and reporting of the trial

For each clinical trial for which information about the actual services delivered by the CRO are not provided on a regular current basis, the Group reviews the approved budgets for the clinical trial from the original executed agreements and categorizes the individual costs according to the three stages described above. The start-up activities, which include site recruitment, regulatory applications and investigator meetings, usually are performed reasonably uniformly throughout the start-up stage and the related costs are expensed ratably over this stage, which reflects the manner in which related services are rendered by the CRO.

The start-up stage is followed by the treatment stage, during which patients are dosed with the drug under study and results are monitored and measured. The costs incurred in this stage of the trial, which comprises the major portion of the total cost of the clinical trial, is mainly driven by the number of enrolled patients undergoing treatment. The Group estimates the costs attributable to activities performed in this stage of the trial on a per-patient basis. These costs are expensed over the treatment stage as patients are enrolled and undergo treatment, as reported by the CRO. After the last patient has been treated, the trial begins to be closed down and activities are performed related to data quality assurance and analysis. These activities are performed reasonably uniformly throughout the wrap-up stage and are expensed ratably over this last stage. Other costs, such as central laboratory costs and drug supply costs, are expensed as incurred, which is typically when the service has been rendered or the goods delivered.

CROs invoice the Group upon the occurrence of predetermined milestones (such as the enrollment of patients); however, the timing of these invoices and the Group's related payments often do not correspond directly to the level of performance of contracted activities. To the extent payments are made by the Group in advance of the related activities performed by the CROs, they are included in prepayments to vendors (see Note 3.4) and expensed in accordance with the expense model discussed above. To the extent that the payments are made by the Group following the performance of the related activities, the expense is reflected as an accrual (see Note 3.7) in accordance with the expense model.

Research and development expenses include costs arising from research and clinical development activities including employee costs for research and development personnel (i.e. salaries, bonuses, employer contributions to pension schemes, share-based compensation), legal expenses related to the protection, defense and enforcement of the Group's intellectual property, depreciation of right-of-use assets associated with facilities and equipment used for research and development purposes, as well as close down and restructuring

costs for clinical trial close-out costs and employee redundancies. The following table presents research and development expenses recognized for the years ended December 31:

DKK 000	2021	2020	2019
External costs	229,942	261,136	218,143
Employee costs (Note 2.5)	86,329	96,108	64,167
Depreciation, amortization and impairment (Notes 3.1, 3.2, 3.3)	13,709	4,040	3,103
Total research and development expenses	329,980	361,284	285,413

External costs comprise mainly expenses related to third party vendors providing services related to our research and development activities and facility costs. External costs in 2021 include expense from write-down of pre-launch inventory of DKK 80.2m (Note 3.6). Further, research and development expenses include costs relating to products sold under the French early access compassionate use program.

Included in total research and development expenses is an amount of DKK 34.5 million attributable to restructuring activities, i.e. clinical trial close-out costs and impairment of intangible assets (DKK 33.1 million) and employee redundancies (DKK 1.4 million).

Estimate of research and development expenses associated with clinical trials

Accounting for clinical trial costs related to activities performed by Contract Research Organizations (CROs) and other external vendors requires Management to make significant estimates regarding the timing of the expense recognition of these costs. The diverse nature of services being provided by CROs, the different compensation arrangements that exist for each type of service, and the limitation in the availability of information related to when certain clinical activities are performed add complexity to the estimation of the timing of expense recognition for services rendered by CROs and other vendors in connection with clinical trials. In addition, the COVID-19 pandemic has increased the estimation uncertainty of clinical trial costs, in particular the timing of the expense recognition due to potential delays in services being performed.

2.3 GOVERNMENT GRANTS

§ ACCOUNTING POLICIES

Government grants are recognized when there is reasonable assurance that the funding will be received, and all underlying conditions will be fulfilled. Income from grants is recognized in the Statement of Profit or Loss as a reduction of the related expenses being reimbursed in the period when the related expenses are incurred.

Government grants comprise research funding from the Danish government and the European Union. The grants received by Orphazyme provide reimbursement for certain project-specific research and development expenses, including wages and salaries. During the year ended December 31, 2020, Orphazyme was awarded a new government grant that provides for the reimbursement of one-third of research costs incurred in connection with the new molecular entity project over the next two years. The maximum amount to be reimbursed under this grant is DKK 5 million, which is to be reimbursed on a quarterly basis. During 2021, Orphazyme has received grant and other funding of DKK 1.4 million (2020: DKK 0.0 million; 2019: 0.1 million).

As of the year ended December 31, 2021, the total amount receivable under government grants is DKK 0.0 million (2020: DKK 0.1 million) and is classified as Current Other Receivables in the Statement of Financial Position, as all remaining funding from grants is receivable within the next year (Note 3.4). One grant had been paid to Orphazyme in advance in a previous year and income related to this grant had been deferred in 2020 (DKK 0.0 million) and 2019 (DKK 0.1 million). The deferred income was presented in the Statement of Financial Position as current other liabilities (Note 3.7). As of December 31, 2021, all income related to that grant has been recognized.

All the grants received are subject to repayment clauses upon breach of conditions to maintain the terms under which the grant was awarded. Orphazyme has complied with and anticipates continuing to fully comply with all such terms.

With reference to note 1.6, significant events have occurred after the reporting period, which is relevant in the understanding of this item. The events disclosed in note 1.6 have not been taking into account in the current reporting period.

2.4 GENERAL AND ADMINISTRATIVE EXPENSES

§ ACCOUNTING POLICIES

General and administrative expenses include salaries for our employees working on pre-launch preparation activities as well as administrative employees and Executive Management; remuneration to the Board of Directors; share-based compensation costs related to such employees and the Board; depreciation of right-of-use assets associated with facilities not used for research and development purposes, investor relations, and accounting and legal fees. In addition, we include costs incurred in pre-launch preparation activities such as market access, marketing, and medical affairs in general and administrative expenses, including the costs associated with the Early Access Program for NPC in the U.S., tradename costs, market and pricing studies and related costs.

The following table presents general and administrative expenses for the years ended December 31:

DKK 000	2021	2020	2019
External costs	192,913	118,971	23,847
Employee costs (Note 2.5)	142,201	127,120	25,995
Depreciation (Notes 3.2 and 3.3)	4,402	1,159	699
Total general and administrative expenses	339,516	247,250	50,541

External costs comprise expenses related to third party vendors providing assistance with establishing a commercial organization and the escalation of launch preparation activities, including hiring a commercial team in our subsidiaries in the U.S. and Switzerland, market access activities, and medical affairs activities to further engage with the scientific community through communication and education programs. In addition, external costs comprise expenses related to administrative services such as legal and accounting support, IT, and investor relations. In 2021 costs as well comprise expenses related to restructuring activities, i.e. employee redundancies and termination of onerous contracts.

Included in these amounts is DKK 12.1 million attributable to restructuring activities, i.e. employee redundancies (DKK 11.4 million) and termination of onerous contracts (DKK 0.7 million).

2.5 EMPLOYEE COSTS

§ ACCOUNTING POLICIES

Employee costs primarily comprise salaries, bonuses, social security contributions, share-based compensation, vacation and sick leave as well as the employer portion of pension contributions. In addition, severance payments or termination benefits are also included under Employee Costs. The cost of these benefits is recognized as an expense as services are received. All employee pension plans are defined contribution plans and not defined benefit plans.

Employees are eligible to receive a discretionary bonus subject to certain predefined and individual goals as determined by the Board of Directors. Employees are also eligible to receive an extraordinary bonus at the discretion of the Board of Directors.

The following table presents Employee Costs, including remuneration to the Board of Directors and Executive Management, for the years ended December 31, 2021, 2020 and 2019. Refer to note 4.5 for more discussion on remuneration of Board of Directors and Executive Management.

DKK 000			
Employee costs	2021	2020	2019
Salaries	167,316	131,606	68,719
Cash bonus	16,663	40,481	8,707
Share-based compensation (Note 2.6)	15,576	27,258	2,405
Pension	12,963	11,313	5,561
Other social security contributions	4,113	5,172	875
Other staff costs	8,066	3,083	862
Total employee costs excluding board remuneration	224,696	218,913	87,129
Board remuneration (Note 4.5)	3,391	3,469	2,888
Board share-based compensation (Note 2.6 and Note 4.5)	443	846	145
Total employee costs	228,530	223,228	90,162
<i>Recognized as follows in the statement of Profit or Loss</i>			
Research and development expenses	86,329	96,108	64,167
General and administrative expenses	142,201	127,120	25,995
Total employee costs	228,530	223,228	90,162
Average number of full-time employees	130	117	74
Year-end number of full-time employees	62	141	86

2.6 SHARE-BASED COMPENSATION COSTS

§ ACCOUNTING POLICIES

Equity-settled awards

Shares awarded under the long-term incentive program (“LTIP”) are equity-settled awards. The fair value of these awards is determined at the date of grant, resulting in a fixed fair value at grant date that is not adjusted for future changes in the fair value of the awards that may occur over the service period. The fair value of the LTIP awards has been determined using the Monte-Carlo model. Further details of the valuation models are presented below.

The fair value of equity-settled awards with service conditions and non-market performance conditions is recognized as compensation expense pro rata over the service period to the extent such awards are estimated to vest. The compensation expense is recognized together with a corresponding increase in equity over the period in which the performance and/or service conditions are fulfilled. The cumulative expense for the Group’s share-based compensation awards recognized at each reporting date until the vesting date reflects the extent to which the vesting period has expired and Management’s best estimate of the number of instruments that will ultimately vest. The expense or credit in the Statement of Profit or Loss for a period represents the movement in cumulative expense recognized as at the beginning and end of that period.

In the event that equity instruments are granted conditionally upon an equal number of equity instruments granted in prior periods not being exercised, they are treated as a new grant for the current period and a modification of the equity instruments granted in the prior period.

When the terms of an equity-settled award are modified, the minimum expense recognized is the grant date fair value of the unmodified award, provided that the original terms of the award are met. An additional expense, measured as at the date of modification, is recognized for any modification that increases the total fair value of the share-based payment transaction, or is otherwise beneficial to the employee. Where an award is cancelled by the entity or by the counterparty, any remaining fair value of the award is expensed immediately in the Statement of Profit or Loss.

Cash-settled awards

The phantom share-based incentive programs established by the Group are settled in cash and are treated as cash-settled awards. Similarly, as the Restricted Share Units (RSU) awards to the board of directors may be settled in cash or in shares at the choice of the participant, they are also treated as cash-settled awards. If the RSUs are ultimately exercised by the holder and settled in equity, the amount accrued as a liability is settled by reversing it into equity.

A liability is recognized for the fair value of cash-settled awards, measured initially and at each reporting date up to and including the settlement date, with changes recognized through profit or loss at each reporting date. The fair value is expensed over the period until vesting date with recognition of a corresponding liability. The fair value is determined using the Monte-Carlo model, further details of which are presented below. The fair value of the cash-settled awards, which vest subject to obtaining a specified share price (i.e. market condition), is reported as compensation expense regardless of whether the share price condition is met if all other vesting conditions are met. For these awards, fair value is determined taking into account the probability of meeting the share price target. No expense is recognized for awards that do not ultimately vest. If the RSUs are finally exercised, the related liability is reclassified as equity.

Estimate of inputs and assumptions used in share-based compensation valuation models

Estimating the fair value of the Group's share-based compensation programs requires determination of the most appropriate valuation model, which depends on the terms and conditions of the respective award. This estimate also requires making assumptions to determine the most appropriate inputs to the valuation model, including the expected life of the award, expected volatility, dividend pay-out ratio, and risk-free interest rate.

With reference to note 1.6, significant events have occurred after the reporting period, which is relevant in the understanding of this item. The events disclosed in note 1.6 have not been taking into account in the current reporting period.

All references to share price relate to the Company's share price on Nasdaq Copenhagen.

a) Long-term incentive program (equity-settled)

In connection with the completion of the Company's initial public offering (IPO) on Nasdaq Copenhagen in November 2017, the Executive Management and Key Employees were offered to subscribe for Offer Shares ("Investment Shares") at the Offer Price for a maximum amount corresponding to approximately 15% (CMO) and 20% (CEO, CFO, and CSO) of their respective current annual base salaries.

Under the post-IPO long-term incentive program (2017 LTIP), the Executive Management as well as certain Key Employees of Orphazyme have subscribed to 14,875 ordinary shares (Investment Shares) at the offer price of DKK 80. In April 2018, a Key Employee subscribed to 4,300 Investment Shares at the then-current market price of DKK 67.5.

The participants in the 2017 LTIP may be allocated a number of shares in Orphazyme ("Performance Shares") at a price per Performance Share of DKK 1 at the end of a vesting period of four years from Orphazyme's first day of trading and official listing on Nasdaq Copenhagen. The number of Performance Shares should be proportional to the potential increase in the price of Orphazyme's shares at the time of exercise compared to the offer price. The potential increase in the price of Orphazyme's shares was calculated as the volume-weighted average share price as quoted on Nasdaq Copenhagen during the 10 trading days preceding the vesting date. The maximum allocation of Performance Shares was six shares for the CEO and four shares for the other participants multiplied by the number of Investment Shares subscribed for in connection with the IPO. Performance Shares was allocated on a linear scale with maximum allocation triggered by an 80% increase in share price, whereas no Performance Shares would be allocated if the price of Orphazyme's shares has increased 20% or less at the end of the vesting period. Among other things, vesting was also subject to the participants having maintained ownership of their Investment Shares and continued employment. Based on the number of Investment Shares subscribed for, a total maximum of 86,700 Performance Shares could be issued at the end of the vesting period. The vesting period ended in November 2021 with no performance shares granted as the minimum increase of 20% over the vesting period was not met.

In addition, the participants were allocated a number of shares in Orphazyme (“Matching Shares”) at a price per Matching Share of DKK 1 in connection with the first anniversary of the subscription date of the Investment Shares. The number of Matching Shares was equal to the number of Investment Shares subscribed for and vesting was subject to the participants having maintained ownership of their Investment Shares and continued employment during the one-year vesting period. By March 2019, all 19,175 Matching Shares under the 2017 LTIP vested in full and were issued against a nominal payment of DKK 1 per share.

In July 2019, the Company initiated a 2019 long-term investment program (2019 LTIP) for the Executive Management and certain Key Employees with the same terms and conditions as the 2017 LTIP, i.e. Matching Shares vesting over one year and Performance Shares vesting over four years. In July 2020, 31,250 matching shares fully vested and were issued against a nominal payment of DKK 1 per share. The maximum number of Performance Shares that can vest in July 2023 as part of the 2019 LTIP is 125,000.

In July 2020, the Company initiated a 2020 long-term investment program (2020 LTIP) for the Executive Management and certain other employees with the same terms and conditions as the 2017 LTIP and the 2019 LTIP. However, in case of termination of a participant’s employment and designation as a Good Leaver, the right to receive Matching Shares and Performance Shares will be prorated and calculated through the date of notice of termination. During 2020, awards were granted on four different grant dates shown in the table below. Matching Shares for all awards granted under the 2020 LTIP was fully vested on January 1, 2021. The maximum number of Performance Shares that can vest in January 2024 as part of the 2020 LTIP is 489,757.

In April 2021, the Group initiated a 2021 new long-term share-based incentive program (original 2021 LTIP) for the Executive Management and other employees. The LTIP grants comprise Restricted Share Units (“RSUs”) and Performance Share Units (“PSUs”) which entitle the participants, subject to vesting occurring, to be allocated a number of shares in the Company, equivalent to the number of vested RSUs and/or PSUs, against payment of the par value of each share. The RSUs will have a total vesting period of three years (beginning on January 1 or July 1 in 2021) and with one third of the granted RSUs vesting on each January 1 or July 1 in the following three financial years. The PSUs will have a total vesting period of three years (beginning on January 1 or July 1 in 2021) and with the granted PSUs vesting, in whole or in part, on January 1 or July 1 in the third year. Vesting of RSUs is not conditional upon achieving any financial or non-financial targets, whereas vesting of PSUs is conditional upon an increase in the quoted share price of the Company’s shares, while vesting of both RSUs and PSUs is conditional upon the participant remaining employed with a group member throughout the total vesting period. However, in case of termination of a participant’s employment and designation as a Good Leaver, the right to receive vested RSUs or PSUs will be prorated and calculated through the date of release of the Participant’s work obligations. The vested RSUs and PSUs can only be exercised within four months after the expiration of the total vesting period. However, the delivery period may be extended to the next open trading window in certain circumstances. The original LTIP were expected to comprise up to 607,460 shares in total.

In October 2021, the Group initiated a modified 2021 long-term share-based incentive program (modified 2021 LTIP) for the Executive Management and other employees. The terms of the modified LTIP are the same as the LTIP that was implemented in April 2021, however, the number of RSUs and PSUs and the applicable performance target for the PSUs were reset, calculated based on a share price equal to DKK 31.94 per share, corresponding to the volume weighted average share price of the Company’s shares as quoted on Nasdaq Copenhagen during the ten (10) trading days from September 1, 2021. The exercise of the RSUs and PSUs to be granted under the modified LTIP is conditional upon the participant not exercising his or her RSUs or PSUs granted in April 2021, which will subsequently lapse and no longer be exercisable, and are therefore considered replacement equity instruments for the cancelled equity instruments. The fair value of the originally granted RSUs and PSUs at the date of the modification was determined to be DKK 24.72 and DKK 7.70, respectively. The incremental fair value, calculated based on the number of modified awards granted multiplied with the modified unit fair value less the fair value of the original LTIP granted remeasured at the modification date, will be recognised as an expense over the period from the modification date to the end of the vesting period. The expense for the original LTIP grant will continue to be recognised as if the terms had not been modified. In connection with the modified LTIP, the members of Executive Management received an extraordinary grant of RSUs and PSUs corresponding to 100% of the grant under the modified LTIP and on the same terms as the modified LTIP, and the sign-on RSUs granted to the CEO in April 2021 were also reset after the same principles as the modified LTIP but with immediate vesting upon grant. The modified LTIP including the other share-based retention grants to the Executive Management are expected to comprise up to 595,916 shares in total.

The fair value of RSU awards was estimated using a Black Scholes option valuation model, whereas all other LTIP awards were estimated using a Monte-Carlo simulation model at the respective grant dates, considering the terms and conditions on which the awards were granted.

The risk-free interest rate has been estimated based on Danish government bonds with similar maturities. Since November 2020, expected volatility has been determined based on the Group's own historical volatility, as the Company has been publicly traded for three years. Before November 2020, expected volatility was determined based on the historical volatility of comparable listed companies. The Group does not plan to pay out dividends in the foreseeable future.

The following table presents the fair value of the shares granted in the last three years under each program and the inputs used in the valuation models at the respective grant dates:

Program	2021 RSU	2021 PSU	2021 RSU	2021 PSU
Grant date	Oct 2021	Oct 2021	Apr 2021	Apr 2021
	(modified)	(modified)	(original)	(original)
Fair value at the measurement date (DKK 000)	24.72	12.56	58.04	20.02
Dividend yield (%)	-	-	-	-
Expected volatility (%)	98.6%	98.6%	55.6%	55.6%
Risk-free interest rate (%)	(0.61%)	(0.61%)	(0.53%)	(0.53%)
Expected life of awards (years)	0.23-2.23	2.23	0.69-2.69	2.69
Weighted average share price (DKK)	25.70	25.70	59.05	59.05

Program	2020 LTIP	2020 LTIP	2020 LTIP	2020 LTIP
Grant date	Dec 2020	Oct 2020	Sep 2020	Aug 2020
Fair value at the measurement date (DKK 000)	446	4,464	1,482	44,126
Dividend yield (%)	-	-	-	-
Expected volatility (%)	45.9%	56.5%	56.1%	55.4%
Risk-free interest rate (%)	(0.66%)	(0.60%)	(0.59%)	(0.54%)
Expected life of awards (years)	3.06	3.24	3.24	3.35
Weighted average share price (DKK)	56.30	68.50	83.50	90.10

Program	2019 LTIP
Grant date	Aug 2019
Fair value at the measurement date (DKK 000)	6,214
Dividend yield (%)	—
Expected volatility (%)	51.8%
Risk-free interest rate (%)	(0.70%)
Expected life of awards (years)	3.42
Weighted average share price (DKK)	62.6

The following table presents the weighted average remaining contractual life in years of the Performance Shares of the LTIP awards outstanding at December 31 for the respective year presented. Matching Shares under all of the LTIP programs were fully vested as of January 1, 2021 :

Program	2021	2020
2020 LTIP	2.0	3.0
2019 LTIP	1.7	2.7

The exercise price for each LTIP award outstanding as of December 31, 2020 was DKK 1 (2019: DKK 1; 2018: DKK 1).

The table below summarizes the activity related to the LTIP awards for the years ended December 31:

DKK 000	Executive Management	Key Employees	Total Awards	Awards exercisable
Outstanding at December 31, 2019	6,250	25,000	31,250	
Granted	52,865	119,623	172,488	
Exercised	(6,250)	(25,000)	(31,250)	
Expired	—	—	—	
Forfeited	—	(937)	—	
Outstanding at December 31, 2020	52,865	118,686	172,488	172,488
Granted				
Exercised	(52,865)	(118,686)	(172,488)	
Expired	—	—	—	
Forfeited	—	—	—	
Outstanding at December 31, 2021				

For the year ended December 31, 2021, DKK 17.1 million (2020: DKK 17.9 million; 2019: DKK 2.1 million) was recognized as compensation expense related to the LTIP awards. Of the total expense, DKK 6.8 million (2020: DKK 8.2 million; 2019: DKK 0.7 million) is attributed to the Executive Management.

b) Phantom share-based incentive program (cash-settled)

In June 2018, Orphazyme introduced a four-year phantom share-based incentive program (the “2018 Phantom Shares Program”) for all employees other than the Executive Management and Key Employees under the LTIP. Programs with similar terms and conditions were initiated in August 2019 (2019 Phantom Shares Program) and December 2020 (2020 Phantom Shares Program), respectively.

The Phantom Shares Programs are based on the share price of the Company and entitles the participants to a cash bonus if there has been an increase of at least 20% in Orphazyme’s share price compared to the entry price at the grant date. The Phantom Shares Programs will not have any dilutive effect on the shareholders of Orphazyme as the phantom shares do not constitute or qualify for actual shares in Orphazyme.

The overall objectives of the Phantom Shares Programs are (i) to retain qualified employees, (ii) to create long-term incentive for the participants, and (iii) to align the interests of the employees with those of Orphazyme’s shareholders. Each employee participating in the program earns the right to a certain number of phantom shares per month, depending on the employee’s position. Subject to any adjustments to the Phantom Shares Programs made by the Board of Directors due to, for example, changes in Orphazyme’s share capital structure or other significant events, each employee will be eligible to receive up to a total of 144 or 288 phantom shares under the program. By the end of each calendar year of the four-year program, the participants will have earned phantom shares free of charge.

The entry price per phantom share for the 2018 and 2019 Phantom Programs was DKK 61 and for the 2020 Phantom Program was DKK 71.2. The entry prices were calculated on the basis of the volume-weighted average closing price of Orphazyme’s share on Nasdaq Copenhagen during a period of 10 trading days prior to the introduction of the respective Phantom Shares Program. The phantom shares will automatically be settled in cash at the end of January 2023 for the 2018 Phantom Shares Program, at the end of January 2024 for the 2019 Phantom Shares Program and at the end of January 2024 for the 2020 Phantom Shares Program by subtracting the entry price per share from the market price per share and multiplying the change by the total number of granted phantom shares, presuming the market-based condition (share price increase by 20%) is met. The market price per share will be based on the volume-weighted average closing price of Orphazyme’s shares on Nasdaq Copenhagen during a period of 10 trading days prior to the settlement of the phantom shares.

The employee’s cash award for each program is capped and cannot exceed a gross amount of DKK 37,500 or DKK 75,000 per employee per program, depending on the number of phantom shares allocated to the respective employee under the program. Based on the number of participants in the Phantom Shares Programs as of December 31, 2021 and 2020, the programs are expected to consist of up to a total of 41,351 phantom shares.

As of December 31, 2020, all phantom shares granted under the Phantom Shares Program were only granted to employees of Orphazyme. No phantom shares were forfeited or expired, and none of the phantom shares were eligible for exercise.

As the Phantom Shares Programs are cash-settled, the fair value of the phantom shares granted as part of the program is estimated at each reporting date. For the year ended December 31, 2021, an aggregate amount of DKK (0.3) million (2020: DKK 0.1 million; 2019: DKK 0.3 thousand) was recognized as compensation expense related to the Phantom Shares Programs, with a corresponding amount recognized as a non-current liability as the earliest settlement is in January 2023 (Note 3.7). The risk-free interest rate has been estimated based on Danish government bonds with similar maturities. Since November 2020, expected volatility has been determined based on the Company's own historical volatility, as the Company has been publicly traded for three years. Before November 2020, expected volatility was determined based on the historical volatility of comparable listed companies.

The following table presents the inputs to the Monte-Carlo model used to estimate the fair values of the phantom shares as of year-end, when the cash-settled programs are re-valued:

Valuation date:	December 31, 2021			December 31, 2020		
	2020 Program	2019 Program	2018 Program	2020 Program	2019 Program	2018 Program
Fair value at valuation date (DKK 000)	40	48	12	406	293	160
Dividend yield (%)	—	—	—	—	—	—
Expected volatility (%)	77.4%	101.1%	101.1%	47.1%	47.3%	54.3%
Risk-free interest rate (%)	(0.48%)	(0.60%)	(0.60%)	(0.59%)	(0.61%)	(0.61%)
Expected life of awards (years)	3.08	2.08	1.08	4.08	3.08	2.08
Weighted average share price (DKK)	17.16	17.16	17.16	67.10	67.10	67.10

c) Restricted Share Units (cash-settled)

According to the terms and conditions of the Restricted Share Units program (RSU), directors may annually be granted a number of RSUs with a value corresponding to up to 50% of the participant's fixed annual base fee as member of the Board of Directors, not including committee membership fees. The value is calculated on the basis of the volume-weighted average share price of Orphazyme's shares as quoted on Nasdaq Copenhagen during the ten trading days preceding the grant date. The RSUs vest from the grant date to the date of the next annual general meeting. Upon vesting, RSUs may be exercised within a period of twelve months from vesting (Exercise Period) at a price corresponding to the volume-weighted average share price during the ten trading days preceding the grant date (Exercise Price). In the event of a participant's resignation from the Board of Directors, any unvested RSUs will lapse without any rights of compensation. A decision not to be re-elected is not a resignation from the Board of Directors.

The RSUs are classified as a cash-settled program, as the Board of Directors may choose to settle any vested RSUs in cash. In such event, the cash settlement amount is based on the difference between the Exercise Price and the volume-weighted average share price as quoted on Nasdaq Copenhagen during the ten trading days preceding the first day of the Exercise Period.

In August 2019, Restricted Share Units (2019 RSUs) were granted to members of the Board of Directors. During 2021 and 2020 certain board members exercised their RSUs. As these RSUs were not cash-settled, the corresponding liability of DKK 35 thousand and DKK 156 thousand, respectively, was reversed into equity and treated as equity-settled. The remaining 1,927 RSUs expired in March 2021 resulting a positive impact on the Statement of Profit or Loss of DKK 38 thousands.

In March 2020, the 2020 RSU program was announced, granting the Board of Directors an aggregate of 15,177 RSUs under similar terms and conditions as the 2019 RSUs. The RSUs fully vests on the date of the general meeting in the following year.

In September 2020, a new RSU incentive program was announced (2020-2 RSU program), which comprised 22,993 RSUs in total, including an on-boarding grant to a new board member in accordance with the

Group's remuneration policy. The 2020-2 RSU program runs in parallel with the 2020 RSU program and board members can only exercise RSUs under one of the programs.

In December 2020, 4,351 RSUs (2020-3 RSU program) were granted to the Chairman of the Board as part of a consultancy agreement (see Note 4.6).

In May 2021, the 2021 RSU program was announced, granting the Board of Directors an aggregate of 30,450 RSUs under similar terms and conditions as the 2020-1 RSUs.

The fair value of all RSUs was calculated using a Black-Scholes valuation model with the inputs shown in the following table. As the RSUs may be settled in cash, we have re-valued them as of year-end with updated inputs and recognized a cumulative share-based compensation expense in the amount of DKK (0.7) million (2020: 0.8 million; 2019: 0.1) and a corresponding short-term liability as of December 31, 2020. The Exercise Period for all 2020 RSUs is one year following full vesting and for valuation purposes we have assumed exercise three months upon full vesting.

As of December 31, 2021, 1,927 RSUs were expired, and 1,927 RSUs were eligible for exercise.

The following table presents the inputs to the Black-Scholes model used to estimate the fair value of the 2020 RSUs at year-end, as they are classified as cash-settled:

Program	December 31, 2021 2021 RSUs	December 31, 2020 2020 RSUs
Fair value at valuation date (DKK 000)	16	1,913
Dividend yield (%)	—	—
Expected volatility (%)	146.9%	45.9%
Risk-free interest rate (%)	(0.63%)	(0.57%)
Expected life of awards (years)	0,75	0.50
Weighted average share price (DKK)	17.16	67.10

d) Sign-on bonus shares to former CEO

As part of the former CEO service agreement, Kim Stratton was granted 58,000 ordinary shares, which would vest if the Company's share price increased to DKK 125 per share within three years from the date of employment. The total award consisted of (i) 6,000 shares provided that our share price increased to DKK 75 per share, (ii) 12,000 shares provided that our share price increased to DKK 100 per share, and (iii) 40,000 shares provided that our share price increased to DKK 125 per share. The target prices were achieved and the 58,000 ordinary shares were issued to Ms. Stratton in February 2021 (see Note 4.8).

The shares were valued at grant date, October 2019, using a Monte Carlo model due to the market conditions for vesting. The risk-free interest rate used in the model has been estimated based on Danish government bonds with similar maturities; expected volatility has been determined based on the historic volatility of comparable listed companies; the expected life of the award was 3 years, equal to the term of the award; the estimated dividend yield was zero; and the weighted average share price was DKK 55.60. The total valuation of the award at grant date was DKK 1.9 million. The total share-based compensation expense was classified as administrative and it was recognized in full during 2020, as the target prices were achieved in 2020.

e) Sign-on bonus shares to new CEO

As part of the new CEO service agreement, Christophe Bourdon was granted 34,941 RSUs in connection with the on-boarding, which will have a total vesting period of three years (beginning on January 1, 2021) and with one third of the granted RSUs vesting on each January 1 in the following three financial years. Vesting is not conditional upon achieving any financial or non-financial targets. However, in case of termination of employment and designation as a Good Leaver, the right to receive vested RSUs will be prorated and calculated through the date of release of the Participant's work obligations. The vested RSUs can only be exercised after the expiration of the total vesting period. However, the delivery period may be extended to the next open trading window in certain circumstances. The RSUs were valued at grant date, April 2021, using a Black Scholes option valuation model similar to the original 2021 LTIP. The valuation of the award at grant date was DKK 2.0 million. The share-based compensation expense was classified as administrative and with recognition from January 2021.

In October 2021, the grant of onboarding RSUs to was modified similar to the other long-term incentive programs for 2021. The terms of the modified grant are unchanged except for the immediate vesting upon grant and number of RSUs calculated based on a share price equal to DKK 31.94 per share, corresponding to the volume weighted average share price of the Company's shares as quoted on Nasdaq Copenhagen during the ten (10) trading days from September 1, 2021. The exercise of the RSUs to be granted under the modified LTIP is conditional upon the participant not exercising the RSUs granted in April 2021, which will subsequently lapse and no longer be exercisable, and are therefore considered replacement equity instruments for the cancelled equity instruments. The fair value of the originally granted RSUs at the date of the modification was determined to be DKK 24.72. The incremental fair value, calculated as the number of modified awards granted multiplied with the modified unit fair value less the fair value of the original LTIP granted remeasured at the modification date. The remaining expenses related to the original LTIP grant were similar recognised at the modification date. The incremental fair value of the modified award was DKK 0.9 million. The share-based compensation expense was classified as administrative recognised in October 2021.

f) Bonus shares issued to KLSDC and UCL in connection with the license agreement

Please see Note 3.1.

Summary of share-based compensation

The following amounts were recognized as share-based compensation for the years ended December 31

DKK 000	2021	2020	2019
Share-based compensation included in R&D	3,879	7,260	635
Share-based compensation included in G&A	12,140	20,845	1,914
Total share-based compensation expense recognized	16,019	28,105	2,549

2.7 FINANCIAL INCOME AND FINANCIAL EXPENSES

§ ACCOUNTING POLICIES

Financial income and expenses include interest income and expense, gains and losses due to changes in foreign exchange rates, interest expense related to the right-of-use assets, interest expense related to the Loan Agreement and other immaterial miscellaneous items.

The following table presents the various items of financial income and expense recognized for the years end December 31:

DKK 000	2021	2020	2019
Interest income on cash balances	37	45	316
Foreign currency exchange gains	11,849	1,649	—
Gain on embedded call option (Note 3.7)	546	750	—
Total financial income	12,432	2,444	316
Interest expense on Loan Agreement (Note 3.7)	7,350	9,921	3,239
Write-off of transaction costs for Loan Agreement tranche 2 (Note 3.7)	—	—	1,678
Loss on embedded call option (Note 3.7)	—	—	354
Interest expense on lease liabilities (Note 3.2)	624	567	351
Gain on lease modification (Note 3.2)	(365)	—	216
Interest expense on cash balances	1,484	3,626	1,213
Foreign currency exchange loss	1,369	14,805	229
Bank fees and other charges	147	152	79
Total financial expenses	10,609	29,071	7,359

2.8 INCOME TAXES

§ ACCOUNTING POLICIES

Income tax benefit includes the current benefit due from the current period's taxable loss and deferred tax adjustments. The benefit is comprised primarily of refundable tax credits for costs incurred in connection

with research and development activities under the Danish Tax Credit Regime. Income tax expense relates to tax imposed on income recognized in our subsidiaries in the United States and Switzerland as a result of our transfer pricing agreements.

Corporation tax receivable is recognized in the balance sheet as the tax benefit computed on the taxable loss for the year, adjusted for any changes to the prior year benefit due to changes in the taxable loss of prior years and for any taxes already paid or refunded.

Deferred tax is measured using the balance sheet liability method on all temporary differences between the carrying amount and the tax value of assets and liabilities, with the exception of temporary differences occurring at the time of acquisition and liabilities neither affecting the result of operation nor the taxable income.

As of December 31, 2021, 2020 and 2019, there were no tax audits in process nor has management been notified of any pending tax audit.

Judgement regarding the recognition of the deferred tax assets related to taxable losses to be carried forward

Orphazyme is subject to income taxes in Denmark, Switzerland and the U.S.A. The Group recognizes deferred income tax assets if it is probable that sufficient taxable income will be available in the future against which the temporary differences and unused tax losses can be utilized. Significant judgment is required to determine the amount of deferred tax assets that may be recognized, based upon the likely timing and the level of future taxable profits together with future tax planning strategies. This judgment is made periodically after considering current facts and circumstances, budgets and business plans as well as the risks and uncertainty associated with the Group's ability to successfully commercialize and defend its intellectual property. After consideration of these factors, Management has concluded that as regulatory approval has not yet been obtained as of December 31, 2021, the deferred income tax assets related to taxable losses carried forward in Denmark do not meet the criteria for being recognized as assets in the Statement of Financial Position.

The Company's tax losses can be carried forward infinitely subject to the general rules on limited deductibility due to ownership changes. In Denmark, the Company's ability to use tax loss carry forwards in any one year is limited to 100% of the first DKK 8.4 million of taxable income plus 60% of taxable income above DKK 8.8 million.

For the years ended December 31, 2021, 2020 and 2019, the Company has unrecognized net tax loss carry-forwards in the Danish entity in the amount of DKK 1,454 million, DKK 877 million, and DKK 425 million, respectively.

Management periodically evaluates positions taken in tax returns with respect to situations in which applicable tax regulations are subject to interpretation or uncertainty and establishes provisions, where appropriate. To date, there have not been any provisions established for uncertain tax positions.

The following table presents the total income tax benefit for the years ended December 31:

DKK 000	2021	2020	2019
Current tax benefit on net loss	144,379	136,845	75,459
Adjustments prior years	(385)	(1,065)	—
Tax credit research and development expenses	5,500	5,500	5,500
Change in unrecognized deferred tax before tax credit	(155,700)	(142,115)	(74,961)
Permanent differences	11,147	2,750	(498)
Total income tax benefit for the year	4,941	1,915	5,500

The following table presents the reconciliation of the effective tax rate to the statutory corporate income tax rate in Denmark.

DKK 000	2021	2020	2019
Net loss before tax	(631,480)	(635,161)	(342,997)
Corporate income tax rate in Denmark	22%	22%	22%
Computed income tax benefit	138,926	139,735	75,459
<i>Tax effect of:</i>			
Adjustments prior years	(385)	(1,065)	—
Other non-deductible expenses, including US listing-related costs and share-based compensation	11,147	2,750	(498)
Effect of different tax rate	892	(673)	—
Deferred tax asset not recognized after tax credit	(145,639)	(138,832)	(69,461)
Total income tax benefit for the year	4,941	1,915	5,500

The following table presents the carrying amount of deferred tax in the Statement of Financial Position:

DKK 000	2021	2020	2019
Tax deductible losses	319,811	192,837	93,484
Deferred tax on intangible assets	132,310	112,192	74,050
Other temporary differences	6,069	8,174	758
	458,190	313,203	168,292
Deferred tax asset not recognized	458,190	311,138	168,292
Carrying amount included in the Statement of Financial Position	—	2,065	—

SECTION 3 Assets and liabilities

3.1 INTANGIBLE ASSETS

§ ACCOUNTING POLICIES

Intangible assets comprise software development costs and license rights to develop and commercialize products and are acquired separately and measured on initial recognition at cost. Software assets consist of implementation costs to get cloud computing arrangements ready for use, as long as they meet the requirements of IAS 38, Intangible Assets. These cloud computing arrangements begin to be amortized when they are ready for intended use and are amortized over seven years.

For acquisition of intangible rights involving equity-settled share-based payment transactions, Management measures the fair value of the rights received and the corresponding increase in equity by reference to the fair value of the rights received, unless that fair value cannot be estimated reliably. If Management cannot estimate reliably the fair value of the rights received, it measures the fair value and the corresponding increase in equity by reference to the fair value of the equity instruments granted.

Following initial recognition, intangible assets are carried at cost less any accumulated amortization and accumulated impairment losses. The useful lives of intangible assets are assessed as either finite or indefinite. Intangible assets with finite lives such as software and license rights to develop and commercialize products are amortized over the useful economic life and assessed for impairment whenever there is an indication that the intangible asset may be impaired.

The amortization period and the amortization method for an intangible asset with a finite useful life are reviewed at least at the end of each reporting period. Changes in the expected useful life or the expected pattern of consumption of future economic benefits embodied in the asset are considered to modify the amortization period or method, as appropriate, and are treated as changes in accounting estimates. The amortization expense on intangible assets with finite lives is recognized in the Statement of Profit or Loss in the expense category that is consistent with the function of the intangible assets.

Assets with finite useful lifetime are assessed for impairment indicators. Each year, the assets are reviewed in order to assess whether there are indications of impairment. If such indications exist, the recoverable amount, determined as the higher amount of the fair value of the asset adjusted for expected costs to sell and the value in use of the asset, is calculated. The impairment expense on intangible assets with finite lives is recognized in the Statement of Profit or Loss in the expense category that is consistent with the function of the intangible assets.

Estimate of the fair value of licenses

Licenses contains an agreement entered into with the University of Kansas and University College London, in which the Company will obtain access to data and knowhow generated in the course of research in connection with the IBM trial. Consideration for the license is to be paid out by issuing new shares to the contract partners for a value corresponding to the costs incurred during the preceding calendar year. The valuation of the license upon the execution of the agreement involves uncertainty and was estimated by Management based on the expected costs over the contract period. In addition, the estimation of the duration of a license agreement at times involves uncertainty if termination is dependent on a time limit after successful commercialization. Management has considered potential commercialization dates and will re-assess this estimate on an ongoing basis.

Estimate related to recoverable amounts

Through the assessment of impairment indicators and impairment tests, Management identifies the recoverable amount. This is determined based on a value in use calculation, using cash flow and projections for subsequent years, equivalent to the expected useful life of the intangible asset.

With reference to note 1.6, significant events have occurred after the reporting period, which is relevant in the understanding of this item. The events disclosed in note 1.6 have not been considered in the current reporting period.

CytRx Asset Purchase Agreement

In May 2011, Orphazyme entered into an Asset Purchase Agreement with the US biopharmaceutical company CytRx. Pursuant to this agreement, CytRx sold and transferred certain preclinical and clinical data, patents and other intellectual property rights, and other assets, including contractual rights and obligations relating to a portfolio of chemical compounds, including arimoclomol, to Orphazyme. Under the terms of the Asset Purchase Agreement, Orphazyme agreed to make future payments to CytRx that were contingent upon the achievement of specified clinical, regulatory, and sales milestones. These payments are further disclosed in Note 3.9.

In 2016, the Company paid CytRx USD 0.1 million (DKK 0.6 million) for achievement of a clinical milestone for the first product candidate. In August 2018, the Company made a milestone payment of USD 250,000 (DKK 1.6 million) upon the enrollment of the first patient in the ALS clinical trial. The Company capitalizes amounts paid to CytRx as an acquired license right if the recognition criteria under IAS 38 is met. Management assesses that the consideration paid reflects market expectations about the probability that future economic benefits will flow to the Company. The acquired license is not being amortized until approval of the underlying asset has been received from regulatory authorities.

The Asset Purchase Agreement further includes sales milestones and royalty payments to be made by Orphazyme based on a specified percentage of any eventual net sales of products containing one of the compounds purchased. In addition, under the terms of the Asset Purchase Agreement, the Company was assigned and became party to a royalty agreement with ALS Charitable Remainder Trust pursuant to which the Company is obliged to pay a 1% royalty to the ALS Charitable Remainder Trust on global net sales of products to treat ALS. Orphazyme has no liabilities prior to the occurrence of future sales of products and accordingly neither such liabilities nor contingent consideration have been recognized as part of the license agreement.

Remaining life for this intangible asset is not possible to determine until approval of the underlying asset has been received from regulatory authorities.

License Agreement with KLSDC and UCL

In 2017, the Company entered into a license agreement with KU Center for Technology Commercialization Inc., University of Kansas, Kansas Life Sciences Development Company, Inc., ("KLSDC") and UCL Business PLC ("UCL") granting Orphazyme the right to develop and commercialize products under

all data generated in the course of the on-going Phase 2/3 clinical trial on arimoclomol for the treatment of IBM worldwide. The total consideration for the license is to be paid out in bonus shares to KLSDC and UCL up to an aggregate value of USD 2.5 million (DKK 15.8 million), depending on the amount of grants awarded to KLSDC and UCL for use in the trial. At the time the license agreement was executed, Management estimated the aggregate amount of the funding to be received by KLSDC and UCL to be USD 1.6 million (DKK 10 million), which has been recognized as an intangible asset (License) with a corresponding increase in equity reserves (Share-based compensation - acquisition of intangible assets).

Consideration to KLSDC and UCL is payable in shares of the Company ("Bonus Shares") each January and is based on incurred costs reported by KLSDC and UCL for the previous year. In January 2020, 20,650 (2019: 26,060) Bonus Shares were issued to KLSDC and UCL based on aggregate costs incurred by KLSDC and UCL in the amount of USD 0.3 million (DKK 2.2 million) (2018: USD 0.2 million (DKK 1,197 million)). The Bonus Shares were derived based on the average 30-day closing price of Orphazyme's shares at the date of issuance. At the time of the share issuance the equity reserve was decreased by DKK 2.1 million, which represents the market value of the shares issued. See Note 4.8 for Bonus Shares to be issued in 2021 related to the incurred costs reported by KLSDC and UCL for the year 2020.

Under the terms of the license agreement, Orphazyme shall furthermore pay an aggregate royalty of a low single-digit percentage of net sales of products sold for the treatment of IBM. Orphazyme expects to generate income from such products sold for the treatment of IBM which will exceed any royalty payments due. Orphazyme has no liabilities prior to the occurrence of future sales of products sold for the treatment of IBM and accordingly, neither such liabilities nor contingent considerations have been recognized as part of the rights acquired.

The license is being amortized over the duration of the license agreement, which has been estimated to be approximately 14 years. Amortization expense for the years ended December 31, 2020, 2019 amounts to DKK 0.7 million each year and is recognized within research and development expenses.

In March 2021, it was announced, that the phase 2/3 trial evaluating arimoclomol for the treatment of inclusion body myositis (IBM), a progressively debilitating muscle-wasting disease, did not meet its primary and secondary endpoints. As a result, Orphazyme has recognized an impairment loss of DKK 7.6 million corresponding to the remaining carrying amount of the license agreement. The impairment loss was recognized under research and development expenses.

License Agreement with the University of Miami

In September 2019, the Company entered into an exclusive license agreement with the University of Miami. Pursuant to the exclusive license agreement, the Company was granted a global royalty-bearing, exclusive license to all data, know-how, inventions and technology generated by the University of Miami and certain other institutions in a Phase 2 clinical trial of arimoclomol in ALS with the A4V SOD1 mutation to research, develop, make, use or sell certain pharmaceutical products or processes containing arimoclomol.

Under the terms of the exclusive license agreement, the Company made an up-front cash payment of \$75,000 (DKK 0.5 million) and further agreed to make certain future payments, including (i) a development milestone payment of \$1,150,000 (DKK 7.7 million) upon receiving regulatory approval for a pharmaceutical product containing arimoclomol for which the intended indication is ALS if the institution's Phase 2 clinical trial results were used in support of such regulatory approval, (ii) annual license fees from 2023 until the earlier of 2033 or termination of the agreement for a maximum aggregate amount of \$570,000 (DKK 3.8 million), and, (iii) beginning on the date of first commercial sale by the Company, its affiliates or sublicensees of a licensed product or licensed process in a country, a low single-digit royalty on net sales of licensed products or licensed processes on a product-by-product and country-by-country basis for a period of ten years thereafter unless the agreement is terminated earlier. Any annual license fees will be creditable against other payments due in the same calendar year.

In May 2021, it was announced, the ORARIALS-01 pivotal trial of arimoclomol in amyotrophic lateral sclerosis (ALS) did not meet its primary and secondary endpoints to show benefit in people living with ALS. As a result, Orphazyme has recognized an impairment loss of DKK 0.5 million corresponding to the remaining carrying amount of the license agreement. The impairment loss was recognized under research and development activities.

Orphazyme has no liabilities prior to the occurrence of future sales of products and accordingly neither such liabilities nor contingent consideration have been recognized as part of the license agreement.

The up-front cash payment was capitalized as an acquired license right, which is not being amortized until approval of the underlying asset has been received from regulatory authorities.

The following table presents the cost and respective amortization of software and licenses held by Orphazyme. The foreign currency effect is immaterial:

DKK 000	Software	Licenses	Total
Cost at December 31, 2019	—	12,083	12,083
Additions	2,736	—	2,736
Cost at December 31, 2020	2,736	12,083	14,819
Additions	902		902
Cost at December 31, 2021	3,638	12,083	15,721
Accumulated amortization at December 31, 2019	—	1,544	1,544
Amortization expense	109	712	821
Accumulated amortization at December 31, 2020	109	2,256	2,365
Amortization expense	322	119	441
Impairment expense	2,658	8,105	10,763
Accumulated amortization at December 31, 2021	3,089	10,480	13,569
Net carrying value at			
December 31, 2020	2,627	9,827	12,454
December 31, 2021	549	1,603	2,152

3.2 LEASES

§ ACCOUNTING POLICIES

On January 1, 2019, Orphazyme adopted IFRS 16, Leases, using the modified retrospective method. At contract inception, the Group assesses whether a contract is, or contains, a lease. A contract is, or contains, a lease if the contract conveys the right to control the use of an identified asset for a period of time in exchange for consideration. The Group is party to lease agreements only in which it is a lessee and not a lessor.

As a lessee, the Group applies a single recognition and measurement approach for all leases, except for short-term leases and leases of low-value assets. At inception or on reassessment of a contract that contains a lease component, the Group allocates the consideration in the contract to each lease component on the basis of their relative stand-alone prices.

The Group recognizes lease liabilities to make lease payments and right-of-use assets representing the right to use the underlying assets.

Right-of-use assets

The Group recognizes right-of-use assets at the commencement date of the lease (i.e., the date the underlying asset is available for use). Right-of-use assets are measured at cost, less any accumulated depreciation and impairment losses, and adjusted for any remeasurement of lease liabilities. The cost of right-of-use assets includes the amount of lease liabilities recognized, initial direct costs incurred, and lease payments made at or before the commencement date less any lease incentives received.

Right-of-use assets are depreciated on a straight-line basis over the shorter of the lease term and the estimated useful life of the underlying asset. If ownership of the leased asset transfers to the Group at the end of the lease term or the cost reflects the exercise of a purchase option, depreciation is calculated using the estimated useful life of the asset, which for the operating equipment under lease is ten years. The right-of-use assets are also subject to impairment.

Lease liabilities

At the commencement date of the lease, the Group recognizes lease liabilities measured at the present value of lease payments to be made over the lease term. The lease payments include fixed payments less any lease incentives receivable, variable lease payments that depend on an index or a rate, and amounts expected to be paid under residual value guarantees. The lease payments also include the exercise price of a purchase option reasonably certain to be exercised by the Group and payments of penalties for terminating the lease, if the lease term reflects the Group exercising the option to terminate. Variable lease payments that do not depend on an index or a rate are recognized as expenses in the period in which the event or condition that triggers the payment occurs. In calculating the present value of lease payments, the Group uses its incremental borrowing rate at the lease commencement date because the interest rate implicit in the lease is not readily determinable. After the commencement date, the amount of lease liabilities is increased to reflect the accretion of interest and reduced for the lease payments made. In addition, the carrying amount of lease liabilities is remeasured if there is a modification, a change in the lease term, a change in the lease payments (e.g., changes to future payments resulting from a change in an index or rate used to determine such lease payments) or a change in the assessment of an option to purchase the underlying asset. The Group's non-current lease liabilities are included as a separate line item on the Group's consolidated balance sheet and the current portion of lease liabilities is included in Other current liabilities.

Short-term leases and leases of low-value assets

The Group applies the short-term lease recognition exemption to its short-term leases of machinery and equipment (i.e., those leases that have a lease term of 12 months or less from the commencement date and do not contain a purchase option). It also applies the lease of low-value assets recognition exemption to leases of office equipment that are considered to be low value. Lease payments on short-term leases and leases of low-value assets are recognized as expense on a straight-line basis over the lease term.

Lease modifications

Lease modifications are accounted for at the effective date of modification, which is the date when both parties agree to the lease modification. Modifications are accounted for either as a separate lease or as a remeasurement of the initial lease. A modification is accounted for as a separate lease if both of the following conditions are met: (a) the modification increases the scope of the lease by adding the right to use one or more underlying assets; and (b) the consideration for the lease increases by an amount equivalent to the stand-alone price for the underlying asset. For a modification that is not a separate lease, the lease liability is remeasured using a discount rate determined at the effective date of the modification.

Estimate relating to the incremental borrowing rate

The Group cannot readily determine the interest rate implicit in its leases, therefore it uses its incremental borrowing rate to measure lease liabilities. This is the rate of interest that the Group would have to pay to borrow over a similar term, and with a similar security, the funds necessary to obtain an asset of a similar value to the right-of-use asset in a similar economic environment. As there are no observable rates available for such a rate, the Group estimates its incremental borrowing rate using observable inputs, such as market interest rates, and is required to make certain entity-specific estimates.

With reference to note 1.6, significant events have occurred after the reporting period, which is relevant in the understanding of this item. The events disclosed in note 1.6 have not been considered in the current reporting period.

The Group has lease contracts for its headquarters in Copenhagen, for its office in Chicago and for machinery used in its operations. The lease terms range from three to five years. During 2020, the lease contract in Copenhagen was modified to include additional space, which was accounted for as a separate lease addition. Furthermore, in June 2020 the Company entered a new lease contract for its US office premises in Chicago that expires September 2025. Following the Sale of Assets to KemPharm, Orphazyme has, as of June 22, 2022, terminated its lease agreement with its premises in Chicago.

During 2021, the lease contract in Copenhagen was modified to downscale the space, a significant change in the lease terms. The modification was accounted for as a change in the scope of the existing lease and therefore the initial lease was remeasured on the effective date of the modification at the weighted average incremental borrowing rate of 8.6%. The effect on the right-of-use assets, lease liabilities and the Statement of Profit or Loss is disclosed in the tables below. Furthermore during 2021 Orphazyme has impaired leased laboratory equipment in the amount of DKK 3.2 million. This amount is recognized under research and development activities.

The following table presents the carrying amounts of right-of-use assets recognized and the movements during the period:

DKK 000	Office buildings	Operating equipment	Total
At December 31, 2019	10,095	3,808	13,903
Additions	3,963	—	3,963
Depreciation expense	(2,606)	(401)	(3,007)
Modifications	0	—	0
At December 31, 2020	11,452	3,407	14,859
Additions	—	—	—
Disposals	(1,177)	—	(1,177)
Depreciation expense	(2,824)	(167)	(2,991)
Impairment expense	—	(3,239)	(3,239)
Modifications	(2,166)	—	(2,166)
Exchange rate adjustments	149	(1)	148
At December 31, 2021	5,434	—	5,434

The following table presents the carrying amounts of lease liabilities and the movements during the period:

DKK 000	2021	2020
At January 1	13,534	12,689
Additions	—	3,963
Accretion of interest	624	567
Disposals	(1,212)	—
Payments	(4,127)	(3,678)
Exchange rate adjustments	180	(7)
Modifications	(2,496)	—
At December 31	6,503	13,534
Current	2,578	3,657
Non-current	3,925	9,877

The maturity analysis of lease liabilities is disclosed in Note 3.7.

The following amounts are recognized in the Statement of Profit or Loss:

DKK 000	2021	2020	2019
Depreciation and impairment expense of right-of-use assets (R&D)	5,386	2,441	1,847
Depreciation and impairment expense of right-of-use assets (G&A)	844	566	211
Interest expense on lease liabilities	624	567	351
Gain on lease modification and disposals	(365)	—	216
Total amount recognized in the Statement of Profit or Loss	6,489	3,574	2,625

3.3 PROPERTY, PLANT, AND EQUIPMENT

§ ACCOUNTING POLICIES

Property, plant, and equipment includes IT, lab and other equipment, furniture and leasehold improvements that are measured at cost less accumulated depreciation and impairment losses. Cost includes the acquisition price and costs directly related to the acquisition until the time the asset is ready for use. The residual value of equipment is not material. Depreciation is calculated on a straight-line basis over the expected useful life of the asset, being 3-5 for equipment and furniture. Leasehold improvements are depreciated over the shorter of the useful life of the improvement or the remaining lease term. The useful life of assets and method of depreciation are reviewed by management at least each year-end or more often based on changes in facts and

circumstances. Changes in useful lives or residual values are adjusted prospectively as changes in accounting estimates. In addition, the Company has fully depreciated equipment still in use.

Property, plant, and equipment is required to be tested for impairment when there are impairment indicators present. Impairment tests are conducted at the individual asset level, or at the lowest level for which separately identifiable cash flows for groups of assets exist. Impaired assets or asset groups are written down to their recoverable amount, which is the higher of the value in use and the net realizable value of the asset or asset group, with impairment charges allocated proportionately to the assets within the impaired asset group.

With reference to note 1.6, significant events have occurred after the reporting period, which is relevant in the understanding of this item. The events disclosed in note 1.6 have not been considered in the current reporting period.

Gross carrying amount of any fully depreciated property, plant and equipment that is still in use is DKK 3.7 million.

The following table presents the Company's Property, plant and equipment as of the years presented:

DKK 000	Furniture and equipment	Leasehold improvements	Total
Cost at December 31, 2019	5,532	2,066	7,598
Additions	1,840	525	2,365
Disposals	—	—	—
Cost at December 31, 2020	7,372	2,591	9,963
Additions	48	44	92
Disposals	(597)	—	(597)
Cost at December 31, 2021	6,823	2,635	9,458
Accumulated depreciation at December 31, 2019	3,613	300	3,913
Depreciation expense	1,004	367	1,371
Exchange rate adjustments	(8)	0	(8)
Accumulated depreciation at December 31, 2020	4,609	667	5,276
Depreciation expense	1,156	429	1,585
Depreciation reversed on disposals during the year	(389)	0	(389)
Exchange rate adjustments	1	0	1
Accumulated depreciation at December 31, 2021	5,377	1,096	6,473
Net carrying value at			
December 31, 2020	2,763	1,924	4,687
December 31, 2021	1,446	1,539	2,985

There has been no impairment of property, plant and equipment for the years ended December 31, 2021 and 2020. Depreciation expense is included within operating loss as follows:

DKK 000	2021	2020	2019
Research and development expenses	1,301	887	544
General and administrative expenses	285	484	489
Total depreciation expense	1,585	1,371	1,033

3.4 PREPAYMENTS, DEPOSITS, AND OTHER RECEIVABLES

§ ACCOUNTING POLICIES

Prepayments

Prepayments include advance payments made to vendors that will be incurred and expensed in subsequent financial reporting periods. When the period for full expense recognition is longer than one year from the balance sheet date, the portion to be expensed subsequent to one year is classified as non-current.

Deposits

Deposits include advance payments made to vendors to be settled upon completion of the underlying contract. When the contract term is longer than one year from the balance sheet date, the deposit is classified as non-current.

Other receivables

Other receivables include current and non-current amounts due to the Company.

Sales tax

Expenses and assets are recognized net of the amount of sales tax, except:

- when the sales tax incurred on a purchase of assets or services is not recoverable from the taxation authority, in which case, the sales tax is recognized as part of the cost of acquisition of the asset or as part of the expense item, as applicable
- when receivables and payables are stated with the amount of sales tax included

The net amount of sales tax recoverable from, or payable to, the taxation authority is included as part of receivables or payables in the Statement of Financial Position.

Estimate of prepayments related to clinical trial development costs

As explained in Note 2.2, Orphazyme incurs substantial costs associated with clinical trials related to its development programs and there is a high degree of estimation involved in accounting for clinical trial development costs. In particular, certain CROs and vendors are paid upfront in connection with clinical activities and Management is required to estimate the timing of the prepayment release to expense. This expense for the year is estimated by using an expense model, as described in Note 2.2.

With reference to note 1.6, significant events have occurred after the reporting period, which is relevant in the understanding of this item. The events disclosed in note 1.6 have not been considered in the current reporting period.

The following items comprised non-current prepayments and deposits as of December 31:

DKK 000	2021	2020
Deposits with vendors	215	500
Prepayments to vendors	749	280
Leasehold deposit	—	1,234
Total non-current prepayments and deposits	964	2,014

Non-current prepayments and deposits mainly includes a deposit with a CRO for advance payment of pass-through costs in connection with a clinical trial, prepaid insurance, and the lease deposit on our headquarters in Copenhagen.

Current prepayments and other receivables are specified below:

DKK 000	2021	2020
Prepayments to vendors	12,872	38,281
Grant income receivable	0	81
VAT receivable, net	2,903	10,333
Foreign VAT receivable	1,627	1,304
Other current receivables	2,790	1,236
Total current prepayments and other receivables	20,192	51,235

Current prepayments to vendors include prepayments made to CROs for clinical trial costs of DKK 2.4 million (2020: DKK 5.2 million).

3.5 TRADE RECEIVABLES

§ ACCOUNTING POLICIES

Trade receivables are recognized and derecognized on a settlement date basis. They are measured at nominal value less expected credit losses based on historical experience. Orphazyme applies the simplified approach for determining expected credit losses.

At December 31, 2021 trade receivables in the amount of DKK 29.3 million are recognized in the balance sheet at the total invoiced amount less any expected credit losses. Due to the nature of the revenue transactions, expected credit losses are very limited.

There are no overdue receivables and the write-down for expected credit losses is not material.

3.6 PRE-LAUNCH INVENTORY

§ ACCOUNTING POLICIES

The Company may scale-up and make commercial quantities of its product candidate prior to the date it anticipates that such product will receive final regulatory approval. The scale-up and commercial production of pre-launch inventory involves the risk that such products may not be approved for marketing on a timely basis, or ever. This risk notwithstanding, the Company may scale-up and build pre-launch inventory of product that have not yet received final regulatory approval when the Company believes that such action is appropriate in relation to the commercial value of the product launch opportunity.

Inventory manufactured prior to regulatory approval (pre-launch inventory) is capitalized but immediately provided for, until regulatory approval for the product is obtained. A write-down is made against inventory, and the cost is recognized in the statement of profit or loss and other comprehensive loss as research and development costs. Once regulatory approval is obtained, the write-down is reversed, up to no more than the original cost.

With reference to note 1.6, significant events have occurred after the reporting period, which is relevant in the understanding of this item. The events disclosed in note 1.6 have not been considered in the current reporting period.

Pre-launch inventory intended for commercial sale

As of December 31, 2021 and 2020, the Company did not have pre-launch inventory that qualified for capitalization. As of December 31, 2021, the Company had pre-launch inventory of approximately DKK 92.6m (2020: DKK 12.4m) intended for commercial sale following regulatory approval of arimoclomol for the treatment of Niemann-Pick Disease Type C (NPC). This amount is fully provided for and recognized under research and development expenses in the statement of profit or loss and other comprehensive loss along with the production costs for drug substance and drug products used in clinical trials and early access programs that are not eligible for reversal at a later time. As the nATU sale in France is not commercial sale, cost for the related inventory has not been recognized on inventory.

3.7 FINANCIAL ASSETS AND LIABILITIES

§ ACCOUNTING POLICIES

Financial assets

Initial recognition and measurement

Financial assets that meet certain criteria are classified at initial recognition as subsequently measured at amortized cost, fair value through other comprehensive income (OCI), or fair value through profit or loss. The Group does not hold any financial assets meeting these classification criteria except cash and certain types of other receivables, which are valued at amortized cost. Generally, the Company's financial assets are available to support current operations and amounts expected to be realized within the next twelve months are classified in the Statement of Financial Position as current assets.

The Group's financial assets are recognized initially at fair value plus, in the case of financial assets not carried at fair value through profit and loss, transaction costs that are attributable to the acquisition of the financial asset, if any. Financial instruments recognized at fair value are allocated to one of the following valuation hierarchy levels:

- Level 1: Quoted (unadjusted) prices in active markets for identical assets or liabilities.
- Level 2: Other techniques for which all inputs that have a significant effect on the recorded fair value are observable, either directly or indirectly.
- Level 3: Techniques that use inputs that have a significant effect on the recorded fair value that are not based on observable market data.

Subsequent measurement

Historically, the Group's receivables are due within a twelve-month period and therefore the impact of using the effective interest rate method on the Group's financial statements has been immaterial.

Financial asset impairment

The Group assesses at the end of each reporting period whether there has been objective evidence that a financial asset may be impaired. Impairment losses are recognized if there is objective evidence of impairment and the evidence indicates that estimated future cash flows will be negatively impacted. The Group did not assess an impairment of a financial asset for either of the years ended December 31, 2021 or 2020.

Financial liabilities

Borrowings

Financial liabilities, including borrowings, are initially measured at fair value less transaction costs incurred. Subsequently, borrowings are measured at amortized cost. Amortized cost is calculated as original cost less instalments plus/less the accumulated amortization of the difference between cost and nominal value, so that the effective interest rate is recognized in the income statement over the loan period. Financial liabilities are derecognized when settled.

The Facilitation Fee in our Loan Agreement, defined below, is accounted for as an embedded derivative. The variability arising from the change in Orphazyme's share price is not closely related to the host debt instrument characterized mainly by interest rate and credit risk. Therefore, the embedded equity-linked amount is separated from the host debt instrument and accounted for as an embedded written call option at fair value through profit and loss.

The portion of the debt maturing after one year is presented as non-current debt and the remainder as current debt.

Trade payables and accruals

Trade payables and accruals relate to the Group's purchase of products and services from various vendors in the normal course of business.

Other liabilities

Other payables are measured at amortized cost. The amount payable to employees for the Phantom Shares Program (Note 2.6) is classified as non-current and is measured at fair value, at Level 2 in the fair value hierarchy.

Discount and rebate liabilities

Discount and rebate liabilities is classified as both current and non-current liabilities based on an existing legal or constructive obligation as a result of events occurring prior to or on the balance sheet date, and it is probable that the utilization of economic resources will be required to settle the obligation and is measured at management's best estimate of the expenses required to settle the obligation.

Estimate of accruals related to clinical trial development costs

As explained in Note 2.2, Orphazyme incurs substantial costs associated with clinical trials related to its development programs and there is a high degree of estimation involved in accounting for clinical trial development costs. As described in Note 2.2, Management uses an expense model to estimate the timing of expenses recognition in each period and related accruals at the end of the year.

With reference to note 1.6, significant events have occurred after the reporting period, which is relevant in the understanding of this item. The events disclosed in note 1.6 have not been considered in the current reporting period.

The Group's financial assets include mainly cash (Note 3.8). The Group has no derivative financial assets nor has there been a change in classification of a financial asset after initial recognition and measurements as discussed herein. Financial assets are not acquired for trading or speculative purposes, nor has the Group placed any assets as security for loans at either December 31, 2021 or 2020.

The Group's financial liabilities comprise the following as of the years ended December 31:

DKK 000	2021	2020
Borrowings	33,465	57,180
Lease liabilities (Note 3.2)	6,503	13,534
Trade payables	41,780	29,937
Accruals	15,743	42,198
Total liabilities measured at amortized cost	97,491	142,849

Kreos Debt Facility

In August 2019, Orphazyme entered into a structured debt facility ("Loan Agreement") with Kreos Capital to secure funding of €9 million (Tranche 1") to be repaid over forty-two months ("Loan Term"), with the first twelve months requiring interest only payments at nominal annual fixed interest rate of 9.75% and the remaining thirty months requiring equal installments comprising principal and interest. Early repayment of the borrowed amounts may be made in whole but not in part, with the repayment amount being equal to the principal outstanding plus the sum of all the interest repayments that would have been paid throughout the remainder of the loan discounted at an annual rate of 4.0%.

In addition, the lender may, at any time in its sole discretion in eight years, depending on certain events defined in the Loan Agreement, notify the Company that a Facilitation Fee is due and payable ("Notification").

The Facilitation Fee is an amount equal to the greater of (i) 10% of the aggregate amount of the amount borrowed and (ii) the percentage increase in the Company's share price on Nasdaq Copenhagen between the 30-day volume-weighted average share price on the date of the Loan Agreement and the closing share price on the day immediately preceding the date of the notification applied to the aggregate amount of amounts borrowed. The variability arising from the change in Orphazyme's share price is not closely related to the host debt instrument characterized mainly by interest rate and credit risk. Therefore, the embedded equity-linked amount is separated from the host debt instrument and accounted for as an embedded written call option at fair value through profit and loss.

Fair value on inception of the Loan Agreement is included as part of the transaction costs. The call option is measured at fair value at level 2 in the fair value hierarchy.

The written call option is measured at fair value using a Black-Scholes option valuation model. In measuring the fair value, various observable and unobservable inputs are required. Observable input mainly relates to the market price of Orphazyme's shares, and risk-free interest rate. Unobservable inputs mainly relate to the expected volatility of Orphazyme's share price, which was determined based on the Company's own historical volatility, and the term. The table below shows the inputs used in the valuation of the call option and the estimated fair value at year-end December 31.

Call option on Facilitation Fee	Dec 2021	Dec 2020
Fair value of call option	326	838
Dividend yield (%)	—	—
Expected volatility (%)	147%	54%
Risk-free interest rate (%)	(0.63)%	(0.61)%
Expected life (years)	1.2	2.2
Share price (DKK)	17.2	67.1

The change in fair value of the call option is recognized as a finance income or expense in the statement of profit or loss. For the year ended December 31, 2021, the Company recognized a gain of DKK 0.5 million (2020: DKK 0.8 million).

The structured debt facility included a potential second tranche available to Orphazyme, however as of December 31, 2019 conditions allowing for the drawdown of the second tranche were not met and it expired unused. In connection with the drawdown of Tranche 1, Orphazyme incurred transaction costs in the amount of €0.5 million (DKK 3.4 million). As the transaction costs secured a potential financing of two tranches, half of the transaction costs, or €0.2 million (DKK 1.7 million) are being amortized with the first tranche and upon expiration of the second tranche, the other half of the transaction costs were written off as finance expense in the statement of profit or loss (Note 2.7).

As part of the closing of the Loan Agreement, Orphazyme made a payment in the amount of €0.4 million (DKK 2.5 million) as a deposit for the last cash payment to be made on the borrowing ("Advance Payment").

The total liability for the Loan Agreement is being amortized net of the transaction costs, the Facilitation Fee and the call option; and it is being presented net of the Advance Payment.

Maturities of financial liabilities

The table below presents the Group's financial liabilities by relevant maturity groupings based on their contractual maturities for all non-derivative financial liabilities and derivative financial instruments for which the contractual maturities are essential for an understanding of the timing of the cash flows.

As the Facilitation Fee is due upon demand, it is shown as current Borrowings under non-derivatives. The call option on the Facilitation Fee is shown as current under derivatives.

The amounts disclosed in the table are the contractual undiscounted cash flows. Balances due within 12 months equal their carrying balances as the impact of discounting is not significant.

DKK 000	Less than 12 months	Between 1 and 2 years	Between 2 and 5 years	Total contractual cash flows	Carrying amount
Non-derivatives					
Trade payables and accruals	57,524	—	—	57,524	57,524
Borrowings	30,031	4,510	—	34,541	33,465
Lease liabilities	2,973	2,145	2,101	7,219	6,503
Total non-derivatives	90,528	6,655	2,101	99,284	97,492
Derivatives (Borrowings)	326	—	—	326	326
Total derivatives	326	—	—	326	326

Total changes in liabilities arising from financing activities are comprised as follows:

DKK 000	December 31, 2020	Cash flows	Non-cash changes				December 31, 2021
			Disposals	Adjustments and modifications	Accumulated interest	Exchange rate adjustments	
Borrowings	57,179	(30,904)	—	—	7,350	(160)	33,465
Lease liabilities	13,534	(4,127)	(1,212)	(2,496)	624	180	6,503
Total liabilities from financing activities	70,713	(35,031)	(1,212)	(2,496)	7,974	20	39,968

DKK 000	December 31, 2019	Cash flows	Non-cash changes				December 31, 2020
			Additions	Adjustments and modifications	Accumulated interest	Exchange rate adjustments	
Borrowings	62,824	(16,349)	—	750	9,921	33	57,179
Lease liabilities	12,689	(3,678)	3,963	—	567	(7)	13,534
Total liabilities from financing activities	75,513	(20,027)	3,963	750	10,488	26	70,713

Liabilities from accrued discount and rebates are calculated based on specific terms in the individual agreements. Please refer to note 2.1 further information on the accrued discount and rebates and managements estimates and judgements.

The clawback liability comprise both a current and non-current portion. Current clawback liability of DKK 7.9 million is settled annually, as a rebate, in accordance with nATU program. Non-current clawback liability of DKK 28.2 million will expected to be settled as the final price has been negotiated. As mentioned in note 2.1, the limited available relevant market information for directly comparable commercialized drugs within rare deceases increases the uncertainty in managements estimate of the clawback liability.

As mentioned in note 1.6 and 3.7, the clawback liability has been transferred to KemPharm at the carrying amount subsequently to the balance sheet date and on the basis of the amounts estimated by management. Thus, while the liability has been transferred after the balance sheet date to KemPharm, this is considered a non-adjusting event and has not impacted current vs. non-current presentation.

Total changes in liabilities arising from accrued discounts and rebates are comprised as follows:

DKK 000	December 31, 2020	Cash flows	Accruals	December 31, 2021
Discount and rebate liabilities	—	—	36,193	36,193
Total liabilities from accrued discount and rebates	—	—	36,193	36,193

Total current other liabilities are comprised of the following as of the years ended December 31:

DKK 000	2021	2020
Remuneration to the Board of Directors	293	2,840
Payroll and employee-related costs	29,230	50,487
Total current other liabilities	29,523	53,327

In addition, the Group has the following total other non-current liabilities as of the years ended December 31:

DKK 000	2021	2020
Accrual for milestone payment to vendor	—	1,179
Phantom shares liability to employees	98	455
Total non-current other liabilities	98	1,634

3.8 CASH

§ ACCOUNTING POLICIES

Cash includes cash on hand and in banks. Please see Financial Risks discussed in Note 4.4.

The Group's cash balance denominated in foreign currencies were as follows as of the years ended December 31:

DKK 000	2021	2020
DKK	53,291	483,862
USD	43,340	241,353
EUR	4,070	644
CHF	662	18
GBP	892	1,052
Total cash	102,255	726,929

3.9 COMMITMENTS AND CONTINGENCIES

Pledges and securities for loans.

In connection with a loan agreement in the amount up to €18.0 million entered into on August 27, 2019 with Kreos Capital VI (UK) Ltd., the Company has granted security in favor of Kreos Capital VI (UK) Ltd. over (i) certain of its assets, including its intellectual property rights, pursuant to a floating charge agreement registered with the Danish personal register in the initial principal amount of €9.0 million, (ii) its patents registered in Germany, the UK and the US pursuant to a patent pledge agreement and (iii) its shares in its US subsidiary, Orphazyme US, Inc. Furthermore, Orphazyme US, Inc. has granted in favor of Kreos Capital VI (UK) Ltd. (i) a guarantee for the Company's obligations under the loan agreement pursuant to a guaranty agreement and (ii) security over certain of its assets, including its intellectual property rights, pursuant to a security agreement governed under US law.

On July 9, 2021, a putative class action lawsuit was filed against the company and certain of its current and former directors and officers in the United States District Court for the Northern District of Illinois. This lawsuit alleges that certain representations about arimoclomol in the Company's U.S. IPO offering documents and in subsequent public statements were false and misleading, in violations of U.S. securities. Management does not believe these claims have any merit and believe that the outcome will not materially affect the Company's financial position.

With reference to note 1.6, significant events have occurred after the reporting period, with substantially all assets and business activities sold to KemPharm, Inc. The transaction included Orphazyme assets, including those relating to the development and approval of arimoclomol and the full claw back liability related to the French early access program. As a result, such commitments and contingencies related to the French early access program have been transferred to KemPharm Inc.

SECTION 4 Other disclosures

4.1 CAPITAL MANAGEMENT

For the purpose of the Group's capital management, capital includes issued capital, share premium and all other equity reserves attributable to the equity holders of the Group. The primary objective of the Group's capital management is to maximize shareholder value while limiting the financial risk. The Board of Directors' policy is to maintain needed capital base in order to maintain investor, creditor and market confidence.

As of December 31, 2021, the Group held cash totaling DKK 102.3 million (2020: DKK 726.9 million). With reference to note 1.6 management therefore considers it appropriate to prepare these financial statements on a going concern basis.

As of December 31, 2021 the Group has lost more than 50% of its subscribed share capital. On the ordinary general meeting of shareholders on June 29, 2022, the Board of Directors will give an account of the Group's financial position.

4.2 EQUITY

The following table summarizes the Company's share activity:

	Ordinary shares
December 31, 2018	19,939,564
Issuance of bonus shares as part of license agreement (note 3.1)	26,060
Issuance of Matching Shares (Note 2.6)	19,175
December 31, 2019	19,984,799
Issuance of bonus shares as part of license agreement (note 3.1)	20,650
Issuance of Matching Shares (Note 2.6)	31,250
Issuance of shares due to exercise of restricted share-units	11,921
Issuance of shares related to directed issue and private placement, February 2020	7,032,937
Issuance of shares related to US listing, September 2020	7,616,146
December 31, 2020	34,697,703
Capital increase, issuance of Matching Shares, net of costs	170,131
Capital increase, Bonus Shares	22,553
Capital increase, issuance of sign-on bonus shares to former CEO	58,000
Capital increase, exercise of RSUs	3,854
December 31, 2021	34,952,241

The Company has never declared or paid any cash dividends on its ordinary shares and does not anticipate doing so in the foreseeable future. The Company intends to use all available financial resources as well as revenue, if any, for purposes of the Company's current and future business.

In March 2019, the Company issued 19,175 Matching Shares to participants in the 2017 LTIP (see Note 2.6)

In January 2020, the Company issued 20,650 (2019: 26,060) bonus shares to KLSDC and UCL under the terms of the license agreement discussed in Note 3.1.

In February 2020, the Company completed an offering of 7,032,937 shares in a directed issue and private placement and raised gross proceeds of approximately DKK 745 million and net proceeds of approximately DKK 694 million.

The transaction consisted of a directed issue and private placement of up to 3,961,264 new shares of a nominal value of DKK 1 each (the "New Shares") and private placement of up to 3,071,673 existing shares of a nominal value of DKK 1 each (the "Existing Shares" and together with the New Shares, the "Offer Shares") at an offer price of DKK 106 per Offer Share, as determined by the Board of Directors of the Company through a book-building process (the "Offering"). The New Shares will be issued without pre-emption rights for existing shareholders.

The offering of Existing Shares was facilitated by a share loan from Novo Holdings A/S and Orpha Pooling B.V. (the “Lending Shareholders”) to the Company pursuant to a stock lending and subscription agreement with an obligation for the Company to redeliver new shares of an equivalent number as the Existing Shares borrowed by the Company from each of the Lending Shareholders (the “Replacement Shares”), which were issued without pre-emption rights for existing shareholders. The Lending Shareholders did not participate in the Offering and only facilitated the loan of the Lending Shares for purposes of the Company’s offering of Existing Shares in the Offering.

In April 2020, the Company issued 5,378 new shares to board members following the exercise of fully vested RSUs under the 2019 RSU program (see Note 2.5).

In July 2020, the Company issued 31,250 Matching Shares to participants in the 2019 LTIP (see Note 2.6)

In September 2020, the Company listed American Depositary Shares (ADSs) on the Nasdaq Global Select Market. In connection with this listing, we issued and sold 3,650,000 ordinary shares and 3,966,146 ADSs, each representing one ordinary share. Aggregate gross proceeds from the offering amounted to DKK 534.5 million, or USD 83.7 million translated at the exchange rate on the date the transaction closed. Orphazyme incurred transaction costs in the amount of DKK 56.6 million in connection with the US listing, which were accounted for as a deduction from equity.

In January 2021, the Company issued 22,553 (2020: 20,650 and 2019: 26,060) bonus shares to KLSDC and UCL under the terms of the license agreement discussed in Note 3.1.

In March 2021, the Company issued 3,854 new shares to board members following the exercise of fully vested RSUs under the 2020 RSU program (see Note 2.6).

In January 2021, the Company issued 170,131 Matching Shares to participants in the 2020 LTIP (see Note 2.6)

In February 2021, the Company issued 58,000 new shares to former CEO, Kim Stratton following the service agreement (see Note 2.6)

As a result of the above transactions, the total nominal share capital of the Company as of December 31, 2021 was DKK 34,952,241, divided into 34,952,241 ordinary shares each with a nominal value of DKK 1.

Pursuant to Section 3 of the Company’s articles of association, the Board of Directors was at December 31, 2021 authorized to increase the Company’s share capital by:

- Issue of new shares at market price without pre-emption rights by up to a nominal amount of DKK 6,989,767 in the period until 25 March 2026 (Article 3.1 of the Articles of Association)
- Issue of new shares against cash payment at a subscription price, which may be below the market price, to members of the Board of Directors, executives and/or employees of the Company without pre-emption rights by up to a nominal amount of DKK 1,300,000 in the period until 2 November 2022 (Article 3.2 of the Articles of Association). The authorization in article 3.2 has been partly exercised following which a nominal value of DKK 294,331 of the authorization has been issued.
- Directed issues of bonus shares, and/or directed issues of new shares effected by cash payment, to Kansas Life Sciences Development Inc. and UCL Business PLC (or entities designated by them), respectively, without pre-emption rights by up to a nominal amount of DKK 15,750,000 in the period until 2 November 2022 (Article 3.3 of the Articles of Association). The capital increase shall take place at par value (i.e. below market price). The value of such new shares to be issued can in any case not exceed a maximum of USD 2.5 million with a fixed exchange rate of DKK 6.30 per 1 USD based on the average closing price of the Company’s shares on Nasdaq Copenhagen A/S for the 30 days immediately prior to the date of issuance. The authorization in article 3.3 has been partly exercised following which a nominal value of DKK 80,643 of the authorization has been issued.

- Issue of new shares at a subscription price which may be below the market price with pre-emption rights by up to a nominal amount of DKK 25,000,000 in the period until 25 January 2025 (Article 3.4 of the Articles of Association).
- Issue of new shares against cash payment at a subscription price, which may be below the market price to members of the Board of Directors, executives and/or employees of the Company without pre-emption rights by up to a nominal amount of DKK 1,300,000 in the period until 25 March 2026. The new shares shall be issued (Article 3.5 of the Articles of Association).

The authorisations granted to the Board of Directors at December 31, 2021 pursuant to Articles 3.2 and

3.5 of the Articles of Association could in the aggregate only be exercised to increase the share capital by a maximum nominal amount of DKK 2,000,000.

4.3 LOSS PER SHARE

Basic loss per share for the year is calculated by dividing the net loss for the year by the weighted average number of ordinary shares outstanding during the year. The diluted loss per share is calculated by dividing the net loss for the year by the weighted average number of ordinary shares outstanding during the period increased by the dilutive effect of the assumed issuance of outstanding share-based awards. As a result of the Group incurring losses for each of the years ended December 31, 2021, 2020 and 2019, the potential shares issuable related to outstanding share-based awards have been excluded from the calculation of diluted per share amounts, as the effect of such shares is anti-dilutive.

Basic and diluted loss per share for the years presented have been adjusted retrospectively to include the 2019 Bonus Shares, the 2020 Bonus Shares and the 2021 Bonus Shares discussed in Note 3.1 in the number of weighted average shares outstanding for the years ended December 31, 2021, 2020 and 2019. This results in the comparative figures for 2020 and 2019 being updated accordingly.

The following reflects the net loss attributable to shareholders and share data used in the basic and diluted earnings/(loss) per share computations for the years ended December 31:

	2021	2020	2019
Net loss for the year (DKK 000)	(626,539)	(633,246)	(337,497)
Weighted-average shares outstanding	34,924,702	28,366,469	20,024,692
Loss per share	(17.94)	(22.32)	(16.85)

4.4 FINANCIAL RISKS

The Group's activities expose it to a number of financial risks whereby future events, which can be outside the control of the Group, could have a material effect on its financial position and results of operations. The known risks include foreign currency, interest and credit risk and there could be other risks currently unknown to Management. The Group has not historically hedged its financial risks.

Liquidity Risk

At December 31, 2021, the Group's liquidity risk was assessed to be high. Management continuously assesses the Group's capital structure in order to evaluate whether its liquidity reserves allow it to achieve its business objectives. At December 31, 2021, the available liquidity reserves, including funded capital in subsequent period, were assessed to be sufficient for the Group to meet its planned operating activities, including decreased levels of research and development activities, in the normal course of business for at least the next twelve months.

Foreign Currency Risk

The Group's foreign currency risk is assessed to be high. The Group conducts cross border transactions where the functional currency of the respective group entity is not always used. Accordingly, future changes in the exchange rates of the DKK against the EUR, the USD, the CHF and/or the GBP will expose the Group to currency gains or losses that will impact the reported amounts of assets, liabilities, income and expenses and the impact could be material.

Interest Rate Risk

The Group's interest rate risk is assessed to be low. The Group has a borrowing on which it incurs a fixed rate of interest (see Note 3.6). In addition, due to the current interest level in Denmark, the Group incurs negative interest on bank deposits.

Credit Risk

The Group's credit risk is assessed to be low. The Group's credit risk is associated with cash held in banks. The Company does not trade financial assets for speculative purposes and invests with the objective of preserving capital. The Company's cash is held primarily at two banks in Denmark with Moody's long-term credit ratings exceeding of A1.

The Group has prepared a sensitivity analysis in order to assess the potential impact on the Group's net loss for possible fluctuations in the EUR and USD exchange rates against the DKK and the impact for the possible fluctuations in the interest rate on bank deposits in Denmark and in the USA. The methods and assumptions used are consistent with prior year and consider increases and decreases in the Group's three main currencies, as well as reasonable fluctuations in the interest rate on its bank deposits. Based on these analyses, if interest rates on our cash deposits would have fluctuated by +/- 1%, the impact on the Group's net loss for the year ended December 31, 2021 would have been approximately DKK 14 thousand (2020: DKK 36 thousand; 2019: DKK 8 thousand).

The impact of currency fluctuations on the Group's net loss is shown in the table below:

Currency	Currency fluctuation	Effect 2021 TDKK	Effect 2020 TDKK	Effect 2019 TDKK
EUR	+/- 2%	284	538	503
USD	+/-10%	19,239	22,178	21
CHF	+/-10%	508	611	-
GBP	+/-10%	22	199	461

4.5 REMUNERATION OF BOARD OF DIRECTORS AND EXECUTIVE MANAGEMENT

Executive Management consists of the Company's Chief Executive Officer and the Chief Financial Officer, also the registered management of the Company. In July 2019, Orphazyme announced that the Board of Directors appointed Kim Stratton as the new Chief Executive Officer, succeeding Anders Hinsby on October 1, 2019.

Ms. Stratton resigned from her position at Orphazyme on December 10, 2020. As part of the separation agreement, Ms. Stratton will continue to receive her monthly base salary during 2021 and on December 31, 2021 she will receive severance pay equal to one year's base salary. Therefore, as of December 31, 2020, two times her base salary is additionally recognized as salary expense. Subsequent to December 31, 2020, Ms. Stratton received 35,304 Matching Shares as part of the 2020 LTIP program and will receive 58,000 ordinary shares as part of the sign-on bonus described in Note 2.5. As of December 31, 2020, 52,956 Performance Shares had vested as part of the 2020 LTIP program. These awards will be settled in January 2024 based on the development of the Company's share price (Note 2.5).

In March 2021, Orphazyme announced that the Board of Directors appointed Christophe Bourdon as the new Chief Executive Officer, succeeding Interim Chief Executive Officer Anders Fink Vadsholt April 1, 2021.

The Executive Management is eligible to receive an annual performance-based cash bonus subject to certain predefined corporate and individual goals as determined by the Board of Directors on an annual basis. A cash bonus received under the short-term incentive program may not exceed 100% of the annual fixed salary of the participants. As part of the separation agreement, Ms. Stratton is entitled to the annual performance-based cash bonus. The Executive Management is also eligible to receive an extraordinary bonus at the discretion of the Board of Directors.

The following table presents remuneration to the Executive Management for the years ended December 31, 2021, 2020 and 2019.

REMUNERATION TO INDIVIDUAL MEMBERS OF EXECUTIVE MANAGEMENT (DKK 000)	2021	2020	2019
Anders Vadsholt (CFO and interim CEO)			
Salary	2,376	2,324	1,803
Bonus	585	2,491	1,250
Share-based compensation	2,164	2,805	406
Other employee benefits	277	983	260
Total	5,402	8,603	3,719
Christophe Bourdon (CEO from April 1, 2021)			
Salary	2,925	—	—
Bonus	1,330	—	—
Share-based compensation ⁽¹⁾	4,967	—	—
Other employee benefits	107	—	—
Total	9,329	—	—
Kim Stratton (CEO through December 31, 2020)			
Salary	—	11,001	962
Bonus	—	3,500	1,025
Share-based compensation	—	7,359	—
Other employee benefits	—	2,542	215
Total	—	24,402	2,202
Anders Hinsby (former CEO)			
Salary	—	—	2,424
Bonus	—	—	1,038
Share-based compensation	—	—	294
Other employee benefits	—	—	270
Total	—	—	4,026
Total remuneration to the Executive Management	14,731	33,005	9,947

(1) includes two times share based compensation. Both sign-on bonus and LTIP 2021 program, both described in note 2.6

Remuneration paid to members of the Board of Directors is made up of board and committee fees, a travel allowance, ad-hoc fees for additional services provided as described in Note 4.6, and share-based compensation related to the Restricted Share Units (RSUs) as described in Note 2.6. Board remuneration is

recognized as general and administrative expenses in the Statement of Profit or Loss. The following table lists Board of Directors remuneration for the years ended December 31:

**REMUNERATION TO INDIVIDUAL MEMBERS
OF THE BOARD OF DIRECTORS (DKK 000)**

	2021	2020	2019
Georges Gemayel (Chairman of the Board)			
Board and committee fees	659	565	470
Ad hoc board fees (1)	1,312	186	—
Travel allowance	30	27	64
Share-based compensation	164	161	28
Total	2,165	939	562
Bo Jesper Hansen (Deputy Chairman of the Board)			
Board and committee fees	447	421	395
Ad hoc board fees	—	112	—
Travel allowance	97	34	46
Share-based compensation	68	102	21
Total	612	669	462
Martin Bonde			
Board and committee fees	318	276	259
Travel allowance	—	—	—
Share-based compensation	48	73	16
Total	366	349	275
Martijn Kleijwegt			
Board and committee fees	161	304	285
Travel allowance	—	35	46
Share-based compensation	25	73	16
Total	186	412	347
Rémi Droller (2)			
Board and committee fees	(828)	288	270
Travel allowance	(90)	25	46
Share-based compensation	(89)	73	16
Total	(1,007)	386	332
Sten Verland			
Board and committee fees	81	327	309
Travel allowance	—	1	—
Share-based compensation	25	73	16
Total	106	401	325
Anders Hedegaard			
Board and committee fees	153	288	270
Travel allowance	—	13	46
Share-based compensation	25	73	16
Total	178	374	332
Catherine Moukheibir			
Board and committee fees	353	355	336
Ad hoc board fees	—	112	—
Travel allowance	—	23	46
Share-based compensation	25	73	16
Total	378	563	398
Carolee Barlow			
Board and committee fees	320	77	—
Travel allowance	—	—	—
Share-based compensation	80	145	—
Total	400	222	—
Stephanie Okey			
Board and committee fees	247	—	—
Ad hoc board fees	107	—	—
Share-based compensation	72	—	—
Total	426	—	—
Andrew Mercieca			
Board and committee fees	24	—	—
Total	24	—	—
Total remuneration to the Board of Directors	3,834	4,315	3,033

- (1) George Gemayel received ad-hoc fee in connection with a consultancy agreement for support during the interim period until Christophe Bourdon joined as CEO in April 2021
- (2) Rémi Droller resigned effective as of June 30, 2021. At the same time all historical board fee was reversed.

4.6 RELATED PARTIES

Orphazyme A/S, incorporated in Denmark, is the ultimate parent company of the Group, which wholly owns Orphazyme US, Inc and Orphazyme Switzerland GmbH. These three entities are considered related parties. Orphazyme A/S is not ultimately controlled by any of its investors. Major investors owning more than 10% of the Company are considered related parties.

For the years ended December 31, 2021, 2020 and 2019, the following related party transactions were identified:

- Remuneration to Executive Management (Note 4.5)
- Remuneration to the Board of Directors (Note 4.5)
- Participation of Executive Management in the 2017 LTIP, the 2019 LTIP and the 2020 LTIP (Note 2.5)
- Participation of the Board members in the 2019 RSU and 2020 RSU programs (Note 2.5)
- Share lending arrangement in connection with the directed issue and private placement in February 2020 (Note 4.2). We entered into a Stock Lending and Subscription Agreement on February 6, 2020 with Danske Bank A/S, Orpha Pooling B.V. and Novo Holdings A/S, pursuant to which we borrowed 3,071,673 existing ordinary shares (the Lending Shares) from Orpha Pooling B.V. and Novo Holdings A/S, major investors at the time, through Danske Bank A/S as settlement agent in order for us to place such ordinary shares in a private placement. The Lending Shares were borrowed subject to an obligation for us to issue new ordinary shares of an equivalent number as the Lending Shares placed in this private placement, or the Listing Shares, and for Danske Bank A/S to use the proceeds from the sale of Lending Shares in the private placement to subscribe for the Listing Shares and deliver the Listing Shares to the Orpha Pooling B.V. and Novo Holdings A/S. The Listing Shares were issued and delivered, as agreed, on February 11, 2020.
- Ad-hoc fees paid to certain members of the Board of Directors in connection for their support during the US listing process in 2020. Total ad-hoc fees amounted to EUR 55,000 (DKK 0.4 million) and was recorded in the statement of profit or loss and other comprehensive income.
- Ad-hoc fees paid to the Chairman of the Board in 2020 connection with a consultancy agreement for support during the interim period until a new CEO is hired. As part of this agreement, Orphazyme has paid the Chairman of the Board an up-front payment of EUR 88,605 (DKK 0.7 million) in December 2020, which is recognized as a prepayment in the statement of financial position. An additional payment of 100% of his aggregate annual board and committee fees is payable in June 2021. In addition to cash compensation, the Chairman has been granted 4.351 RSUs under the 2020-2 RSU program (see Note 3.5). The full remuneration to the Chairman of the Board under this consultancy agreement is subject to the approval of the shareholders at the Company's annual general meeting in March 2021.

As of December 31, 2021 and 2020, the Company did not have any amounts receivable from related parties and therefore recorded no related impairment. The Company has not granted any loans, guarantees, or other commitments to or on behalf of any of the members of the Board of Directors or Executive Management. For amounts payable to the Board of Directors, please see Note 3.6.

Executive Management and members of the Board of Directors had the following shareholding in Orphazyme A/S for the years ended December 31:

	Number of shares owned 2021	Number of shares owned 2020	Number of shares owned 2019
Anders Vadsholt	160,717	143,156	132,595
Christophe Bourdon	—	—	—
Kim Stratton	—	50,600	—
Anders Hinsby (Former CEO)	—	—	209,596

MEMBERS OF THE BOARD OF DIRECTORS:	December 31, 2021			December 31, 2020		December 31, 2019
	Number of shares owned	Number of Unvested RSUs 2021	Number of Unexercised RSUs 2020	Number of shares owned	Number of Unvested RSUs 2019	Number of shares owned
Georges Gemayel	100,809	9,222	—	100,809	4,351	97,358
Bo Jesper Hansen	143,234	2,849	—	143,234	2,689	100,545
Martijn Kleijwegt	1,927	—	—	—	1,927	—
Martin Bonde	47,936	2,042	—	47,936	1,927	46,009
Rémi Droller	—	—	—	—	1,927	—
Sten Verland	—	—	—	—	1,927	—
Anders Hedegaard	15,677	—	—	15,677	1,927	13,750
Catherine Moukheibir	9,907	—	—	7,980	1,927	7,980
Carolee Barlow	—	2,042	—	—	4,391	—

4.7 FEES TO AUDITORS

The following table presents the fees to our independent registered public accounting firm, EY Godkendt Revisionspartnerselskab ("EY"), recognized in general and administrative expenses in the Statement of Profit or Loss for the years ended December 31. This note was prepared in accordance with the requirements of the Danish Financial Statements Act:

DKK 000	2021	2020	2019
Audit services	2,743	2,416	2,244
Audit-related services	430	803	882
Other assistance	753	3,795	—
Total fees to auditors	3,926	7,014	3,126

Audit services

Audit services consist of fees billed for professional services rendered by EY for the audit of our annual consolidated financial statements or services that are normally provided by the accountant in connection with statutory and regulatory filings or engagements for those fiscal years. With reference to the events disclosed in note 1.6, the fee for audit services have increased compared to 2020.

Audit-Related services

Audit-related services consist of assurance and related services performed by EY that are reasonably related to the performance of the audit or review of our consolidated financial statements and are not reported under "Audit services".

Other assistance

In 2020, other assistance consists of services provided by EY for other permitted services, including fees for work performed by EY in connection with the U.S. listing in September 2020

In 2021, other assistance consists of services provided by EY for other permitted services, including fees for work performed by EY in connection with the U.S. At -the-Market offering program in November 2021

Pre-approval policies

The Audit Committee assesses and pre-approves all services provided by the statutory auditors. The pre-approval includes the type of service and a fee budget.



Articles of Association

Orphazyme A/S, CVR no. 32 26 63 55

Gorrissen Federspiel



1 Name and objects

1.1 The Company's name is Orphazyme A/S.

1.2 The Company's objects are research, development, production, marketing, sales and/or licensing of medicinal products for treatment of various disorders, including lysosomal storage diseases (LSD), neuromuscular disorders and other related diseases, as well as to carry out associated activities. Furthermore, the Company may, within its line of business, participate in partnerships or co-operate with other businesses.

2 Share capital and shares

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

2.2 The share capital has been fully paid up.

2.3 The shares shall be issued in the name of the holder and shall be recorded in the name of the holder in the Company's register of shareholders.

2.4 The register of shareholders is kept by Computershare A/S, CVR no. 27 08 88 99.

2.5 The shares are negotiable instruments. No restrictions shall apply to the transferability of the shares.

2.6 No shares shall carry special rights.

2.7 No shareholder shall be under an obligation to have his/her shares redeemed in whole or in part by the Company or by any third party.

2.8 The shares are registered with and issued in dematerialised form through VP SECURITIES A/S, CVR no. 21 59 93 36. Dividend is paid out through VP SECURITIES A/S and is deposited at the registered dividend accounts at VP SECURITIES A/S. Rights concerning the shares shall be notified to VP SECURITIES A/S in accordance with applicable rules.

3 Increase of share capital

3.1 In the period until 25 March 2026, the Board of Directors is authorised to increase the Company's share capital in one or more issues of new shares without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 6,989,767. The capital increase shall take place at market price as determined by the Board of Directors and shall be effected by cash payment, debt conversion or contribution in kind.

3.1.1 By decision of February 11, 2022, the Board of Directors has partly exercised the authorisation in Article 3.1 to increase the Company's share capital following which a nominal value of DKK 360,000 of the authorisation has been issued.

3.2 In the period until 2 November 2022, the Board of Directors is authorised to increase the Company's share capital in one or more issues without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 1,300,000 in



connection with the issue of new shares to members of the Board of Directors, executives and/or employees of the Company. The new shares shall be issued against cash payment at a subscription price to be determined by the Board of Directors, which may be below the market price.

- 3.2.1 By decision of 4 March 2019, 27 March 2020, 29 July 2020, 1 February 2021 and 25 February 2021, the Board of Directors has partly exercised the authorisation in Article 3.2 to increase the Company's share capital following which a nominal value of DKK 294,331 of the authorisation has been issued.
- 3.3 In the period until 2 November 2022, the Board of Directors is authorised to increase the Company's share capital in one or more issues without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 15,750,000 in connection with directed issues of bonus shares, and/or directed issues of new shares effected by cash payment, to Kansas Life Sciences Development Inc. and UCL Business PLC (or entities designated by them), respectively. The capital increase shall take place at par value (i.e. below market price). The value of such new shares to be issued can in any case not exceed a maximum of USD 2.5 million with a fixed exchange rate of DKK 6.30 per 1 USD based on the average closing price of the Company's shares on Nasdaq Copenhagen A/S for the 30 days immediately prior to the date of issuance.
- 3.3.1 By decision of 29 January 2018, 31 January 2019, 31 January 2020 and 25 February 2021, the Board of Directors has partly exercised the authorisation in Article 3.3 to increase the Company's share capital following which a nominal value of DKK 80,643 of the authorisation has been issued.
- 3.4 In the period until 25 January 2025, the Board of Directors is authorised to increase the Company's share capital through one or more issues of new shares with pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 25,000,000. The capital increase may be effected by cash payment or conversion of debt and shall take place at subscription price as determined by the Board of Directors which may be below the market price.
- 3.5 In the period until 25 March 2026, the Board of Directors is authorised to increase the Company's share capital in one or more issues without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 1,300,000 in connection with the issue of new shares to members of the Board of Directors, executives and/or employees of the Company. The new shares shall be issued against cash payment at a subscription price to be determined by the Board of Directors, which may be below the market price.
- 3.6 In the period until 1 January 2027, the Board of Directors is authorised to increase the Company's share capital in one or more issues of new shares without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 20,000,000. The capital increase shall take place at market price as determined by the Board of Directors and shall be effected by cash payment, debt conversion or contribution in kind.



- 3.7 In the period until 1 January 2027, the Board of Directors is authorised to increase the Company's share capital through one or more issues of new shares without pre-emption rights for the Company's existing shareholders by up to a nominal amount of DKK 20,000,000. The capital increase may be effected by cash payment or conversion of debt and shall take place at market price as determined by the Board of Directors.
- 3.8 In the period until 1 January 2027, the Board of Directors is authorised to issue convertible bonds on one or more occasions without pre-emption rights for the existing shareholders and with a total principal amount of up to DKK 70,000,000 which are convertible into shares in the Company. The convertible bonds shall be effected by cash payment. The conversion price as determined by the Board of Directors may be above or at the market price at the time of issuance of the convertible bonds. The issuance of convertible bonds may be directed at qualified investors. The Board of Directors is authorised in the period until 1 January 2027 to increase the Company's share capital by up to nominally DKK 20,000,000 by conversion of convertible bonds issued pursuant to this Article 3.8 and to effect the associated capital increases.
- 3.9 In the period until 1 January 2027, the Board of Directors is authorised to issue warrants on one or more occasions without pre-emption rights for the existing shareholders granting the holders right to subscribe for shares in the Company for a total amount of up to nominally DKK 20,000,000. The Board of Directors is entitled to determine the exercise price for the warrants upon issue given that the exercise price may be above or at the market price at the time of issuance. The Board of Directors is authorised in the period until 1 January 2027 to increase the Company's share capital by up to nominally DKK 20,000,000 resulting from the exercise of warrants pursuant to this Article 3.9 and to effect the associated capital increases.
- 3.10 The authorisations granted to the Board of Directors pursuant to Articles 3.1, 3.6 and 3.7 can in the aggregate only be exercised to increase the share capital by a maximum nominal amount of DKK 40,000,000.
- 3.11 The authorisations granted to the Board of Directors pursuant to Articles 3.2 and 3.5 can in the aggregate only be exercised to increase the share capital by a maximum nominal amount of DKK 2,000,000.
- 3.12 New shares issued pursuant to Articles 3.1, 3.2, 3.3, 3.4, 3.5, 3.6, 3.7, 3.8 and 3.9 shall be fully paid up, issued in the name of the holder and shall be recorded in the name of the holder in the Company's register of shareholders, shall be negotiable instruments and shall in every respect carry the same rights as the existing shares. The Board of Directors is authorised to lay down the terms and conditions for capital increases pursuant to the above authorisations and to make any such amendments to the Company's Articles of Association as may be required as a result of the Board of Directors' exercise of said authorisations.
- 4 General meeting, venue and notice**
- 4.1 The general meetings of the Company shall be held in the Capital Region of Denmark.
- 4.2 The annual general meeting of the Company shall be held each year in due time for the audited and approved annual report to be received by the relevant authorities before



the applicable statutory time limit. The Company shall no later than eight weeks before the contemplated date of the annual general meeting publish the date of the general meeting and the deadline for submitting requests for specific proposals to be included on the agenda.

- 4.3 If the Board of Directors finds it appropriate, and if the general meeting can be conducted in a technically safe manner, the Board of Directors may decide that the general meeting shall be held fully or partially as an electronic general meeting. If so decided, shareholders will be able to attend, express their opinion and vote at the general meeting by electronic means. Detailed information on the procedures for electronic attendance and participation will be made available on the company's website and in the relevant notices convening the general meetings, and written information on the subject will also be sent to shareholders registered in the Company's register of shareholders if so requested.
- 4.4 Extraordinary general meetings shall be held when determined by the Board of Directors or requested by the Company's auditor. Furthermore, an extraordinary general meeting shall be held when requested by shareholders possessing no less than five per cent of the share capital. Such request shall be submitted in writing to the Board of Directors and be accompanied by a specific proposal for the business to be transacted. The Board of Directors convenes an extraordinary general meeting no later than two weeks after such request has been made.
- 4.5 General meetings shall be convened by the Board of Directors with at least three weeks' and not more than five weeks' notice. The notice shall be published on the Company's website. Furthermore, a notice of the general meeting shall be sent to all shareholders recorded in the Company's register of shareholders who have so requested.
- 4.6 For a period of three weeks prior to the general meeting, including the date of the general meeting, the following information shall be available on the Company's website:
- a. The notice convening the general meeting
 - b. The aggregate number of shares and voting rights as at the date of the notice
 - c. The documents to be presented at the general meeting
 - d. The agenda and the complete proposals as well as, for annual general meetings, the audited annual report
 - e. The forms to be used for voting by proxy or by postal vote
- 4.7 General meetings shall be held in English. The Board of Directors may decide to offer simultaneous interpretation into Danish. Documents prepared in connection with or following a general meeting shall be in English and to the extent required by law or if decided by the Board of Directors, in Danish.



4.8 The general meeting shall be presided over by a chairman elected by the Board of Directors who shall ensure that the general meeting is conducted in a proper and efficient manner.

5 Agenda for the annual general meeting

5.1 The agenda for the annual general meeting shall include the following:

- a. The Board of Directors' report on the Company's activities in the past financial year
- b. Presentation and adoption of the annual report
- c. Distribution of profit or covering of loss according to the adopted annual report
- d. Resolution to grant discharge of liability to the Board of Directors and the Executive Management
- e. Presentation of the remuneration report
- f. Approval of remuneration of the Board of Directors for the current financial year
- g. Election of members to the Board of Directors
- h. Election of auditor
- i. Authorisation to acquire treasury shares
- j. Any proposals from the Board of Directors or shareholders
- k. Any other business

5.2 Every shareholder shall be entitled to have a specific subject considered at the annual general meeting. Such proposals must be submitted in writing to the Board of Directors not later than six weeks prior to the annual general meeting.

6 Shareholders' attendance and voting rights at the general meeting

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

6.2 A shareholder who is entitled to attend the general meeting pursuant to Article 6.1 and who wants to attend the general meeting shall notify the Company of his/her attendance not later than three days prior to the date of the general meeting.

6.3 A shareholder may attend in person or by proxy, and the shareholder or the proxy may attend together with an adviser.



- 6.4 The right to vote may be exercised by a written and dated instrument of proxy in accordance with applicable laws.
- 6.5 A shareholder who is entitled to participate in the general meeting pursuant to Article 6.1 may vote by postal vote in accordance with the provisions of the Danish Companies Act. Such postal votes shall be received by the Company not later than the business day before the general meeting. Postal votes cannot be withdrawn.
- 6.6 Each share of the nominal value of DKK 1 shall carry 1 vote. A person registered as a holder of shares of the Company in VP Securities A/S and acting in a professional capacity on behalf of other natural or legal persons, including holders of American Depositary Shares representing shares of the Company, may exercise voting rights attached to any such shares in a manner that is not identical to the exercise of the voting rights attached to other shares of the Company held by such person.
- 7 Resolutions at general meetings**
- 7.1 Resolutions by the general meeting shall be passed by a simple majority of votes cast unless otherwise prescribed by law or by these Articles of Association.
- 7.2 Adoption of changes to these Articles of Association, dissolution of the Company, merger or demerger requires that the decision is adopted with at least 2/3 of the votes cast as well as the share capital represented at the general meeting, unless applicable laws prescribe stricter or less strict adoption requirements or applicable laws confer specific authority to the Board of Directors or other bodies.
- 8 Board of Directors**
- 8.1 The Board of Directors consists of not less than six and not more than nine members elected by the general meeting.
- 8.2 The members of the Board of Directors elected by the general meeting are elected for a term of one year. Re-election of board members may take place.
- 8.3 The Board of Directors elects a Chairman and, if so decided by the Board of Directors, a Deputy Chairman among its members. If the Chairman of the Board of Directors resigns during a term of election, the Deputy Chairman (if elected) shall take up the position as Chairman until a new Chairman is elected among the members of the Board of Directors.
- 8.4 Any employee representatives on the Board of Directors and their alternates, if any, are elected in accordance with applicable law thereon in force from time to time.
- 8.5 Resolutions of the Board of Directors are passed by simple majority. In the event of equal votes, the Chairman or, in his/her absence, the Deputy Chairman shall have a casting vote.
- 8.6 The Board of Directors forms a quorum when more than half of its members are represented, including the Chairman or the Deputy Chairman.



9 Executive Management

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

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

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

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

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

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



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

10 Rules of signature

10.1 The Company shall be bound (i) by the joint signatures of the Chairman and a member of the Executive Management, (ii) by the joint signatures of the Chairman and two members of the Board of Directors or (iii) by the joint signatures of two members of the Executive Management.

11 Electronic communication

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

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

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

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

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

12 Annual report

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

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

13 Financial year

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

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



Company's annual general meeting held on 25 March 2021, by the Board of Directors on 11 February 2022, and on the Company's extraordinary general meeting held on 15 February 2022.

Gorrissen Federspiel

DESCRIPTION OF SHARE CAPITAL AND ARTICLES OF ASSOCIATION

Securities registered under Section 12(b) of the Securities Exchange Act of 1934 (the “Exchange Act”), as of December 31, 2021:

Title of each class	Trading Symbol(s)	Name of each exchange on which registered
American Depositary Shares, each representing one ordinary share, nominal value DKK 1 per share	ORPH	The Nasdaq Global Select Market**
Ordinary shares, nominal value DKK 1 per share*	ORPH	The Nasdaq Global Select Market*

* Not for trading, but only in connection with the registration of the American Depositary Shares (“ADSs”) on The Nasdaq Global Select Market.

** The ADSs were delisted from The Nasdaq Global Select Market on March 31, 2022. The Deposit Agreement governing the ADSs will be terminated on July 6, 2022.

This exhibit contains a description of the rights of (i) the holders of ordinary shares and (ii) the holders of ADSs as of December 31, 2021.

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 Danish Companies Act (Selskabsloven) (the “DCA”). The following summary is subject to and qualified in its entirety by our articles of association and by applicable Danish law, particularly the DCA. This is not a summary of all the significant provisions of the articles of association or of Danish law and does not purport to be complete. 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.

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 this Annual Report on Form 20-F.

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. We were admitted to trading and official listing on Nasdaq Copenhagen in November 2017. Our company was established with the objectives of engaging in research, development, production, marketing, sales and/or licensing of medicinal products for treatment of various disorders, including lysosomal storage diseases (LSD), neuromuscular disorders and other related diseases, as well as to carry out associated activities.

On March 11, 2022 the Board of Directors of Orphazyme initiated in-court restructuring proceedings whereby we pursued potential opportunities for a sale of our assets and operations, a sale of the Company and/or a refinancing (potentially in combination with a compulsory composition). The ADSs were delisted from The Nasdaq Global Select Market on March 31, 2022.

In May 2022, substantially all of the Company’s assets and business activities, were sold to KemPharm Denmark A/S (“KemPharm”), a wholly-owned subsidiary of KemPharm Inc. KemPharm (i) acquired substantially all of our employees and most of our assets, except for certain exempted assets; (ii) took over certain liabilities, including employee liabilities, liabilities relating to an early access patient program and certain milestone payments linked to approvals of medicinal products; and (iii) took over certain of our

continued agreements relevant to the continued operations of the business operated by us. KemPharm paid a total of USD 12.8 million in cash and assumed liabilities estimated to equal approximately USD 5.2 million as consideration.

Our headquarters and principal executive office is located at Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark, and our telephone number is +45 39 17 82 72. We have vacated our laboratory space and assigned the remaining office facility to KemPharm Denmark ApS as of June 1, 2022. Our website address is www.orphazyme.com. The information contained on, or accessible through, our website is not incorporated by reference into this Annual Report on Form 20-F, and you should not consider any information contained in, or that can be accessed through, our website as part of this Annual Report on Form 20-F or in deciding whether to purchase our ordinary shares or ADSs. We have included our website address as an inactive textual reference only.

Development of Share Capital and Our Shares

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

As of December 31, 2021, our registered, issued and fully paid outstanding share capital was DKK 34,952,241 distributed into 34,952,241 shares of nominal value of DKK 1 each.

As of December 31, 2021, 19,886,184 shares were represented by ADS. Each ADS represents one ordinary share. Our ADSs were listed on the Nasdaq Global Select Market under the symbol "ORPH". The underlying shares are listed on Nasdaq Copenhagen under the symbol "ORPHA".

In February 2022, the Company issued new shares as a result of the utilization of the Company's U.S. At-the-Market Offering Program with Cowen and Company, LLC ("Cowen"). On February 11, 2022, a total of 360,000 ordinary shares of nominally DKK 1 each, represented by ADSs, were issued by the Company and sold in the market by Cowen as the sales agent at market price as determined by the Company's Board of Directors in accordance with the authorization in article 3.1 of the Company's Articles of Association.

As a result of the share issuance in February 2022, the Company's nominal share capital is DKK 35,312,241 divided into shares of DKK 1 each or multiples thereof.

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, unless adopted in accordance with the procedures set out above.

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 and/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 ADS holders and other beneficial holders of our shares.

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 with 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.

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 (the "FSA") of 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 and/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 natural or legal person;
- (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 natural or legal person; or
- (viii) exercisable by that natural or legal 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.

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)

It should be noted that during the period from 16 March 2020 to 19 March 2021, ESMA decided to temporarily lower the notification threshold to 0.1% in light of the market unrest following the COVID-19 pandemic. From 20 March 2021 to 28 January 2022, the reporting limit was 0.2 %, until the European Commission decided to permanently lower the threshold for notification of net short positions to 0.1 %. Since 31 January 2022, the reporting limit has been 0.1 %. As a result, when a natural or legal person reaches, exceeds or falls below a net, short position of 0.1% 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 0.1% threshold. In addition, when a natural or legal person reaches or exceeds 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 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.

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.

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.

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 the board of directors or executive management or refrain from bringing law suits against, among other persons, a member of the board of directors or executive management, a shareholder may bring a derivative action on behalf of the company against, among other persons, a member of the 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.

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. The company may only pay out dividends from the company’s 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. The 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 the company’s financial condition or be to the detriment of the company’s 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 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.

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.



Asset Purchase Agreement

regarding certain assets in Orphazyme A/S in restructuring

Gorrissen Federspiel



This Asset Purchase Agreement (the "**Agreement**") is on this day entered into between:

- Seller:** Orphazyme A/S in restructuring, CVR no. 32 26 63 55, a company incorporated and registered under the laws of Denmark having its registered address at Ole Maaløes Vej 3, 2200 København, Denmark (the "**Seller**"); and
- Buyer:** KemPharm Denmark A/S, CVR no. 43259326, a company incorporated and registered under the laws of Denmark having its registered address at C/O Moalem Weittemeyer Advokatpartnersels Amaliegade 3, 1256 Copenhagen, Denmark (the "**Buyer**"); and
- Parent:** KemPharm Inc, a company incorporated and registered under the laws of the state of Delaware having its principal executive office at 1180 Celebration Blvd., Suite 103, Celebration, Florida 34747, USA of Delaware having its principal executive office at 1180 Celebration (the "**Parent**"), as owner of the total share capital in the Buyer.

The Seller and the Buyer are collectively referred to as the "**Parties**" and/or each a "**Party**".

WHEREAS

- (i) the Seller has agreed to sell, assign and transfer to the Buyer and the Buyer has agreed to purchase, accept assignment and assume from the Seller the Business (as defined below), excluding the Retained Assets, upon the terms and subject to the conditions herein set forth;
- (ii) the Seller is a late-stage biopharmaceutical company developing Arimoclomol for Niemann-Pick disease type C (NPC);
- (iii) Arimoclomol is an investigational drug candidate that amplifies the production of heat shock proteins (HSPs), which can rescue defective misfolded proteins and improve the function of lysosomes;
- (iii) the Buyer has been informed that the Seller is in restructuring pursuant to the Danish Bankruptcy Act (in Danish: "konkursloven");
- (iv) the purpose of this Agreement and the intention of the Parties are that the Buyer shall take over all relevant Assets of the Seller and – if possible and accepted by the relevant third parties – continue all early access programs on Arimoclomol and all processes of approval of Arimoclomol for commercial sale; and
- (v) on 4 September 2019 the Seller has granted a floating charge covering certain of the Assets (the "**Floating Charge**") (in Danish "virksomhedspant") to Kreos Capital VI (UK) Limited ("**Kreos**") of EUR 9,000,000.
- (vi) the Parent guarantees certain obligations of the Buyer under this Agreement.

The Parties have entered into the following Agreement regarding the purchase of the Business:

Gorrissen Federspiel



1 Certain Definitions

For the purpose of this business transfer agreement, unless the context otherwise requires or separately defined herein, the term

- 1.1 "Business" means the Seller's business activities as defined in this Agreement, including activities within the field of research, development, production, marketing, sale and/or licencing of medicinal products for the treatment of various diseases, including Lysosomal Storage Disease (LSD), neuromuscular diseases and related diseases, and other associated activities.
- 1.2 "Closing" means the completion of the sale and purchase of the Business and the other transactions provided for herein in accordance with Clause 9;
- 1.3 "Closing Date" means the date on which Closing occurs;
- 1.4 "EMA" means the European Medicines Agency.
- 1.5 "FDA" means the US Food and Drug Administration.
- 1.6 "French Revenue Liabilities" means any obligation to pay back revenue or other liability of the Seller and/or its Subsidiaries under the French Early Access Compassionate Use Program (ATU) relating to any sale performed under the French Early Access Compassionate Use Program performed both prior to the Closing Date by the Buyer and after the Closing Date by the Seller. The obligation relating to the time prior to the Closing Date is estimated to be maximum of \$5,200,000.
- 1.7 "Office Locations" means Ole Maaløes Vej 3, 2200 København, Denmark;
- 1.8 "Restructuring Proposal" means a restructuring proposal issued pursuant to section 13 b of the Danish Bankruptcy Act;
- 1.9 "Signing" means the signing of this Agreement on the date of this Agreement;
- 1.10 "Subsidiaries" means Orphazyme Schweiz GmbH and Orphazyme US Inc; and
- 1.11 "Transaction" means the actions and agreements contemplated by this Agreement.

2 Sale and Purchase of assets and assumption of liabilities and obligations

- 2.1 Subject to the terms and conditions of this Agreement, on the Closing Date, the Seller sells, assigns and transfers to the Buyer, and the Buyer purchases, accepts assignment and assumes from the Seller all of the Seller's right, title and interest in and to all assets free and clear of any charges, pledges and similar third party rights other than the Retained Assets of the Seller as the same exists on the Closing Date (the "**Assets**"), including, but not limited to the following assets and rights:



- 2.1.1 Fixed Tangible Assets: All fixed tangible assets owned by the Seller and relating to the Business as of the Closing Date, including machinery, operating equipment, installations, fixtures, fittings, office equipment and furniture, IT hardware (e.g. desktops, laptops, printers, servers, etc.), spare parts, tools, consumables and fittings, and including the fixed tangible assets specified in **Schedule 2.1.1** (the "**Fixed Tangible Assets**").
- 2.1.2 Inventory: All inventory of the Business owned by the Seller and which are located at the Office Locations, with suppliers, warehouses or in transit as of the Closing Date, including raw materials, ancillary materials, work in progress (including work in progress on behalf of third parties), finished goods and goods for sale, and including the inventory specified in **Schedule 2.1.2** (the "**Inventory**").
- 2.1.3 Records: Documentation and information of the Seller concerning the Business as of the Closing Date, including databases, customer and supplier lists, files, correspondence and other documentation, and including the documentation and information concerning the Business as of the Closing Date located at the locations specified in **Schedule 2.1.3** (the "**Records**"), save for as set out in clause 2.2.2. For the avoidance of doubt, the Seller is allowed to keep a copy of the Records.
- 2.1.4 Intellectual Property: All intellectual property rights related to NME Diaoxazines, NME Oximes, NME Pyridines (jointly "**Discovery Compounds**") and Arimoclomol, or in any other way relating to the Business, including trademarks, show-how, know-how, patents, copyrights, rights in business names, trade names, websites, other domain names, social media pages/accounts, telephone numbers (if assignable), historical and planned product developments for products forming part of the Business, trade secrets and know-how, and including the intellectual property specified in **Schedule 2.1.4** (the "**Intellectual Property**"). For the purpose of evidencing the transfer of the Intellectual Property from the Seller to the Buyer at public authorities such as the USPTO and/or EPO, the Seller is on the Closing Date obligated to execute and deliver to the Buyer the trademark and patent assignments as specified in clause 9.2.2.
- 2.1.4.1 The Buyer is, to a level consistent with the customary practices in the biotechnology and pharmaceutical industries, obliged to continue commercially reasonable efforts on developing Discovery Compounds for the purpose of having the drug approved and commercialised. If such efforts cannot be evidenced by the Buyer towards Seller no later than 48 months after Closing, the Buyer is obliged to reassign to the Seller all intellectual property rights relating to the Discovery Compounds, which were taken over from the Seller. To the extent the Buyer after Closing generates any new intellectual property rights related to Discovery Compounds, the Buyer retains full ownership to such new intellectual property, and such new intellectual property shall not be assigned to Seller. The Seller (or a third-party designated by the Seller) shall, however, for a period of 48 months after Closing be entitled to license such new intellectual property rights related to Discovery



Compounds from the Buyer on reasonable terms negotiated in good faith between the parties at such relevant point in time.

- 2.1.5 Goodwill and Reputation: The goodwill and reputation related to the Business, but not the name/trademark “Orphazyme”. The Seller acknowledges that the Buyer on a need to have basis and solely for the purpose of communication with patients, physicians, treatment centers, and for the purpose of distribution of Arimoclomol under early access programmes is entitled to use the name and trademark “Orphazyme” for a period of 24 month after Closing without the Seller being entitled to receive any royalty payment for such use, provided that such use is permitted under applicable law including regulatory requirements. The name may not be used by the Buyer in a way that could have an adverse effect for the Seller and the Buyer is liable towards the Seller for any unauthorized use of the name and/or any discredit caused by the Buyer’s use of the name. When/if using the name “Orphazyme” the Buyer shall clearly state that the Buyer does not represent or act on behalf of the Seller.
- 2.1.6 FDA and EMA applications on approval of Arimoclomol and patients early access program: To the extent possible, which is not guaranteed by the Seller in any way, the Buyer shall take over and continue all patients early access programs for the Seller worldwide and shall continue the ongoing approval processes at the FDA and EMA, subject to applicable consents and permissions. With respect to the FDA application, the Parties acknowledge that both Parties and their respective counsels, consultants and employees shall cooperate on regulatory matters pertaining to Arimoclomol prior to the meeting with the FDA on 14 July 2022. This cooperation includes to the extent necessary the sharing of data between the Parties, including confidential and privileged information. All costs for the Parties’ consultants and counsels assisting in this respect after Closing, on request of the Buyer, must be paid by the Buyer.
- 2.1.7 Permits: All of the Seller’s, if any, transferable permits, licenses, approvals, certifications, accreditations and other authorizations relating to the Business and/or the property located at the Office Locations, including those specified in **Schedule 2.1.7** to the extent assignable (the “Permits”).
- 2.1.8 The Assets mentioned in this clause 2.1 shall not be regarded as complete if the Parties before or after the Closing Date become aware of further assets which are not described in this Agreement, but which nevertheless form part of the Business. In that case such further assets shall on or after the Closing Date without any notice from the Buyer be transferred and delivered to the Buyer without this triggering further payments to the Seller, unless otherwise stated in the Agreement or agreed in writing between the Parties.
- 2.1.9 The Seller shall use all reasonable commercial efforts to assist the Buyer with the Business transfer in order for the Buyer to operate the Business immediately after the Closing Date as previously carried out by the Seller prior to Closing. The Parties both acknowledge the importance of start working closely together immediately after Signing on important matters, including but not limited to resubmissions. In addition, the Buyer acknowledges that Employees working with legal, finance and bookkeeping to a reasonable extent will have to assist the Seller in paying obligations of the Seller, communication with suppliers, preparation of annual report, disclosures etc. in expectedly the first 2 months after



Closing. The Seller shall not compensate the Buyer for this financially, unless the extent exceeds what can reasonably be expected of the Buyer. Similarly, the CEO of the Seller will assist the Buyer to the same extent for the first 2 months after Closing without any compensation from the Buyer. If further assistance is needed from the Seller's CEO, the Buyer and the CEO will have to discuss entering into a consultancy agreement. The Seller's CEO will not act as CEO for the Buyer during the term of his employment with the Seller.

2.1.9.1 The Seller shall, subject to any legal, regulatory or listing requirements of the Seller, after Closing procure that all references and data related to any of the transferred Assets will be removed from the Seller's website and similar public platforms/medias. The Seller shall also after Closing procure that all traffic on the Seller's website related to the early access programs in a reasonable way is redirected to the Buyer's website. However, Seller is not to remove issued company announcements and annual reports (even though information on transferred Assets appear from these) or other information required to be kept publicly available under any laws or regulations or listing requirement applicable to the Seller.

2.2 Notwithstanding the generality of Clause 2.1, the following assets etc. of the Seller (the "**Retained Assets**") shall not be transferred from the Seller to the Buyer. All costs, risks, liabilities and expenses related to the Business, attributable to or accrued before or on Closing shall, with the qualifications and limitations provided for in this Agreement, be for the account of the Seller and all costs, risks, liabilities and expenses related to the Business due, attributable to or accrued after Closing shall be for the account of the Buyer, provided that, for the avoidance of doubt, that Employee Related Liabilities shall, as of Closing, be for the account of the Buyer subject to the terms set out in clause 3:

2.2.1 The Seller's Good Distribution Practice (GDP) licence/permit(s) issued by the Danish Medicines Agency (in Danish "Lægemiddelstyrelsen"), Certificate No: DK GDP 10000177.

2.2.2 Bookkeeping and other documents, including the entire ERP-system relevant for the Seller to continue operating an active company and/or to perform solvent liquidation and/or to comply with applicable law, remains with the Seller; provided that the Seller will give the Buyer access to information necessary for the Buyer to continue the Business and early access programs and the application processes with the FDA and EMA. The e-mail domain and accounts "@ophazyme.com" and all e-mails send to and from any e-mail under this domain is not to be transferred to the Seller, unless these e-mails are stored in the cloud database to be transferred to the Seller, where all data relevant for the early access programs, studies and approval processes with EMA and FDA is store. The Seller will disclose any such information to the Buyer, provided that such disclosure shall be subject to clause 12.2 on confidentiality and the Buyer is obliged not to use the information in any way that could have an adverse effect for the Seller, including but not limited to the Seller's defense in law suits and Seller's compliance with stock exchange regulation.

2.2.3 The Seller's receivables for deliveries performed under the French Early Access Compassionate Use Program prior to the Closing Date are not transferred to the Buyer. Hereby, the Seller shall (from Clinigen Ireland Ltd) retain rights to all revenue for any sale under the French Early Access Compassionate Use Program performed before (but not including) the Closing Date (to be used to repay Kreos). On the contrary, revenue and receivables for sale performed after (and including) the Closing Date is to be received by



the Buyer (provided that the Buyer is allowed to continue the French Early Access Compassionate Use Program), which the Seller does not guarantee in any way.

- 2.2.4 Any other receivable, refund amount including but not limited to VAT returns, tax credit payments and tax deficits, or other claim for money of the Seller existing on the Closing Date, including but not limited to any amount of the Seller retained by or in other way owned by Clinigen Ireland Ltd. to the Seller relating to deliveries performed under the French Early Access Compassionate Use Program prior to the Closing Date.
- 2.2.5 Cash and money in bank accounts of the Seller.
- 2.2.6 The Seller's shares in Orphazyme Schweiz GmbH and Orphazyme US Inc. However, post Closing the Parties will discuss if some contracts are to be transferred from Orphazyme US Inc to the Buyer.
- 2.2.7 Internet and e-mail-domains/accounts including the name "Orphazyme". However, as for Employees, the Seller will keep their e-mail-accounts active for the first 4 months after Closing and hereby allow the Employees to use the e-mail accounts when working for the Buyer, until the Buyer has set up new e-mail-accounts for the Employees provided that Employee's signatures clearly state that the Buyer and not the Seller is the sender.
- 2.2.8 It is emphasised by the Seller that the Seller's lab room at Ole Maaløes Vej 3 and many office working stations are to be vacated before Closing. The lab equipment in the lab room and office equipment at vacated office stations are not transferred to the Buyer, as the Sellers will sell these assets to third party buyers prior to Closing when vacating the premises, which causes no adjustment of the Purchase Price. However, it is emphasised that the content of the Seller's freezer in the basement of Ole Maaløes Vej 3 and at any other third-party's storage facility shall also be transferred to the Buyer provided that the content relates to the transferred Business and Assets.
- 2.2.9 The laptops, telephones and other equipment specifically listed in **Schedule 2.2.9** and the telephone number/subscription of the Seller's CEO.
- 2.3 Subject to the terms and conditions of this Agreement, on the Closing Date, the Seller assigns and transfers to the Buyer, and the Buyer accepts assignment and assumes from the Seller the following liabilities and obligations of the Seller (the "**Assumed Liabilities**"):
 - 2.3.1 **Employee Related Liabilities:** The Seller's liabilities and obligations related to the Employees as described in Clause 3 (the "**Employee Related Liabilities**");
 - 2.3.2 **The French Revenue Liabilities:** The Buyer expects from a commercial perspective to have to satisfy all the Seller's French Revenue Liabilities to enable the Buyer to continue French Early Access Compassionate Use Program. On this basis, it has been a requirement of the Buyer to take over from the Seller and pay the French Revenue Liabilities directly to the receiver. The French Revenue Liabilities are expected to amount to a maximum of \$5,200,000 per Closing. However, no adjustment of the Purchase Price shall be made, if



the French Revenue Liabilities turns out to be of a higher or lower amount (or not payable at all).

- 2.3.3 All Seller's obligations under asset purchase agreement between CytRx Corporation and Seller of May 13, 2011 (**Schedule 2.3.3**), including milestone payments to CytRx Corporation pursuant to section 2.7 therein.
- 2.3.4 The Seller's obligation to perform a potential milestone payment of USD 1,000,000 to Hyman, Phelps & McNamara, P.C. in case of FDA Approval as defined in letter of July 7, 2021 (**Schedule 2.3.4**).
- 2.3.5 All rights and obligations under the lease agreement regarding Ole Maaløes Vej 3, 2200 København, Denmark, as the lease agreement is assigned from the Seller to the Buyer in all aspects. The Seller shall prior to or on the Closing Date deliver evidence that the landlord has approved the assignment of the lease agreement to the Buyer.
- 2.3.6 All rights and obligations of the Seller under any agreement relating to patients early access programs, studies, developments and approval of Arimoclomol and/or Discovery Compounds (including but not limited to leasing agreements, consultancy agreements, studies, licenses), and including those specified in **Schedule 2.3.6** (the "**Contracts**"), provided that (i) the Buyer has not in writing to the Seller declined to accept the assignment of a specific Contract no later than 28 June 2022 (any such declined Contracts shall be retained by the Seller) and (ii) that the agreements are assignable according to their terms and/or clause 14 c (2) of the Danish Bankruptcy Act or that the relevant contracting parties accepts assignment. The Buyer and the Seller shall both exercise reasonable efforts to ensure that the Contracts accepted by the Buyer are assigned and relevant consents are obtained and hereby that the Seller is released from any obligation of such Contract as per the Closing Date in accordance with Clause 2.5. However, the Buyer shall in good faith handle the dialogue, negotiation etc. with relevant contracting parties as the Buyer from Closing will have taken over the Employees from the Seller, whereby the Seller will have limited resources to assist in relation to such matters. However, the board and management of the Seller will assist by signing necessary documents etc. as set out in Clause 2.5.
- 2.4 Notwithstanding Clauses 2.1 and 2.3, the Buyer only acquires – except from sensitive personally information related to the Employees – non sensitive personal information, and no sensitive patient information is transferred to Buyer. The Parties are aware that the transfer of the Assets set out in this Agreement may constitute a transfer pursuant to the Danish Data Protection Act. The Parties agree that the transfer of the Assets can take place with title in the Danish Data Protection Act since there will only be transferred non-personally sensitive identity information. In the case there has been transferred personally sensitive information to the Buyer, and such transfer cannot take place with title in the Danish Data Protection Act or other relevant statutory law, then the Buyer shall not be authorised to use such sensitive personal information and shall be obliged to promptly delete such information as the time where the Buyer becomes aware hereof.
- 2.5 The ownership of, title to and risk of the Assets shall pass from the Seller to the Buyer on the Closing Date, and the Buyer shall accept from the Seller assignment of and assume the



Assumed Liabilities on the Closing Date. Hereby, from (and including) the Closing Date all obligations under the Assumed Liabilities rests with the Buyer. The Seller shall, however, remain liable for any actual or potential liability or obligation under the Assumed Liabilities relating to the period prior to the Closing Date, except from Employee Related Liabilities which are regulated by Clause 3.3. In addition, earnings on the Assets until the Closing Date belongs to the Seller (regardless if paid after the Closing Date), while earnings on the Assets (including and) after the Closing Date will belong to the Buyer. The Seller undertakes to assist to allow for transfer of any of the Assets and the Assumed Liabilities, including signing all required transfer documents and forms to effect the transfer. The payment of fees and registration duties shall be paid by the Buyer exclusively. The Buyer will handle the formalities and potential negotiations in relation to transfer of Assets and contracts as the Buyer will have taken over Seller's Employees.

2.6 The Assets shall be transferred to the Buyer without any charges or pledges. The Seller informs and warrants that the Seller will satisfy all of the Seller's obligations towards Kreos under the Floating Charge from the Purchase Price, meaning that the Buyer will take over the Assets free of Kreos' Floating Charge. As for potential liens in Inventory, the Seller will pay for storage and freight up to the Closing Date, while the Buyer will be obliged to pay for storage etc. after the Closing Date and/or to negotiate own terms and pay relevant costs for continues storage or release of the Inventory free from liens etc.

3 Employees

3.1 On the Closing Date, the Buyer shall assume the Seller's employment rights and duties towards the employees specified in **Schedule 3.1** (the "**Employees**"). The Employees will transfer in accordance with the Danish Business Transfer Act (in Danish: "*Lov om lønmodtageres retsstilling ved virksomhedsoverdragelse*").

3.2 In respect of any Employee listed in Schedule 3.1 not transferred pursuant to the provisions under the Danish Act on Transfers of Undertakings, the Parties shall in good faith cooperate to facilitate the transfer of such Employee(s) from the Seller to the Buyer, such transfer ultimately being subject to the acceptance by such Employee.

3.3 The employment relationships of the Employees shall be transferred to the Buyer on the same employment terms and conditions as applicable to the Employees at the Closing Date. As concerns the direct relation to the Employees, all obligations towards the Employees shall be taken over and assumed by the Buyer, including but not limited to earned salary, holiday and pensions pay accrued – but not settled – before the Closing Date to the extent that such obligations concern the period after the initiation of the restructuring process (in Danish: "*indledningen af rekonstruktionsbehandlingen*"). Notwithstanding aforesaid, the Seller shall reimburse the Buyer all incurred Employee costs to the extent that these costs concern – in whole or in part - the period before the Closing Date. Moreover, any retention bonus and bonus for a successful transfer of the Business offered to the Employees by the Seller from 11 March to the Closing Date is to be paid by the Seller (or reimbursed from the Seller to the Buyer if paid by the Buyer). Hence, the only obligation of the Seller towards the Employees after the Closing Date is to pay such bonus, as all other obligations rest with the Buyer and causes no reimbursement of the Seller towards the Buyer.



- 3.4 The Buyer has been informed that the Employees may be subject to the Seller's Long Term Incentive Program, which will terminate as of the Closing Date. The Buyer acknowledges that the Buyer after the Closing Date will seek to replace the Long Term Incentive Program with an incentive program on terms negotiated between the relevant Employees and the Buyer.
- 3.5 The Seller shall retain all liability in relation to employees of the Seller that are not listed in Schedule 3.1, including dismissed employees, save to the extent that the Buyer is liable for a dismissal executed in contravention of section 3(1) of the Danish Act on the Transfer of Undertakings ("Virksomhedsoverdragelsesloven").
- 4 Purchase Price**
- 4.1 The Purchase Price for the Assets (and taking into account the Assumed Liabilities) has been determined at \$ 12.8 million (the "**Purchase Price**"). On the Closing Date, the Purchase Price shall be paid by way of a payment of \$12.8 million in cash transferred by the Buyer to a bank account designated by the Seller prior to Closing.
- 4.2 The purchase price shall be paid in the currency of USD.
- 4.3 In accordance with section 45 (2) of the Danish Act on Depreciation (in Danish "Afskrivningslovens § 45, stk. 2") the allocation of the Purchase Price on the Assets appears from **Schedule 4.3**.
- 5 Representations and Warranties**
- 5.1 The Buyer is aware that the Seller is in restructuring pursuant to the Danish Bankruptcy Act. In that connection the Buyer has accepted that the Buyer cannot direct any claims or remedies towards the Seller in the event of any breach or lack of conformity, except for any defect in title relating to the Assets subject, however, to the disclosed material in the virtual data room, cf. Clause 6. The Parties agree that this has been taken into consideration when determining the Purchase Price.
- 5.2 The Seller does not undertake any representations or warranties, neither directly, implied nor written or oral, in respect of the Assets, which are transferred on an as-is basis, or in connection with this Agreement.
- 5.2.1 Notwithstanding Clause 5.2, the Seller represents and warrants that to the best of the Seller's knowledge (understood as the actual knowledge of the CEO and the general counsel of the Seller as per the date of Signing) without having carried out inquiries other than a review carried out by the Seller's external IP advisor, the information contained in Schedule 2.1.4 represents a full and complete list of the Seller's Intellectual Property related to the Business as transferred to the Buyer as part of the Assets. However, it is emphasised by Seller that some of the intellectual property rights listed in schedule 2.4.1 have expired.
- 5.3 If the Buyer notwithstanding the provisions of this Clause 5 is entitled to direct a claim or remedy towards the Seller, the Parties agree that such claim or remedy only can be made in the form of a claim for a proportional reduction of the Purchase Price by deduction of the value of the Asset in question under this Agreement from the purchase price and



subject to the rules under Danish law on pro rata reduction (in Danish: “forholdsmæssigt afslag”). Such claims are in the aggregate maximized to a sum corresponding to the Purchase Price and as effectively paid to the Seller.

5.4 Any and all claims pursuant to or arising out of this Agreement will lapse, if the Buyer does not take documented legal actions in respect of such claims and notifies the Seller hereof in writing within 6 months of the Closing Date.

5.5 The remedy in this Clause 5 is the exclusive remedy available to the Buyer. In particular, the Buyer is not entitled to cancel or rescind the Agreement or to claim damages.

6 Due diligence

6.1 The Buyer has been given the opportunity to conduct a due diligence investigation from the virtual data room Gamora at the data site Datasite.com.

6.2 The Parties agree that all material in the data room is considered disclosed and to the Buyer’s knowledge as of the Closing Date to the extent that the matter was disclosed in a way which a prudent buyer should reasonable be expected to understand the repercussions of. The Buyer confirms that the Seller has answered all questions raised by the Buyer during the diligence investigation and that such answers to the Buyer’s knowledge have been adequate and sufficient. The Buyer further confirms as per Signing not to be aware of any matter which could constitute a breach under this Agreement.

7 Covenants between Signing and Closing

7.1 Conduct of Business

7.1.1 During the period from Signing until Closing the Seller shall:

7.1.1.1 Operate the Business in the ordinary course of business consistent with past practice and in accordance with current legislation;

7.1.1.2 Not without the prior written consent of the Buyer terminate or materially amend any terms in the Contracts, including renegotiating or prolonging such agreements with terms less attractive to the Business than applicable as per Signing;

7.1.1.3 Not without the prior written consent of the Buyer materially change the employment terms or terminate the employment of or make redundant any of the Employees; and

7.1.1.4 Not without the prior written consent of the Buyer dispose of any of the Assets existing as of the Signing Date.

7.1.1.5 Procure that any annuities, prosecution matters and any other intellectual activity, that needs to be effectuated to prevent any harm, damage or loss of rights with respect to the Intellectual Property listed in schedule 2.1.4 is duly handled by the Seller.

8 Conditions to Closing

8.1 The Closing shall not occur as set forth in Clause 9 unless the restructuring proceedings of the Seller has been completed on one of following two ways:



- (a) The Restructuring Proposal has been approved by a simple majority (measured by amounts) of the creditors of the Seller attending and able to vote on the Restructuring Proposal at a creditor's meeting, cf. Section 13 d of the Danish Bankruptcy Act (konkurslovens § 13 d) and the subsequent affirmation of the Restructuring Proposal by the Danish Bankruptcy Court, cf. Section 13 e of the Danish Bankruptcy Act (konkurslovens § 13 e), or
- (b) The bankruptcy court has formally determined that the restructuring proceedings are terminated pursuant to section 15(1)(i) of the Danish Bankruptcy Act, allowing the restructuring proceedings to stop if the Seller is no longer insolvent.

8.2 If the conditions in Clause 8.1 has not been satisfied at Closing, none of the Parties are obliged by this Agreement and the Parties shall not have any claims of any kind towards each other based on this Agreement (other than as set forth in Clause 9.4), and this Agreement shall terminate. However, the Parties will in this situation in good faith seek to negotiate on potential alternative solutions.

9 Closing

9.1 Closing shall take place at 0.01 (CET), on 1 June 2022, or at such other place, date or time as may be agreed between the Parties.

9.2 On the Closing Date, the Seller shall deliver evidence to the Buyer that the landlord has approved the assignment of the lease agreement to the Buyer, cf. clause 2.3.5.

9.2.1 On the Closing Date, the Seller shall deliver a USB-stick to the Buyer with a copy of the virtual data room, together with written confirmation that the virtual data room has been closed and that no new documentation has been included in the virtual data room after Signing.

9.2.2 On Closing Date, the Seller shall deliver to the Buyer executed trademark and patent assignments in the form attached as **Schedule 9.2.2(a)** and **Schedule 9.2.2(b)**. The executed trademark and patent assignments shall be deliver by an international delivery service to Buyer's legal intellectual property representative, McAndrews, Held & Malloy, 500 West Madison Street, 34th Floor, Chicago, Illinois 60661, United States to the attention of Mr. Troy Groetken.

9.3 On the Closing Date, immediately following receipt by the Seller of the Purchase Price, the Seller shall satisfy all of the Seller's obligations towards Kreos under the Floating Charge, meaning that the Buyer shall take over the Assets free of Kreos' Floating Charge.

9.4 In case of any termination of this Agreement pursuant to Clause 8.2, this Agreement (other than Clauses 10 (Expenses), 12 (Confidentiality) and 19 (Governing Law and Venue)) shall terminate. Nothing in this Clause 9.4 shall be deemed to release either Party



from any liability for any breach by such Party of the terms of this Agreement prior to termination.

10 Financial Statements

- 10.1 After Closing, the Seller shall deliver to the Buyer as promptly as practicable, (i) final audited financial statements for the year ended December 31, 2021¹, (ii) unaudited financial statements as of March 31, 2022, taking into account adjustments required by Regulation S-X (which shall have been reviewed by Seller's independent accountants in accordance with the Statement on Auditing Standards No. 100) (the "Financial Statements").
- 10.2 The Financial Statements shall be prepared in accordance with IFRS applied on a consistent basis throughout the periods covered thereby and the books and records of Seller, in a manner consistent with the preparation of the historical financial statements for the equivalent periods, including with respect to (i) footnote disclosure (except, in the case of interim financial statements, such exceptions and omitted footnotes as are customary in the preparation of interim financial statements) and (ii) the audit or review, as applicable, of Seller's independent accountants.
- 10.3 The Seller agrees to consent to the inclusion of any financial statements or information obtained by the Buyer in any filings by the Buyer with the SEC or any other securities regulatory authority or exchange.
- 10.4 The Seller agrees that it will use reasonable best efforts to cause its independent accountants to provide to the Buyer with a consent for the use of its report relating to any financial statements or information obtained by the Buyer pursuant to this section in any SEC filings by or on behalf of the Buyer.
- 10.5 The Seller shall reasonably cooperate with the Buyer, and shall cause its affiliates to reasonably cooperate with the Buyer, with respect to any financial statements (including pro forma financial statements) that the Buyer deems reasonably necessary in order to make any filing required by applicable Law or stock exchange requirements, including filings with the SEC.
- 10.6 The Seller's obligations under this clause 10 are in all aspects subject to restrictions and limitations of applicable law and stock exchange regulation of the Seller. The principles of clause 2.2.2 last sentence equally apply to information disclosed pursuant to this clause 10.
- 10.7 Information etc. pursuant to this clause 10 are disclosed from the Seller to the Buyer free of charge, if such information in any case are to be prepared by the Seller to comply with applicable law and stock exchange regulation of the Seller. To the extent the Seller is to prepare additional information to satisfy the terms of this clause 10, the Buyer will be obliged to compensate the Seller for any reasonable related costs.



11 Expenses

11.1 Each Party shall bear its own costs and expenses, including but not limited to fees to legal, financial and other advisors and representatives, in relation to the negotiation, preparation, execution and carrying into effect of this Agreement and other agreements referred to hereby.

12 Confidentiality

12.1 Pursuant to Section 13 b of the Danish Bankruptcy Act the terms of this Agreement will to a certain extent be disclosed to the creditors of the Seller in the Restructuring Proposal (unless the restructuring process is terminated pursuant to Clause 8.1(b) without presenting a Restructuring Proposal to the creditors). Such information shall not be considered confidential information. The Seller shall provide the Buyer with a copy of the Restructuring Proposal at the same time as such proposal is sent to the creditors of the Seller to allow the Buyer immediately to send out a stock exchange announcement, if and as required.

12.2 Except from information disclosed in the Restructuring Proposal, the Parties agree that this Agreement and all information related to the transfer is to be handled in confidence and shall not be disclosed to any third party without the prior written consent of the other Party, cf. however Clause 13, unless required by law or applicable stock exchange regulation.

13 VAT

13.1 The Parties agree that the Asset Transfer is a transfer of an existing business, and hence VAT exempt. Consequently, the Seller may notify the transfer to the Danish tax authorities, stating the name and address of the Buyer and the purchase price for the Assets in accordance with Section 1(5) of the Danish VAT Act (in Danish: "momsloven").

13.2 If the Danish Tax Authority finds that the transfer is subject to VAT, the Buyer pays VAT on the transfer.

13.3 The Buyer takes over any VAT adjustment obligations (in Danish: "momsreguleringsforpligtelser") on the Assets without this giving rise to a deduction in the purchase price and signs all documents necessary in relation hereto.

14 Severability

14.1 Should one or more of the provisions of this Agreement cease to apply or be modified as a result of invalidity, voidability or for other reason, this shall not affect the validity of the remaining provisions of this Agreement.

14.2 If one or more of the provisions of this Agreement are held to be contrary to Danish Law, the Parties agree that such provision(s) shall, to the extent possible, be modified and shall apply with such contents as may be validly agreed (seeking to maintain as much of the original intentions as possible) and that the remaining provisions of this Agreement shall still apply.

15 No assignment

15.1 Except as otherwise specifically set forth in this Agreement, this Agreement and any right or obligation hereunder may not be assigned, in whole or in part, by any Party without the consent of the other Party.



16 Waiver and amendments

16.1 This Agreement may be amended and the terms hereof may be waived only by written instrument signed by the Parties or in the case of a waiver, by the Party waiving its rights under this Agreement.

17 Headings

17.1 The headings inserted are for convenience and reference only and shall not be used to construe or interpret this Agreement.

18 Interpretation

18.1 The Parties have participated jointly in the negotiation and drafting of this Agreement. In the event of any ambiguity or question of intent or interpretation arises, this Agreement shall be construed as if drafted jointly by the Parties and no presumption or burden of proof shall arise favouring or disfavouing any Party by virtue of the authorship of any of the provisions of this Agreement.

19 Guarantee from Parent

19.1 The Parent, as a primary obligor, guarantees in all aspects the Buyer's obligations under clauses 2.3, 3, 4 and 13, but not other obligations of the Buyer under this Agreement.

20 Governing Law and Venue

20.1 This Agreement shall be governed by, and shall be construed in accordance with, the laws of Denmark, excluding its rules on choice of law.

20.2 Any dispute in connection with this Agreement shall be finally settled by arbitration in accordance with the Rules of Procedure of the Danish Institute of Arbitration (Danish Arbitration) and the Danish Arbitration Act (in Danish: voldgiftsloven). The place of the arbitration shall be Copenhagen and the language of the arbitration shall be English. The arbitration tribunal shall be composed of three arbitrators. The Buyer and the Seller shall each appoint one arbitrator. The Danish Institute of Arbitration shall appoint the third arbitrator, who shall be the chairman of the arbitration tribunal.

20.3 The arbitration award and the arbitration proceedings are confidential, and the Parties must not make any statements to the public as to the dispute, its outcome or the arbitration proceedings, save for as provided in Clause 12.

-oOo-

[Signatures on separate pages]



For Orphazyme A/S in restructuring as the Seller:

Name: Anders Vadsholt

Name: Georges Gemayel

Approved by the reconstructor:

Name: John Sommer Schmidt

Gorrissen Federspiel



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For KemPharm Denmark A/S as the Buyer:

Name:

Name:

For KemPharm Inc as Parent pursuant to clause 19:

Name:

Name:

Gorrissen Federspiel

21 April 2021 | U.S. participants

Orphazyme A/S – Long-Term Incentive Programme

General Terms and Conditions for Restricted Share Units and Performance Share Units

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These are the general terms and conditions (the “General Terms and Conditions”) for the 2021 long-term incentive program (the “LTIP”) of Orphazyme A/S, CVR no. 32266355 (the “**Company**”) and its subsidiary undertakings (the Company and its subsidiary undertakings are referred to as the “**Group**” and each a “**Group Member**”).

The General Terms and Conditions apply to grants of restricted share units (“**Restricted Share Units**” or “**RSUs**”) and performance share units (“**Performance Share Units**” or “**PSUs**”) under the LTIP awarded to members of the executive management, registered as such with the Danish Business Authority, and certain key employees of the Group (such executives and employees referred to as “**Participants**” and each a “**Participant**”).

1 Introduction

1.1 The LTIP has been established by the Company’s board of directors (the “**Board of Directors**”) and has been adopted in accordance with the Company’s remuneration policy adopted by the Company’s annual general meeting on 25 March 2021.

1.2 Participation in the Group’s LTIP and in each individual grant thereunder is resolved by the Board of Directors at its sole discretion from time to time in connection with each grant.

1.3 The LTIP comprises the following documents:

- (a) These General Terms and Conditions;
- (b) Individual Grant Letter (as defined below);
- (c) Agreement on the application of section 7P of the Danish Tax Assessment Act; and
- (d) Danish Share Option Act statement for Participants comprised by this act.

In case of any discrepancy between the above documents, these General Terms and Conditions shall prevail. To the extent any of the documents exist in other languages than English, the English version shall prevail, except for the Danish Share Option Act statement where the Danish version shall prevail.

1.4 If eligible for participation under the LTIP, the Participant will receive an individual grant letter setting out the individual terms applicable to the grant of RSUs and PSUs (as defined below) (the “**Individual Grant Letter**”). The Individual Grant Letter will, inter alia, state the number of RSUs and PSUs granted and the conditions for vesting of the RSUs and PSUs, in whole or in part, including any performance targets for the PSUs as defined by the Board of Directors (each a “**Performance Target**” and, if more than one, collectively the “**Performance Targets**”). If a Participant is entitled to a fraction of a share in the Company as part of the LTIP, the number of shares to such Participant will be rounded down to the nearest integer number.

1.5 The purpose of the LTIP is mainly, but not only, to align the interests of the Participants with the interests of the Company’s shareholders while also encouraging Participants to remain employed in the Group. Further, the granting of PSUs under the LTIP aims to support the performance culture in the Group by rewarding the Participants in proportion to the achievement of one or more Performance Targets. Any Performance Target will be set prior to any grants and communicated to Participants in the Individual Grant Letters. Assuming the other vesting conditions are met, achieving one or more Performance Targets will contribute to the vesting of PSUs by the proportion set out in the Individual Grant Letters.

- 1.6 The LTIP is forward-looking in that grants of RSUs and PSUs are not based on previous performance of the Company or the Participant, and vesting is dependent on satisfaction of certain vesting criteria, including one or more Performance Targets, which are forward-looking.
- 1.7 These General Terms and Conditions govern grants made under the LTIP in 2021. Grants made after 1 January 2022 will be governed by the general terms and conditions applicable at the time of such grants.
- 2 Restricted Share Units (RSUs)**
- 2.1 When vested and not lapsed pursuant to these General Terms and Conditions, RSUs will, subject to fulfilment of certain conditions set out in Clause 2.3 below, entail that the Participant receives a number of shares equivalent to the number of RSUs vested against payment (in a manner and within a period decided by the Board of Directors) of the par value of each share in accordance with the terms and conditions outlined herein and in the Individual Grant Letter.
- 2.2 The Participant will be entitled to receive RSUs based on the volume weighted average price (VWAP) of the Company's shares as quoted on Nasdaq Copenhagen during a period of ten (10) trading days preceding the date stipulated in the Individual Grant Letter.
- 2.3 Conditions for vesting of RSUs
- 2.3.1 As stipulated in the Individual Grant Letter, the Participant will be entitled to receive RSUs that have a total vesting period of approximately three (3) years from the date of the grant or such other date as stipulated in the Individual Grant Letter with one third of the granted RSUs vesting on each such date as stipulated in the Individual Grant Letter in the following three (3) financial years (each period is referred to as a "**RSU Vesting Period**" and the total continuous period of approximately three (3) years from the date of the grant as the "**Total RSU Vesting Period**"), subject to the satisfaction of each of the following conditions:
- (a) The Participant remaining employed with a Group Member throughout the Total RSU Vesting Period, or the Participant becoming a Good Leaver (as defined below) during the Total RSU Vesting Period in which case the Participant will be entitled to keep any vested RSUs and receive a pro rata allocation in accordance with Clause 2.5.1 below; and
 - (b) The Participant having complied in all respects with the General Terms and Conditions including Clause 2.5.3.
- 2.3.2 Any grant of RSUs or delivery of shares underlying such RSUs upon vesting must comply with applicable statutory requirements and regulations, and no such grant may be made if the Board of Directors determines, at its sole discretion, that such grant constitutes a breach of current statutory requirements or regulations.
- 2.3.3 In the event and to the extent any of the above conditions are not satisfied, any and all rights to receive shares underlying RSUs shall terminate and lapse without further notice and without any rights of compensation, unless the Board of Directors resolves otherwise at its sole discretion.
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- 2.4 Vesting of RSUs
- 2.4.1 Subject to satisfaction of the conditions set out in Clause 2.3.1, during and upon expiration of the Total RSU Vesting Period, the Participant will receive a number of shares in the Company equivalent to the number of vested RSUs against payment of the par value of each share in accordance with and to the extent set out in these General Terms and Conditions.
- 2.4.2 The delivery of shares by the Company to the Participant after the Total RSU Vesting Period will be effectuated in a manner determined by the Board of Directors.
- 2.4.3 In order to be eligible to receive the vested RSUs, as set out in Clause 2.4.2, the Participant shall comply with the practical instructions provided by the Company. If the Participant does not timely comply with the instructions provided by the Company, the Company will be entitled to consider the right to receive shares underlying vested RSUs as having lapsed without further notice and without compensation to the Participant. Neither the Company nor any other Group Member or their representatives may be held liable for any failure to deliver shares underlying vested RSUs to the Participant due to the Participant not complying with the instructions provided.
- 2.4.4 Delivery by the Company to the Participant of the shares underlying vested RSUs will be effected on or after the date following the satisfaction of the Total RSU Vesting Period but not later than sixty (60) days thereafter (the “**RSU Delivery Period**”). The RSU Delivery Period is automatically extended to the next open trading window if the settlement of the RSUs or delivery of shares underlying vested RSUs would contravene applicable laws, rules or regulations, including, but not limited to, situations where the delivery of shares underlying vested RSUs cannot occur due to the Company being in possession of inside information, but in no event shall such delivery, with respect to any United States taxpayer, occur beyond 15 March of the calendar year following the year in which the Total RSU Vesting Period is satisfied. If the Participant has not paid the applicable subscription price, or has not complied with the General Terms and Conditions and the applicable Individual Grant Letter, in any case, as of the last day of the RSU Delivery Period, then the Participant’s right to receive shares underlying vested RSUs shall be forfeited with no compensation or other payment due to the Participant or any other person or entity.
- 2.4.5 The Participant will not be deemed to be the owner or holder of ownership rights or any other rights in respect of the RSUs in the Company until (i) the RSUs have vested and the instructions to receive the RSUs have been complied with, and (ii) the Participant’s ownership of the shares underlying the vested RSUs has been registered in the Company’s shareholders’ register.
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- 2.5 Continued employment or Good Leaver
- 2.5.1 In case of termination of the Participant's employment with the Group Member due to the Participant's resignation due to the Group Member's material breach of contract; (ii) the Group Member's termination of the Participant without such termination being due to the Participant's breach of obligations towards the Group Member; or (iii) the Participant being deceased (the Participant will in each situation mentioned under (i)-(iii) be considered a "**Good Leaver**"), the Participant (or the deceased Participant's estate, as the case may be) will remain entitled to the Participant's RSUs vested at the date of release of the Participant's work obligations (in Danish: "*fritstillingstidspunktet*") subject to and in accordance with the Individual Grant Letter and the General Terms and Conditions. For any ongoing RSU Vesting Period, a Good Leaver will be entitled to receive a pro rata allocation of RSUs for the period from commencement of the ongoing RSU Vesting Period and until the date of release of the Participant's work obligations (in Danish: "*fritstillingstidspunktet*").
- 2.5.2 In the event of termination of employment for any other reason than stated in Clause 2.5.1, including due to the Participant's own resignation or material breach of obligations towards the Group Member then any vested and/or unvested RSUs will lapse without further notice and without any rights of compensation as of the date of release of the Participant's work obligations (in Danish: "*fritstillingstidspunktet*"), unless otherwise decided by the Board of Directors.
- 2.5.3 Further, any vested and unvested RSUs will lapse without further notice and without any rights of compensation if, during the Participant's employment with the Group Member, or after the termination of the Participant's employment while the Participant holds vested and/or unvested RSUs, the Participant (i) violates a non-competition and/or non-solicitation clause in the Participant's employment contract or service agreement or otherwise breaches the duty of loyalty towards the Group Member, (ii) discloses or otherwise misuses any confidential information, whether written or oral, including, without limitation, financial information, trade secrets and other proprietary business information regarding the Group Member, (iii) violates the Group Member's compliance policies, or (iv) violates the Group Member's accounting rules including the financial reporting rules.
- 3 Performance Share Units (PSUs)**
- 3.1 When vested and not lapsed pursuant to these General Terms and Conditions, PSUs will, subject to fulfilment of certain conditions set out in Clause 3.3 below and in the Individual Grant Letter, entail that the Participant receives a number of shares equivalent to the number of PSUs vested against payment (in a manner and within a period decided by the Board of Directors) of the par value of each share in accordance with the terms and conditions outlined herein and in the Individual Grant Letter.
- 3.2 The Participant will be entitled to receive PSUs based on the volume weighted average price (VWAP) of the Company's shares as quoted on Nasdaq Copenhagen during a period of ten (10) trading days preceding the date stipulated in the Individual Grant Letter.
- 3.3 Conditions for vesting of PSUs
- 3.3.1 As stipulated in the Individual Grant Letter, the Participant will be entitled to receive PSUs that have a vesting period of approximately three (3) years from the date of the grant or such other date as stipulated in the Individual Grant Letter, with the granted PSUs vesting, in whole or in part, on such date as stipulated in the Individual Grant
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Letter in the third financial year following the date of the grant (the “**PSU Vesting Period**”), subject to the satisfaction of each of the following conditions:

- (a) The Participant remaining employed with a Group Member throughout the PSU Vesting Period, or the Participant becoming a Good Leaver (as defined below) during the PSU Vesting Period in which case the Participant may be entitled to keep a proportion of the PSUs in accordance with Clause 3.5.1 below;
- (b) Achievement of one or more Performance Targets, during the PSU Vesting Period, as set out in the Individual Grant Letter (if more than one Performance Target has been defined by the Board of Directors, achievement of each Performance Target will contribute to the vesting of PSUs by the proportion set out in the Individual Grant Letter); and
- (c) The Participant having complied in all respects with the General Terms and Conditions.

3.3.2 Any grant of PSUs or delivery of shares underlying such PSUs upon vesting must comply with applicable statutory requirements and regulations, and no such grant may be made if the Board of Directors determines, at its sole discretion, that such grant constitutes a breach of current statutory requirements or regulations.

3.3.3 In the event and to the extent any of the above conditions are not satisfied, any and all rights to receive shares underlying PSUs shall terminate and lapse without further notice and without any rights of compensation, unless the Board of Directors resolves otherwise at its sole discretion.

3.4 Vesting of PSUs

3.4.1 Subject to satisfaction of the conditions set out in Clause 3.3.1 and in the Individual Grant Letter, during and upon expiration of the PSU Vesting Period, the Participant will receive a number of shares in the Company equivalent to the number of vested PSUs against payment of the par value of each share in accordance with and to the extent set out in these General Terms and Conditions and in the Individual Grant Letter. The Board of Directors will determine if and when any Performance Target has been met and make an announcement to that effect no later than four (4) weeks after the expiration of the PSU Vesting Period.

3.4.2 The delivery of shares by the Company to the Participant after the PSU Vesting Period will be effectuated in a manner determined by the Board of Directors.

3.4.3 In order to be eligible to receive the vested PSUs, as set out in Clause 3.4.2, the Participant shall comply with the practical instructions provided by the Company. If the Participant does not timely comply with the instructions provided by the Company, the Company will be entitled to consider the right to receive shares underlying vested PSUs as having lapsed without further notice and without compensation to the Participant. Neither the Company nor any other Group Member or their representatives may be held liable for any failure to deliver shares underlying vested PSUs to the Participant due to the Participant not complying with the instructions provided.

- 3.4.4 Delivery by the Company to the Participant of the shares underlying vested PSUs will be effected on or after the date following the satisfaction of the PSU Vesting Period but not later than sixty (60) days thereafter (the “**PSU Delivery Period**”). The PSU Delivery Period is automatically extended to the next open trading window if the settlement of the PSUs or delivery of shares underlying vested PSUs would contravene applicable laws, rules or regulations, including, but not limited to, situations where the delivery of shares underlying vested PSUs cannot occur due to the Company being in possession of inside information, but in no event shall such delivery, with respect to any United States taxpayer, occur beyond 15 March of the calendar year following the year in which the PSU Vesting Period is satisfied. If the Participant has not paid the applicable subscription price, or has not complied with the General Terms and Conditions and the applicable Individual Grant Letter, in any case, as of the last day of the PSU Delivery Period, then the Participant’s right to receive shares underlying vested PSUs shall be forfeited with no compensation or other payment due to the Participant or any other person or entity.
- 3.4.5 The Participant will not be deemed to be the owner or holder of ownership rights or any other rights in respect of the PSUs in the Company until (i) the PSUs have vested and the instructions to receive the PSUs have been complied with, and (ii) the Participant’s ownership of the shares underlying the vested PSUs has been registered in the Company’s shareholders’ register.
- 3.5 Continued employment or Good Leaver
- 3.5.1 In case of termination of the Participant’s employment with the Group Member due to (i) the Participant’s resignation due to the Group Member’s material breach of contract; (ii) the Group Member’s termination of the Participant without such termination being due to the Participant’s breach of obligations towards the Group Member; or (iii) the Participant being deceased (the Participant will in each situation mentioned under (i)-(iii) be considered a “**Good Leaver**”), the Participant (or the deceased Participant’s estate, as the case may be) will remain entitled to keep such proportion of the Participant’s PSUs which have one or more Performance Targets that have been achieved at the date of release of the Participant’s work obligations (in Danish: “*fritstillingstidspunktet*”) and such portion of the Participant’s PSUs shall vest subject to and in accordance with the Individual Grant Letter and the General Terms and Conditions.
- 3.5.2 In the event of termination of employment for any other reason than stated in Clause 3.5.1, including due to the Participant’s own resignation or material breach of obligations towards the Group Member then all PSUs will lapse without further notice and without any rights of compensation as of the date of release of the Participant’s work obligations (in Danish: “*fritstillingstidspunktet*”), unless otherwise decided by the Board of Directors.
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- 3.5.3 Further, any vested and unvested PSUs will lapse without further notice and without any rights of compensation if, during the Participant's employment with the Group Member, or after the termination of the Participant's employment while the Participant holds vested but not settled PSUs, the Participant (i) violates a non-competition and/or non-solicitation clause in the Participant's employment contract or service agreement or otherwise breaches the duty of loyalty towards the Group Member, (ii) discloses or otherwise misuses any confidential information, whether written or oral, including, without limitation, financial information, trade secrets and other proprietary business information regarding the Group Member, (iii) violates the Group Member's compliance policies, or (iv) violates the Group Member's accounting rules including the financial reporting rules.

4 Cash settlement

- 4.1 In cases where the Board of Directors assesses that the issue or transfer of shares would have an adverse effect on the Company and/or the Participant, the Board of Directors may choose to cash settle the RSUs and/or PSUs instead of allowing the Participant to subscribe for or receive shares upon expiry of the Total RSU Vesting Period and the PSU Vesting Period, respectively. In such event, the Company or the Group Member, as applicable, shall pay a cash settlement amount based on the volume weighted average price of the Company's shares as quoted on Nasdaq Copenhagen during the ten (10) trading days preceding the date of expiry of the Total RSU Vesting Period and the PSU Vesting Period, respectively, with a deduction of the subscription price. The Company and/or the Group Member, as applicable, is entitled to deduct any tax withholding amounts in the cash settlement amount. Cash settlement shall be made at the same time as shares underlying vested RSUs and PSUs (as applicable) would have been delivered, as described in Sections 2.4.4 and 3.4.4, respectively.

5 Claw back

- 5.1 If the Board of Directors concludes in good faith that the basis for granting the RSUs and/or PSUs or that the conditions for vesting of any RSUs and/or PSUs are based on fraud, willful misconduct, gross negligence, incorrect or misleading information, or that the conditions have otherwise not been fulfilled, the Company shall be entitled (a) to consider the RSUs and/or PSUs as lapsed without compensation to the Participant, (b) to recalculate and/or adjust accordingly the number of RSUs and/or PSUs to be granted and/or (c) to require redelivery to the Company of any shares having been delivered to the Participant after vesting of the RSUs and/or PSUs or repayment of the cash settlement amount, as applicable.

6 Adjustments in case of changes to the Company's capital structure

- 6.1 In order to ensure that the value of the rights under the RSUs and PSUs is duly protected in the event of changes to the Company's capital structure, the Board of Directors may, at its sole discretion, adopt changes to the number of granted RSUs and/or PSUs, inter alia, in the case of the following:

- (a) after changes have been made to the nominal value of the shares of the Company;
 - (b) after the Company's share capital has been increased at a price lower than market price other than capital increases at a price lower than market price offered to board members, executives or other employees of the Company in connection with the LTIP or the share-based incentive program for the Board of Directors or any existing or subsequent incentive plans, including general employee share purchase plans;
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- (c) after the Company's share capital has been increased with pre-emption rights for the Company's existing shareholders allowing them to purchase shares at a price lower than market price;
- (d) after the Company has issued or granted convertible bonds or other convertible loans, stock options (except for RSUs, PSUs and Performance Shares under the LTIP or the share-based incentive program for the Board of Directors, or any existing or subsequent share based incentive plans, including general employee share purchase plans), and such issue or grant has been made with pre-emption rights for the existing shareholders at a lower price than market price;
- (e) after the Company's share capital has been reduced for any other purpose than to cover losses or through distribution to the Company's shareholders in the ordinary course of business through cancellation of shares acquired in treasury through share buy-back programmes established by the Company from time to time and in line with the Company's dividend policy (as amended from time to time); or
- (f) after distribution of extraordinary dividends during a financial year in excess of the Company's dividend policy (as amended from time to time).

Any such adoption of changes to the number of RSUs and/or PSUs or shares to be received in the Individual Grant Letters will seek to achieve that the Participant receives a reasonable compensation for the adjustment of the value of the RSUs and/or PSUs caused by the situation in question. The Board of Directors may decide that the aforementioned compensation is made by adjusting the number of RSUs and/or PSUs granted or by cash-settling the difference in value caused by the changes to the Company's capital structure outlined in 6.1(a)-(f) above.

Any adjustment made from time to time pursuant to these General Terms and Conditions will be notified to the Participant in writing.

If events affecting the share capital in the Company occur which are comparable in nature to the events outlined in 6.1(a)-(f) above, and with similar effect, the Board of Directors may decide to treat the event as if comprised by 6.1(a)-(f) and adjust the number of RSUs and/or PSUs set out in the Individual Grant Letters accordingly.

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7.1

Takeover offer or delisting

If

- (a) a mandatory takeover bid concerning the Company's shares is to be made according to the rules of the Danish Capital Markets Act (as amended, supplemented or replaced from time to time), or



- (b) a voluntary takeover bid is made concerning the Company's shares, in which connection there is a change in the controlling interest (as defined in section 44 of the Danish Capital Markets Act (as amended, supplemented or replaced from time to time)) to a third party independent of the Company, or
- (c) otherwise any transaction whereby a third party independent of the Company obtains control of more than 50% of the Company's shares, or
- (d) the shares in the Company are delisted,

the Board of Directors shall be required to accelerate vesting of the RSUs at a time determined by the Board of Directors in relation to acceptance of such takeover offer or other transaction. Any such accelerated RSUs shall be settled within sixty (60) days after vesting. To the extent necessary to prevent the imposition of taxes or penalties under Section 409A (as defined below), in no event will the consummation of any such takeover or other transaction be deemed to be a takeover or other transaction for purposes of this Section 7 if such transaction is not also a "change in the ownership or effective control of" the Company or "a change in the ownership of a substantial portion of the assets of" the Company as determined under Treasury Regulation Section 1.409A-3(i)(5) (without regard to any alternative definition thereunder).

- 7.2 In the situations listed under Section 7.1(a)-(d) above, the Board of Directors shall further in relation to acceptance of such takeover offer or other transaction be required to accelerate vesting of the PSUs to the extent that any applicable Performance Targets have been achieved. In case a Performance Target has not been achieved at the time of such accelerated vesting, the PSUs pertaining hereto will lapse, unless otherwise decided by the Board of Directors. Any such accelerated PSUs shall be settled within sixty (60) days after vesting.

8 Assignment

- 8.1 The RSUs and PSUs or the right to receive shares upon vesting of the RSUs and/or PSUs may not be transferred or assigned to a third party or pledged, encumbered or placed as collateral with a third party, except from transfers to the Company or a third party designated by the Company. Furthermore, any rights to receive RSUs and PSUs that are transferred or sought to be transferred to a third party as a consequence of creditor suit or statutory execution will lapse automatically without further notice.
- 8.2 Shares received by the Participant upon vesting of the RSUs and/or PSUs shall not be subject to any restrictions on transferability and may be sold by the Participant in accordance with the Company's internal rules on insider trading and applicable laws, rules and regulation.

9 Tax consequences

- 9.1 Any tax matter or liability affecting the Participant, including, but not limited to, tax liability in case of: (i) expatriation; (ii) repatriation; (iii) grants of RSUs and PSUs; (iv) vesting of RSUs and/or PSUs; (v) receipt of shares underlying vested RSUs and/or PSUs; (vi) cash settlement; adjustment of the terms of the LTIP; and (vii) any claw back pursuant to Clause 5, is of no concern to the Company or any other Group Member, and the Company and/or other Group Member, as applicable, cannot without its expressed written consent be liable for any tax or tax reporting in connection thereto.
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- 9.2 The Participant agrees to make appropriate arrangements with the Company and/or other Group Member, as applicable, for the satisfaction of all state, local and foreign income and employment tax withholding requirements applicable to the granting of the RSUs and PSUs.
- 9.3 The Participant agrees that, if deemed necessary by a Group Member, the Group Member may withhold an appropriate proportion of the shares underlying vested RSUs and/or PSUs to ensure that any tax liability and relevant selling costs in connection with the grant of RSUs and/or PSUs are met.
- 9.4 The Participant agrees that a Group Member may satisfy all federal, state, local and foreign income and employment tax withholding and/or information disclosure requirements in connection with vesting and granting of the RSUs and/or PSUs.
- 10 Amendment of the LTIP**
- 10.1 The Board of Directors is at its sole discretion entitled to amend the LTIP, including these General Terms and Conditions and the Individual Grant Letter, subject to applicable law. This includes, but is not limited to, changes in order to comply with local legislation and adjusting the method for granting RSUs and/or PSUs; adjusting any Performance Target and the weighting thereof; providing for an extraordinary access to cash settlement; accelerated vesting or extraordinary grants of RSUs and/or PSUs. Further, amendments can be made by the Board of Directors in case any Performance Target will be affected by material and/or unforeseen events, inter alia, in case the Group acquires substantial business or assets or divests a substantial part of its business, or in case a non-recurring one-off event relating to the current or past financial years is deemed to have an unintended effect on any Performance Target.
- 10.2 The Board of Directors may at its sole discretion and by giving written notice to a Participant amend the number of RSUs and/or PSUs and/or other terms of an individual grant under the LTIP, including acceleration of vesting, in case of extraordinary, material or unforeseen events or circumstances.
- 11 Choice of law and venue**
- 11.1 The Individual Grant Letters and these General Terms and Conditions are governed by Danish law, however, the Danish Share Option Act shall only apply to Participants who are otherwise and generally subject to mandatory Danish law applicable to salaried employees.
- 11.2 Any dispute shall be finally and exclusively settled by the Danish courts.
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12 Personal data

12.1 As part of the LTIP, the Company will process personal data concerning the Participant for the purposes of the establishment and administration of the LTIP. The processing of personal data is required for the Company to fulfil its obligations in relation to the LTIP. The personal data will include name and other identification data of the Participant, information regarding employment relationship which is relevant for administration of the LTIP and information on number of RSUs and PSUs each Participant is eligible to receive. The data may be transferred to public authorities, if required by law or regulations. Personal data will be stored by the Company for a period of five (5) years following the point in time when the Participant is no longer covered by the LTIP. The Participant has the right to request access to and rectification of the data relating to the Participant. Provision by the Participants of the above personal data is a prerequisite for participating in the LTIP. Questions regarding the processing of personal data in relation to the LTIP may be addressed to Chief Financial Officer, Anders Vadsholt, afv@orphazyme.com, and Participants may also lodge complaints with the Danish Data Protection Agency.

13 Miscellaneous

- 13.1 By signing the Individual Grant Letter, the Participant confirms having received and read these General Terms and Conditions. At the same time the Participant confirms, if relevant, having received and read the statement construed in accordance with section 3 of the Danish Share Option Act.
- 13.2 The Participant is not ensured any economic benefit when participating in the LTIP.
- 13.3 Where local legislation prevents the enforcement of one or more Clauses of these General Terms and Conditions, such particular Clause(s) will be void while the remaining provisions of these General Terms and Conditions shall remain valid to the extent possible.
- 13.4 Unless otherwise stated in these General Terms and Conditions and the Company's internal guidelines, any costs incurred in connection with the grant and/or vesting of RSUs and/or PSUs shall be paid by the Company.
- 13.5 The value of the RSUs and/or PSUs shall not be included in the calculation of any compensation, including, but not limited to, severance pay and holiday pay, to which the Participant may be entitled to during the employment or in connection with termination of the employment.
- 13.6 The LTIP, these General Terms and Conditions, all Individual Grant Letters, the RSUs and PSUs are, with respect to each Participant who is a United States taxpayer, intended to be exempt from Section 409A of the United States Internal Revenue Code of 1986, as amended ("**Section 409A**"), and shall be interpreted in a manner consistent therewith. In all events, RSUs and PSUs granted to a Participant who is a United States taxpayer shall be settled within sixty (60) days after vesting; provided that the RSU Delivery Period or PSU Delivery Period (as applicable) is automatically extended to the next open trading window if the settlement of the RSUs or PSUs or delivery of shares underlying vested RSUs or PSUs would contravene applicable laws, rules or regulations, including, but not limited to, situations where the delivery of shares underlying vested RSUs or PSUs cannot occur due to the Company being in possession of inside information, but in no event shall such delivery occur beyond 15 March of the calendar year following the year in which the Total RSU Vesting Period or PSU Vesting Period, respectively, is satisfied. To the extent that any United States taxpayer is required to pay a subscription price to acquire shares underlying vested RSUs and PSUs, such Participant shall be required to pay the applicable subscription price by the last day of the RSU Delivery Period or PSU Delivery Period (as applicable). Notwithstanding any contrary provision
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of these General Terms and Conditions, if the vesting of any RSUs and/or PSUs held by a United States taxpayer is accelerated, any applicable subscription price required to be paid by such Participant shall be paid only during a period immediately following such acceleration, with such period to be specified by the Company in its sole discretion but not to exceed ten (10) days after the date of the event causing such acceleration. If the subscription price is not paid in the required time period, such RSUs and/or PSUs (as applicable) will be forfeited for no compensation or other payment due to the Participant or any other person or entity. If the Participant has not paid the applicable subscription price, or has not complied with the General Terms and Conditions and the applicable Individual Grant Letter, in any case, as of the settlement date of the RSUs and PSUs (as applicable), then the Participant's right to receive shares underlying such RSUs and PSUs (as applicable) shall be forfeited with no compensation or other payment due to the Participant or any other person or entity. Notwithstanding the foregoing or anything to the contrary contained in the LTIP, in these General Terms and Conditions or in any Individual Grant Letter, neither the Company nor any Group Member shall have any liability or obligation to any Participant or to any other person or entity for any taxes, interest, penalties or fines resulting from the failure of the LTIP, these General Terms and Conditions, any Individual Grant Letter or any award of RSUs or PSUs to be exempt from or compliant with Section 409A. If any RSU Delivery Period or PSU Delivery Period spans two calendar years, then, to the extent necessary to prevent the imposition of taxes or penalties under Section 409A, the delivery of shares underlying such vested RSUs or PSUs shall occur no earlier than 1 January of the second of such calendar years.

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These General Terms and Conditions have been adopted by the Board of Directors on 21 April 2021.

21 April 2021 | Danish and Swiss participants

Orphazyme A/S – Long-Term Incentive Programme

General Terms and Conditions for Restricted Share Units and Performance Share Units

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These are the general terms and conditions (the “General Terms and Conditions”) for the 2021 long-term incentive program (the “LTIP”) of Orphazyme A/S, CVR no. 32266355 (the “**Company**”) and its subsidiary undertakings (the Company and its subsidiary undertakings are referred to as the “**Group**” and each a “**Group Member**”).

The General Terms and Conditions apply to grants of restricted share units (“**Restricted Share Units**” or “**RSUs**”) and performance share units (“**Performance Share Units**” or “**PSUs**”) under the LTIP awarded to members of the executive management, registered as such with the Danish Business Authority, and certain key employees of the Group (such executives and employees referred to as “**Participants**” and each a “**Participant**”).

1 Introduction

- 1.1 The LTIP has been established by the Company’s board of directors (the “**Board of Directors**”) and has been adopted in accordance with the Company’s remuneration policy adopted by the Company’s annual general meeting on 25 March 2021.
- 1.2 Participation in the Group’s LTIP and in each individual grant thereunder is resolved by the Board of Directors at its sole discretion from time to time in connection with each grant.
- 1.3 The LTIP comprises the following documents:
 - (a) These General Terms and Conditions;
 - (b) Individual Grant Letter (as defined below);
 - (c) Agreement on the application of section 7P of the Danish Tax Assessment Act; and
 - (d) Danish Share Option Act statement for Participants comprised by this act.

In case of any discrepancy between the above documents, these General Terms and Conditions shall prevail. To the extent any of the documents exist in other languages than English, the English version shall prevail, except for the Danish Share Option Act statement where the Danish version shall prevail.

- 1.4 If eligible for participation under the LTIP, the Participant will receive an individual grant letter setting out the individual terms applicable to the grant of RSUs and PSUs (as defined below) (the “**Individual Grant Letter**”). The Individual Grant Letter will, inter alia, state the number of RSUs and PSUs granted and the conditions for vesting of the RSUs and PSUs, in whole or in part, including any performance targets for the PSUs as defined by the Board of Directors (each a “**Performance Target**” and, if more than one, collectively the “**Performance Targets**”). If a Participant is entitled to a fraction of a share in the Company as part of the LTIP, the number of shares to such Participant will be rounded down to the nearest integer number.
 - 1.5 The purpose of the LTIP is mainly, but not only, to align the interests of the Participants with the interests of the Company’s shareholders while also encouraging Participants to remain employed in the Group. Further, the granting of PSUs under the LTIP aims to support the performance culture in the Group by rewarding the Participants in proportion to the achievement of one or more Performance Targets. Any Performance Target will be set prior to any grants and communicated to Participants in the Individual Grant Letters. Assuming the other vesting conditions are met, achieving one or more Performance Targets will contribute to the vesting of PSUs by the proportion set out in the Individual Grant Letters.
 - 1.6 The LTIP is forward-looking in that grants of RSUs and PSUs are not based on previous performance of the Company or the Participant, and vesting is dependent on satisfaction of certain vesting criteria, including one or more Performance Targets, which are forward-looking.
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- 1.7 These General Terms and Conditions govern grants made under the LTIP in 2021. Grants made after 1 January 2022 will be governed by the general terms and conditions applicable at the time of such grants.
- 2 Restricted Share Units (RSUs)**
- 2.1 When vested and not lapsed pursuant to these General Terms and Conditions, RSUs will, subject to fulfilment of certain conditions set out in Clause 2.3 below, entail that the Participant receives a number of shares equivalent to the number of RSUs vested against payment (in a manner and within a period decided by the Board of Directors) of the par value of each share in accordance with the terms and conditions outlined herein and in the Individual Grant Letter.
- 2.2 The Participant will be entitled to receive RSUs based on the volume weighted average price (VWAP) of the Company's shares as quoted on Nasdaq Copenhagen during a period of ten (10) trading days preceding the date stipulated in the Individual Grant Letter.
- 2.3 Conditions for vesting of RSUs
- 2.3.1 As stipulated in the Individual Grant Letter, the Participant will be entitled to receive RSUs that have a total vesting period of approximately three (3) years from the date of the grant or such other date as stipulated in the Individual Grant Letter with one third of the granted RSUs vesting on each such date as stipulated in the Individual Grant Letter in the following three (3) financial years (each period is referred to as a "**RSU Vesting Period**" and the total continuous period of approximately three (3) years from the date of the grant as the "**Total RSU Vesting Period**"), subject to the satisfaction of each of the following conditions:
- (a) The Participant remaining employed with a Group Member throughout the Total RSU Vesting Period, or the Participant becoming a Good Leaver (as defined below) during the Total RSU Vesting Period in which case the Participant will be entitled to keep any vested RSUs and receive a pro rata allocation in accordance with Clause 2.5.1 below; and
- (b) The Participant having complied in all respects with the General Terms and Conditions including Clause 2.5.4.
- 2.3.2 Any grant of RSUs or delivery of shares underlying such RSUs upon vesting must comply with applicable statutory requirements and regulations, and no such grant may be made if the Board of Directors determines, at its sole discretion, that such grant constitutes a breach of current statutory requirements or regulations.
- 2.3.3 In the event and to the extent any of the above conditions are not satisfied, any and all rights to receive shares underlying RSUs shall terminate and lapse without further notice and without any rights of compensation, unless the Board of Directors resolves otherwise at its sole discretion.
- 2.4 Vesting of RSUs
- 2.4.1 Subject to satisfaction of the conditions set out in Clause 2.3.1, during and upon expiration of the Total RSU Vesting Period, the Participant will be entitled to receive a number of shares in the Company equivalent to the number of vested RSUs against payment of the par value of each share in accordance with and to the extent set out in these General Terms and Conditions.
- 2.4.2 The delivery of shares by the Company to the Participant after the Total RSU Vesting Period will be effectuated in a manner determined by the Board of Directors.
- 2.4.3 In order to be eligible to receive the vested RSUs, as set out in Clause 2.4.2, the Participant shall comply with the practical instructions provided by the Company. If the Par-
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Participant does not timely comply with the instructions provided by the Company, the Company will be entitled to consider the right to receive shares underlying vested RSUs as having lapsed without further notice and without compensation to the Participant. Neither the Company nor any other Group Member or their representatives may be held liable for any failure to deliver shares underlying vested RSUs to the Participant due to the Participant not complying with the instructions provided.

- 2.4.4 Subject to compliance with the instructions provided, delivery by the Company to the Participant of the shares underlying vested RSUs will be effected at the latest four (4) months after expiration of the Total RSU Vesting Period (the “**RSU Delivery Period**”). The RSU Delivery Period is automatically extended to the next open trading window if the exercise of the RSUs or delivery of shares underlying vested RSUs would contravene applicable laws, rules or regulations, including, but not limited to, situations where the delivery of shares underlying vested RSUs cannot occur due to the Company being in possession of inside information.
- 2.4.5 The Participant will not be deemed to be the owner or holder of ownership rights or any other rights in respect of the RSUs in the Company until (i) the RSUs have vested and the instructions to receive the RSUs have been complied with, and (ii) the Participant’s ownership of the shares underlying the vested RSUs has been registered in the Company’s shareholders’ register.
- 2.5 Continued employment or Good Leaver
- 2.5.1 In case of termination of the Participant’s employment with the Group Member due to (i) the Participant’s resignation due to the Group Member’s material breach of contract; (ii) the Group Member’s termination of the Participant without such termination being due to the Participant’s breach of obligations towards the Group Member; (iii) the Participant’s retirement; or (iv) the Participant being deceased (the Participant will in each situation mentioned under (i)-(iv) be considered a “Good Leaver”), the Participant (or the deceased Participant’s estate, as the case may be) will remain entitled to the Participant’s RSUs vested at the date of release of the Participant’s work obligations (in Danish: “*fritstillingstidspunktet*”) subject to and in accordance with the Individual Grant Letter and the General Terms and Conditions. For any ongoing RSU Vesting Period, a Good Leaver will be entitled to receive a pro rata allocation of RSUs for the period from commencement of the ongoing RSU Vesting Period and until the date of release of the Participant’s work obligations (in Danish: “*fritstillingstidspunktet*”).
- 2.5.2 In the event that the Participant does not effectively retire from the labour market upon resignation from the Group Member, as provided under Clause 2.5.1 above, but takes employment with a different employer or works as a self-employed after retirement from the Group Member, the Company may, in its discretion and taking into account the further definitions for a Good Leaver as set out in Clause 2.5.1, decide that vested and/or unvested RSUs shall lapse without any rights of compensation.
- 2.5.3 In the event of termination of employment for any other reason than stated in Clause 2.5.1, including due to the Participant’s own resignation or material breach of obligations towards the Group Member then any vested and/or unvested RSUs will lapse without further notice and without any rights of compensation as of the date of release of the Participant’s work obligations (in Danish: “*fritstillingstidspunktet*”), unless otherwise decided by the Board of Directors.
- 2.5.4 Further, any vested and unvested RSUs will lapse without further notice and without any rights of compensation if, during the Participant’s employment with the Group Member, or after the termination of the Participant’s employment while the Participant holds vested and/or unvested RSUs, the Participant (i) violates a non-competition and/or non-solicitation clause in the Participant’s employment contract or service agreement or otherwise breaches the duty of loyalty towards the Group Member, (ii) discloses or otherwise misuses any confidential information, whether written or oral, including, without limitation, financial information, trade secrets and other proprietary business information regarding the Group Member, (iii) violates the Group Mem-
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ber's compliance policies, or (iv) violates the Group Member's accounting rules including the financial reporting rules.

3 Performance Share Units (PSUs)

- 3.1 When vested and not lapsed pursuant to these General Terms and Conditions, PSUs will, subject to fulfilment of certain conditions set out in Clause 3.3 below and in the Individual Grant Letter, entail that the Participant receives a number of shares equivalent to the number of PSUs vested against payment (in a manner and within a period decided by the Board of Directors) of the par value of each share in accordance with the terms and conditions outlined herein and in the Individual Grant Letter.
- 3.2 The Participant will be entitled to receive PSUs based on the volume weighted average price (VWAP) of the Company's shares as quoted on Nasdaq Copenhagen during a period of ten (10) trading days preceding the date stipulated in the Individual Grant Letter.
- 3.3 Conditions for vesting of PSUs
- 3.3.1 As stipulated in the Individual Grant Letter, the Participant will be entitled to receive PSUs that have a vesting period of approximately three (3) years from the date of the grant or such other date as stipulated in the Individual Grant Letter, with the granted PSUs vesting, in whole or in part, on such date as stipulated in the Individual Grant Letter in the third financial year following the date of the grant (the "**PSU Vesting Period**"), subject to the satisfaction of each of the following conditions:
- (a) The Participant remaining employed with a Group Member throughout the PSU Vesting Period, or the Participant becoming a Good Leaver (as defined below) during the PSU Vesting Period in which case the Participant may be entitled to keep a proportion of the PSUs in accordance with Clause 3.5.1 below;
 - (b) Achievement of one or more Performance Targets, during the PSU Vesting Period, as set out in the Individual Grant Letter (if more than one Performance Target has been defined by the Board of Directors, achievement of each Performance Target will contribute to the vesting of PSUs by the proportion set out in the Individual Grant Letter); and
 - (c) The Participant having complied in all respects with the General Terms and Conditions.
- 3.3.2 Any grant of PSUs or delivery of shares underlying such PSUs upon vesting must comply with applicable statutory requirements and regulations, and no such grant may be made if the Board of Directors determines, at its sole discretion, that such grant constitutes a breach of current statutory requirements or regulations.
- 3.3.3 In the event and to the extent any of the above conditions are not satisfied, any and all rights to receive shares underlying PSUs shall terminate and lapse without further notice and without any rights of compensation, unless the Board of Directors resolves otherwise at its sole discretion.
- 3.4 Vesting of PSUs
- 3.4.1 Subject to satisfaction of the conditions set out in Clause 3.3.1 and in the Individual Grant Letter, during and upon expiration of the PSU Vesting Period, the Participant will be entitled to receive a number of shares in the Company equivalent to the number of vested PSUs against payment of the par value of each share in accordance with and to the extent set out in these General Terms and Conditions and in the Individual Grant Letter. The Board of Directors will determine if and when any Performance Target has been met and make an announcement to that effect no later than four (4) weeks after the expiration of the PSU Vesting Period.
- 3.4.2 The delivery of shares by the Company to the Participant after the PSU Vesting Period will be effectuated in a manner determined by the Board of Directors.
- 3.4.3 In order to be eligible to receive the vested PSUs, as set out in Clause 3.4.2, the Participant shall comply with the practical instructions provided by the Company. If the Participant does not timely comply with the instructions provided by the Company, the Company will be entitled to consider the right to receive shares underlying vested PSUs
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as having lapsed without further notice and without compensation to the Participant. Neither the Company nor any other Group Member or their representatives may be held liable for any failure to deliver shares underlying vested PSUs to the Participant due to the Participant not complying with the instructions provided.

- 3.4.4 Subject to compliance with the instructions provided, delivery by the Company to the Participant of the shares underlying vested PSUs will be effected at the latest four (4) months after expiration of the PSU Vesting Period (the “**PSU Delivery Period**”). The PSU Delivery Period is automatically extended to the next open trading window if the exercise of the PSUs or delivery of shares underlying vested PSUs would contravene applicable laws, rules or regulations, including, but not limited to, situations where the delivery of shares underlying vested PSUs cannot occur due to the Company being in possession of inside information.
- 3.4.5 The Participant will not be deemed to be the owner or holder of ownership rights or any other rights in respect of the PSUs in the Company until (i) the PSUs have vested and the instructions to receive the PSUs have been complied with, and (ii) the Participant’s ownership of the shares underlying the vested PSUs has been registered in the Company’s shareholders’ register.
- 3.5 Continued employment or Good Leaver
- 3.5.1 In case of termination of the Participant’s employment with the Group Member due to (i) the Participant’s resignation due to the Group Member’s material breach of contract; (ii) the Group Member’s termination of the Participant without such termination being due to the Participant’s breach of obligations towards the Group Member; (iii) the Participant’s retirement; or (iv) the Participant being deceased (the Participant will in each situation mentioned under (i)-(iv) be considered a “**Good Leaver**”), the Participant (or the deceased Participant’s estate, as the case may be) will remain entitled to keep such proportion of the Participant’s PSUs which have one or more Performance Targets that have been achieved at the date of release of the Participant’s work obligations (in Danish: “*fritstillingstidspunktet*”) and such portion of the Participant’s PSUs shall vest subject to and in accordance with the Individual Grant Letter and the General Terms and Conditions.
- 3.5.2 In the event that the Participant does not effectively retire from the labour market upon resignation from the Group Member, as provided under Clause 3.5.1 above, but takes employment with a different employer or works as a self-employed after retirement from the Group Member, the Company may, in its discretion and taking into account the further definitions for a Good Leaver as set out in Clause 3.5.1, decide that all PSUs shall lapse without any rights of compensation regardless of whether one or more Performance Targets have been met at the date of release of the Participant’s work obligations (in Danish: “*fritstillingstidspunktet*”).
- 3.5.3 In the event of termination of employment for any other reason than stated in Clause 3.5.1, including due to the Participant’s own resignation or material breach of obligations towards the Group Member then all PSUs will lapse without further notice and without any rights of compensation as of the date of release of the Participant’s work obligations (in Danish: “*fritstillingstidspunktet*”), unless otherwise decided by the Board of Directors.
- 3.5.4 Further, any vested and unvested PSUs will lapse without further notice and without any rights of compensation if, during the Participant’s employment with the Group Member, or after the termination of the Participant’s employment while the Participant holds vested but not exercised PSUs, the Participant (i) violates a non-competition and/or non-solicitation clause in the Participant’s employment contract or service agreement or otherwise breaches the duty of loyalty towards the Group Member, (ii) discloses or otherwise misuses any confidential information, whether written or oral, including, without limitation, financial information, trade secrets and other proprietary business information regarding the Group Member, (iii) violates the Group Member’s
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compliance policies, or (iv) violates the Group Member's accounting rules including the financial reporting rules.

4 Cash settlement

4.1 In cases where the Board of Directors assesses that the issue or transfer of shares would have an adverse effect on the Company and/or the Participant, the Board of Directors may choose to cash settle the RSUs and/or PSUs instead of allowing the Participant to subscribe for or receive shares upon expiry of the Total RSU Vesting Period and the PSU Vesting Period, respectively. In such event, the Company or the Group Member, as applicable, shall pay a cash settlement amount based on the volume weighted average price of the Company's shares as quoted on Nasdaq Copenhagen during the ten (10) trading days preceding the date of expiry of the Total RSU Vesting Period and the PSU Vesting Period, respectively, with a deduction of the exercise price. The Company and/or the Group Member, as applicable, is entitled to deduct any tax withholding amounts in the cash settlement amount.

5 Claw back

5.1 If the Board of Directors concludes in good faith that the conditions for vesting of any RSUs and/or PSUs are based on fraud, willful misconduct, gross negligence, incorrect or misleading information, or that the conditions have otherwise not been fulfilled, the Company shall be entitled (a) to consider the RSUs and/or PSUs as lapsed without compensation to the Participant, (b) to recalculate and/or adjust accordingly the number of RSUs and/or PSUs to be granted and/or (c) to require redelivery to the Company of any shares having been delivered to the Participant after vesting of the RSUs and/or PSUs or repayment of the cash settlement amount, as applicable.

6 Adjustments in case of changes to the Company's capital structure

6.1 In order to ensure that the value of the rights under the RSUs and PSUs is duly protected in the event of changes to the Company's capital structure, the Board of Directors may, at its sole discretion, adopt changes to the number of granted RSUs and/or PSUs, inter alia, in the case of the following:

- (a) after changes have been made to the nominal value of the shares of the Company;
 - (b) after the Company's share capital has been increased at a price lower than market price other than capital increases at a price lower than market price offered to board members, executives or other employees of the Company in connection with the LTIP or the share-based incentive program for the Board of Directors or any existing or subsequent incentive plans, including general employee share purchase plans;
 - (c) after the Company's share capital has been increased with pre-emption rights for the Company's existing shareholders allowing them to purchase shares at a price lower than market price;
 - (d) after the Company has issued or granted convertible bonds or other convertible loans, stock options (except for RSUs, PSUs and Performance Shares under the LTIP or the share-based incentive program for the Board of Directors, or any existing or subsequent share based incentive plans, including general employee share purchase plans), and such issue or grant has been made with pre-emption rights for the existing shareholders at a lower price than market price;
 - (e) after the Company's share capital has been reduced for any other purpose than to cover losses or through distribution to the Company's shareholders in the ordinary course of business through cancellation of shares acquired in treasury through share buy-back programmes established by the Company from time to time and in line with the Company's dividend policy (as amended from time to time); or
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- (f) after distribution of extraordinary dividends during a financial year in excess of the Company's dividend policy (as amended from time to time).

Any such adoption of changes to the number of RSUs and/or PSUs or shares to be received in the Individual Grant Letters will seek to achieve that the Participant receives a reasonable compensation for the adjustment of the value of the RSUs and/or PSUs caused by the situation in question. The Board of Directors may decide that the aforementioned compensation is made by adjusting the number of RSUs and/or PSUs granted or by cash-settling the difference in value caused by the changes to the Company's capital structure outlined in 6.1(a)-(f) above.

Any adjustment made from time to time pursuant to these General Terms and Conditions will be notified to the Participant in writing.

If events affecting the share capital in the Company occur which are comparable in nature to the events outlined in 6.1(a)-(f) above, and with similar effect, the Board of Directors may decide to treat the event as if comprised by 6.1(a)-(f) and adjust the number of RSUs and/or PSUs set out in the Individual Grant Letters accordingly.

7 Takeover offer or delisting

7.1

- (a) a mandatory takeover bid concerning the Company's shares is to be made according to the rules of the Danish Capital Markets Act (as amended, supplemented or replaced from time to time), or
- (b) a voluntary takeover bid is made concerning the Company's shares, in which connection there is a change in the controlling interest (as defined in section 44 of the Danish Capital Markets Act (as amended, supplemented or replaced from time to time)) to a third party independent of the Company, or
- (c) otherwise any transaction whereby a third party independent of the Company obtains control of more than 50% of the Company's shares, or
- (d) the shares in the Company are delisted,

the Board of Directors shall be required to accelerate vesting of the RSUs at a time determined by the Board of Directors in relation to acceptance of such takeover offer or other transaction.

- 7.2 In the situations listed under Section 7.1(a)-(d) above, the Board of Directors shall further in relation to acceptance of such takeover offer or other transaction be required to accelerate vesting of the PSUs to the extent that any applicable Performance Targets have been achieved. In case a Performance Target has not been achieved at the time of such accelerated vesting, the PSUs pertaining hereto will lapse, unless otherwise decided by the Board of Directors.

8 Assignment

- 8.1 The RSUs and PSUs or the right to receive shares upon vesting of the RSUs and/or PSUs may not be transferred or assigned to a third party or pledged, encumbered or placed as collateral with a third party, except from transfers to the Company or a third party designated by the Company. Furthermore, any rights to receive RSUs and PSUs that are transferred or sought to be transferred to a third party as a consequence of creditor suit or statutory execution will lapse automatically without further notice.
 - 8.2 Shares received by the Participant upon vesting of the RSUs and/or PSUs shall not be subject to any restrictions on transferability and may be sold by the Participant in accordance with the Company's internal rules on insider trading and applicable laws, rules and regulation.
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9 Tax consequences

9.1 Any tax matter or liability affecting the Participant, including, but not limited to, tax liability in case of: (i) expatriation; (ii) repatriation; (iii) grants of RSUs and PSUs; (iv) vesting of RSUs and/or PSUs; (v) receipt of shares underlying vested RSUs and/or PSUs; (vi) cash settlement; adjustment of the terms of the LTIP; and (vii) any claw back pursuant to Clause 5, is of no concern to the Company or any other Group Member, and the Company and/or other Group Member, as applicable, cannot without its expressed written consent be liable for any tax or tax reporting in connection thereto.

9.2 The Participant agrees to make appropriate arrangements with the Company and/or other Group Member, as applicable, for the satisfaction of all state, local and foreign income and employment tax withholding requirements applicable to the granting of the RSUs and PSUs.

9.3 The Participant agrees that, if deemed necessary by a Group Member, the Group Member may withhold an appropriate proportion of the shares underlying vested RSUs and/or PSUs to ensure that any tax liability and relevant selling costs in connection with the grant of RSUs and/or PSUs are met.

The Participant agrees that a Group Member may satisfy all federal, state, local and foreign income and employment tax withholding and/or information disclosure requirements in connection with vesting and granting of the RSUs and/or PSUs.

10 Amendment of the LTIP

10.1 The Board of Directors is at its sole discretion entitled to amend the LTIP, including these General Terms and Conditions and the Individual Grant Letter, subject to applicable law. This includes, but is not limited to, changes in order to comply with local legislation and adjusting the method for granting RSUs and/or PSUs; adjusting any Performance Target and the weighting thereof; providing for an extraordinary access to cash settlement; accelerated vesting or extraordinary grants of RSUs and/or PSUs. Further, amendments can be made by the Board of Directors in case any Performance Target will be affected by material and/or unforeseen events, inter alia, in case the Group acquires substantial business or assets or divests a substantial part of its business, or in case a non-recurring one-off event relating to the current or past financial years is deemed to have an unintended effect on any Performance Target.

10.2 The Board of Directors may at its sole discretion and by giving written notice to a Participant amend the number of RSUs and/or PSUs and/or other terms of an individual grant under the LTIP, including acceleration of vesting, in case of extraordinary, material or unforeseen events or circumstances.

11 Choice of law and venue

11.1 The Individual Grant Letters and these General Terms and Conditions are governed by Danish law, however, the Danish Share Option Act shall only apply to Participants who are otherwise and generally subject to mandatory Danish law applicable to salaried employees.

11.2 Any dispute shall be finally and exclusively settled by the Danish courts.

12 Personal data

12.1 As part of the LTIP, the Company will process personal data concerning the Participant for the purposes of the establishment and administration of the LTIP. The processing of personal data is required for the Company to fulfil its obligations in relation to the LTIP. The personal data will include name and other identification data of the Participant, information regarding employment relationship which is relevant for administration of the LTIP and information on number of RSUs and PSUs each Participant is eligible to receive. The data may be transferred to public authorities, if required by law or regulations. Personal data will be stored by the Company for a period of five (5) years following the point in time when the Participant is no longer covered by the LTIP. The

Participant has the right to request access to and rectification of the data relating to the Participant. Provision by the Participants of the above personal data is a prerequisite for participating in the LTIP. Questions regarding the processing of personal data in relation to the LTIP may be addressed to Chief Financial Officer, Anders Vadsholt, afv@orphazyme.com, and Participants may also lodge complaints with the Danish Data Protection Agency.

13 Miscellaneous

- 13.1 By signing the Individual Grant Letter, the Participant confirms having received and read these General Terms and Conditions. At the same time the Participant confirms, if relevant, having received and read the statement construed in accordance with section 3 of the Danish Share Option Act.
- 13.2 The Participant is not ensured any economic benefit when participating in the LTIP.
- 13.3 Where local legislation prevents the enforcement of one or more Clauses of these General Terms and Conditions, such particular Clause(s) will be void while the remaining provisions of these General Terms and Conditions shall remain valid to the extent possible.
- 13.4 Unless otherwise stated in these General Terms and Conditions and the Company's internal guidelines, any costs incurred in connection with the grant and/or vesting of RSUs and/or PSUs shall be paid by the Company.
- 13.5 The value of the RSUs and/or PSUs shall not be included in the calculation of any compensation, including, but not limited to, severance pay and holiday pay, to which the Participant may be entitled to during the employment or in connection with termination of the employment.

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These General Terms and Conditions have been adopted by the Board of Directors on 21 April 2021.

Orphazyme A/S – Share-based incentive program for the Board of Directors for 2021

General Terms and Conditions

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These are the general terms and conditions (the “**General Terms and Conditions**”) for the 2021 board incentive program (the “**Board Incentive Program**”) of Orphazyme A/S, CVR no. 32266355 (the “**Company**”).

The General Terms and Conditions apply to grants of restricted share units (“**Restricted Share Units**” or “**RSUs**”) under the Board Incentive Program awarded to members of the Company’s board of directors (the “**Board of Directors**”) (such members of the Board of Directors referred to as “**Participants**” and each a “**Participant**”).

1 Introduction

- 1.1 The Board Incentive Program has been established by the Board of Directors and has been adopted in accordance with the Company’s remuneration policy adopted by the Company’s annual general meeting on 25 March 2021.
- 1.2 Participation in the Board Incentive Program and each individual grant thereunder is resolved by the Board of Directors and approved by the Company’s annual general meeting in connection with approval of the remuneration of the Board of Directors.
- 1.3 The Board Incentive Program comprises the following documents:
- (a) These General Terms and Conditions; and
 - (b) Individual Grant Letter (as defined below).

In case of any discrepancy between the above documents, these General Terms and Conditions shall prevail.

- 1.4 The purpose of the Board Incentive Program is to attract and retain qualified members of the Board of Directors as share-based incentives are commonly used among competing international biotech and pharmaceutical companies. Further, the Board Incentive Program reflects the objective of a motivated and lasting value creation for the Company and the shareholders.

2 Grant of Restricted Share Units

- 2.1 Participants may annually be granted a number of RSUs (a “**Grant**”) with a value corresponding to up to 50% of the Participant’s fixed annual base fee as member of the Board of Directors, such base fee to include additional base fee to the Chairman and Deputy Chairman, respectively, but excluding any additional fees for committee membership. The number of RSUs granted shall be determined by calculating the value of the RSUs and by applying a reference share price calculated on the basis of the volume weighted average share price of the Company’s shares as quoted on Nasdaq Copenhagen during the ten (10) trading days preceding 1 January in the year of Grant.
- 2.2 A Participant may also be eligible to receive additional grants of RSUs in accordance with the Company’s remuneration policy as approved by the general meeting for time to time.

2.3 Each Grant will be effected pursuant to an individual grant letter setting out the individual terms applicable to the Grant (the “**Individual Grant Letter**”). The Individual Grant Letter will, inter alia, state the number of RSUs granted and the conditions for vesting of the RSUs, if any. If a Participant is entitled to a fraction of a share in the Company as part of the Board Incentive Program, the number of shares to such Participant will be rounded down to the nearest integer number.

2.4 Any Grant will occur as soon as possible after the Company’s annual general meeting (the “**Grant Date**”). Each Grant is communicated to each of the Participants as soon as possible following the Grant Date.

2.5 When vested and not lapsed pursuant to the General Terms and Conditions, each RSU entitles the Participant to be allocated one (1) share in the Company against payment of the Exercise Price (as defined below).

3 Vesting of Restricted Share Units

3.1 RSUs granted under the Board Incentive Program will have a vesting period from the Grant Date and until the date of the next annual general meeting in the following year (the “**Vesting Period**”).

3.2 Vesting of the RSUs are not conditional on any financial performance criteria, however vesting will be conditional upon the Participant’s continued membership of the Board of Directors during the entire Vesting Period.

4 Exercise of Restricted Share Units

4.1 Upon vesting in accordance with Clause 3, RSUs may be exercised within a period of four (4) weeks from vesting or the date of publication of the Company’s interim report for the first six months in 2022 (the “**Exercise Period**”). The Exercise Period is automatically extend to the next open trading window if the exercise of the RSUs or delivery of shares underlying vested RSUs would contravene applicable laws, rules or regulations, including, but not limited to, situations where the delivery of shares underlying vested RSUs cannot occur due to the Company being in possession of inside information.

4.2 A Participant may be allocated a number of shares equivalent to the number of RSUs vested at a price per RSU equal to the par value of one share issued by the Company (the “**Exercise Price**”). The allocation of shares upon vesting of the RSUs may be made, in the Company’s discretion, through (a) new shares of the Company, in which case the Participant will be offered to subscribe for such amount of new shares equal to the number of RSUs vested at a subscription price per RSU equal to the Exercise Price, or (b) existing shares of the Company held by the Company in treasury, in which case the Participant will be offered to purchase such amount of treasury shares equal to the number of RSUs vested at a purchase price per RSU equal to the Exercise Price.

4.3 The Participant will not be deemed to be the owner or holder of ownership rights or any other rights in respect of the RSUs until (i) the RSUs have been exercised, and (ii) the Participant’s ownership of the shares acquired or subscribed for has been registered in the Company’s shareholders’ register.

4.4 When exercising vested RSUs, the Participant shall observe the Company's internal rules for trading in the Company's shares as well as applicable laws.

4.5 RSUs that have not been exercised during the Exercise Period will automatically lapse without compensation when the Exercise Period has ended.

5 Lapse of Restricted Share Units

5.1 In the event of a Participant's resignation from the Board of Directors during a term which for the avoidance of doubt shall not comprise a decision not to be re-elected, any unvested RSUs will lapse without further notice and without any rights of compensation, unless otherwise decided by the Board of Directors.

5.2 Further, any unvested RSUs will lapse without further notice and without any rights of compensation if, during the Participant's membership of the Board of Directors, the Participant (i) discloses or otherwise misuses any confidential information, whether written or oral, including, without limitation, financial information, trade secrets and other proprietary business information regarding the Company, (ii) violates the Company's compliance policies, or (iii) violates the Company's accounting rules including the financial reporting rules.

6 Cash settlement

6.1 Notwithstanding Clause 4, in cases where the Board of Directors assesses that the issue or transfer of shares would have an adverse effect on the Company and/or the Participant, the Board of Directors may choose to cash settle the RSUs instead of allowing the Participant to receive shares upon expiry of the Vesting Period. In such event, the Company shall pay a cash settlement amount based on the volume weighted average price of the Company's shares as quoted on Nasdaq Copenhagen during the ten (10) trading days preceding the first day of the Exercise Period, with a deduction of the Exercise Price. The Company is entitled to deduct any tax withholding amounts in the cash settlement amount.

7 Claw back

7.1 If the Company can demonstrate that the basis for granting the RSUs or that the conditions for vesting of any RSUs are based on fraud, willful misconduct, gross negligence, incorrect or misleading information, or that the conditions have otherwise not been fulfilled, the Company shall be entitled (a) to consider the RSUs as lapsed without further notice or compensation to the Participant, (b) to recalculate and/or adjust accordingly the number of RSUs to be granted and/or (c) to require redelivery to the Company of any shares having been delivered to the Participant after vesting of the RSUs or repayment of the cash settlement amount, as applicable.

8 Adjustments in case of changes to the Company's capital structure

8.1 In order to ensure that the value of the rights under the RSUs is duly protected in the event of changes to the Company's capital structure, the Board of Directors may, at its sole discretion, adopt changes to the number of granted RSUs, inter alia, in case of the following:

- (a) after changes have been made to the nominal value of the shares of the Company;
- (b) after the Company's share capital has been increased at a price lower than market price other than capital increases at a price lower than market price offered to board members, executives or other employees of the Company in connection with the Board Incentive Program, the long term incentive program for executive management and other employees or any existing or subsequent incentive plans, including general employee share purchase plans;
- (c) after the Company's share capital has been increased with pre-emption rights for the Company's existing shareholders allowing them to purchase shares at a price lower than market price;
- (d) after the Company has issued or granted convertible bonds or other convertible loans, stock options (except for RSUs, PSUs and Performance Shares under the Board Incentive Program, the long term incentive program for the executive management and other employees or any existing or subsequent share based incentive plans, including general employee share purchase plans), and such issue or grant has been made with pre-emption rights for the existing shareholders at a lower price than market price;
- (e) after the Company's share capital has been reduced for any other purpose than to cover losses or through distribution to the Company's shareholders in the ordinary course of business through cancellation of shares acquired in treasury through share buy-back programs established by the Company from time to time and in line with the Company's dividend policy (as amended from time to time); or
- (f) after distribution of extraordinary dividends during a financial year in excess of the Company's dividend policy (as amended from time to time).

Any such adoption of changes to the number of RSUs or shares to be received in the Individual Grant Letters will seek to achieve that the Participant receives a reasonable compensation for the adjustment of the value of the RSUs caused by the situation in question. The Board of Directors may decide that the aforementioned compensation is made by adjusting the number of RSUs granted or by cash-settling the difference in value caused by the changes to the Company's capital structure outlined in 8.1(a)-(f) above.

Any adjustment made from time to time pursuant to these General Terms and Conditions will be notified to the Participant in writing.

If events affecting the share capital in the Company occur which are comparable in nature to the events outlined in 8.1(a)-(f) above, and with similar effect, the Board of Directors may decide to treat the event as if comprised by 8.1(a)-(f) and adjust the number of RSUs set out in the Individual Grant Letters accordingly.

9 Takeover offer or delisting

9.1 If

- (a) a mandatory takeover bid concerning the Company's shares is to be made according to the rules of the Danish Capital Markets Act (as amended, supplemented or replaced from time to time), or
- (b) a voluntary takeover bid is made concerning the Company's shares, in which connection there is a change in the controlling interest (as defined in section 44 of the Danish Capital Markets Act (as amended, supplemented or replaced from time to time)) to a third party independent of the Company, or
- (c) otherwise any transaction whereby a third party independent of the Company obtains control of more than 50% of the Company's shares, or
- (d) the shares in the Company are delisted,

the Board of Directors shall be required to accelerate vesting of the RSUs at a time determined by the Board of Directors in relation to acceptance of such takeover offer or other transaction.

10 Assignment

10.1 The RSUs or the right to receive shares upon vesting of the RSUs may not be transferred or assigned to a third party or pledged, encumbered or placed as collateral with a third party, except from transfers to the Company or a third party designated by the Company. Furthermore, any rights to receive RSUs that are transferred or sought to be transferred to a third party as a consequence of creditor suit or statutory execution will lapse automatically without further notice.

10.2 Shares received by the Participant upon vesting of the RSUs shall not be subject to any restrictions on transferability and may be sold by the Participant in accordance with the Company's internal rules on insider trading and applicable laws, rules and regulation.

11 Tax consequences

11.1 Any tax matter or liability affecting the Participant, including, but not limited to, tax liability in case of (i) expatriation; (ii) repatriation; (iii) grants of RSUs; (iv) vesting of RSUs; (v) receipt of shares underlying vested RSUs; (vi) cash settlement; adjustment of the terms of the Board Incentive Program; and (vii) any claw back pursuant to Clause 7 is of no concern to the Company or any subsidiary undertaking of the Company (the Company including its subsidiary undertakings are collective referred to as the "**Group**" and each a "**Group Member**"), and a Group Member cannot without its expressed written consent be liable for any tax or tax reporting in connection thereto.

11.2 The Participant agrees to make appropriate arrangements with the Company and/or other Group Member, as applicable, for the satisfaction of all state, local and foreign income and employment tax withholding requirements applicable to the granting of the RSUs.

- 11.3 The Participant agrees that, if deemed necessary by a Group Member, the Group Member may withhold an appropriate proportion of the shares underlying vested RSUs to ensure that any tax liability and relevant selling costs in connection with the grant of RSUs are met.
- 11.4 The Participant agrees that a Group Member may satisfy all federal, state, local and foreign income and employment tax withholding and/or information disclosure requirements in connection with vesting and granting of the RSUs.

12 Amendment of the General Terms and Conditions etc.

- 12.1 The Board of Directors is at its sole discretion entitled to amend the Board Incentive Program, including these General Terms and Conditions and the Individual Grant Letter, subject to applicable law. This includes, but is not limited to, changes in order to comply with local legislation and adjusting the method for granting RSUs. Amendments must be in compliance with the Company's remuneration policy in force at the time of amendment.
- 12.2 The Board of Directors may at its sole discretion and by giving written notice to a Participant amend the number of RSUs or other terms of an individual grant under the Board Incentive Program, including acceleration of vesting, in case of extraordinary, material or unforeseen events or circumstances.

13 Choice of law and venue

- 13.1 The Individual Grant Letters and these General Terms and Conditions are governed by Danish law.
- 13.2 Any dispute shall be finally and exclusively settled by the Danish courts.

14 Personal data

- 14.1 As part of the Board Incentive Program, the Company will process personal data concerning the Participant for the purposes of the establishment and administration of the Board Incentive Program. The processing of personal data is required for the Company to fulfil its obligations in relation to the Board Incentive Program. The personal data will include name and other identification data of the Participant, information regarding board membership which is relevant for administration of the Board Incentive Program and information on number of RSUs each Participant is eligible to receive. The data may be transferred to public authorities, if required by law or regulations. Personal data will be stored by the Company for a period of five (5) years following the point in time when the Participant is no longer covered by the Board Incentive Program. The Participant has the right to request access to and rectification of the data relating to the Participant. Provision by the Participants of the above personal data is a prerequisite for participating in the Board Incentive Program. Questions regarding the processing of personal data in relation to the Board Incentive Program may be addressed to Chief Financial Officer, Anders Vadsholt, afv@orphazyme.com, and Participants may also lodge complaints with the Danish Data Protection Agency.

15 Miscellaneous

- 15.1 By signing the Individual Grant Letter, the Participant confirms having received and read these General Terms and Conditions.
- 15.2 The Participant is not ensured any economic benefit when participating in the Board Incentive Program.
- 15.3 Where local legislation prevents the enforcement of one or more Clauses of these General Terms and Conditions, such particular Clause(s) will be void while the remaining provisions of these General Terms and Conditions shall remain valid to the extent possible.
- 15.4 Unless otherwise stated in in these General Terms and Conditions and the Company's internal guidelines, any costs incurred in connection with the grant and/or vesting of RSUs shall be paid by the Company.

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These General Terms and Conditions have been adopted by the Board of Directors on March 25, 2021.

**CERTIFICATION PURSUANT TO
18 U.S.C. SECTION 1350, AS ADOPTED PURSUANT TO
SECTION 906 OF THE SARBANES-OXLEY ACT OF 2002**

The certification set forth below is being submitted in connection with the Annual Report on Form 20-F for the year ended December 31, 2021 (the "Report") for the purpose of complying with Rule 13a-14(b) or Rule 15d-14(b) of the Securities Exchange Act of 1934 (the "Exchange Act") and Section 1350 of Chapter 63 of Title 18 of the United States Code.

I, Anders Fink Vadsholt, certify that:

1. the Report fully complies with the requirements of Section 13(a) or 15(d) of the Exchange Act; and
2. The information contained in the Report fairly presents, in all material respects, the financial condition and results of operations of the Company.

Date: June 28, 2022

By: _____ /s/ Anders Fink Vadsholt
Anders Fink Vadsholt
Chief Executive Officer and Chief Financial Officer
(Principal Executive Officer)

**CERTIFICATION PURSUANT TO
18 U.S.C. SECTION 1350, AS ADOPTED PURSUANT TO
SECTION 906 OF THE SARBANES-OXLEY ACT OF 2002**

The certification set forth below is being submitted in connection with the Annual Report on Form 20-F for the year ended December 31, 2021 (the "Report") for the purpose of complying with Rule 13a-14(b) or Rule 15d-14(b) of the Securities Exchange Act of 1934 (the "Exchange Act") and Section 1350 of Chapter 63 of Title 18 of the United States Code.

I, Anders Fink Vadsholt, certify that:

1. the Report fully complies with the requirements of Section 13(a) or 15(d) of the Exchange Act; and
2. The information contained in the Report fairly presents, in all material respects, the financial condition and results of operations of the Company.

Date: June 28, 2022

By: _____
/s/ Anders Fink Vadsholt
Anders Fink Vadsholt
Chief Executive Officer and Chief Financial Officer
(Principal Financial Officer)

Code of business conduct and ethics

Introduction Purpose and Scope

The Board of Directors of Orphazyme A/S (together with its subsidiaries as existing from time- to-time, “**Orphazyme**”) have established this Code of Business Conduct and Ethics (the “**Code**”) to aid Orphazyme’s directors, officers and employees in making ethical and legal decisions when conducting Orphazyme’s business and performing their day-to-day duties.

Orphazyme’s Board of Directors, or any committee of the Board delegated by the Board, shall be responsible for administering this Code. The Board of Directors has delegated day-to-day responsibility for administering and interpreting the Code to a Compliance Officer. Orphazyme’s Chief Financial Officer has been appointed as our Compliance Officer under this Code.

Orphazyme expects its directors, officers and employees to exercise reasonable judgment when conducting its business. Orphazyme encourages its directors, officers and employees to refer to this Code frequently to ensure that they are acting within both the letter and the spirit of this Code, and to promptly report any suspected violations hereof to the Compliance Officer.

Orphazyme also understands that this Code will not contain the answer to every situation you may encounter or every concern you may have about conducting Orphazyme’s business ethically and legally. In these situations, or if you otherwise have questions or concerns about this Code, we encourage each officer and employee to speak with his or her supervisor (if applicable) or, if you are uncomfortable doing that, with the Compliance Officer under this Code.

Contents of this Code

This Code has two sections which follow this Introduction. The first section, “*Standards of Conduct,*” contains the actual guidelines that Orphazyme’s directors, officers and employees are expected to adhere to in the conduct of Orphazyme’s business. The second section, “*Compliance Procedures,*” contains specific information about how this Code functions, including who administers the Code, who can provide guidance under the Code and how violations may be reported, investigated and punished. This section also contains a discussion about waivers of and amendments to this Code.

A Note About Other Obligations

Orphazyme’s directors, officers and employees generally have other legal and contractual obligations to Orphazyme. This Code is not intended to reduce or limit the other obligations that

you may have to Orphazyme. Instead, the standards in this Code should be viewed as the *minimum standards* that Orphazyme expects from its directors, officers and employees in the conduct of its business.

1. Standards of conduct Conflicts of Interest

Orphazyme recognizes and respects the right of its directors, officers and employees to engage in outside activities, unless otherwise specifically agreed, which they may deem proper and desirable, provided that these activities do not impair or interfere with the performance of their duties to Orphazyme or their ability to act in Orphazyme's best interests. In most, if not all, cases this will mean that our directors, officers and employees must avoid situations that present a potential or actual conflict between their personal interests and Orphazyme's interests.

A "conflict of interest" occurs when a director's, officer's or employee's personal interest, including financial interest interferes with Orphazyme's interests. Conflicts of interest may arise in many situations. For example, conflicts of interest can arise when a director, officer or employee takes an action or has an outside interest, responsibility or obligation that may make it difficult for him or her to perform the responsibilities of his or her position objectively and/or effectively in Orphazyme's best interests. Conflicts of interest may also occur when a director, officer or employee or his or her immediate family member receives a personal benefit (whether improper or not) as a result of the director's, officer's or employee's position with Orphazyme. Each individual's situation is different and in evaluating his or her own situation, a director, officer or employee will have to consider many factors.

Further, no director or officer may participate in the transaction of business involving any agreement between Orphazyme and the director or officer or any legal proceedings against the director or officer or any agreement between Orphazyme and a third party or any legal proceedings against a third party if the director or officer has a material interest in such business that may conflict with the interests of Orphazyme.

Any material transaction or relationship that reasonably could be expected to give rise to a conflict of interest should be reported promptly to the Compliance Officer. The Compliance Officer may notify the Board of Directors or a committee thereof as he or she deems appropriate.

Compliance with Laws, Rules and Regulations

Orphazyme seeks to conduct its business in compliance with applicable laws, rules and regulations. No director, officer or employee shall engage in any unlawful activity in conducting Orphazyme's business or in performing his or her day-to-day company duties, nor shall any director, officer or employee instruct others to do so.

Anti-Bribery and Anti-Corruption

Orphazyme has a zero tolerance policy against bribery and corruption and condemns it in all forms. Orphazyme recognises that corruption and other unethical practices are undermining to its

business and therefore takes active measures to ensure that bribery and corruption does not occur in its business activities.

Orphazyme seeks to conduct its business in compliance with the U.S. Foreign Corrupt Practices Act of 1977 (the “FCPA”), Sections 122 and 299(2) of the Danish Criminal Code consolidated by Act no. 976 of 17 September 2019 (in Danish: *Straffeloven*) and other applicable anti-bribery and anti-corruption laws, rules and regulations, including the laws of such countries in which Orphazyme A/S has subsidiaries. To ensure compliance with such anti-corruption and anti-bribery laws, Orphazyme has adopted an Anti-Corruption Policy containing guidelines, standards and procedures applicable to directors, officers, and employees as well as business partners. Please direct any questions or concerns related hereto to Orphazyme’s Compliance Officer.

Protection and Proper Use of Orphazyme’s Assets

Loss, theft and misuse of Orphazyme’s assets has a direct impact on Orphazyme’s business and its profitability. Employees, officers and directors are expected to protect Orphazyme’s assets that are entrusted to them and to protect Orphazyme’s assets in general. Employees, officers and directors are also expected to take steps to ensure that Orphazyme’s assets are only used for legitimate business purposes.

Corporate Opportunities

Employees, officers and directors owe a duty to Orphazyme to act loyally towards Orphazyme and to advance its legitimate business interests when the opportunity to do so arises. Each employee, officer and director is prohibited from:

- diverting to himself or herself or to others any opportunities that are discovered through the use of Orphazyme’s property or information as a result of his or her position with Orphazyme unless such opportunity has first been presented to, and rejected by, Orphazyme;
- using Orphazyme’s property or information or his or her position for improper personal gain; or
- competing with Orphazyme.

Confidentiality

Confidential information generated and gathered in Orphazyme’s business plays a vital role in Orphazyme’s business, prospects and ability to compete. “Confidential information” includes all non-public information that might be of use to competitors or harmful to Orphazyme, its partners or collaborators or its customers if disclosed. Directors, officers and employees may not disclose or distribute Orphazyme’s confidential information, except when disclosure is authorized by Orphazyme or required by applicable law, rule or regulation or pursuant to an applicable legal proceeding. Directors, officers and employees shall use confidential information solely for legitimate business purposes. Directors, officers and employees must return or provide evidence

of destruction of all of Orphazyme's confidential and/or proprietary information in their possession to Orphazyme when they cease to be employed by or to otherwise serve Orphazyme.

Fair Dealing

Competing vigorously, yet lawfully, with competitors and establishing advantageous, but fair, business relationships with business partners is a part of the foundation for long-term success. However, unlawful and unethical conduct, which may lead to short-term gain, may damage a company's reputation and long-term business prospects. Accordingly, it is Orphazyme's policy that directors, officers and employees must endeavor to deal ethically and lawfully with Orphazyme's suppliers and other business partners, competitors and employees in all business dealings on Orphazyme's behalf. No director, officer or employee should take unfair advantage of another person in business dealings on Orphazyme's behalf through the abuse of privileged or confidential information or through improper manipulation, concealment or misrepresentation of material facts.

Accuracy of Records

The integrity, reliability and accuracy in all material respects of Orphazyme's books, records and financial statements is fundamental to Orphazyme's continued and future commercial success. No director, officer or employee may cause us to enter into a transaction with the intent to document or record it in a deceptive or unlawful manner. In addition, no director, officer or employee may create any false or artificial documentation or book entry for any transaction entered into by Orphazyme. Similarly, officers and employees who have responsibility for accounting and financial reporting matters have a responsibility to accurately record all funds, assets and transactions on our books and records.

Quality of Public Disclosures

Orphazyme is committed to providing its shareholders with information about its financial condition and results of operations as required by the securities laws of Denmark, the United States and all other relevant jurisdictions. It is Orphazyme's policy that the reports and documents it files with or submits to the U.S. Securities and Exchange Commission (the "SEC"), Nasdaq Copenhagen and (if relevant) the Danish Financial Supervisory Authority, and the earnings releases and similar public communications made by Orphazyme, include fair, timely, understandable, full and accurate disclosure. Officers and employees who are responsible for these filings and disclosures, including Orphazyme's principal executive, financial and accounting officers, must use reasonable judgment and perform their responsibilities honestly, ethically and objectively in order to ensure that this disclosure policy is fulfilled. Orphazyme's senior management is primarily responsible for monitoring Orphazyme's public disclosure.

Code of Ethics for the CEO and Senior Financial Officers

The CEO and all senior financial officers, including the CFO, the Director of Finance and persons performing similar functions– (the “**Senior Financial Officers**” and together with the CEO, the “**Named Officers**”) shall:

- engage in and promote honest and ethical conduct, including the ethical handling of actual or apparent conflicts of interest between personal and professional relationships;
- avoid conflicts of interest and disclose to the chairman of the Audit Committee any material transaction or relationship that reasonably could be expected to give rise to such a conflict;
- take all reasonable measures to protect the confidentiality of non-public information about Orphazyme and its customers obtained or created in connection with their activities and prevent the unauthorized disclosure of such information unless required by applicable law or regulation or legal or regulatory process;
- take all reasonable measures to achieve responsible use of and control over Orphazyme’s assets and resources;
- promote full, fair, accurate, timely, and understandable disclosure in material respects in reports and documents that Orphazyme files with, or submits to, the SEC, Nasdaq Copenhagen, the Danish Financial Supervisory Authority and other regulators and in other public communications made by Orphazyme in accordance with the following guidelines:
 - o all accounting records, and the reports produced from such records, must be in accordance with all applicable laws;
 - o all accounting records must fairly and accurately reflect the transactions or occurrences to which they relate;
 - o all accounting records must fairly and accurately reflect in reasonable detail in accordance with generally accepted accounting principles Orphazyme’s assets, liabilities, revenues and expenses;
 - o all accounting records must not contain any materially false or intentionally misleading entries;
 - o no transactions should be intentionally misclassified as to accounts, departments or accounting periods;
 - o all transactions must be supported by accurate documentation in reasonable detail and in all material respects to be recorded in the proper account and in the proper accounting period;

- no information should be concealed from the auditors; and compliance with Orphazyme’s system of internal controls is required;
- comply with all governmental laws, rules and regulations applicable to Orphazyme’s business, including taking necessary steps to avoid and, where possible, prevent any violations of the securities laws; and
- promptly report to the Chairman of the Audit Committee (or, if the Chairman is unavailable, to all other members of the Audit Committee) any fraud, whether or not material, involving management or other employees of Orphazyme who have a significant role in Orphazyme’s disclosures or internal controls over financial reporting; and promptly report any possible violation of this Section to the Chairman of the Audit Committee.

2. Compliance procedures Communication of Code

All directors, officers and employees will be supplied with a copy of the Code upon beginning service at Orphazyme and will, on a periodic basis, be asked to review and sign an acknowledgment that he or she has: (i) received a copy of the Code; (ii) read and understood the Code; (iii) not violated the Code; and (iv) agreed to comply with the Code. Updates of the Code will be provided from time-to-time. A copy of the Code is also available to all directors, officers and employees by requesting one from the Compliance Officer.

Monitoring Compliance and Disciplinary Action

Orphazyme’s management, under the supervision of its Board of Directors or a committee thereof or, in the case of accounting, internal accounting controls, auditing or securities law matters, the Audit Committee, shall take reasonable steps from time to time to (i) monitor compliance with the Code, (ii) when appropriate, impose and enforce appropriate disciplinary measures for violations of the Code, and (iii) when appropriate, report violators to the appropriate authorities.

Any failure to comply with this Code may result in Orphazyme taking disciplinary actions against those involved in such non-compliance.

Orphazyme’s management shall periodically report to the Board of Directors or a committee thereof on these compliance efforts including, without limitation, periodic reporting of alleged violations of the Code and the actions taken with respect to any such violation.

Reporting Concerns/Receiving Advice - Communication Channels

Be Proactive

Every employee is expected to act proactively by asking questions, seeking guidance and reporting suspected violations of the Code and other policies and procedures of Orphazyme, as well as any violation or suspected violation of applicable law, rule or regulation arising in the conduct of Orphazyme's business or occurring on its property. **If any employee believes that actions have taken place, may be taking place, or may be about to take place that violate or would violate the Code or any law, rule or regulation applicable to Orphazyme, he or she must bring the matter to the attention of Orphazyme.**

Seeking Guidance

The best starting point for an officer or employee seeking advice on ethics-related issues or reporting potential violations of the Code will usually be his or her supervisor. However, if the conduct in question involves his or her supervisor, if the employee has reported the conduct in question to his or her supervisor and does not believe that he or she has dealt with it properly, or if the officer or employee does not feel that he or she can discuss the matter with his or her supervisor, the employee may raise the matter with the Compliance Officer.

Communication Alternatives

Any officer or employee may communicate with the Compliance Officer by any of the following methods:

- in writing (which may be done anonymously as set forth below under "Reporting; Anonymity; Retaliation"), addressed to the Compliance Officer by mail to Orphazyme A/S, Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark; or
- via the independent external whistleblower portal (Link to whistleblower portal: <https://orphazyme.whistleblowernetwork.net>).

Reporting by an officer or employee through the above methods may be done anonymously, and the reporting should preferably be made in the company languages (English and Danish).

Reporting Accounting, Securities Law and Similar Concerns

Any concerns or questions regarding potential violations of the Code, any other company policy or procedure or applicable law, rules or regulations involving accounting, internal accounting controls, auditing or securities law (including the Foreign Corrupt Practices Act) matters should be directed to (i) the Audit Committee or a designee of the Audit Committee. Officers and employees may communicate with the Audit Committee or its designee: in writing to: Audit Committee, Orphazyme A/S, Ole Maaløes Vej 3, DK-2200 Copenhagen N, Denmark, or (ii) the independent

external whistleblower portal (<https://orphazyme.whistleblownetwork.net>), in accordance with Orphazyme's Whistleblower Policy.

Reporting by an officer or employee through the above methods may be done anonymously, and the reporting should preferably be made in the company languages (English and Danish).

Cooperation

Within the parameters of applicable law, employees are expected to cooperate with Orphazyme in any investigation of a potential violation of the Code, any other company policy or procedure, or any applicable law, rule or regulation.

Misuse of Reporting Channels

Employees must not use these reporting channels in bad faith or in a false or unreasonable manner.

Reporting; Anonymity; Retaliation

When reporting suspected violations of the Code, Orphazyme prefers that officers and employees identify themselves in order to facilitate its ability to take appropriate steps to address the report, including conducting any appropriate investigation. However, we also recognize that some people may feel more comfortable reporting a suspected violation anonymously.

If an officer or employee wishes to remain anonymous, he or she may do so, and Orphazyme will use reasonable efforts to protect the confidentiality of the reporting person subject to applicable law, rule or regulation or to any applicable legal proceedings. In the event the report is made anonymously, however, Orphazyme may not have sufficient information to look into or otherwise investigate or evaluate the allegations. Accordingly, persons who make reports anonymously should provide as much detail as possible to permit Orphazyme to evaluate the matter(s) set forth in the anonymous report and, if appropriate, commence and conduct an appropriate investigation.

No Retaliation

Orphazyme expressly forbids any retaliation against any officer or employee who, acting in good faith on the basis of a reasonable belief, reports suspected misconduct. Specifically, Orphazyme will not discharge, demote, suspend, threaten, harass or in any other manner discriminate against, such an officer or employee in the terms and conditions of his or her employment. Any person who participates in any such retaliation is subject to disciplinary action, including termination.

Waivers and Amendments

No waiver of any provisions of the Code for the benefit of a director or an executive officer (which

includes, without limitation, for purposes of this Code, Orphazyme's principal executive, financial and accounting officers) shall be effective unless (i) approved by the Board of Directors or, if permitted, a committee thereof, and (ii) if applicable, such waiver is promptly disclosed to Orphazyme's shareholders in accordance with applicable U.S. securities laws and/or the rules and regulations of the exchange or system on which Orphazyme's shares are traded or quoted, as the case may be.

Any waivers of the Code for other employees may be made by the Compliance Officer, the Board of Directors or, if permitted, a committee thereof.

All amendments to the Code must be approved by the Board of Directors or a committee thereof and, if applicable, must be promptly disclosed to Orphazyme's shareholders in accordance with applicable U.S. securities laws and/or the rules and regulations of the exchange or system on which Orphazyme's shares are traded or quoted, as the case may be.

Approved on August 20, 2020