

ANNUAL REPORT

2022

Orphazyme A/S

Company registration no.: 32266355

Ole Maaløes Vej 3, DK-2200

Copenhagen N, Denmark

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Management Review

Introduction

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).

In light of this, and considering Orphazyme's financial situation at the time, in March 2022, the Board of Directors of Orphazyme filed a petition to initiate 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, the Company 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 Orphazyme's operations to continue, including a basis for injecting further capital, and/or a basis for a sale of all or parts of its assets.

Also in March 2022, the Company voluntarily delisted American Depositary Shares (ADSs) representing Orphazyme's ordinary shares from the Nasdaq Global Select Market in the United States, effective March 31, 2022. The Deposit Agreement among the Company, The Bank of New York Mellon, as depositary, and owners and American Depositary Receipt (ADRs) holders was terminated, effective July 6, 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. (now known as Zevra Therapeutics¹) 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). 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, Orphazyme 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 provided full coverage to creditors with undisputed claims based on the claims filed during the restructuring. All known claims related to the time prior to restructuring have been paid in full, including all obligations outstanding under the Company's debt facility with Kreos Capital.

Since the Sale of Assets, the focus of the Board of Directors and management has been to ensure the smooth transition of assets to KemPharm and manage our limited business operations in as lean and cost-efficient manner as possible. The Company has also continued to assist its legal representatives with a class action lawsuit in the United States. In April 2023, the Company and the other parties to the class action lawsuit in the United States reached an agreement in principle to settle the action in its entirety. More details on the class action lawsuit can be found in the following sections in the Management Review: 'Letter from the CEO', 'Risk Management' and in Note 3.9 to the Financial Statements.

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

¹ On February 22, 2023 KemPharm Inc. changed its name to Zevra Therapeutics. However, KemPharm will be used throughout this annual report as the Sale of Assets was completed prior to this, within the reporting period.

Key Figures & Ratios

Key figures & ratios for 2022 and 2021 are presented with a split of continuing and discontinued operations for P&L and cash flow to reflect restructuring and the KemPharm transaction. 2020, 2019 and 2018 figures have not been restated since they represent a significantly different stage of the business and such presentation is not meaningful to an understanding of Orphazyme's current business.

| TDKK | 2022 | 2021 | 2020 | 2019 | 2018 |
|--|-----------------|------------------|------------------|------------------|------------------|
| Statement of Profit and Loss and other comprehensive income | | | | | |
| Net revenue | - | - | - | - | - |
| R&D expenses | - | - | (361,284) | (285,413) | (196,525) |
| G&A expenses | (41,241) | (83,472) | (247,250) | (50,541) | (35,127) |
| Operating Loss | (41,241) | (83,472) | (608,534) | (335,954) | (231,652) |
| Net Financial items | 193 | 37 | (26,627) | (7,043) | (3,448) |
| Loss Before Tax | (41,048) | (83,436) | (635,161) | (342,997) | (235,100) |
| Income tax benefit | 2,736 | 4,941 | 1,915 | 5,500 | 5,500 |
| Loss from continuing operations | (38,312) | (78,495) | - | - | - |
| Net result from discontinued operations | 64,382 | (548,044) | - | - | - |
| Net result for the period | 26,070 | (626,539) | (633,246) | (337,497) | (229,600) |
| Total comprehensive profit/ loss | 25,165 | (626,841) | (632,641) | (337,430) | (229,558) |
| Total profit/loss per share, basic (DKK) | 0.74 | (17.94) | (22.32) | (16.87*) | (11.5*) |
| * adjusted retrospectively, see Note 4.3 in consolidated financial statements. See also footnote (4) | | | | | |
| Statement of financial position | | | | | |
| Intangible Assets | - | 2,152 | 12,454 | 10,539 | 10,744 |
| Right-of-use assets | - | 5,434 | 14,859 | 13,903 | - |
| Property, plant, and equipment | - | 2,985 | 4,687 | 3,685 | 1,940 |
| Other non-current assets | - | 3,714 | 6,829 | - | - |
| Total non-current assets | - | 14,285 | 38,829 | 32,529 | 17,965 |
| Cash | 42,464 | 102,255 | 726,929 | 123,588 | 394,706 |
| Other current assets | 15,658 | 56,689 | 56,735 | 19,137 | 28,678 |
| Total assets | 58,122 | 173,229 | 822,493 | 180,754 | 441,349 |
| Share capital | 35,312 | 34,952 | 34,698 | 19,984 | 19,939 |
| Total equity | 41,667 | 9,339 | 620,525 | 52,969 | 388,249 |
| Non-current borrowings | - | 2,482 | 23,830 | 51,606 | - |
| Non-current lease liabilities | - | 3,925 | 9,877 | 9,813 | - |
| Other non-current liabilities | 98 | 98 | 1,634 | - | - |
| Total current liabilities | 16,357 | 129,092 | 166,627 | 65,988 | 52,995 |
| Cash flow statement | | | | | |
| Cash flow from operating activities | (117,945) | (602,571) | (539,076) | (326,818) | (234,764) |
| Cash flow from investing activities | 90,347 | 46 | (5,101) | (3,285) | (2,346) |
| Cash flow from financing activities | (35,078) | (30,344) | 1,159,422 | 58,939 | - |
| Net cash flow from discontinued operations | 32,862 | (549,447) | - | - | - |
| Other | | | | | |
| Share Price (DKK) | 0.88 | 17.16 | 67.10 | 72.40 | 43.35 |
| Total outstanding shares | 35,312,241 | 34,952,241 | 28,514,047 | 19,984,799 | 19,939,564 |
| Market capitalization (MDKK) ¹ | 31.1 | 599.8 | 1,913.3 | 1,446.9 | 864.4 |
| Equity ratio ² | 72% | 5% | 75% | 29% | 88.0% |
| Equity per share (DKK) ³ | 1.18 | 0.27 | 21.76 | 2.65 | 19.47 |
| Average number of employees | 21 | 130 | 117 | 74 | 46 |
| Number employees at year end | 1 | 62 | 141 | 86 | 57 |

(1) Market cap is calculated as the share price multiplied with the total outstanding shares as of the balance sheet date; (2) Equity ratio is calculated as the equity divided by the total assets as of the balance sheet date; (3) Equity per share is calculated as the total equity divided by the total outstanding shares as of the balance sheet date.

Letter from the CEO

2022 was a challenging year for Orphazyme which culminated, in May 2022, with the sale of substantially all of our assets and business activities to KemPharm Denmark A/S (KemPharm), a wholly owned subsidiary of KemPharm Inc (the Sale of Assets). As of December 31, 2022, the Company had one employee.

Prior to the Sale of Assets, Orphazyme's focus was on developing our investigational product candidate arimoclomol for the treatment of Niemann-Pick disease type-C (NPC), an ultra-rare disease with limited available treatment options. Arimoclomol was under regulatory review in Europe but in February 2022, we were notified by the Committee for Medicinal Products for Human Use (CHMP) of the European Medicines Agency (EMA) of a negative trend vote relating to our marketing authorisation application (MAA). We withdrew our MAA in March 2022, ahead of the CHMP's final vote. We were also working towards registration of arimoclomol in the United States, where we had received a Complete Response Letter (CRL) from the U.S. Food and Drug Administration (FDA) for our New Drug Application (NDA) in June 2021.

In light of our financial situation at the time, and the negative trend vote from the CHMP, in March 2022, the Board of Directors of Orphazyme commenced in-court restructuring proceedings of Orphazyme, further details of which are outlined in the Introduction. As a result of this process, we sold substantially all of our assets and business activities, including those relating to the development and approval of arimoclomol and the early access programs, to KemPharm for a total of USD 12.8 million in cash and assumed liabilities estimated to equal approximately USD 5.2 million. Under the terms of the agreement, KemPharm agreed to also retain a majority of Orphazyme's remaining Danish employees. The Sale of Assets was completed May 31, 2022 and it provided full coverage to creditors with undisputed claims filed during the restructuring as well as all (undisputed and unconditional) debts related to the time prior to restructuring, including all obligations outstanding under the Company's debt facility with Kreos Capital.

Considering the limited nature of our business following the Sale of Assets, Stephanie Okey, Carrolee Barlow and Martin Bonde stepped down from their positions as members of the Board of Directors of Orphazyme as of May 23, 2022. At the Annual General Meeting (AGM) in June 2022, the Board of Directors was further reduced to three members: Bo Jesper Hansen assumed the role of Chairman, and John Sommer Schmidt and myself, Anders Vadsholt, were appointed as new Board members.

Since the Sale of Assets the focus of the Board of Directors and Management has been to ensure the smooth transition of assets to KemPharm. This has included the transfer of data, contracts and other obligations that were part of the agreement. We have also continued to assist our legal representatives with a previously communicated putative class action lawsuit, filed in July 2021 in the United States. While Management does not believe the class action lawsuit claims have any merit, the Company decided, for cost control reasons and because of the risks inherent in any litigation, to engage in settlement discussions. In April 2023, the parties reached an agreement in principle to settle this action in its entirety. The Court has stayed all proceedings in the action pending the filing of the parties' settlement documents. The parties currently anticipate that a motion seeking the Court's preliminary approval of the settlement will be filed in May 2023 and that, if preliminary approval is granted, final court approval will be sought after members of the proposed settlement class have been afforded an opportunity to object to or opt out of the settlement. As the legal process is ongoing and no settlement has yet been concluded or approved by the Court, material uncertainty remains with regards to the final outcome. Whether or not the parties' proposed settlement is ultimately concluded and approved by the Court, the Company believes it will be able to continue its operations until at least December 31, 2023 and therefore is regarded as a going concern. We expect to end 2023 with DKK 6 – 10 million in cash.

The Board of Directors continues to assess ways to maximize value for shareholders, including exploring ways to realize value from the Company's remaining assets and listing on Nasdaq Copenhagen.

Our limited business activities have been operated in as lean and cost-efficient manner as practicable in order to conserve our limited capital. We ended year with DKK 42.5 million in cash as of as of December 31, 2022.

I would like to extend my sincere gratitude to Orphazyme's alumni and to our long-standing shareholders for their commitment during a challenging year for Orphazyme.

Anders Vadsholt, Chief Executive Officer & Chief Financial Officer

2022 Financial Review

Introduction

Substantially all of Orphazyme's assets and business activities, including those relating to the development and approval of arimoclomol, the full claw back liability related to the French early access program and costs associated with certain employees, were transferred to KemPharm Denmark A/S (KemPharm), a wholly owned subsidiary of KemPharm, Inc., from June 1, 2022, in line with the terms of the Sale of Assets agreement which was completed on May 31, 2022. These activities have been reclassified as discontinued operations, cf note 1.7, in the presentation of the consolidated Financial Statements. Please note the figures below are presented on a continuing basis, unless otherwise described.

Income statement

The net result for the full year ended December 31, 2022 (from continuing and discontinued operations) was a profit of DKK 26.1 million compared to a net loss of DKK 626.5 million for the same period in 2021.

Loss from continuing operations was DKK 38.3 million compared to a loss of DKK 78.5 million for the full year 2021. The reduction in loss from continuing operations of DKK 40.2 million compared to the prior year is primarily due to a reduction in operating expenses following the restructuring process and the Sale of Assets to KemPharm. Note that the loss from continuing operations for 2022 of DKK 38.3 million is marginally better than our previously communicated outlook of a loss from continuing operations of DKK 40 – 45 million for the period.

The net result from discontinued operations was a profit of DKK 64.4 million for the full year compared to a net loss of DKK 548.0 million for the same period in 2021. The change primarily relates to the net effect of income received from the Sale of Assets to KemPharm and higher costs during the same period the prior year associated with preparations for commercial launch of arimoclomol in the U.S., accelerated close out of certain clinical trials and impairment costs following unfavorable results in two late-stage clinical trials in amyotrophic lateral sclerosis (ALS) and inclusion body myositis (IBM), initiation of a restructuring program and termination of onerous contracts. Note that the net result from discontinued operations of DKK 64.4 million was lower than our previously communicated guidance of DKK 87.4 million as a result of higher-than-expected expenses relating to discontinued operations in the second half of 2022.

Net revenue

As substantially all Orphazyme's business activities were sold to KemPharm, including any revenue from sales of arimoclomol, net revenue from continuing operations for the full year 2022 was DKK 0 million (DKK 0 million for 2021). Net revenue from discontinued operations was DKK 17.9 million in 2022 compared to DKK 36.2 million in the same period in 2021. The reduction in net revenue compared to the prior year is due to the sale of assets to KemPharm resulting in only a partial year of net revenues recorded in 2022.

Research and development expenses

Following the Sale of Assets to KemPharm, research and development (R&D) expenses have also been reclassified as discontinued operations. As such, R&D expenses from continuing operations totaled DKK 0 million for the full year 2022 (DKK 0 million for the same period in 2021). R&D expenses from discontinued operations totaled DKK 36.7 million for the year ended December 31, 2022 compared to DKK 330.0 million in 2021. The difference primarily relates to higher costs in 2021 associated with manufacturing, accelerated close out of certain clinical trials following unfavorable results and associated impairment costs, initiation of a restructuring program and termination of onerous contracts.

General and administrative expenses

General and administrative (G&A) expenses from continuing operations totaled DKK 41.2 million in 2022, a reduction of approx. DKK 42 million compared to the prior year (DKK 83.5 million in 2021). G&A expenses include costs associated with employees and Board of Directors, service providers and external assistance, legal and technology expenses. G&A expenses from discontinued operations were DKK 59.5 million for the year ended December 31, 2022, compared to DKK 256.0 million for the same period in 2021. The difference primarily relates to lower costs in 2022 following the Sale of Assets to KemPharm against costs in 2021 associated with the build-up of a commercial organization, including commercial launch preparation activities,

as well as expenses related to support functions and an impairment charge for software and the employee restructuring program.

Net financial items

Net financial income from continuing operations for the financial period ended December 31, 2022, was DKK 193 thousand compared to DKK 37 thousand for the same period in 2021. Net financial items from discontinued operations were an expense of DKK 2.8 million in 2022 compared to an income of DKK 1.8 million in 2021. The difference primarily relates to fluctuations in foreign exchange rates year over year, particularly the USD / DKK exchange rate.

Income tax benefit

Income tax benefit from continuing operations totaled DKK 2.7 million for the full year ended December 31, 2022, compared to DKK 4.9 million for the same period in 2021. This includes income tax benefit related to tax credits for R&D expenses at the applicable tax rate under the Danish Corporate Income Tax Act. The reduction compared to the prior year is due to cessation of eligible R&D activities following the Sale of Assets to KemPharm resulting in only a partial year of tax benefit being recorded in 2022. Income tax benefit from discontinued operations was DKK 0 million during 2022 compared to DKK 0 million during 2021.

Statement of financial position

Cash

As of December 31, 2022, Orphazyme held cash of DKK 42.5 million as compared to DKK 102.3 million as of December 31, 2021. Note the cash position at the end of 2022 is in line with our previously communicated outlook for cash of >DKK 30 million for the period.

Equity

As of December 31, 2022, total equity amounted to DKK 41.7 million compared to DKK 9.3 million as of December 31, 2021. The increase relates to a positive impact from the Sale of Assets transaction with KemPharm.

Cash flows

Cash flow from operating activities

Net cash flow from operating activities (continuing and discontinued operations) amounted to an outflow of DKK 117.9 million for the full year ended December 31, 2022, compared to an outflow of DKK 602.6 million for the same period in 2021. The difference in cash flow from operating activities during the period was due to higher costs in the prior year relating to clinical, manufacturing and pre-commercial activities relating to arimoclomol, among other operating costs. Net cash flow from discontinued operating activities was an outflow of DKK 17.1 million in 2022, compared to an outflow of DKK 520.3 million in 2021.

Cash flow from investing activities

Net cash inflow from investing activities amounted to DKK 90.3 million in 2022 (cash inflow of DKK 46 thousand in 2021). The change compared to the prior year primarily relates to proceeds for the Sale of Assets to KemPharm.

Cash flow from financing activities

Net cash flow from financing activities amounted to an outflow of DKK 35.1 million in 2022 compared to an outflow of DKK 30.3 million in 2021. The difference relates to the net effect of cash inflow relating to the U.S. At-the-market program share issue in H1 2022 and repayment of the debt facility with Kreos Capital, also in H1 2022, compared to cash outflows in H1 2021 relating to bonus share issues due to the license agreement with KLSDC and UCL (note 3.1).

Other disclosures

Uncertainty regarding recognition and measurement, unusual matters and going concern

We have continued to assist our legal representatives with a previously communicated putative class action lawsuit, filed in July 2021 in the United States. While Management does not believe the class action lawsuit claims have any merit, the Company decided, for cost control reasons and because of the risks inherent in any litigation, to engage in settlement discussions. In April 2023, the parties reached an agreement in principle to settle this action in its entirety. The parties are now undertaking to prepare mutually agreeable settlement documents, and the Court has stayed all proceedings in the action pending the filing of the parties' settlement documents. The parties currently anticipate that a motion seeking the Court's preliminary approval of the settlement will be filed in May 2023 and that, if the Court grants preliminary approval, final court approval will be sought after members of the proposed settlement class have been afforded an opportunity to object to or opt out of the settlement. As the legal process is ongoing and no settlement has yet been concluded or approved by the Court, uncertainty remains with regards to the final outcome. Whether or not the parties' proposed settlement is ultimately concluded and approved by the Court, the Company believes it will be able to continue its operations until at least December 31, 2023. For further information reference is made to note 3.9 to the consolidated Financial Statements.

Outlook

For the full-year 2023 we anticipate an operating loss in the range of DKK 30 – 35 million. We expect to end 2023 with DKK 6 - 10 million in cash. There are inherent risks and uncertainties in our Outlook for 2023 including the limited nature of our business activities, the class action lawsuit in the United States and our future prospects. For further information please refer to note 3.9 of the consolidated Financial Statements and for further information on risks please see the Risk Management section in the Management Review.

Shareholder Information

The share

Orphazyme's shares are listed on Nasdaq Copenhagen (since November 16, 2017) under the ticker symbol ORPHA. Orphazyme previously had ADSs listed on Nasdaq Global Select Market under the ticker symbol ORPH until the Company's voluntary delisting of the ADSs, which became effective on March 31, 2022.

We conduct our communications in accordance with the applicable rules and regulations required under Danish, and EU laws, including as set forth by the Danish Financial Supervisory Authority.

Ownership and share capital

On November 4, 2021, Orphazyme established a U.S. At-the-Market Offering Program (ATM Program) with Cowen and Company, LLC, pursuant to which Orphazyme may issue and sell ADSs having an aggregate offering price of up to USD 50,000,000, each ADS representing one ordinary share of Orphazyme, to be sold in the United States at market price, from time to time, at its option, in "at the market" transactions on Nasdaq Global Select Market. On February 11, 2022, Orphazyme issued and sold a total of 360,000 shares of nominally DKK 1 each, represented by ADSs, under the ATM Program entailing an increase of the share capital of a total of 360,000 shares of nominally DKK 1 each. Following the share capital increase, Orphazyme's share capital amounts to a nominal value of DKK 35,312,241 divided into 35,312,241 ordinary shares of DKK 1 per share. Reference is made to company announcements as published by Orphazyme on November 4, 2021 and February 18, 2022.

As of December 31, 2022, Orphazyme has a total share capital of nominally DKK 35,312,241 divided into 35,312,241 shares and representing a total of 35,312,241 voting rights. As of December 31, 2022, 11,714,720 shares were represented by American Depositary Shares (ADS) held by The Bank of New York Mellon, as depositary.

As of December 31, 2022 the Company had approximately 11,783 registered shareholders. Please refer to note 4.2 for further information on our share capital.

Shareholders rights and ADS holders

All ordinary shareholders have the same rights, including in respect of eligibility to receive dividends under the Danish Companies Act.

Orphazyme voluntarily delisted the Company's ADSs representing its shares from Nasdaq Global Select Market in the US, effective March 31, 2022. In connection with this, the Deposit Agreement with the Bank of New York Mellon, as depositary, was terminated July 6, 2022 and holders of ADRs were asked to surrender ADRs for delivery of the underlying shares by July 11, 2022. If the shares were not surrendered then, under the terms of the Deposit Agreement, the Depositary may attempt to sell the underlying shares. If the Depositary has sold such shares, ADR Holders must surrender their ADRs to obtain payment of the sale proceeds, 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 ADRs.

Dividend policy

Orphazyme has not declared or made any dividend payments for the last two financial years. The Company has limited ongoing operational business activities and, as of the date of this annual report, does not expect to make dividend payments within the foreseeable future.

Major Shareholders (>5% share capital)

As of the publication of this Annual Report, LSP V Cooperatieve U.A., Johannes Vermeer, Plein 9, 1071 DV Amsterdam, Netherlands owns 2,426,711 shares representing 6.87% of total share capital.

Corporate Governance

Orphazyme is committed to ensuring transparent and good corporate governance. As a Danish company listed on Nasdaq Copenhagen, Orphazyme is subject to the Danish Recommendations on Corporate Governance. The Recommendations on Corporate Governance are best practice guidelines for the management of companies admitted to trading on a regulated market.

Orphazyme complies with the Recommendations on Corporate Governance where deemed relevant given Orphazyme's current situation and focus. Therefore, the Company has opted to deviate in the following areas:

- Given the Company's current situation, and as the Company has focused its limited resources on handling the Company's restructuring proceedings, the Company has decided only to publish annual reports and half-yearly financial reports.
- The Company has limited resources and has therefore not provided and does not intend to provide a webcast or other digital transmission of the general meeting.
- The Company has focused on the restructuring proceedings, as well as assisting its legal representatives with a putative class action lawsuit in the United States, and therefore the Company's purpose is not currently being used actively as part of the Company's strategy.
- Given the Company's current situation, including limited resources and the size of the Board of Directors, at least half of the board members elected by the general meeting are not considered independent. Moreover, the majority of the members of the board committees are not considered independent.
- The Company has not included information in the annual report on board members' individual participation in board meetings and committee meetings. Any board member unable to attend a board or committee meeting has the opportunity to present his or her views or engage in dialogue with the Chairmanship regarding the items of the agenda prior to the board or committee meeting. Accordingly, a board member may contribute to the discussions at a board or committee meeting, even if he or she is not present.
- Given the Company's current situation, the Board of Directors has chosen not to adopt a policy for the Company's corporate social responsibility and tax, as the Company has focused its limited resources on handling the Company's restructuring proceedings.
- Given the Company's current situation, including limited resources, a member of the Executive Management is temporarily also a member of the Board of Directors.
- Given the Company's current situation, the Company has focused its limited resources on handling the Company's restructuring proceedings and legal matters. Therefore, the Company has not included information in the management commentary on the board committees' most significant activities and number of meetings in the past year.
- Given the Company's current situation, and as the Company has focused its limited resources on handling the Company's restructuring proceedings and legal matters, the Board of Directors has not engaged external assistance in the evaluation of the Board of Directors.
- The general conclusions of the latest evaluation of the Board of Directors are not described in the management commentary, however, the Chairman will account for the evaluation process and the general conclusions at the annual general meeting.
- Given the Company's current situation, the Company has not used its limited resources on considering the potential value of the variable remuneration at the time of exercise under pessimistic, expected, and optimistic scenarios.
- Share-based compensation, e.g. shares, share options, performance shares or warrants, constitutes a common part of the board remuneration in international biotech companies. Due to the Company's status as a biotech company, Orphazyme has programs in place which offers share-based incentives to the Board of Directors in the form of Restricted Share Units. As members of the Board of Directors are elected for a term of one year, the share-based instruments granted to board members have a vesting period of one year. No share-based incentives were granted to the Board of Directors in 2022.
- Orphazyme believes that share-based remuneration may serve shareholders' long-term interests as share-based incentives together with the base fee support the objective of lasting value creation for the shareholders.

Orphazyme’s corporate governance statement includes a summary of the Company’s governance structure, a description of internal control and financial reporting procedures, Orphazyme’s position on the Recommendation on Corporate Governance as well as a complete list of the Company’s comments to recommendations that the Company opted to deviate from.

The corporate governance statement is available under “Corporate Governance” in the *Investors & Media* section of our website: investors.orphazyme.com/corporate-governance.

Board of Directors

The Board of Directors is responsible for the overall management and strategic direction of Orphazyme’s business and operations and it supervises the Company’s activities, management, and organization. The Board of Directors appoints and dismisses the members of the Executive Management, who are responsible for the day-to-day management of the Company.

Meetings

The Board of Directors normally holds at least five regular meetings annually, including a strategy review, plus ad-hoc meetings as required. Extraordinary board meetings are convened by the Chairman when necessary or when requested by a member of the Board of Directors, a member of the Executive Management, or by the Company’s auditor. There was a higher frequency of meetings in H1 2022 due to the in-court restructuring process, and three Board meetings in H2 2022. The Board of Directors forms a quorum when more than half of its members are represented, including the Chairman or the Deputy Chairman. Resolutions of the Board of Directors are passed by a simple majority of the votes present at the meeting. In the event of equal votes, the Chairman or, in his absence, the Deputy Chairman shall have the casting vote. The Board of Directors conducts an annual evaluation of the effectiveness, performance, achievements, and competencies of the Board of Directors and of the individual members as well as the collaboration with the Executive Management.

The members of the Board of Directors elected by the general meeting are elected for a term of one year. Members of the Board of Directors may be re-elected.

Orphazyme Board of Directors

| Name | Position | Independent ⁽¹⁾ | Year of first appointment | Expiration of term |
|---------------------|-----------------|----------------------------|---------------------------|--------------------|
| Bo Jesper Hansen | Chairman | Non-independent | 2010 | 2023 |
| John Sommer Schmidt | Deputy Chairman | Independent | 2022 | 2023 |
| Anders Vadsholt | Member | Non-independent | 2022 | 2023 |

(1) According to the Danish Recommendations on Corporate Governance at least half of the members of the Board of Directors should be independent.

Board Committees

To support the Board of Directors in its duties, the Board of Directors has established and appointed an Audit Committee, a Nomination Committee, and a Remuneration Committee. These committees are charged with reviewing issues pertaining to their respective fields that are due to be considered at board meetings. The Board of Directors discontinued a Science Committee in May 2022.

Audit Committee

Members

John Sommer Schmidt
(Chairman)

Bo Jesper Hansen
(Member)

Purpose & Key Roles

- Reviews and evaluates certain accounting and audit matters that by decision of the Board of Directors or the Audit Committee require a more thorough evaluation;
- Assesses internal controls and risk management systems;
- Supervises the Company’s auditors and review the audit process;

- The Audit Committee Chairman monitors Orphazyme’s Whistleblower Hotline.

Key Requirements

- No less than two members appointed by and among the Board of Directors, including the Chairman of the Audit Committee.
- The Chairman of the Board of Directors may not also be the Chairman of the Audit Committee.
- The members of the Audit Committee are required to meet the independence requirements set out in the Corporate Governance Recommendations. Currently, one member does not meet such independence requirements.
- At least one member shall have accounting or audit qualifications and between them, the members shall possess such expertise and experience to be able to provide an updated insight into, and experience in, the financial, accounting, and audit aspects of companies with shares admitted to and trading on a regulated market.
- The Company’s external auditor shall participate in meetings of the Audit Committee if requested by the Audit Committee.
- The external auditor shall attend at least one meeting per year, of the relevant part thereof, where the Executive Management is not present.

Nomination Committee

Members

Bo Jesper Hansen
(Chairman)

John Sommer Schmidt
(Member)

Anders Vadsholt
(Member)

Purpose & Key Roles

- Assists the Board of Directors in ensuring that appropriate plans and processes are in place for the nomination of candidates to the Board of Directors and the board committees;
- Evaluates the composition of the Board of Directors, including making recommendations for nomination or appointment of members of (a) the Board of Directors and (b) board committees.
- Evaluates the composition of the Executive Management annually, including making recommendations for nomination or appointment of members of the Executive Management;
- Assists the Board of Directors with ensuring that appropriate plans and processes are in place for nomination of candidates to the Executive Management.

Key Requirements

- No less than three members appointed by and among the Board of Directors.
- The members of the Nomination Committee are required to meet the independence requirements set out in the Corporate Governance Recommendations. Currently, two members do not meet such independence requirements.

Remuneration Committee

Members

Bo Jesper Hansen
(Chairman)

John Sommer Schmidt
(Member)

Purpose & Key Roles

- Ensures the Company maintains a Remuneration Policy for the members of the Board of Directors and the Executive Management;
- Evaluates and makes recommendations for the remuneration of the members of the Board of Directors and the Executive Management;
- Assist the review and preparation of the Company’s Remuneration Report.

Key Requirements

- No less than two members appointed by and among the Board of Directors.
- The members of the Remuneration Committee are required to meet the independence requirements set out in the Corporate Governance Recommendations. Currently, one member does not meet such independence requirements.

Visit the Corporate Governance section under *Investors & Media* at www.orphazyme.com for more information.

Internal controls and financial reporting procedures

The Board of Directors, the Audit Committee, and the Executive Management are responsible for risk management and internal controls over its financial reporting and approve general policies in that regard. The Audit Committee assists the Board of Directors in overseeing the reporting process and the most important risks involved in this respect. The Executive Management is responsible for the effectiveness of the internal controls and risk management and for the implementation of such controls aimed at mitigating the risk associated with the financial reporting.

The Board of Directors and Executive Management assess risks on an on-going basis, including risks related to financial reporting, and assess measures to manage, reduce, or eliminate identified risks. The Audit Committee reviews selected key risk areas on a frequent basis, including significant accounting estimates and material changes to accounting policies. At least once a year, the Audit Committee oversees a review of current internal controls to determine whether they are effective in relation to the risks identified in the financial reporting process.

Orphazyme has adopted and defined an internal control framework that identifies key processes, inherent risks, and control procedures in order to secure appropriate accounting processes. The control procedures include a variety of processes in order to prevent any misrepresentation, significant errors, omissions, or fraudulent behavior.

Orphazyme's independent auditors are appointed for a term of one year by the shareholders at the Company's annual general meeting upon recommendation from the Audit Committee. The Board of Directors assesses the independence and competencies and other matters pertaining to the auditors. The framework for the auditors' compensation and duties, including audit and non-audit tasks, is agreed annually between the Board of Directors and the auditors based on recommendations from the Audit Committee.

Risk Management

Our financial situation and risks are assessed on an ongoing basis and reported to the Audit Committee and the Board of Directors. The risks presented below relate to the limited ongoing business operations of Orphazyme following the sale of substantially all its assets and business activities to KemPharm Denmark A/S (KemPharm), a wholly owned subsidiary of KemPharm Inc. The following significant risks and uncertainties have been identified:

Risks relating to Securities litigation:

Risks: We have been and may in the future be the target of securities litigation which could result in substantial costs and divert our management's attention from other business concerns, which could seriously harm our financial position and business.

Actions to mitigate the risks: 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. While Management does not believe the class action lawsuit claims have any merit, the Company decided, for cost control reasons and because of the risks inherent in any litigation, to engage in settlement discussions. In April 2023, the parties reached an agreement in principle to settle this action in its entirety. The parties are now undertaking to prepare mutually agreeable settlement documents, and the Court has stayed all proceedings in the action pending the filing of the parties' settlement documents. The parties currently anticipate that a motion seeking the Court's preliminary approval of the settlement will be filed in May 2023 and that, if the Court grants preliminary approval, final court approval will be sought after members of the proposed settlement class have been afforded an opportunity to object to or opt out of the settlement.

As the legal process is ongoing and no settlement has yet been concluded or approved by the Court, uncertainty remains with regards to the final outcome. As a result, the Company has not recorded any provisions related to the class action lawsuit as of December 31, 2022. Whether or not the parties' proposed settlement is ultimately concluded and approved by the Court, the Company believes it will be able to continue its operations until at least December 31, 2023.

Retention of key personnel:

Risks: We may not be able to retain or attract key personnel and advisors to enable us to operate our business. Specifically, we rely on the efforts of our Chief Executive Officer and Chief Financial Officer, Anders Vadsholt. The loss of key personnel and advisors 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 our executive officer, Anders Vadsholt, we can make no assurances that he will continue to be employed.

Our actions to mitigate the risks: 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. Anders 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.

Risks relating to financial position and business prospects:

We may need to adjust our gross to net revenue estimate relating to rebates granted to co-financing the healthcare authorities, which could impact our financial position. Further information is provided in note 2.1 to the consolidated Financial Statements. Following the sale of substantially all our assets and business activities to KemPharm, we have limited operating business activities and employees. As such our future prospects are uncertain. We may choose to wind down our business, which could result in additional costs associated with any distribution which would further limit funds to shareholders.

Risks Related to the Sale of Assets:

We sold substantially all of Orphazyme's assets and business activities to KemPharm (the "Sale of Assets") 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.

Non-compliance with legislation and industry standards:

We are subject to regulatory and legislative obligations in order to conduct business. These requirements are subject to change and if we do not remain abreast of the regulations and actively work to comply, we are at risk of receiving penalties or fines. There is also a risk that cybersecurity attacks could compromise data privacy or cause interruption to our limited operations.

Currency risk:

Financial risks may arise from changes in exchange rates. We are most exposed to foreign exchange movements relating to the DKK, USD, EUR.

Political and geopolitical conflict:

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.

Corporate Social Responsibility

This section constitutes the Company's statutory reporting according to Section 99a of the Danish Financial Statements Act.

Our business

Until recently, Orphazyme A/S was a biopharmaceutical company involved in the research and development of novel therapeutics for the treatment of neurodegenerative rare diseases. In May 2022, substantially all our assets and business activities were sold to KemPharm Denmark A/S, a wholly owned subsidiary of KemPharm Inc., and we now have limited ongoing operational business activities and employees. Our headquarters is in Copenhagen, and we have non-operating subsidiaries in the U.S. and Switzerland.

Our responsibility

Before the Sale of Assets, our primary purpose was working towards a common vision: To profoundly impact the lives of patients with underserved diseases. We established a team connected by a set of core values, focused on courage, integrity, care and perseverance, which underpinned our corporate culture and guided our responsibility towards society, patients, employees, and our stakeholders.

In May 2022, the majority of our Danish employees were transferred to KemPharm, leaving us with minimal business operations and a couple of employees. As at December 31, 2022 there was only one permanent employee remaining at Orphazyme, CEO and CFO, Anders Vadsholt.

In light of such limited business operations, Orphazyme's Corporate and Social Responsibility (CSR) risks are considered very limited. Our CSR activities for 2022 were commensurate with the size and limited operations of the Company, though we continued to strive to uphold our values and responsibilities towards society, employees, and our stakeholders. For 2023 we will continue, where possible, to fulfill our CSR obligations as we execute our strategy.

2022 CSR reporting areas

Human Rights

Risk

- Very Limited: Orphazyme has limited operations, employees and suppliers.

Actions

- Continued to respect internationally declared human rights and did not employ child labor.

Policies in place

Orphazyme acknowledges and supports the maintenance of internationally declared human rights and bases its work on the UN Universal Declaration of Human Rights and the interpretation that it is the responsibility of the State to protect, and the companies' responsibility to respect, these rights. Orphazyme interprets human rights to comprise respect for diversity.

- Diversity policy.
- Whistleblower policy.

Results

- No diversity related incidents reported in 2022.
- No human rights violations reported in 2022.
- Employee composition: Not meaningful given company structure (One employee as of December 31, 2022; male).
- Leadership (director level and above): Not meaningful given company structure (One employee as of December 31, 2022; male).
- Due to the changes to the board of directors that occurred in connection with the Sale of Assets to KemPharm we no longer have an equal representation of men and women on the board of directors. Thus, the Company has set a target of increasing the representation of women on the Board of Directors

to 40% by 2026 in accordance with the guidelines from the Danish Business Authority. As at year end 2022 the Board of Directors was comprised of three members, none of which were women.

Future Plans

- Continue to support and respect internationally declared human rights and will not employ child labor. Aim to improve Board diversity in the future, subject to expansion of current business operations.

Anti-Corruption & Bribery

Risk

- Very Limited: We do not tolerate the use of bribery or corruption to achieve business objectives. Given that we have limited operations, employees and suppliers our anti-corruption and bribery risk is very limited.

Actions

- The Company is committed to maintaining the highest standards of conduct and will not tolerate the use of bribery or corruption to achieve its business objectives.
- Anti-corruption and bribery training conducted when employees start.
- Legal & Compliance training refreshers, including anti-corruption and bribery

Policies in place

- Our policies on bribery and corruption are clearly set out in our anti-corruption policy and our employee handbook.

Results

- No bribery and corruption violations identified or reported in 2022.

Future Plans

- Continue to maintain the highest standards of conduct and not tolerate the use of bribery or corruption to achieve business objectives.

Environment & Climate

Risk

- Very limited: We have a very limited number of employees, minimal physical office presence and use external suppliers for certain activities such as administration, finance and legal activities which we believe have a low potential risk for impact on the environment & climate. Prior to the Sale of Assets, a potential environment & climate risk was in regard to the use of hazardous substances in our laboratories. The Company conducted its research and development activities in a highly regulated industry and followed applicable rules on hazardous substances in order to minimize such risks. The Company closed its laboratory space in Q1 2022 and therefore the potential risk from such hazardous substances to the environment & climate is now zero.

Actions

- Followed established procedures both during use and at disposal of hazardous substances.

Policies in place

- Considering the business of the Company, and its limited operations and employees, Orphazyme's general potential impact on the environment and climate and the impact of the climate on Orphazyme's business is viewed as minimal. Applicable rules and procedures were followed regarding the use of hazardous substances (no longer relevant due to closure of our laboratory space early in 2022) and we continue to endeavor to protect the environment and climate through mindful business practices such as, e.g. careful use of office materials and energy consumption.

Results

- Continued to keep records of all accidents in 2022.
- Recorded no records of spill of hazardous substances (monitored until our laboratory space was vacated in Q1 2022).
- Continued to focus on efficient energy use and management of office materials in 2022.

Future Plans

- Orphazyme is no longer active in research and development activities associated with the use of hazardous substances and as such these potential risks have been minimized. Further, with only limited operating activities, employees and suppliers focusing on administration, finance and legal activities, the general potential impact on the environment and climate and the impact of the environment and climate on Orphazyme's business is viewed as minimal. We will continue to endeavor to protect the environment and climate through mindful business practices such as, e.g. careful use of office materials and energy consumption.

Social / Employees

Risk

- Very Limited: As of December 31, 2022, Orphazyme had only one employee. The company continues to value diversity in gender, age, ethnicity, nationality, religion, education, sexual orientation, work history, perspectives, opinions, and skills at all levels of our business however given its limited employees it currently does not have a diverse workforce. Further, the Company's limited operations, such as office space and support network, could impact the working conditions of remaining employees.

Actions

- The health and safety of our employees is of utmost importance and Orphazyme continually works to ensure that all systems and processes meet strict international standards.
- Continued to train laboratory employees in the systems, processes and workplace safety until closure of our laboratory space in Q1 2022.
- Continued to conduct regular mandatory Health and Safety surveys (APVs) assessing several aspects of the working environment such as psychological, ergonomics, and chemical working environment until employees transferred to KemPharm in June 2022.
- Continued to foster an open, trusting and inclusive workplace committed to freedom from discrimination, harassment, and bullying.
- We went through a significant business transformation in 2022 which resulted in some redundancies. We conducted our processes in accordance with applicable laws and regulations in the relevant jurisdictions and the processes were well planned with a clear focus on transparency and support.
- Continued to provide health insurance as standard for all our employees.

Policies in place

- Diversity Policy.
- Health and Safety Policies.

Results

- Assisted staff with IT and health support both in-office and at home working.
- Established a resilient culture centered on trust and collaboration.
- Due to the changes in the Company following the Sale of Assets Orphazyme ended the financial year December 31, 2022 with only one male employee, the CEO/CFO. Therefore, we believe diversity metrics for the period are not meaningful at this time.
- Leadership (director level and above): Not meaningful given company structure (One employee as of December 31,2022; male).

Future Plans

- As of the date of this annual report, the Company has limited ongoing business activities and only one employee. Our future social / employee activities will be commensurate with the size and limited operations of the Company. We will continue to strive to uphold our values and responsibilities and promote a healthy, diverse and inclusive workplace, as we execute our strategy.

Data Ethics

Orphazyme operates in a highly regulated industry and the importance of responsible data handling is appreciated and followed across our organization. We currently do not have a data ethics policy but we have a Global Data Privacy Policy. Given the Company has limited operational business activities, it is no longer an integrated part of the Company's business strategy or activities to process data or use algorithms for data

analysis in connection with clinical trials, etc. However, our practices will be evaluated on an ongoing basis in order to ensure they align with the statutory requirements set forth in Section 99d of the Danish Financial Statements Act.

Diversity in management cf. §99b

As of December 31, 2021, Orphazyme had 62 employees (FTEs), of which 58% were female and 42% were male. Of our employees at director level and above, 53% were female and 47% were male and, below director level, 63% of employees were female and 37% were male.

In March 2022, Orphazyme entered in-court restructuring proceedings, resulting in a reduction in its workforce of approximately 50%. In May 2022, the majority of the remaining Danish employees were transferred to KemPharm, following the Sale of Assets. As at December 31, 2022, there was only one employee, Anders Vadsholt CEO and CFO. Given the size of our workforce, it is not meaningful to set out diversity figures nor is it required in accordance with the Danish Companies Act.

The Board of Directors also saw some changes in 2022. At year-end 2021, the Board of Directors was comprised of five members, of which two (40%) were women. Due to the evolution of the business, there were only three Board members at the end of 2022, all of which were men. The company remains committed to promoting a diverse and inclusive workplace and has therefore set a target of increasing the representation of women on the Board of Directors to 40% by 2026 in accordance with the guidelines from the Danish Business Authority.

Board of Directors and Executive Management

Board of Directors

Bo Jesper Hansen, Chairman of the Board

- Member since: 2010 (Chairman, 2022)
- Born in: 1958
- Nationality: Danish
- Committees: Remuneration (Chair); Nomination (Chair); Audit

Special competencies: Dr Hansen has extensive experience in orphan drugs, both from the operations and supervisory point of view and has broad and current know-how of the biotechnology environment. He holds an MD and a PhD in Medicine from the University of Copenhagen.

Current positions: Deputy Chairman of SOBI AB, member of the Board of directors of Laborie Inc., Innoventa Medica ApS, and Reaplix A/S. Venture Partner at Wellington Partners; Advisory Consultant for Aescap 2.0, Nordic Capital, EQT AB and Broad Street Principal Investments Europe Ltd. & senior business advisor for HBM Ventures Ltd.

John Sommer Schmidt, Deputy Chairman

- Member since: 2022
- Born in: 1973
- Nationality: Danish
- Committees: Audit (Chair), Remuneration, Nomination

Special competencies: John is a partner at Gorrissen Federspiel Advokatpartnerselskab where he heads up the Restructuring & Insolvency practice group. He holds a Law degree from Aarhus University, a LL.M. from University of California, Los Angeles School of Law, and a B.Sc. (jur.) from Aarhus School of Business 1998. He was appointed lawyer by the Danish Bar and law Society in 2004.

Current positions: Partner at Gorrissen Federspiel Advokatpartnerselskab.

Anders Vadsholt

- Member since: 2022
- Born in: 1969
- Nationality: Danish
- Committees: Nomination

Special competencies: Anders has 25+ years' experience from biotech and corporate finance. Previously he was at Topotarget, BankInvest Biomedical Venture and Carnegie Investment Bank. Anders holds an MBA in Finance and Strategy from the University of Melbourne, and an MSc in Corporate Law and Economics from Copenhagen Business School.

Current positions: Anders is currently owner of Alpha Healthcare Investments ApS, through which he holds a position as Board member of Amplify Therapeutics ApS.

Executive Management

Anders Vadsholt, Chief Executive Officer, Chief Financial Officer

- Born in: 1969
- Nationality: Danish

Anders joined Orphazyme in May 2016 as Chief Financial Officer, and took the position of Chief Executive Officer, in addition to his role as Chief Financial Officer, March 1, 2022.

Corporate information

Commercial bankers

Danske Bank A/S

Holmens Kanal 2-12

DK-1092 Copenhagen K

Annual Report

This annual report will be available on www.orphazyme.com and printed copies are available upon request.

Annual General Meeting

Information about our Annual General Meeting can be found in the section for Investors & Media at www.orphazyme.com under *Events & Presentations* and *General Meetings*.

Financial Statements

2022 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 | 2022 | 2021 |
|--|----------|-----------------|------------------|
| Net revenue | 2.1 | - | - |
| Research and development expenses | 2.2, 2.3 | - | - |
| General and administrative expenses | 2.4 | (41,241) | (83,472) |
| Operating loss | | (41,241) | (83,472) |
| Financial income | 2.7 | 193 | 37 |
| Financial expenses | 2.7 | - | - |
| Loss before tax | | (41,048) | (83,436) |
| Income tax benefit | 2.8 | 2,736 | 4,941 |
| Loss from continuing operations | | (38,312) | (78,495) |
| Net result from discontinued operations | 1.7 | 64,382 | (548,044) |
| Net result for the year | | 26,070 | (626,539) |
| Items that will be reclassified subsequently to the Statement of Profit or Loss: | | | |
| Exchange difference from translation of foreign operations | | (905) | (302) |
| Other comprehensive Profit/loss | | (905) | (302) |
| Total comprehensive Profit/loss | | 25,165 | (626,841) |
| Weighted-average shares outstanding | | 35,268,725 | 34,924,702 |
| Profit/ loss per share from continuing operations, basic and diluted (DKK) | | (1.08) | (2.25) |
| Profit/ loss per share from discontinued operations, basic and diluted (DKK) | | 1.82 | (15.69) |
| Profit/ loss per share, basic and diluted (DKK) | 4.3 | 0.74 | (17.94) |

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 | 2022 | 2021 | |
| Non-current assets | | | | |
| Intangible assets | 3.1 | — | 2,152 | |
| Right-of-use assets | 3.2 | — | 5,434 | |
| Property, plant, and equipment | 3.3 | — | 2,985 | |
| Corporation tax receivable | 2.8 | — | 2,750 | |
| Prepayments and deposits | 3.4 | — | 964 | |
| Total non-currents assets | | — | 14,285 | |
| Current assets | | | | |
| Corporation tax receivable | 2.8 | 7,338 | 7,229 | |
| Trade receivables | 3.5 | 4,103 | 29,268 | |
| Prepayments and other receivables | 3.4 | 4,217 | 20,192 | |
| Inventory | 3.6 | — | — | |
| Cash | 3.8 | 42,464 | 102,255 | |
| Total current assets | | 58,122 | 158,944 | |
| Total assets | | 58,122 | 173,229 | |
| EQUITY AND LIABILITIES | | | | |
| | Note | 2022 | 2021 | |
| Equity | | | | |
| Share capital | 4.2 | 35,312 | 34,952 | |
| Share premium | | 2,087,436 | 2,082,486 | |
| Other reserves | | (493) | 2,899 | |
| Accumulated deficit | | (2,080,588) | (2,110,998) | |
| Total equity | | 41,667 | 9,339 | |
| Non-current liabilities | | | | |
| Borrowings | 3.7 | — | 2,482 | |
| Lease liabilities | 3.2 | — | 3,925 | |
| Discount and rebate liabilities | 3.7 | — | 28,293 | |
| Other non-current liabilities | 3.7 | 98 | 98 | |
| Total non-current liabilities | | 98 | 34,798 | |
| Current liabilities | | | | |
| Borrowings | 3.7 | — | 30,983 | |
| Lease liabilities | 3.2 | — | 2,578 | |
| Trade payables and accruals | 3.7 | 10,540 | 57,524 | |
| Corporation tax payables | 2.8 | 284 | 584 | |
| Discount and rebate liabilities | 3.7 | 4,457 | 7,900 | |
| Other liabilities | 3.7 | 1,075 | 29,523 | |
| Total current liabilities | | 16,357 | 129,092 | |
| Total equity and liabilities | | 58,122 | 173,229 | |

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

CONSOLIDATED STATEMENTS OF CHANGES IN SHAREHOLDERS' EQUITY

| DKK 000 | | | Other reserves | | | | |
|--|------|---------------|------------------|--------------------------------------|---|---------------------|------------------|
| | Note | Share capital | Share premium | Foreign currency translation reserve | Share-based compensation – acquisition of intangible assets | Accumulated deficit | Total |
| Balance as of December 31, 2020 | | 34,698 | 2,082,254 | 714 | 5,780 | (1,502,921) | 620,525 |
| 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 | | 34,952 | 2,082,486 | 412 | 2,487 | (2,110,998) | 9,339 |
| Net loss for the year | | – | – | – | – | 26,070 | 26,070 |
| Other comprehensive income | | – | – | (905) | – | – | (905) |
| Total other comprehensive income (loss) | | – | – | (905) | – | 26,070 | 25,165 |
| Transactions with owners: | | | | | | | |
| Capital increase | 4.2 | 360 | 5,091 | – | – | – | 5,451 |
| Transaction cost related to capital increase | 4.2 | – | (141) | – | – | – | (141) |
| Reserve for bonus shares reversed | 4.2 | – | – | – | (2,487) | 2,487 | – |
| Share-based compensation expense | 2.6 | – | – | – | – | 1,853 | 1,853 |
| Total transactions with owners | | 360 | 4,950 | – | (2,487) | 4,340 | 7,163 |
| Balance as of December 31, 2022 | | 35,312 | 2,087,436 | (493) | – | (2,080,588) | 41,667 |

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 | 2022 | 2021 |
|---|----------|------------------|------------------|
| Operating result from continuing operations | | (41,241) | (83,472) |
| Operating result from discontinued operations | | (78,309) | (549,831) |
| Adjustments to reconcile operating result to cash flows from operating activities: | | | |
| Equity-settled share-based compensation expense | 2.6 | 1,773 | 16,019 |
| Depreciation and amortization | 2.2,2.4 | 1,820 | 18,111 |
| Change in prepayments, deposits, and other receivables | 3.4, 3.5 | 42,103 | 2,826 |
| Change in trade payables, accruals, and other liabilities | 3.7 | (48,652) | (3,758) |
| Corporation taxes received | | 5,500 | 5,500 |
| Corporation taxes paid | | — | (1,738) |
| Interest received | | 193 | 37 |
| Interest paid | | (1,132) | (6,263) |
| Cash flow from operating activities | | (117,945) | (602,571) |
| Investing activities | | | |
| Purchase of intangible assets | 3.1 | — | (902) |
| Purchase of property, plant, and equipment | 3.3 | — | (92) |
| Proceeds from sale of property, plant and equipment | | 1,460 | 1,040 |
| Proceeds from sale of activity | 1.7 | 88,887 | — |
| Cash flow from investing activities | | 90,347 | 46 |
| Financing activities | | | |
| Repayment of borrowings | | (39,155) | (25,657) |
| Repayment of lease obligations | 3.2 | (1,233) | (3,503) |
| Proceeds from issuance of shares | | 5,451 | 464 |
| Transaction cost related to issuance of shares | | (141) | — |
| Cash settlement of Bonus Shares | | — | (1,648) |
| Cash flow from financing activities | | (35,078) | (30,344) |
| Net change in cash and cash equivalents | | (62,676) | (632,869) |
| Effects of changes in exchange rates | | (2,885) | 8,195 |
| Cash at the beginning of the year | | 102,255 | 726,929 |
| Cash at the end of the year | | 42,464 | 102,255 |

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

Notes to Financial Statements

SECTION 1 Basis of preparation and significant accounting policies

1.1 CORPORATE INFORMATION

Orphazyme A/S (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 initiated voluntary delisting of the ADSs representing its shares from 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").

Orphazyme was previously involved in the research and development of novel therapeutics for the treatment of neurodegenerative rare diseases, including Niemann-Pick disease type C, or NPC. 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, arimocloamol, 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. The disposed assets and business activities are presented as discontinued operations.

KemPharm Inc. and associated entities, including KemPharm Denmark A/S, changed their name in February 2023 to Zevra Inc. and Zevra A/S respectively. For the purposes of this Annual Report the name of the Company at the time of the Sale of Assets in March 2022, KemPharm Denmark A/S (KemPharm) is being used.

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

These consolidated financial statements were approved and authorized for issuance by the Board of Directors on April 25, 2023.

1.2 BASIS OF PREPARATION

The consolidated financial statements of the Group and parent company financial statements have been prepared in accordance with International Financial Reporting Standards (IFRS) as adopted by the EU and additional requirements of the Danish Financial Statements Act.

The consolidated financial statements and parent company 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 result 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 and sale of activity.

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

Assets held for sale and discontinued operations

The criteria for held for sale classification is regarded as met only when the sale is highly probable, and the asset or disposal group is available for immediate sale in its present condition. Actions required to complete the sale should indicate that it is unlikely that significant changes to the sale will be made or that the decision to sell will be withdrawn. Management must be committed to the plan to sell the asset and the sale expected to be completed within one year from the date of the classification.

Property, plant and equipment and intangible assets are not depreciated or amortised once classified as held for sale.

Assets and liabilities classified as held for sale are presented separately as current items in the statement of financial position.

Discontinued operations are excluded from the results of continuing operations and are presented as a single amount as profit or loss after tax from discontinued operations in the statement of profit or loss.

Additional disclosures are provided in Note 1.7. All other notes to the financial statements include amounts for continuing operations, unless indicated otherwise.

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, 2022 and 2021 the Danish entity generated revenue which is disclosed in a separate note. For the years ended December 31, 2022 and 2021 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 inputs and assumptions used in share-based compensation valuation models (Note 2.6)
- 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)
- Judgement regarding management's assessment of the outfall of the class action lawsuit (Note 3.9)
- Estimate of rebates granted to co-financing healthcare authorities (Note 2.1)
- Judgement regarding management's accounting for discontinued operations (Note 1.7)

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

New accounting policies and disclosures

The Group has implemented the standards and amendments that are effective for the financial year 2022. The new standards and amendments have not affected the Group's recognition or measurement for 2022, nor are they expected to have significant future impact.

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, 2023 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

No significant events after the reporting period to disclose, besides that already disclosed in note 3.9 to the consolidated Financial Statements on the class action lawsuit.

1.7 DISCONTINUED OPERATIONS

In May 2022, Orphazyme 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.

The Sale of Assets agreement 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). Under the terms of the agreement, KemPharm agreed to also retain a majority of Orphazyme's employees.

The business operations and activities that were part of the Sale of Assets agreement with KemPharm have been reclassified as discontinued operations. The net result related to the above-mentioned discontinued operations are therefore presented separately in the income statement and the statement of cash flows. The comparative figures have been restated accordingly.

Judgement regarding management's accounting for discontinued operations

Significant judgment is required to determine the presentation of discontinued operations in profit and loss and cash flow, for both the current and prior year. Management has done the judgement based on the Purchase agreement from the sale of assets to KemPharm.

Substantially all of Orphazyme's assets and business activities was sold to KemPharm (the "Sale of Assets") on an "as-is" basis and there have not been given any substantial representations or warranties in favor of KemPharm. While the management are not aware of any outstanding matters that would reasonably form a basis for a claim related to the Sale of Assets, if the Group become subject to liability based upon our contractual obligations to KemPharm or otherwise, it could have a material adverse effect on our financial position.

Net result and net cash flow from discontinued operations are specified below:

For the years ended December 31,

| DKK 000 | Note | 2022 | 2021 |
|--|-------------|-----------------|------------------|
| Net revenue | 2.1 | 17,867 | 36,193 |
| Research and development expenses | 2.2, 2.3 | (36,660) | (329,980) |
| General and administrative expenses | 2.4 | (59,516) | (256,044) |
| Operating loss | | (78,309) | (549,831) |
| Financial income | 2.7 | 6,354 | 12,395 |
| Financial expenses | 2.7 | (9,184) | (10,608) |
| Loss before tax | | (81,139) | (548,044) |
| Income tax benefit | 2.8 | — | — |
| Net result for the period | | (76,682) | (548,044) |
| Gain from disposal of discontinued operations | | 145,520 | — |
| Net result from discontinued operations | | (64,382) | (548,044) |

| DKK 000 | Note | 2022 | 2021 |
|---|-------------|---------------|------------------|
| Cash flow from operating activities | | (17,097) | (520,333) |
| Cash flow from investing activities | | 90,347 | 46 |
| Cash flow from financing activities | | (40,388) | (29,160) |
| Net cash flow from discontinued operations | | 32,862 | (549,447) |

Gain on disposal/carrying amount of disposed assets

| DKK 000 | Note | 2022 | 2021 |
|--|------|-----------------|------|
| Carrying values of intangible assets | | 1,603 | — |
| Carrying values of property, plan and equipment | | 134 | — |
| Carrying values of liabilities | | (58,370) | — |
| Carrying values of assets and liabilities | | (56,633) | — |
| Gain on disposal – net | | 145,520 | — |
| Gain on disposal | | 145,520 | — |
| Net cash inflow from disposal of business | | 88,887 | — |

As the Company has sold substantially all assets and business activities, additional information is provided for revenue (note 2.1), research and development expenses (note 2.2), government grants (note 2.3), general and administration expenses (note 2.4), employee costs (note 2.5), share-based compensation costs (note 2.6) and financial income and financial expenses (note 2.7), to which references are made.

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.

The revenue recognized reflects the various types of price reductions. Products sold in France are covered by government programs under which products are sold at a discount. Rebates are granted to co-financing the healthcare authorities. Rebates and discounts as described, are recognized in the period in which the underlying sales are recognized as a reduction of gross sale.

The amount of rebates granted to co-financing the healthcare authorities are estimated by management on the basis of the specific terms of the relevant regulations or agreements, and accrued as each of the underlying sale transactions is recognized. The estimated amounts described above are recognized in the income statement within Net revenue as a reduction of gross sales, and within Rebate and discount liability in the balance sheet 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.

As of June 2022, the remunerated early access compassionate use program (“nATU”) in France, was transferred to KemPharm as part of the sale of substantially all of the Company’s assets and business activities. All revenue is therefore presented as discontinued operations (Note 1.7).

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

| DKK 000 | 2022 | 2021 |
|----------------------------|--------|--------|
| Revenue by type | | |
| Revenue from sale of goods | 17,867 | 36,193 |
| Revenue by partner | | |
| Clinigen Health Limited | 17,867 | 36,193 |
| Geographical areas | | |
| France | 17,867 | 36,193 |

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 Group'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 | 2022 | 2021 |
|---|----------------------|-----------------------|
| External costs | 18,385 | 229,942 |
| Employee costs (Note 2.5) | 17,926 | 86,329 |
| Depreciation, amortization and impairment (Notes 3.1, 3.2, 3.3) | 348 | 13,709 |
| Total research and development expenses | <u>36,660</u> | <u>329,980</u> |

External costs comprise mainly expenses related to third party vendors providing services related to our research and development activities and facility costs. Research and development expenses include costs relating to products sold under the French early access compassionate use program.

As of June 2022, all research and development activity were transferred to KemPharm as part of the sale of substantially all of the Company's assets and business activities. All research and development expenses are therefore presented as discontinued operations (Note 1.7).

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 2022, Orphazyme has received grant and other funding of DKK 0.0 million (2021: DKK 1.4 million).

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.

As of June 2022, substantially all of the Company's assets and business activities was transferred to KemPharm as part of the sale. Governments grants are therefore presented as discontinued operations (Note 1.7).

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 | 2022 | 2021 |
|--|-----------------------|-----------------------|
| External costs | 70,737 | 192,913 |
| Employee costs (Note 2.5) | 28,548 | 142,201 |
| Depreciation (Notes 3.2 and 3.3) | 1,471 | 4,402 |
| Total general and administrative expenses | <u>100,756</u> | <u>339,516</u> |

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 2022 costs as well comprise expenses related to restructuring activities, i.e. employee redundancies and termination of onerous contracts.

As of June 2022, general and administrative activity was partly transferred to KemPharm as part of the sale of substantially all of the Company's assets and business activities. General and administrative expenses are therefore presented as both continued operations, DKK 41.2 million (2021: DKK 83.5 million) and discontinued operations (Note 1.7), DKK 59.5 million (2021: DKK 256.0 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, 2022, 2021. Refer to note 4.5 for more discussion on remuneration of Board of Directors and Executive Management.

| DKK 000 | 2022 | 2021 |
|---|---------------|----------------|
| Employee costs | | |
| Salaries | 28,185 | 167,316 |
| Cash bonus | 9,046 | 16,663 |
| Share-based compensation (Note 2.6) | 1,773 | 15,576 |
| Pension | 2,184 | 12,963 |
| Other social security contributions | 402 | 4,113 |
| Other staff costs | 3,004 | 8,066 |
| Total employee costs excluding board remuneration | 44,594 | 224,696 |
| Board remuneration (Note 4.5) | 2,211 | 3,391 |
| Board share-based compensation (Note 2.6 and Note 4.5) | (331) | 443 |
| Total employee costs | 46,474 | 228,530 |
| <i>Recognized as follows in the statement of Profit or Loss</i> | | |
| Research and development expenses | 17,926 | 86,329 |
| General and administrative expenses | 28,548 | 142,201 |
| Total employee costs | 46,474 | 228,530 |
| Average number of full-time employees | 21 | 130 |
| Year-end number of full-time employees | 1 | 62 |

As of June 2022, substantially all of the Company's assets and business activities were transferred to KemPharm as part of the sale. Employee costs are therefore presented as both continued operations, DKK 10.5 million (2021: DKK 24.1 million) and discontinued operations (Note 1.7), DKK 36.0 million (2021: DKK 204.4 million).

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 Black-Scholes or Monte-Carlo model depending on the terms and conditions of the respective award. 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

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").

Under the post-IPO long-term incentive program (2017 LTIP), the Executive Management as well as certain Key Employees of Orphazyme had subscribed to 14,875 ordinary shares (Investment Shares). In 2018, a Key Employee subscribed to 4,300 Investment Shares.

The participants in the 2017 LTIP had the opportunity to 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. 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. 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 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, respectively, and vesting among other things also being subject to the participants having maintained ownership of their Investment Shares and continued employment. 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.

No long-term share-based incentive program was announced in 2022 because of the restructuring of the Company and subsequent sale of substantially all of its assets and business activities. 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 Oct 2021 (modified) | 2021 PSU Oct 2021 (modified) | 2021 RSU Apr 2021 (original) | 2021 PSU Apr 2021 (original) |
|--|---|---|---|---|
| Grant date | | | | |
| 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 |

Matching Shares under all of the LTIP programs were fully vested as of January 1, 2021.

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 :

| | 2022 | 2021 |
|------------------|------|------|
| Program | | |
| 2020 LTIP | 1.0 | 2.0 |
| 2019 LTIP | 0.7 | 1.7 |

The exercise price for each LTIP award outstanding as of December 31, 2022 was DKK 1 (2021: 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, 2020 | 52,865 | 118,686 | 172,488 | 172,488 |
| Granted | — | — | — | — |
| Exercised | (52,865) | (118,686) | (172,488) | — |
| Expired | — | — | — | — |
| Forfeited | — | — | — | — |
| Outstanding at December 31, 2021 | — | — | — | — |
| Granted | — | — | — | — |
| Exercised | — | — | — | — |
| Expired | — | — | — | — |
| Forfeited | — | — | — | — |
| Outstanding at December 31, 2022 | — | — | — | — |

For the year ended December 31, 2022, DKK 1.9 million (2021: DKK 17.1 million) was recognized as compensation expense related to the LTIP awards. Of the total expense, DKK 1.4 million (2021: DKK 6.8 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. Leavers are entitled to keep already granted phantom shares and will receive a pro-rata grant compared to the time of employment in the applicable granting year.

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, 2022 and 2021, the programs are expected to consist of up to a total of 41,351 phantom shares.

As of December 31, 2022, 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, 2022, an aggregate amount of DKK 0.1 million (2021: DKK 0.3 million) was recognized as compensation income 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. The expected volatility is determined based on the Company's own historical volatility over a period corresponding to the remaining lifetime of the option.

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, 2022 | | | December 31, 2021 | | |
|--|-------------------|--------------|--------------|-------------------|--------------|--------------|
| | 2020 Program | 2019 Program | 2018 Program | 2020 Program | 2019 Program | 2018 Program |
| Fair value at valuation date (DKK 000) | — | — | — | 40 | 48 | 12 |
| Dividend yield (%) | — | — | — | — | — | — |
| Expected volatility (%) | 86.6% | 105.9% | 86.6% | 77.4% | 101.1% | 101.1% |
| Risk-free interest rate (%) | (2.70%) | (2.56%) | (2.52%) | (0.48%) | (0.60%) | (0.60%) |
| Expected life of awards (years) | 2.08 | 1.08 | 0.08 | 3.08 | 2.08 | 1.08 |
| Weighted average share price (DKK) | 17.16 | 17.16 | 17.16 | 17.16 | 17.16 | 17.16 |

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 certain board members exercised their RSUs. As these RSUs were not cash-settled, the corresponding liability of DKK 35 thousand 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 thousand.

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. 13,525 RSUs lapsed in 2021 in connection with the exercise of grants under the new 2020-2 RSU program, while 1,652 RSUs expired in 2022. Neither the lapses in 2021 nor the expiry in 2022 had any impact on the Statement of Profit or Loss as Management had assessed that none of the options under the 2020 RSU program would ultimately vest as the grants under the 2020-2 RSU program were more beneficial to exercise for the participants.

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). The RSUs fully vested on the date of the general meeting in 2021. All RSUs except for 1,927 were exercised in 2021. The remaining 1,927 RSUs expired in March 2022 resulting a positive impact on the Statement of Profit or Loss of DKK 31 thousand.

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. 10,211 and RSUs 8,168 RSUs lapsed during 2022 and 2021, respectively, as the participants resigned from the Board of Directors forfeiting the vesting condition resulting a positive impact on the Statement of Profit or Loss of DKK 180 thousand and 96 thousand, respectively.

No RSU program was announced in 2022 because of the restructuring of the Company and subsequent sale of substantially all of its assets and business activities.

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 income in the amount of DKK 0.3 million (2021: 0.7 million) and a corresponding short-term liability as of December 31, 2021. The Exercise Period for all 2021 RSUs is one year following full vesting and for valuation purposes we have assumed exercise three months upon full vesting.

As of December 31, 2022, 1,927 RSUs were expired, and 12,071 RSUs were eligible for exercise.

The following table presents the inputs to the Black-Scholes model used to estimate the fair value of the 2021 RSUs at year-end, as they are classified as cash-settled:

| Program | December 31, 2022 2021 RSUs | December 31, 2021 2021 RSUs |
|--|--------------------------------|--------------------------------|
| Fair value at valuation date (DKK 000) | 0.2 | 16 |
| Dividend yield (%) | — | — |
| Expected volatility (%) | 152.3% | 146.9% |
| Risk-free interest rate (%) | (2.52%) | (0.63%) |
| Expected life of awards (years) | 0,00 | 0.75 |
| Weighted average share price (DKK) | 17.16 | 17.16 |

d) Sign-on bonus shares to previous 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 the share price increased to DKK 75 per share, (ii) 12,000 shares provided that the share price increased to DKK 100 per share, and (iii) 40,000 shares provided that the 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 expense was recognized in 2020 when the target price was achieved.

e) Sign-on bonus shares to former CEO

As part of the CEO service agreement made in 2021 with the former CEO, Christophe Bourdon was granted 34,941 RSUs in connection with the on-boarding, which had 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 was 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 would be prorated and calculated through the date of release of the Participant's work obligations. The vested RSUs could only be exercised after the expiration of the total vesting period. 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 was 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 was 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 was classified as general and administrative expenses recognised in October 2021.

Summary of share-based compensation

The following amounts were recognized as share-based compensation for the years ended December 31. All share-based compensation costs are presented as discontinued operations (Note 1.7).

| DKK 000 | 2022 | 2021 |
|--|--------------|---------------|
| Share-based compensation included in R&D | — | 3,879 |
| Share-based compensation included in G&A | 1,773 | 12,140 |
| Total share-based compensation expense recognized | 1,773 | 16,019 |

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 | 2022 | 2021 |
|--|--------------|---------------|
| Interest income on cash balances | 193 | 37 |
| Foreign currency exchange gains | 6,354 | 11,849 |
| Gain on embedded call option (Note 3.7) | — | 546 |
| Total financial income | 6,547 | 12,432 |
| Interest expense on Loan Agreement (Note 3.7) | 5,959 | 7,350 |
| Write-off of transaction costs for Loan Agreement tranche 2 (Note 3.7) | — | — |
| Loss on embedded call option (Note 3.7) | — | — |
| Interest expense on lease liabilities (Note 3.2) | 147 | 624 |
| Loss on lease modification (Note 3.2) | — | (365) |
| Interest expense on cash balances | 158 | 1,484 |
| Foreign currency exchange loss | 2,843 | 1,369 |
| Bank fees and other charges | 77 | 147 |
| Total financial expenses | 9,185 | 10,609 |

As of June 2022, the Group transferred substantially all of the Company's assets and business activities to KemPharm. Financial income and financial expenses are therefore presented as both continued operations, net income DKK 0.2 million (2021: DKK 0.0 million) and discontinued operations (Note 1.7), net expense DKK 2.8 million (2021: DKK 1.8 million).

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, 2022 and 2021, 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 in lack of significant activity in the Group, 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 8.9 million of taxable income plus 60% of taxable income above DKK 8.9 million.

For the years ended December 31, 2022 and 2021, the Company has unrecognized net tax loss carry-forwards in the Danish entity in the amount of DKK 2,186 million, DKK 1,454 million, and DKK 877 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 | 2022 | 2021 |
|---|--------------|--------------|
| Current tax benefit on net result | (5,133) | 144,379 |
| Adjustments prior years | (14) | (385) |
| Tax credit research and development expenses | 2,750 | 5,500 |
| Change in unrecognized deferred tax before tax credit | (27,107) | (155,700) |
| Permanent differences | 32,240 | 11,147 |
| Total income tax benefit for the year | 2,736 | 4,941 |

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

| DKK 000 | 2022 | 2021 |
|--|----------------|----------------|
| Net result before tax from continuing operations | (41,048) | (83,436) |
| Net result before tax from discontinued operations | 64,381 | (548,044) |
| Corporate income tax rate in Denmark | 22% | 22% |
| Computed income tax benefit | (5,133) | 138,926 |
| <i>Tax effect of:</i> | | |
| Adjustments prior years | (14) | (385) |
| Other non-deductible expenses, including US listing-related costs and share-based compensation | 32,240 | 11,147 |
| Effect of different tax rate | (18) | 892 |
| Deferred tax asset not recognized after tax credit | (24,338) | (145,639) |
| Total income tax benefit for the year | 2,736 | 4,941 |

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

| DKK 000 | 2022 | 2021 |
|--|-----------------|-----------------|
| Tax deductible losses | 482,595 | 319,811 |
| Deferred tax on intangible assets | - | 132,310 |
| Other temporary differences | 4,419 | 6,069 |
| | <u>487,015</u> | <u>458,190</u> |
| Deferred tax asset not recognized | 487,015 | 458,190 |
| Carrying amount included in the Statement of Financial Position | <u><u>-</u></u> | <u><u>-</u></u> |

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.

As of June 2022 the all software and licenses was transferred to KemPharm as part of the sale of substantially all of the Company's assets and business activities.

Impairment losses recognized in 2021 corresponding to the, at the time, remaining carrying amount for the license agreement with KLSDC and UCL (DKK 7.6 million), license agreement with the university of Miami (DKK 0.5 million) and software related to research and development (DKK 2,7 million).

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, 2020 | 2,736 | 12,083 | 14,819 |
| Additions | 902 | — | 902 |
| Cost at December 31, 2021 | 3,638 | 12,083 | 15,721 |
| Disposals | (3,638) | (12,083) | (15,721) |
| Cost at December 31, 2022 | — | — | — |
| 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 |
| Amortization expense | 514 | — | 514 |
| Disposal accumulated amortization | (3,603) | (10,480) | (14,083) |
| Accumulated amortization at December 31, 2022 | — | — | — |
| Net carrying value at | | | |
| December 31, 2021 | 549 | 1,603 | 2,152 |
| December 31, 2022 | — | — | — |

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.

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.

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, within discontinued operations.

As of June 2022 the lease contracts were transferred to KemPharm as part of the sale of substantially all of the Company's assets and business 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, 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 |
| Disposals | (3,910) | — | (3,910) |
| Depreciation expense | (1,524) | — | (1,524) |
| At December 31, 2022 | — | — | — |

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

| DKK 000 | 2022 | 2021 |
|---------------------------|-------------|--------------|
| At January 1 | 6,503 | 13,534 |
| Additions | — | — |
| Accretion of interest | — | 624 |
| Disposals | (6,503) | (1,212) |
| Payments | — | (4,127) |
| Exchange rate adjustments | — | 180 |
| Modifications | — | (2,496) |
| At December 31 | — | 6,503 |
| Current | — | 2,578 |
| Non-current | — | 3,925 |

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 | 2022 | 2021 |
|--|--------------|--------------|
| Depreciation and impairment expense of right-of-use assets (R&D) | 1,236 | 5,386 |
| Depreciation and impairment expense of right-of-use assets (G&A) | 288 | 844 |
| Interest expense on lease liabilities | 127 | 624 |
| Gain on lease modification and disposals | — | (365) |
| Total amount recognized in the Statement of Profit or Loss within discontinued operations | 1,651 | 6,489 |

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.

Gross carrying amount of any fully depreciated property, plant and equipment that is still in use is DKK 0.0 million.

As of June 2022 the all property, plant and equipment was transferred to KemPharm as part of the sale of substantially all of the Company's assets and business activities.

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

| DKK 000 | Furniture and equipment | Lease improvements | Total |
|--|--|-------------------------------|--------------|
| 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 |
| Additions | — | — | — |
| Disposals | (6,823) | (2,635) | (9,458) |
| Cost at December 31, 2022 | — | — | — |
| Accumulated depreciation at December 31, 2020 | 4,609 | 667 | 5,276 |
| Depreciation expense | 1,156 | 429 | 1,585 |
| Disposals | (389) | — | (389) |
| Exchange rate adjustments | 1 | — | 1 |
| Accumulated depreciation at December 31, 2021 | 5,377 | 1,096 | 6,473 |
| Depreciation expense | 342 | 122 | 464 |
| Disposals | (5,719) | (1,218) | (6,937) |
| Exchange rate adjustments | — | — | — |
| Accumulated depreciation at December 31, 2022 | — | — | — |
| Net carrying value at | | | |
| December 31, 2021 | 1,446 | 1,539 | 2,985 |
| December 31, 2022 | — | — | — |

There has been no impairment of property, plant and equipment for the years ended December 31, 2022 and 2021. Depreciation expense is included within discontinued operations as follows:

| DKK 000 | 2022 | 2021 |
|---|-------------|--------------|
| Research and development expenses | 357 | 1,301 |
| General and administrative expenses | 108 | 285 |
| Total depreciation expense included within discontinued operations | 464 | 1,585 |

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.

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

| DKK 000 | 2022 | 2021 |
|---|-------------|-------------|
| Deposits with vendors | — | 215 |
| Prepayments to vendors | — | 749 |
| Total non-current prepayments and deposits | — | 964 |

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 | 2022 | 2021 |
|--|--------------|---------------|
| Prepayments to vendors | 1,802 | 12,872 |
| VAT receivable, net | 662 | 2,903 |
| Foreign VAT receivable | 1,275 | 1,627 |
| Other current receivables | 479 | 2,790 |
| Total current prepayments and other receivables | 4,217 | 20,192 |

Current prepayments to vendors include prepayments made to CROs for clinical trial costs of DKK 1.7 million (2021: DKK 2.4 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, 2022 trade receivables in the amount of DKK 4.1 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, due to all receivables at December 31, 2022 have been received at the reporting date.

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.

June 2022 all pre-launch inventory was transferred to KemPharm as part of the sale of substantially all of the Company's assets and business activities.

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

Financial assets 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. The Group did not incur any credit losses on financial assets for either of the years ended December 31, 2022 or 2021.

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.

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, 2022 or 2021.

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

| DKK 000 | 2022 | 2021 |
|---|---------------|---------------|
| Borrowings | — | 33,465 |
| Lease liabilities (Note 3.2) | — | 6,503 |
| Trade payables | 4,362 | 41,780 |
| Accruals | 6,177 | 15,743 |
| Total liabilities measured at amortized cost | 10,539 | 97,491 |

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%.

As of June 2022 the Kreos debt facility was paid off as part of the sale of substantially all of the Company's assets and business activities to KemPharm. The following describes the terms etc. of the repaid debt facility.

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 2022 | Dec 2021 |
|---------------------------------|----------|----------|
| Fair value of call option | — | 326 |
| Dividend yield (%) | — | — |
| Expected volatility (%) | — | 147% |
| Risk-free interest rate (%) | — | (0.63)% |
| Expected life (years) | — | 1.2 |
| Share price (DKK) | — | 17.2 |

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, 2022, the Company recognized a gain of DKK 0.0 million (2021: DKK 0.5 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 | 10,540 | — | — | 10,540 | 10,540 |
| Total non-derivatives | 10,540 | — | — | 10,540 | 10,540 |
| Total derivatives | — | — | — | — | — |

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

| DKK 000 | December 31, 2021 | Cash flows | Non-cash changes | | | | December 31, 2022 |
|--|-------------------|-----------------|------------------|-------------------------------|----------------------|---------------------------|-------------------|
| | | | Disposals | Adjustments and modifications | Accumulated interest | Exchange rate adjustments | |
| Borrowings | 33,465 | (39,155) | — | — | 5,959 | (269) | — |
| Lease liabilities | 6,503 | (1,233) | (5,417) | — | 147 | — | — |
| Total liabilities from financing activities | 39,968 | (40,388) | (5,417) | — | 6,106 | (269) | — |

| DKK 000 | December 31, 2020 | Cash flows | Non-cash changes | | | | December 31, 2021 |
|--|-------------------|-----------------|------------------|-------------------------------|----------------------|---------------------------|-------------------|
| | | | Additions | 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 |

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.

December 31, 2022 accrued discount and rebates relates to rebates granted to co-financing healthcare authorities for 2021 and 2022.

December 31, 2021 accrued discount and rebates relates to the clawback in accordance with nATU program. June 2022 the clawback liability was transferred to KemPharm as part of the sale of substantially all of the Company's assets and business activities.

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

| DKK 000 | December 31, 2021 | Cash flows | Accruals | December 31, 2022 |
|--|------------------------------|-----------------------|-----------------|------------------------------|
| Discount and rebate liabilities | 36,193 | — | (31,736) | 4,457 |
| Total liabilities from accrued discount and rebates | 36,193 | — | (31,736) | 4,457 |

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

| DKK 000 | 2022 | 2021 |
|--|--------------|---------------|
| Remuneration to the Board of Directors | 213 | 293 |
| Payroll and employee-related costs | 862 | 29,230 |
| Total current other liabilities | 1,075 | 29,523 |

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

| DKK 000 | 2022 | 2021 |
|--|-------------|-------------|
| Phantom shares liability to employees | 98 | 98 |
| Total non-current other liabilities | 98 | 98 |

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 | 2022 | 2021 |
|-------------------|---------------|----------------|
| DKK | 14,987 | 53,291 |
| USD | 13,962 | 43,340 |
| EUR | 13,209 | 4,070 |
| CHF | — | 662 |
| GBP | 306 | 892 |
| Total cash | 42,464 | 102,255 |

3.9 COMMITMENTS AND CONTINGENCIES

Class Action Lawsuit

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 laws. While Management does not believe the class action lawsuit claims have any merit, and therefore the Company has not recorded any provisions related to the class action lawsuit as of December 31, 2022, the Company decided, for cost control reasons and because of the risks inherent in any litigation, to engage in settlement discussions. In April 2023, the Parties reached an agreement in principle to settle this action in its entirety. The parties are now undertaking to prepare mutually agreeable settlement documents, and the Court has stayed all

proceedings in the action pending the filing of the parties' settlement documents. The parties currently anticipate that a motion seeking the Court's preliminary approval of the settlement will be filed in May 2023 and that, if the Court grants preliminary approval, final court approval will be sought after members of the proposed settlement class have been afforded an opportunity to object to or opt out of the settlement.

As the legal process is ongoing and no settlement has yet been concluded or approved by the Court, material uncertainty remains with regards to the final outcome. The Company has not recorded any provisions related to the class action lawsuit as of December 31, 2022. Moreover, the Company assesses the criteria of IAS 37.92 as met because it believes that premature disclosure of additional information regarding the parties' agreement in principle would be detrimental to the Company's ability to conclude and obtain final approval of the settlement. Whether or not the parties' proposed settlement is ultimately concluded and approved by the Court, the Company believes it will be able to continue its operations until at least December 31, 2023. The Company expects to end 2023 with DKK 6 – 10 million in cash.

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, 2022, the Group held cash totaling DKK 42.5 million (2021: DKK 102.3 million). With reference to note 3.9 management therefore considers it appropriate to prepare these financial statements on a going concern basis.

For the full-year 2023 the Group anticipates an operating loss in the range of DKK 30 – 35 million. It expects to end 2023 with DKK 6 - 10 million in cash. There are inherent risks and uncertainties in our Outlook for 2023 given the limited nature of the business activities, the class action lawsuit in the United States and the future prospects. Whether or not the parties' proposed settlement is ultimately concluded and approved by the Court, the Company believes it will be able to continue its operations until at least December 31, 2023.

4.2 EQUITY

The following table summarizes the Company's share activity:

| | Ordinary shares |
|--|------------------------|
| 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 |
| Capital increase, U.S. At-the-Market Offering Program | 360,000 |
| December 31, 2022 | 35,312,241 |

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 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.

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)

In February 2022, the Company completed a capital increase of 360,000 shares as a result of the utilization of the U.S. At-the-Market Offering Program.

As a result of the above transactions, the total nominal share capital of the Company as of December 31, 2022 was DKK 35,312,241, divided into 35,312,241 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, 2022 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, 2022 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.

Following the license agreement with Kansas Life Sciences Development Inc. and UCL Business PLC was transferred to KemPharm, as part of the sale of substantially all of the Company's assets and business activities to KemPharm, the reserve for Share-based compensation- acquisition of intangible assets have been reversed in 2022.

4.3 PROFIT/LOSS PER SHARE

Basic profit/loss per share for the year is calculated by dividing the net result for the year by the weighted average number of ordinary shares outstanding during the year. The diluted profit/loss per share is calculated by dividing the net result 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. 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.

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:

| | 2022 | 2021 |
|--|---------------|----------------|
| Net result for the year from continuing operations (DKK 000) | (38,312) | (78,495) |
| Weighted-average shares outstanding | 35,268,725 | 34,924,702 |
| Profit/loss per share from continuing operations | (1.08) | (2.25) |
| Net result for the year from discontinued operations (DKK 000) | 64,382 | (548,044) |
| Weighted-average shares outstanding | 35,268,725 | 34,924,702 |
| Profit/loss per share from discontinued operations | 1.83 | (15.69) |
| Profit/ loss per share, basic and diluted (DKK) | 0.74 | (17.94) |

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, 2022, 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, 2022, 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.

For the full-year 2023 the Group anticipate an operating loss in the range of DKK 30 – 35 million. It expects to end 2023 with DKK 6 - 10 million in cash. There are inherent risks and uncertainties in our Outlook for 2023 given the limited nature of the business activities, the class action lawsuit in the United States and the future prospects. For further information please refer to note 3.9 to the consolidated Financial Statements. Whether or not the parties' proposed settlement is ultimately concluded and approved by the Court, the Company believes it will be able to continue its operations until at least December 31, 2023. The Company expects to end 2023 with DKK 6 – 10 million in cash.

Foreign Currency Risk

The Group's foreign currency risk is assessed to be low. 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 is only of the DKK against the USD exposure for the Group to currency gains or losses that will impact the reported amounts of assets, liabilities, income and expenses.

Interest Rate Risk

The Group's interest rate risk is assessed to be low. The Group has no borrowing of December 31, 2022

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 in one Danish bank 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 USD exchange rates against the DKK and the impact for the possible fluctuations in the interest rate on bank deposits in Denmark. The methods and assumptions used are consistent with prior year and consider increases and decreases in the Group's 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, 2022 would have been approximately DKK 0 thousand (2021: DKK 14 thousand).

The currency fluctuations would have fluctuated by +/- 10%, the impact on the Group's net loss for the year ended December 31, 2022 would have been approximately DKK 2,938 thousand (2021: DKK 19,239 thousand):.

4.5 REMUNERATION OF BOARD OF DIRECTORS AND EXECUTIVE MANAGEMENT

Executive Management consists of the Company's Chief Executive Officer, also the registered management of the Company. In February 2022, Orphazyme announced that the Board of Directors appointed Chief Financial Officer Anders Vadsholt as the new Chief Executive Officer in addition to his existing position as Chief Financial Officer, succeeding Christophe Bourdon on March 1, 2022.

Mr. Bourdon resigned from his position at Orphazyme on February 28, 2022. The resignation entails a forfeiture of the vesting conditions attached to the RSUs and PSUs under the modified and extraordinary granted in October 2021 except for the modified sign-on bonus shares that immediately vested when granted in March 2021. For any ongoing RSU vesting period, Mr. Bourdon was entitled to receive a pro rata allocation of RSUs until the date of release of his work obligations.

The Executive Management was up until and including 2021 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. 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, 2022 and 2021.

REMUNERATION TO INDIVIDUAL

| MEMBERS OF EXECUTIVE MANAGEMENT (DKK 000) | 2022 | 2021 |
|--|---------------|---------------|
| Anders Vadsholt (CEO from March 1, 2022 and CFO since 2016) | | |
| Salary | 2,908 | 2,376 |
| Bonus | 4,134 | 585 |
| Share-based compensation ⁽¹⁾ | 1,657 | 2,164 |
| Other employee benefits ⁽³⁾ | 294 | 277 |
| Total | 8,993 | 5,402 |
| Christophe Bourdon (CEO from April 1, 2021 until February 2022) | | |
| Salary | 650 | 2,925 |
| Bonus | — | 1,330 |
| Share-based compensation ^(1,2) | 231 | 4,967 |
| Other employee benefits | 186 | 107 |
| Total | 1,067 | 9,329 |
| Total remuneration to the Executive Management | 10,060 | 14,731 |

(1) Investing expense on grants in prior years measured at fair value at grant date

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

(3) Board fee shown of DKK 231 thousand not included

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
OF THE BOARD OF DIRECTORS (DKK 000)**

| | 2022 | 2021 |
|---|--------------|----------------|
| Bo Jesper Hansen (Chairman since June 2022) | | |
| Board and committee fees | 606 | 447 |
| Ad hoc board fees | — | — |
| Travel allowance | 33 | 97 |
| Share-based compensation | (50) | 68 |
| Total | 589 | 612 |
| Georges Gemayel (Chairman until June 2022) | | |
| Board and committee fees | 370 | 659 |
| Ad hoc board fees ⁽¹⁾ | — | 1,312 |
| Travel allowance | — | 30 |
| Share-based compensation | (161) | 164 |
| Total | 209 | 2,165 |
| Martin Bonde (resigned in May 2022) | | |
| Board and committee fees | 176 | 318 |
| Travel allowance | — | — |
| Share-based compensation | (24) | 48 |
| Total | 152 | 366 |
| Martijn Kleijwegt (resigned in June 2021) | | |
| Board and committee fees | — | 161 |
| Travel allowance | — | — |
| Share-based compensation | — | 25 |
| Total | — | 186 |
| Rémi Droller (resigned in June 2021)⁽²⁾ | | |
| Board and committee fees | — | (828) |
| Travel allowance | — | (90) |
| Share-based compensation | — | (89) |
| Total | — | (1,007) |
| Sten Verland | | |
| Board and committee fees | — | 81 |
| Travel allowance | — | — |
| Share-based compensation | — | 25 |
| Total | — | 106 |
| Anders Hedegaard (resigned in June 2021) | | |
| Board and committee fees | — | 153 |
| Travel allowance | — | — |
| Share-based compensation | — | 25 |
| Total | — | 178 |
| Catherine Moukheibir (resigned in December 2021) | | |
| Board and committee fees | — | 353 |
| Ad hoc board fees | — | — |
| Travel allowance | — | — |
| Share-based compensation | — | 25 |
| Total | — | 378 |
| Carolee Barlow (resigned in May 2022) | | |
| Board and committee fees | 176 | 320 |
| Travel allowance | — | — |
| Share-based compensation | (24) | 80 |
| Total | 152 | 400 |
| Stephanie Okey (resigned in May 2022) | | |
| Board and committee fees | 176 | 247 |
| Ad hoc board fees | — | 107 |
| Share-based compensation | (72) | 72 |
| Total | 104 | 426 |
| Andrew Mercieca (resigned in June 2022) | | |
| Board and committee fees | 212 | 24 |
| Total | 212 | 24 |
| John Sommer Schmidt (since June 2022) | | |
| Board and committee fees | 231 | - |
| Total | 231 | - |
| Anders Fink Vadsholt (DK) (since June 2022) | | |
| Board and committee fees | 231 | - |
| Total | 231 | - |
| Total remuneration to the Board of Directors | 1,880 | 3,834 |

(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, 2022 and 2021, 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, the 2020 LTIP and the 2021 LTIP (Note 2.5)
- Participation of the Board members in the 2019 RSU, 2020 RSU and 2021 RSU programs (Note 2.5)
- 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, 2022 and 2021, 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 | | Number of shares owned | |
|---|-------------------------------|-------------------------------------|--|-------------------------------|-------------------------------------|
| | | 2022 | | 2021 | |
| Anders Vadsholt | | 159,517 | | 160,717 | |
| | | December 31, 2022 | | December 31, 2021 | |
| MEMBERS OF THE BOARD OF DIRECTORS: | Number of shares owned | Number of Unvested RSUs 2021 | Number of Unexercised RSUs 2020 | Number of shares owned | Number of Unvested RSUs 2021 |
| Bo Jesper Hansen | 143,234 | 2,849 | — | 143,234 | 2,849 |
| John Sommer Schmidt | — | — | — | — | — |

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 | 2022 | 2021 |
|-------------------------------|--------------|--------------|
| Audit services | 692 | 2,743 |
| Audit-related services | 493 | 430 |
| Other assistance | — | 753 |
| Total fees to auditors | 1,185 | 3,926 |

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.

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 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.

2022 Parent Financial Statements

STATEMENTS OF PROFIT OR LOSS AND OTHER COMPREHENSIVE INCOME

For the years ended December 31,

| 'DKK 000, except per share and share data | Note | 2022 | 2021 |
|--|------|-----------------|------------------|
| Net revenue | 2.1 | — | — |
| Research and development expenses | 2.2 | — | — |
| General and administrative expenses | 2.3 | (41,241) | (66,381) |
| Operating loss | | (41,241) | (66,381) |
| Financial income | 2.5 | 185 | 365 |
| Financial expenses | 2.5 | — | — |
| Loss before tax | | (41,056) | (66,017) |
| Income tax benefit | 2.6 | 2,750 | 5,550 |
| Loss from continuing operations | | (38,306) | (60,517) |
| Net result from discontinued operations | 1.3 | 73,378 | (567,607) |
| Net result for the year | 1.3 | 35,072 | (628,123) |
| Items that will be reclassified subsequently to the Statement of Profit or Loss: | | | |
| Exchange difference from translation of foreign operations | | (141) | (57) |
| Other comprehensive Profit/loss | | (141) | (57) |
| Total comprehensive Profit/loss | | 34,931 | (628,181) |

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

STATEMENTS OF FINANCIAL POSITION

DKK 000

| ASSETS | Note | 2022 | 2021 |
|-----------------------------------|------|---------------|----------------|
| Non-current assets | | | |
| Intangible assets | 2.7 | — | 2,152 |
| Right-of-use assets | 2.8 | — | 3,726 |
| Property, plant, and equipment | 2.9 | — | 2,205 |
| Corporation tax receivable | | — | 2,750 |
| Investment in subsidiaries | 2.10 | — | — |
| Prepayments and deposits | 2.11 | — | 593 |
| Total non-currents assets | | — | 11,426 |
| Current assets | | | |
| Corporation tax receivable | 2.6 | 5,500 | 5,500 |
| Trade receivables | | 4,103 | 29,268 |
| Prepayments and other receivables | 2.11 | 8,305 | 25,056 |
| Inventory | | — | — |
| Cash | 2.13 | 38,918 | 85,693 |
| Total current assets | | 56,826 | 145,517 |
| Total assets | | 56,826 | 156,943 |

| EQUITY AND LIABILITIES | Note | 2022 | 2021 |
|--------------------------------------|------|---------------|----------------|
| Equity | | | |
| Share capital | | 35,312 | 34,952 |
| Share premium | | 2,087,437 | 2,082,487 |
| Other reserves | | (303) | 2,324 |
| Accumulated deficit | | (2,081,472) | (2,120,884) |
| Total equity | | 40,974 | (1,121) |
| Non-current liabilities | | | |
| Borrowings | 2.12 | — | 2,482 |
| Lease liabilities | 2.8 | — | 2,427 |
| Discount and rebate liabilities | 2.12 | — | 28,293 |
| Other non-current liabilities | 2.12 | 98 | 98 |
| Total non-current liabilities | | 98 | 33,300 |
| Current liabilities | | | |
| Borrowings | 2.12 | — | 30,983 |
| Lease liabilities | 2.8 | — | 2,122 |
| Trade payables and accruals | 2.12 | 9,206 | 49,445 |
| Discount and rebate liabilities | 2.12 | 4,457 | 7,900 |
| Other liabilities | 2.12 | 2,090 | 34,314 |
| Total current liabilities | | 15,753 | 124,764 |
| Total equity and liabilities | | 56,826 | 156,943 |

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

STATEMENTS OF CHANGES IN SHAREHOLDERS' EQUITY

| | Notes | Share capital | Share premium | Foreign currency translation reserve | Share-based compensation - acquisition of intangible assets | Accumulated deficit | Total |
|--|-------|---------------|------------------|--------------------------------------|---|---------------------|------------------|
| Balance as of December 31, 2020 | | 34,698 | 2,082,255 | (105) | 5,779 | (1,511,221) | 611,406 |
| Net loss for the year | | — | — | — | — | (628,124) | (628,124) |
| Other comprehensive income | | — | — | (57) | — | — | (57) |
| Total other comprehensive income (loss) | | — | — | (57) | — | — | (628,181) |
| Transactions with owners: | | | | | | | |
| Capital increase, issuance of Matching Shares, net of costs | | 170 | — | — | — | — | 170 |
| Capital increase, Bonus Shares | | 22 | — | — | (1,645) | 1,623 | — |
| Cash settlement of Bonus Shares | | — | — | — | (1,648) | — | (1,648) |
| Capital increase, issuance of sign-on bonus shares to former CEO | | 58 | — | — | — | — | 58 |
| Capital increase, exercise of RSUs | | 4 | 232 | — | — | — | 236 |
| Share-based compensation expense | | — | — | — | — | 16,838 | 16,838 |
| Total transactions with owners | | 254 | 232 | — | (3,293) | 18,461 | 15,653 |
| Balance as of December 31, 2021 | | 34,952 | 2,082,487 | (162) | 2,486 | (2,120,884) | (1,121) |
| Net loss for the year | | — | — | — | — | 35,072 | 35,072 |
| Other comprehensive income | | — | — | (141) | — | — | (141) |
| Total other comprehensive income (loss) | | — | — | (141) | — | 35,072 | 34,931 |
| Transactions with owners: | | | | | | | |
| Capital increase, exercise of RSUs | | 360 | 5,091 | — | — | — | 5,451 |
| Transaction cost related to capital increase | | — | (141) | — | — | — | (141) |
| Reserve for bonus shares reversed | | — | — | — | (2,486) | 2,486 | — |
| Share-based compensation expense | | — | — | — | — | 1,853 | 1,853 |
| Total transactions with owners | | 360 | 4,950 | — | (2,486) | 4,339 | 7,163 |
| Balance as of December 31, 2022 | | 35,312 | 2,087,437 | (303) | — | (2,081,472) | 40,974 |

STATEMENTS OF CASH FLOWS

| DKK 000 | Note | 2022 | 2021 |
|---|-------------|------------------|------------------|
| Operating result from continuing operations | | (41,241) | (66,381) |
| Operating result from discontinued operations | | (67,278) | (568,788) |
| Adjustments to reconcile operating result to cash flows from operating activities: | | | |
| Equity-settled share-based compensation expense | | 1,773 | 12,493 |
| Depreciation and amortization | 2.2,2.3 | 782 | 14,870 |
| Change in prepayments, deposits, and other receivables | 2.11 | 41,393 | 2,344 |
| Change in trade payables, accruals, and other liabilities | 2.12 | (37,889) | 12,638 |
| Change in intercompany receivables | 2.11 | 1,117 | 206,578 |
| Change in intercompany payables | 2.12 | (7,794) | 544 |
| Exchange rate adjustments on intercompany balances | | 1,949 | 9,055 |
| Cash generated by operating activities before financial items and tax | | (107,188) | (376,648) |
| Interest paid | | (1,064) | (6,098) |
| Interest received | | 185 | 0 |
| Income taxes received | | 5,500 | 5,500 |
| Cash flow from operating activities | | (102,567) | (377,246) |
| Investment in intangible assets | 2.7 | — | (85) |
| Investment in property, plant and equipment | 2.9 | — | — |
| Proceeds from sale of property, plant and equipment | | 1,460 | 1,040 |
| Proceeds from sale of activity | 1.3 | 88,887 | — |
| Cash flow from investing activities | | 90,347 | 955 |
| Proceeds from issue of shares and exercise of RSUs | | 5,451 | (1,184) |
| Transaction cost related to issuance of shares | | (141) | — |
| Repayment of borrowing | | (39,155) | (25,657) |
| Repayment of leasing liabilities | | (1,233) | (3,090) |
| Cash flow from financing activities | | (35,078) | (29,931) |
| Changes in cash and cash equivalents | | (47,298) | (406,221) |
| Cash and cash equivalents at the beginning of the period | | 85,692 | 487,322 |
| Exchange rate adjustment on cash and cash equivalents | | 523 | 4,592 |
| Cash and cash equivalents at the end of the period | | 38,918 | 85,693 |

SECTION 1 Basis of Preparation

The financial statements of Orphazyme A/S (the "Parent Company") have been prepared in accordance with International Financial Reporting Standards, or IFRS, as adopted by the EU and additional disclosure requirements under the Danish Financial Statements Act.

The Parent Company applies the same disclosed information as disclosed in the Group's consolidated financial statements. Therefore, only disclosed information specific to the Parent Company or that differ from the disclosed information applied by the Group are disclosed in these notes to the parent statements. If a specific information is not mentioned, the disclosed information in the Group is applied.

1.1 COMPANY INFORMATION

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").

1.2 SIGNIFICANT ACCOUNTING POLICIES APPLICABLE TO THE PARENT COMPANY

The Parent Company applies the same accounting policies as disclosed in the Group's consolidated financial statements. Therefore, only accounting policies specific to the Parent Company or that differ from the accounting policies applied by the Group are disclosed in these notes to the parent statements. If an accounting policy is not specifically mentioned, the Group accounting policy is applied.

A description of Management's key accounting estimates and judgements as well as new IFRS standards are disclosed in the Group financial statements and also apply to the Parent Company.

The Parent Company financial statements are presented in Danish Kroner, or DKK, which is both the functional and presentation currency of the Parent Company. Where indicated, amounts are rounded to the nearest thousand, or TDKK.

1.3 SIGNIFICANT EVENTS AFTER THE REPORTING PERIOD

The Parent Company applies the same disclosed information as disclosed in the Group's consolidated financial statements in note 1.6.

1.4 DISCONTINUED OPERATIONS

| DKK 000 | Note | 2022 | 2021 |
|--|----------|-----------------|------------------|
| Net revenue | 2.1 | 17,867 | 36,193 |
| Research and development expenses | 2.2, 2.3 | (36,660) | (331,046) |
| General and administrative expenses | 2.4 | (48,485) | (273,934) |
| Operating loss | | (67,278) | (568,788) |
| Financial income | 2.7 | 4,106 | 11,983 |
| Financial expenses | 2.7 | (9,116) | (10,802) |
| Net result before tax | | (72,288) | (567,607) |
| Income tax benefit | 2.8 | — | — |
| Net result for the period | | (72,288) | — |
| Gain from disposal of discontinued operations | | 145,666 | — |
| Net result from discontinued operations | | 73,378 | (567,607) |

| | | |
|---|---------------|------------------|
| Cash flow from operating activities | (6,973) | (545,926) |
| Cash flow from investing activities | 90,347 | — |
| Cash flow from financing activities | (40,388) | (25,657) |
| Net cash flow from discontinued operations | 42,986 | (571,583) |

Gain on disposal/carrying amount of disposed assets

| DKK 000 | Note | 2022 | 2021 |
|--|------|-----------------|------|
| Carrying values of intangible assets | | 1,603 | — |
| Carrying values of property, plan and equipment | | 134 | — |
| Carrying values of liabilities | | (58,370) | — |
| Carrying values of assets and liabilities | | (56,633) | — |
| Gain on disposal – net | | 145,520 | — |
| Gain on disposal | | 145,520 | — |
| Net cash inflow from disposal of business | | 88,887 | — |

SECTION 2 Notes

2.1 NET REVENUE

| DKK 000 | 2022 | 2021 |
|----------------------------|--------|--------|
| Revenue by type | | |
| Revenue from sale of goods | 17,867 | 36,193 |
| Revenue by partner | | |
| Clinigen Health Limited | 17,867 | 36,193 |
| Geographical areas | | |
| France | 17,867 | 36,193 |

2.2 RESEARCH AND DEVELOPMENT EXPENSES

| DKK 000 | 2022 | 2021 |
|--|---------------|----------------|
| External costs | 18,385 | 229,252 |
| Employee costs | 17,926 | 79,388 |
| Intercompany expenses | — | 8,697 |
| Depreciation and amortization | 349 | 13,709 |
| Total research and development expenses | 36,660 | 331,046 |

2.3 GENERAL AND ADMINISTRATIVE EXPENSES

| DKK 000 | 2022 | 2021 |
|--|---------------|----------------|
| External costs | 59,650 | 136,148 |
| Employee costs | 20,699 | 64,294 |
| Intercompany expenses | 8,944 | 138,713 |
| Depreciation and amortization | 433 | 1,161 |
| Total research and development expenses | 89,726 | 340,316 |

2.4 EMPLOYEE COSTS

DKK 000

| Employee costs | 2022 | 2021 |
|---|---------------|----------------|
| Salaries | 21,488 | 99,110 |
| Cash bonus | 9,046 | 10,886 |
| Share-based compensation | 1,853 | 12,050 |
| Pension | 1,588 | 8,306 |
| Other social security contributions | 108 | 1,900 |
| Other staff costs | 1,474 | 7,596 |
| Total employee costs excluding board remuneration | 35,557 | 139,848 |
| Board remuneration | 3,148 | 3,391 |
| Board share-based compensation | (80) | 443 |
| Total employee costs | 38,625 | 143,683 |
| <i>Recognized as follows in the statement of Profit or Loss</i> | | |
| Research and development expenses | 17,926 | 79,388 |
| General and administrative expenses | 20,699 | 64,294 |
| Total employee costs | 38,625 | 143,683 |
| Average number of full-time employees | 19 | 99 |
| Year-end number of full-time employees | 1 | 49 |

2.5 FINANCIAL INCOME AND FINANCIAL EXPENSES

| DKK 000 | 2022 | 2021 |
|---------------------------------------|--------------|---------------|
| Interest income on cash balances | 185 | — |
| Foreign currency exchange gains | 4,106 | 11,438 |
| Gain on embedded call option | — | 546 |
| Total financial income | 4,291 | 11,983 |
| Interest expense on Loan Agreement | 5,959 | 7,350 |
| Interest expense on lease liabilities | 96 | 490 |
| Interest expense on cash balances | 158 | (330) |
| Foreign currency exchange loss | 2,843 | 1,364 |
| Bank fees and other charges | 60 | 80 |
| Total financial expenses | 9,116 | 10,438 |

2.6 INCOME TAXES

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

| DKK 000 | 2022 | 2021 |
|---|--------------|--------------|
| Current tax benefit on net loss | 7,111 | 139,397 |
| Adjustments prior years | — | — |
| Tax credit research and development expenses | 2,750 | 5,500 |
| Change in unrecognized deferred tax before tax credit | (25,129) | (150,544) |
| Permanent differences | 32,240 | 11,147 |
| Total income tax benefit for the year | 2,750 | 5,500 |

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

| DKK 000 | 2022 | 2021 |
|--|--------------|----------------|
| Net result before tax from continuing operations | (41,056) | (66,017) |
| Net result before tax from discontinued operations | 73,378 | (567,607) |
| Corporate income tax rate in Denmark | 22% | 22% |
| Computed income tax benefit | 7,111 | 139,397 |
| <i>Tax effect of:</i> | | |
| Other non-deductible expenses, including US listing-related costs and share-based compensation | 32,240 | 11,147 |
| Deferred tax asset not recognized after tax credit | (22,379) | (145,044) |
| Total income tax benefit for the year | 2,750 | 5,500 |

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

| DKK 000 | 2022 | 2021 |
|--|-------------|-------------|
| Tax deductible losses | 480,554 | 319,811 |
| Deferred tax on intangible assets | — | 132,310 |
| Other temporary differences | 44 | 1,882 |
| | 480,588 | 454,003 |
| Deferred tax asset not recognized | 480,588 | 454,003 |
| Carrying amount included in the Statement of Financial Position | — | — |

2.7 INTANGIBLE ASSETS

| DKK 000 | Software | Licenses | Total |
|--|-----------------|-----------------|---------------|
| Cost at December 31, 2020 | 896 | 12,083 | 12,979 |
| Additions | 85 | — | 85 |
| Cost at December 31, 2021 | 981 | 12,083 | 13,064 |
| Additions | — | — | — |
| Cost at December 31, 2022 | 981 | 12,083 | 13,064 |
| Accumulated amortization at December 31, 2020 | 42 | 2,256 | 2,298 |
| Amortization expense | 391 | 119 | 510 |
| Impairment expense | — | 8,105 | 8,105 |
| Accumulated amortization at December 31, 2021 | 433 | 10,480 | 10,912 |
| Amortization expense | 464 | — | 464 |
| Depreciation reversed on disposals during the year | (896) | (10,480) | (11,376) |
| Accumulated amortization at December 31, 2022 | — | — | — |
| Net carrying value at | | | |
| December 31, 2021 | 549 | 1,603 | 2,152 |
| December 31, 2022 | — | — | — |

2.8 LEASES

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

| DKK 000 | Office | Operating | Total |
|-----------------------------|--------------|--------------|---------------|
| | buildings | equipment | |
| At December 31, 2020 | 9,456 | 3,406 | 12,862 |
| Disposals | (1,177) | - | (1,177) |
| Depreciation expense | (2,387) | (167) | (2,554) |
| Impairment expense | - | (3,239) | (3,239) |
| Modifications | (2,166) | - | (2,166) |
| At December 31, 2021 | 3,726 | - | 3,726 |
| Disposals | (2,405) | - | (2,405) |
| Depreciation expense | (1,321) | - | (1,321) |
| At December 31, 2022 | - | - | - |

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

| DKK 000 | 2022 | 2021 |
|-----------------------|--------------|---------------|
| At January 1 | 4,549 | 11,346 |
| Accretion of interest | - | 490 |
| Disposals | (4,549) | (1,212) |
| Payments | - | (3,579) |
| Modifications | - | (2,496) |
| At December 31 | - | 4,549 |
| Current | - | 2,122 |
| Non-current | - | 2,427 |

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 | 2022 | 2021 |
|---|--------------|--------------|
| Depreciation and impairment expense of right-of-use assets (R&D) | 1,057 | 5,386 |
| Depreciation and impairment expense of right-of-use assets (G&A) | 264 | 407 |
| Interest expense on lease liabilities | 96 | 490 |
| Gain on lease modification and disposals | - | (330) |
| Total amount recognized in the Statement of Profit or Loss | 1,417 | 5,953 |

2.9 PROPERTY, PLANT, AND EQUIPMENT

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, 2020 | 6,791 | 2,066 | 8,857 |
| Disposals | (597) | - | (597) |
| Cost at December 31, 2021 | 6,194 | 2,066 | 8,260 |
| Disposals | (6,194) | (2,066) | (8,260) |
| Cost at December 31, 2022 | - | - | - |
| Accumulated depreciation at December 31, 2020 | 4,533 | 617 | 5,150 |
| Depreciation expense | 999 | 317 | 1,316 |
| Depreciation reversed on disposals during the year | (411) | - | (411) |
| Accumulated depreciation at December 31, 2021 | 5,121 | 934 | 6,055 |
| Depreciation expense | 266 | 123 | 389 |
| Depreciation reversed on disposals during the year | (5,387) | (1,057) | (6,444) |
| Accumulated depreciation at December 31, 2022 | - | - | - |
| Net carrying value at | | | |
| December 31, 2021 | 1,073 | 1,132 | 2,205 |
| December 31, 2022 | - | - | - |

There has been no impairment of property, plant and equipment for the years ended December 31, 2022 and 2021. Depreciation expense is included within operating loss as follows:

| DKK 000 | 2022 | 2021 |
|-------------------------------------|------------|--------------|
| Research and development expenses | 143 | 1,031 |
| General and administrative expenses | 246 | 285 |
| Total depreciation expense | 389 | 1,316 |

2.10 INVESTMENT IN GROUP COMPANIES

§ ACCOUNTING POLICIES

Investments in subsidiaries are measured in the Parent Company financial statements at the lower of cost or recoverable amount. Any distributed dividends are recognized in the income statement of the Parent Company.

| DKK 000 | 2022 | 2021 |
|--------------------------------------|----------------|----------------|
| Cost at January 1 | 3,942 | 1,346 |
| Additions | - | 2,596 |
| Cost end of year December 31 | 3,942 | 3,942 |
| Adjustment January 1 | (3,942) | (1,207) |
| Impairment | - | (2,735) |
| End of year December 31 | (3,942) | (3,942) |
| Carrying amount of investment | - | - |

| DKK 000 | Registered office | Ownership interest (%) | Share capital | Equity | Net result |
|---------------------|-------------------|------------------------|----------------------|--------|------------|
| Orphazyme US, Inc. | Delaware, USA | 100 % | USD 1 (USD 000) | 1,403 | (227) |
| Orphazyme GmHH (CH) | Zug, Switzerland | 100 % | CHF 20,000 (CHF 000) | 355 | (237) |

2.11 PREPAYMENTS, DEPOSITS, AND OTHER RECEIVABLES

| DKK 000 | 2022 | 2021 |
|---|----------|------------|
| Leasehold deposit | - | 593 |
| Total non-current prepayments and deposits | - | 593 |

| DKK 000 | 2022 | 2021 |
|--|--------------|---------------|
| Prepayments to vendors | 1,706 | 12,436 |
| VAT receivable, net | 662 | 2,903 |
| Foreign VAT receivable | 1,275 | 1,627 |
| Receivables from group entities | 4,183 | 5,300 |
| Other current receivables | 479 | 2,790 |
| Total current prepayments and other receivables | 8,305 | 25,056 |

2.12 FINANCIAL ASSETS AND LIABILITIES

| DKK 000 | 2022 | 2021 |
|---|--------------|---------------|
| Borrowings | - | 33,465 |
| Lease liabilities (Note 3.2) | - | 4,549 |
| Trade payables | 4,357 | 41,395 |
| Accruals | 4,849 | 8,050 |
| Total liabilities measured at amortized cost | 9,206 | 87,459 |

| DKK 000 | 2022 | 2021 |
|--|--------------|---------------|
| Remuneration to the Board of Directors | 213 | 293 |
| Payables to group entities | 657 | 8,451 |
| Payroll and employee-related costs | 1,220 | 25,570 |
| Total current other liabilities | 2,090 | 34,314 |

| DKK 000 | December 31, 2021 | Cash flows | Non-cash changes | | | | December 31, 2022 |
|--|-------------------|-----------------|------------------|-------------------------------|----------------------|---------------------------|-------------------|
| | | | Disposals | Adjustments and modifications | Accumulated interest | Exchange rate adjustments | |
| Borrowings | 33,466 | (39,155) | - | - | 5,959 | (269) | - |
| Lease liabilities | 4,549 | (1,233) | (3,412) | - | 96 | - | - |
| Total liabilities from financing activities | 38,015 | (40,388) | (3,412) | - | 6,055 | (269) | - |

| DKK 000 | December 31, 2021 | Cash flows | Accruals | December 31, 2022 |
|--|----------------------|------------|------------------|----------------------|
| Discount and rebate liabilities | 36,193 | — | (31,736) | 4,457 |
| Total liabilities from accrued discount and rebates | 36,193 | — | (31,736) | 4,457 |

2.13 CASH

| DKK 000 | 2022 | 2021 |
|-------------------|---------------|---------------|
| DKK | 14,794 | 53,291 |
| USD | 10,609 | 27,440 |
| EUR | 13,209 | 4,070 |
| CHF | - | - |
| GBP | 306 | 892 |
| Total cash | 38,918 | 85,693 |

2.14 COMMITMENTS AND CONTINGENCIES

The Parent Company applies the same disclosed information as disclosed in the Group's consolidated financial statements in note 3.9.

2.15 RELATED PARTY DISCLOSURES

Orphazyme A/S' related parties are the parent company's Board of Directors, Executive Management and close members of the family of these persons.

Transactions with subsidiaries

Orphazyme US, Inc. and Orphazyme GmbH (CH) are 100% owned subsidiaries of Orphazyme A/S and are included in the consolidated financial statements. They perform certain research and development, general and administrative and management activities on behalf on the parent company. All intercompany transactions have been eliminated in the consolidated financial statements of the Orphazyme Group.

| DKK 000 | 2022 | 2021 |
|--------------------------------------|-------|---------|
| Transaction with subsidiaries | | |
| Service fee costs | 8,944 | 147,410 |
| Balances with subsidiaries | | |
| Current receivables | 4,183 | 5,300 |
| Current payables | 657 | 8,451 |

Please refer to note 4.6 in the consolidated financial statements for additional information regarding transactions with related parties of the Group.

2.16 CAPITAL MANAGEMENT

The Parent Company applies the same disclosed information as disclosed in the Group's consolidated financial statements in note 4.1.

2.17 REMUNERATION OF BOARD OF DIRECTORS AND EXECUTIVE MANAGEMENT

The Parent Company applies the same disclosed information as disclosed in the Group's consolidated financial statements in note 4.5.

2.18 FEES TO AUDITORS

The Parent Company applies the same disclosed information as disclosed in the Group's consolidated financial statements in note 4.7.

Statements and Signatures

Statements by Board of Directors and Executive Management

The Board of Directors and Executive Management have today considered and approved the annual report of Orphazyme A/S for the financial year January 1-December 31, 2022.

The consolidated financial statements of the Group and the Parent Company's financial statements have been prepared in accordance with International Financial Reporting Standards (IFRS) as issued by the International Accounting Standards Board (IASB) and in accordance with IFRS as endorsed by the EU as well as additional disclosure requirements under the Danish Financial Statements Act.

In our opinion, the Group's consolidated financial statements and the Parent Company financial statements provide a fair presentation of the assets, liabilities, and financial position at December 31, 2022 and the results of the Group's and Parent Company's operations and cash flows for the financial year January 1-December 31, 2022.

In our opinion, Management's Review provides a fair presentation of the development in the Group's operations and financial circumstances, the results of the year, and the overall financial position of the Group as well as a description of the most significant risks and elements of uncertainty facing the Group.

We recommend that the annual report be adopted at the Annual General Meeting scheduled to be held on 17 May 2023.

Copenhagen, 25 April 2023

BOARD OF DIRECTORS

Bo Jesper Hansen
Chairman of the Board

John Sommer Schmidt
Deputy Chairman of the Board

Anders Vadsholt

EXECUTIVE MANAGEMENT

Anders Vadsholt
Chief Executive Officer
and Chief Financial Officer

Independent Auditors' Report

To the Shareholders of Orphazyme A/S

Report on the audit of the Consolidated Financial Statements and Parent Company Financial Statements

Opinion

We have audited the consolidated financial statements and the parent company financial statements of Orphazyme A/S for the financial year 1 January – 31 December 2022, which comprise income statement, statement of comprehensive income, balance sheet, statement of changes in equity, cash flow statement and notes, including accounting policies, for the Group and the Parent Company. The consolidated financial statements and the parent company financial statements are prepared in accordance with International Financial Reporting Standards as adopted by the EU and additional requirements of the Danish Financial Statements Act.

In our opinion, the consolidated financial statements and the parent company financial statements give a true and fair view of the financial position of the Group and the Parent Company at 31 December 2022 and of the results of the Group's and the Parent Company's operations and cash flows for the financial year 1 January – 31 December 2022 in accordance with International Financial Reporting Standards as adopted by the EU and additional requirements of the Danish Financial Statements Act.

Our opinion is consistent with our long-form audit report to the Audit Committee and the Board of Directors.

Basis for opinion

We conducted our audit in accordance with International Standards on Auditing (ISAs) and additional requirements applicable in Denmark. Our responsibilities under those standards and requirements are further described in the "Auditor's responsibilities for the audit of the consolidated financial statements and the parent company financial statements" (hereinafter collectively referred to as "the financial statements") section of our report. We believe that the audit evidence we have obtained is sufficient and appropriate to provide a basis for our opinion.

Independence

We are independent of the Group in accordance with the International Ethics Standards Board for Accountants' International Code of Ethics for Professional Accountants (IESBA Code) and the additional ethical requirements applicable in Denmark, and we have fulfilled our other ethical responsibilities in accordance with these requirements and the IESBA Code.

To the best of our knowledge, we have not provided any prohibited non-audit services as described in article 5(1) of Regulation (EU) no. 537/2014.

Emphasis of matter in the financial statements

We draw attention to note 3.9 to the financial statements, which describes the material uncertainty associated with the outcome of a class action lawsuit that was filed against the Company and certain of its current and former officers and directors. We have not modified our opinion in respect of this matter.

Appointment of auditor

We were initially appointed as auditor of Orphazyme A/S on 4 December 2015 for the financial period 1 July to 31 December 2015. We have been reappointed annually by resolution of the general meeting for a total consecutive period of 8 years up until the financial year 2022.

Key audit matters

Key audit matters are those matters that, in our professional judgement, were of most significance in our audit of the financial statements for the financial year 2022. These matters were addressed during our audit of the financial statements as a whole and in forming our opinion thereon. We do not provide a separate opinion on these matters. For each matter below, our description of how our audit addressed the matter is provided in that context.

We have fulfilled our responsibilities described in the "Auditor's responsibilities for the audit of the financial statements" section, including in relation to the key audit matters below. Accordingly, our audit included the

design and performance of procedures to respond to our assessment of the risks of material misstatement of the financial statements. The results of our audit procedures, including the procedures performed to address the matters below, provide the basis for our audit opinion on the financial statements.

Accounting for discontinued Operations (IFRS 5)

In May 2022, as a part of the restructuring proposal to the Danish Maritime and Commercial High Court and Orphazyme's known creditors, the Company announced that it had 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 for a total of DKK 88.8 million (USD 12.8 million) in cash and assumed liabilities estimated to equal approximately DKK 36.2 million (USD 5.2 million). The sale of assets and business activities was completed on May 31, 2022. The net result of discontinued operations amounts to DKK 64.4 million in 2022, including a gain from disposal of the discontinued operations of DKK 145.5 million.

Management concluded that the sale of the assets and business activities should be reported in accordance with IFRS 5 - Non-Current Assets Held for Sale as discontinued operations in the 2022 financial statements. The application of IFRS 5 is significant to our audit because the transaction and its accounting is non-routine and involves significant management judgements regarding the presentation of the net result as discontinued operations.

As a result of these conclusions, there are requirements around the presentation in the financial statements and disclosure notes, the identification of income and expenses allocated to the discontinued operations and continued operations, including assumptions and estimates made with regard to the allocation and presentation, and adjustments to be recorded, e.g. common cost allocations.

How our audit addressed the matter

Our audit procedures included an evaluation of the Company's conclusions on the classification and presentation of the business activities as discontinued operations. Our audit procedures on the accounting and disclosure of reported net result from discontinued operations focused on the validation with the underlying sales documents and related contracts, agreed the elements of the gain calculations to them, testing of received proceeds per bank statements, performing cut-off procedures on material balance sheet items and income statement items and assessing the assumption applied by management to reassess the potential exposure on the continued operations.

We assessed the presentation of discontinued and continuing operations by reference to its size and nature. We also tested whether the comparative figures in the statement of comprehensive income for the discontinued operations were accurately and fully adjusted. We refer to note 1.7 in the financial statements regarding discontinued operations.

Statement on the Management's review

Management is responsible for the Management's review.

Our opinion on the financial statements does not cover the Management's review, and we do not express any form of assurance conclusion thereon.

In connection with our audit of the financial statements, our responsibility is to read the Management's review and, in doing so, consider whether the Management's review is materially inconsistent with the financial statements or our knowledge obtained during the audit, or otherwise appears to be materially misstated.

Moreover, it is our responsibility to consider whether the Management's review provides the information required under the Danish Financial Statements Act.

Based on the work we have performed, we conclude that the Management's review is in accordance with the financial statements and has been prepared in accordance with the requirements of the Danish Financial Statements Act. We did not identify any material misstatement of the Management's review.

Management's responsibilities for the financial statements

Management is responsible for the preparation of consolidated financial statements and parent company financial statements that give a true and fair view in accordance with International Financial Reporting Standards as adopted by the EU and additional requirements of the Danish Financial Statements Act and for such internal control as Management determines is necessary to enable the preparation of financial statements that are free from material misstatement, whether due to fraud or error.

In preparing the financial statements, Management is responsible for assessing the Group's and the Parent Company's ability to continue as a going concern, disclosing, as applicable, matters related to going concern and using the going concern basis of accounting in preparing the financial statements unless Management either intends to liquidate the Group or the Parent Company or to cease operations, or has no realistic alternative but to do so.

Auditor's responsibilities for the audit of the financial statements

Our objectives are to obtain reasonable assurance as to whether the financial statements as a whole are free from material misstatement, whether due to fraud or error, and to issue an auditor's report that includes our opinion. Reasonable assurance is a high level of assurance, but is not a guarantee that an audit conducted in accordance with ISAs and additional requirements applicable in Denmark will always detect a material misstatement when it exists. Misstatements can arise from fraud or error and are considered material if, individually or in the aggregate, they could reasonably be expected to influence the economic decisions of users taken on the basis of the financial statements.

As part of an audit conducted in accordance with ISAs and additional requirements applicable in Denmark, we exercise professional judgement and maintain professional scepticism throughout the audit. We also:

- Identify and assess the risks of material misstatement of the financial statements, whether due to fraud or error, design and perform audit procedures responsive to those risks and obtain audit evidence that is sufficient and appropriate to provide a basis for our opinion. The risk of not detecting a material misstatement resulting from fraud is higher than for one resulting from error, as fraud may involve collusion, forgery, intentional omissions, misrepresentations or the override of internal control.
- Obtain an understanding of internal control relevant to the audit in order to design audit procedures that are appropriate in the circumstances, but not for the purpose of expressing an opinion on the effectiveness of the Group's and the Parent Company's internal control.
- Evaluate the appropriateness of accounting policies used and the reasonableness of accounting estimates and related disclosures made by Management.
- Conclude on the appropriateness of Management's use of the going concern basis of accounting in preparing the financial statements and, based on the audit evidence obtained, whether a material uncertainty exists related to events or conditions that may cast significant doubt on the Group's and the Parent Company's ability to continue as a going concern. If we conclude that a material uncertainty exists, we are required to draw attention in our auditor's report to the related disclosures in the financial statements or, if such disclosures are inadequate, to modify our opinion. Our conclusions are based on the audit evidence obtained up to the date of our auditor's report. However, future events or conditions may cause the Group and the Parent Company to cease to continue as a going concern.
- Evaluate the overall presentation, structure and contents of the financial statements, including the note disclosures, and whether the financial statements represent the underlying transactions and events in a manner that gives a true and fair view.
- Obtain sufficient appropriate audit evidence regarding the financial information of the entities or business activities within the Group to express an opinion on the consolidated financial statements. We are responsible for the direction, supervision and performance of the group audit. We remain solely responsible for our audit opinion.

We communicate with those charged with governance regarding, among other matters, the planned scope and timing of the audit and significant audit findings, including any significant deficiencies in internal control that we identify during our audit.

We also provide those charged with governance with a statement that we have complied with relevant ethical requirements regarding independence, and to communicate with them all relationships and other matters that may reasonably be thought to bear on our independence, and where applicable, actions taken to eliminate threats or safeguards applied.

From the matters communicated with those charged with governance, we determine those matters that were of most significance in the audit of the consolidated financial statements and the parent company financial statements of the current period and are therefore the key audit matters. We describe these matters in our auditor's report unless law or regulation precludes public disclosure about the matter.

Report on compliance with the ESEF Regulation

As part of our audit of the Consolidated Financial Statements and Parent Company Financial Statements of Orphazyme A/S, we performed procedures to express an opinion on whether the annual report of Orphazyme

A/S for the financial year 1 January – 31 December 2022 with the file name 54930025OZD2GGSQ7L42-2022-12-31-en is prepared, in all material respects, in compliance with the Commission Delegated Regulation (EU) 2019/815 on the European Single Electronic Format (ESEF Regulation) which includes requirements related to the preparation of the annual report in XHTML format and iXBRL tagging of the Consolidated Financial Statements including notes.

Management is responsible for preparing an annual report that complies with the ESEF Regulation. This responsibility includes:

- The preparing of the annual report in XHTML format;
- The selection and application of appropriate iXBRL tags, including extensions to the ESEF taxonomy and the anchoring thereof to elements in the taxonomy, for all financial information required to be tagged using judgement where necessary;
- Ensuring consistency between iXBRL tagged data and the Consolidated Financial Statements presented in human readable format; and
- For such internal control as Management determines necessary to enable the preparation of an annual report that is compliant with the ESEF Regulation.

Our responsibility is to obtain reasonable assurance on whether the annual report is prepared, in all material respects, in compliance with the ESEF Regulation based on the evidence we have obtained, and to issue a report that includes our opinion. The nature, timing and extent of procedures selected depend on the auditor's judgement, including the assessment of the risks of material departures from the requirements set out in the ESEF Regulation, whether due to fraud or error. The procedures include:

- Testing whether the annual report is prepared in XHTML format;
- Obtaining an understanding of the company's iXBRL tagging process and of internal control over the tagging process;
- Evaluating the completeness of the iXBRL tagging of the Consolidated Financial Statements including notes;
- Evaluating the appropriateness of the company's use of iXBRL elements selected from the ESEF taxonomy and the creation of extension elements where no suitable element in the ESEF taxonomy has been identified;
- Evaluating the use of anchoring of extension elements to elements in the ESEF taxonomy; and
- Reconciling the iXBRL tagged data with the audited Consolidated Financial Statements.

In our opinion, the annual report of Orphazyme A/S for the financial year 1 January – 31 December 2022 with the file name 54930025OZD2GGSQ7L42-2022-12-31-en is prepared, in all material respects, in compliance with the ESEF Regulation.

Copenhagen, 25 April 2023

EY Godkendt Revisionspartnerselskab

CVR no. 30 70 02 28

Hans B. Vistisen
State Authorised
Public Accountant
mne23254

Anders Roe Eriksen
State Authorised
Public Accountant
mne46667

Other Information

Disclaimer

This annual report contains certain "forward-looking statements", within the meaning of the "safe harbor" provisions of the U.S. Private Securities Litigation Reform Act of 1995, including, without limitation, statements about the Company's financial outlook, the class action lawsuit in the United States and future prospects. Although the Company believes its expectations are based on reasonable assumptions, all statements other than statements of historical fact included in this Annual Report about future events are subject to (i) change without notice and (ii) factors beyond the Company's control. These statements may include, without limitation, any statements preceded by, followed by, or including words such as "target," "believe," "expect," "aim," "intend," "may," "anticipate," "estimate," "plan," "project," "will," "can have," "likely," "should," "would," "could", and other words and terms of similar meaning or the negative thereof. Forward-looking statements are subject to inherent risks and uncertainties beyond the Company's control that could cause the Company's actual results, performance, or achievements to be materially different from the expected results, performance, or achievements expressed or implied by such forward-looking statements, including the risks and uncertainties that are described in the Risk Management section of this Annual Report. Except as required by law, the Company assumes no obligation to update these forward-looking statements publicly, or to update the reasons actual results could differ materially from those anticipated in the forward-looking statements, even if new information becomes available in the future.

Orphazyme A/S
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Copenhagen N, Denmark