ANNUAL REPORT
2021

Orphazyme A/S
Company registration no.: 32266355
Ole Maaløes Vej 3, DK-2200
Copenhagen N, Denmark

Contents

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) following an oral explanation.

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

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

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

Disclaimer

This annual report may contain certain forward-looking statements, including in respect of the company's outlook for full-year 2022 and financial position as of year-end 2022. Although the Company believes its expectations are based on reasonable assumptions, all statements other than statements of historical fact included in this company announcement 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 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.

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Key Figures & Ratios

TDKK	2021	2020	2019	2018	2017
Statement of Profit and Loss and other comprehensive income					
Net revenue	36,193	0	0	0	0
Research and development expenses	(329,980)	(361,284)	(285,413)	(196,525)	(99,048)
General and administrative expenses	(339,516)	(247,250)	(50,541)	(35,127)	(31,994)
Operating Loss	**(633,303)**	**(608,534)**	**(335,954)**	**(231,652)**	**(131,042)**
Net Financial items	1,823	(26,627)	(7,043)	(3,448)	(662)
Loss Before Tax	**(631,480)**	**(635,161)**	**(342,997)**	**(235,100)**	**(131,704)**
Income tax benefit	4,941	1,915	5,500	5,500	5,500
Net Loss for the period	**(626,539)**	**(633,246)**	**(337,497)**	**(229,600)**	**(126,204)**
Total comprehensive loss	**(626,841)**	**(632,641)**	**(337,430)**	**(229,558)**	**(126,204)**
loss per share, basic (DKK)	-17.94	-22.32	-16.87*	-11.5*	-10.43
* adjusted retrospectively, see Note 4.3 in consolidated financial statements.					
Statement of financial position					
Intangible Assets	2,152	12,454	10,539	10,744	9,853
Right-of-use assets	5,434	14,859	13,903	0	0
Property, plant, and equipment	2,985	4,687	3,685	1,940	1,851
Other non-current assets	3,714	6,829			
Total non-current assets	**14,285**	**38,829**	**32,529**	**17,965**	**14,864**
Cash	102,255	726,929	123,588	394,706	631,735
Other current assets	56,689	56,735	19,137	28,678	16,218
Total assets	**173,229**	**822,493**	**180,754**	**441,349**	**662,817**
Share capital	34,952	34,698	19,984	19,939	19,928
Total equity	9,339	620,525	52,969	388,249	615,702
Non-current borrowings	2,482	23,830	51,606	0	0
Non-current lease liabilities	3,925	9,877	9,813	0	0
Other non-current assets	28,391	1,634	0	0	0
Total current liabilities	129,092	166,627	65,988	52,995	47,115
Cash flow statement					
Cash flow from operating activities	(602,571)	(539,076)	(326,818)	(234,764)	(54,724)
Cash flow from investing activities	46	(5,101)	(3,285)	(2,346)	(238)
Cash flow from financing activities	(30,344)	1,159,422	58,939	0	1,300
Other					
Share Price (DKK)	17.16	67.10	72.40	43.35	76.00
Total outstanding shares	34,952,241	28,514,047	19,984,799	19,939,564	19,928,184
Market captalilazation (MDKK)(1)	599.8	1,913.3	1,446.9	864.4	1,514.5
Equity ratio (2)	5%	75%	29%	88.0%	92.9%
Equity per share (DKK)(3)	0.27	21.76	2.65	19.47	30.90
Average number of employees	130	117	74	46	26
Number of employees at the end of the year	62	141	86	57	34

(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

Management Report

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

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

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

In June 2021, we received a Complete Response Letter (CRL) from the U.S. Food and Drug Administration (FDA) for our New Drug Application (NDA) for arimoclomol in NPC. The CRL outlined the need for additional qualitative and quantitative evidence to further substantiate the validity and interpretation of the 5-domain NPC Clinical Severity Scale (NPCCSS) and, in particular, the swallow domain, in the context of the FDA's preferred and recommended statistical approach. Further, the FDA noted that additional data would be needed to supplement confirmatory evidence beyond the single Phase 2/3 trial for NPC.

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

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

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

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

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

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

Following initiation of the in-court restructuring proceedings, we voluntarily delisted our American Depositary Shares (ADSs) representing Orphazyme's ordinary shares from Nasdaq Global Select Market in the U.S. and filed a Form 25 with the Securities and Exchange Commission (SEC) on March 21, 2022. The delisting became effective on March 31, 2022 and deregistration of the ADSs is expected to become effective approximately 90 days after the filing of the Form 25 with the SEC.

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

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

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

As substantially all of our assets and business activities have been sold to KemPharm, we have limited ongoing operational business activities. Considering this, we announced that Stephanie Okey, Carrolee Barlow and Martin Bonde stepped down from their positions as members of the Board of Directors as of May 23, 2022 with the view to reduce the number of members on our Board of Directors.

I would like to extend my sincere thanks to our stakeholders, including our shareholders, partners, collaborators, employees, patients and the NPC community, who have helped us make important scientific advances in neurodegenerative rare diseases and in developing arimoclomol. I am delighted that KemPharm will continue to move arimoclomol forward in the hope of making it available for NPC patients, and I wish them, and the Orphazyme colleagues joining the company, every success in their mission to bring a new therapeutic option to people living with NPC.

Anders Vadsholt

Chief Executive Officer & Chief Financial Officer

2021 Financial Review

Income statement

The net loss for the financial year ended December 31, 2021 was DKK 626.5 million compared to DKK 633.2 million for the same period in 2020. The slight decrease in net loss was primarily driven by a restructuring of the group during the year, with employee redundancies and termination of onerous contracts.

Net revenue

Net revenue for the year ended December 31, 2021 was DKK 36.2 million compared to DKK 0 million for the year ended December 31, 2020. The increase was driven by the sale of arimoclomol under the remunerated early access compassionate use program (former nATU) in France, that the Group entered during 2021.

R&D expenses

Research and development expenses for the year ended December 31, 2021 were DKK 330.0 million compared to DKK 361.3 million for the year ended December 31, 2020. The decrease of DKK 31.3 million was primarily driven by a restructuring of the group during the year, with employee redundancies and termination of onerous contracts.

G&A expenses

General and administrative (G&A) expenses for the year ended December 31, 2021 were DKK 339.5 million compared to DKK 247.3 million for the year ended December 31, 2020. The increase of DKK 92.2 million was primarily due to the build-up of our commercial organization the first half of 2021 in preparation for potential approval of arimoclomol for the treatment of NPC, as well as expenses related to increased costs for being a listed company in the U.S. In the second half of 2021 the increase was primarily due to costs related to restructuring activities.

Net financial items

Net financial income for the year ended December 31, 2021 was DKK 1.8 million compared to net financial expense DKK 26.6 million for the year ended December 31, 2020. The increase of financial income DKK 28.4 million was mainly related to an increase in net foreign currency exchange gains of DKK 23.6 million primarily due to an increase in the US dollar versus DKK.

Income tax benefit

Income tax benefit for the year ended December 31, 2021 was DKK 4.9 million compared to DKK 1.9 million for the year ended December 31, 2020. Income tax benefit for the two periods include a tax credit for research and development costs at the applicable tax rate under the Danish Corporate Income Tax Act. The amount of the tax benefit in the year 2021 was reduced by an income tax expense in our subsidiaries in the U.S. and Switzerland. Our corporate income tax rate in Denmark was 22%. However, for the year ended December 31, 2021 and 2020, we did not recognize any deferred tax assets in Denmark considering uncertainties surrounding their potential utilization.

Statement of financial position

Cash: As of December 31, 2021, Orphazyme had cash of DKK 102.3 million compared to DKK 726.9 million as of December 31, 2020. The decrease in cash was primarily due to the negative cash flow from the operating activities in 2021.

Equity: As of December 31, 2021, total equity amounted to DKK 9.3 million compared to DKK 620.5 million as of December 31, 2020. The decrease mainly reflects the net loss in 2021.

Cash flows

Cash flow from operating activities:
Net cash used in operating activities for the period ended December 31, 2021 was DKK 602.6 million compared to DKK 539.1 million in the year ended December 31, 2020. The increased use of cash was primarily attributable to the payments for the clinical activities and termination of the onerous contracts.

Cash flow from investing activities:
Net cash provided from investing activities for the period ended December 31, 2021 was DKK 46 thousand compared to net cash used of DKK 5.1 million in the year ended December 31, 2020. The decrease in use of cash in 2021 was mainly due to the high investment in the acquisition of the ERP system in 2020.

Cash flow from financing activities:
Net cash used by financing activities for the period ended December 31, 2021 was DKK 30.3 million compared to net cash provided of DKK 1,159.4 for the year ended December 31, 2020. The decrease of net cash provided DKK 1,189.8 million reflects the net proceeds from our directed issue and private placement in February 2020 and from our global offering and U.S. listing in October 2020.

Outlook

For the full-year 2022 we anticipate an operating profit in the range DKK 10 – 30 million. We expect to end 2022 with more than DKK 30 million in cash and equivalents. Following completion of the sale of substantially all the Company's assets and business activities to KemPharm in May 2022, Orphazyme has limited ongoing operational business activities and only two employees. There are inherent risks and uncertainties in our Outlook for 2022 given the recent closing of the KemPharm transaction, transfer of operating activities from Orphazyme to KemPharm and our future prospects.

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. In addition, deregistration of the ADSs is expected to become effective approximately 90 days after the filing of the Form 25 with the SEC, which occurred on March 21, 2022.

We conduct our communications in accordance with the applicable rules and regulations required under Danish, EU and U.S. law, including as set forth by the Danish Financial Supervisory Authority, and the U.S. Securities and Exchange Commission ("SEC").

Ownership and share capital

As of December 31, 2021, the number of registered Shareholders totaled 12,191 shareholders holding a total of 33,142,640 shares, representing 94.82% of the total share capital of 34,952,241. As of December 31, 2021, 19,886,184 shares were represented by American Depositary Shares (ADS).

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, 2021 the Company has lost more than 50% of its share capital. At the Annual General Meeting to be held on June 29, 2022, the Board of Directors will give an account of the Group's financial position. Please refer to note 1.6 for further information.

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.

Holders of ADSs evidenced by American Depositary Receipts ("ADRs") do not have shareholder rights and may not be able to exercise their right to vote the shares underlying the ADSs.

ADS holders have the contractual rights of an ADS holder, as provided in the deposit agreement among the Company, the depositary and holders and beneficial owners of ADSs from time to time. As a result, ADS holders may only exercise voting rights with respect to the shares underlying the ADSs in accordance with the provisions of the de- posit agreement.

Orphazyme voluntarily delisted the Company's ADSs representing its shares from Nasdaq Global Select Market in the US, effective March 31, 2021. In addition, deregistration of the ADSs is expected to become effective approximately 90 days after the filing of the Form 25 with the SEC, which occurred on March 21, 2022.

In connection with Orphazyme's delisting of the ADSs representing its ordinary shares on Nasdaq Global Select Market in the US, the Bank of New York Mellon, as depositary, has notified holders of ADSs evidenced by

American Depositary Receipts (ADRs) that the Deposit Agreement will be terminated and as a result, the existing ADR facility will be terminated effective at 5:00 PM (Eastern Time) on July 6, 2022. Under the terms of the Deposit Agreement, ADR Holders have until at least July 11, 2022 to surrender their Orphazyme ADRs for delivery of the underlying shares. If ADR Holders surrender ADRs for delivery of the underlying shares, ADR Holders must pay a cable fee of $17.50, a cancellation fee of up to $0.05 per ADRs surrendered and any applicable U.S. or local taxes or governmental charges. Payment should be made payable to the Depositary. Subsequent to July 11, 2022 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)

LSP V Cooperatieve U.A., Johannes Vermeer, Plein 9, 1071 DV Amsterdam, Netherlands.

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 intends to comply with the Recommendations on Corporate Governance in all material respects, however, given Orphazyme's current situation and focus, the Company has opted to deviate from the recommendations in the following areas:

- Orphazyme has decided to only publish annual reports and half-yearly financial reports;
- the Company has not adopted a corporate social responsibility policy;
- the Company has not adopted a tax policy;
- the annual report does not include information on individual board members' participation in board meetings and committee meetings;
- the annual report does not include information on the board committee's most significant activities and number of board committee meetings held in the past year;
- the general conclusion of the latest evaluation of the Board of Directors is not described in the annual report, but is accounted for by the Chairman at the annual general meeting and
- share-based remuneration is offered to the Board of Directors and has a maturity of one year from the date of allocation.

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 in the *Investors & Media* section of our website through this link: 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 2021 due to the unfavorable events experienced in 2021. 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
Georges Gemayel	Chairman	Independent	2012	2022
Bo Jesper Hansen	Deputy Chairman	Independent	2010	2022
Andrew Mercieca	Member	Independent	2022	2023

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 has decided to discontinue the Science Committee with effect from May 2022.

Audit Committee

Members

- Andrew Mercieca (Chairman)
- Bo Jesper Hansen
- Georges Gemayel

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 three 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. The members 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.
- Anders Vadsholt, in his capacity as the CEO and CFO and 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)
- Georges Gemayel

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. The members meet such independence requirements.

Remuneration Committee

Members

- Bo Jesper Hansen (Chairman)
- Georges Gemayel
- Andrew Mercieca

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 three 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. The members meet such independence.

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.

Pervasive risk: Lack of sufficient funding, business operations and human resources could impact our future prospects.

Risks: Following the sale of substantially all our assets and business activities to KemPharm, which completed on May 31, 2022, we have limited operating business activities and only two employees. As such our future prospects are uncertain.

Pervasive risk: Non-compliance with legislation and industry standards.

Risks: 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.

Our actions to mitigate the risks: In recent years, we have strengthened our IT-security procedures in order to reduce the risk of cybercrime. We have in the past had internal training requirements for all employees and contracting external suppliers, to ensure they have adequate measures in place to comply with relevant regulatory requirements.

Pervasive risk: 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 Related to the Sale of Assets

In May 2022, 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.

Risks related 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 business.

Our 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. Orphazyme does not believe these claims have merit and intends to vigorously defend itself against the lawsuit.

Corporate Social Responsibility

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

In 2021, our focus at Orphazyme was on 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.

Below we describe Orphazyme's most significant Corporate Social Responsibility (CSR) risks and how we handled them for the 2021 financial year. As of the date of this annual report, the Company has limited ongoing business activities and only two employees which we believe significantly reduces the company's CSR risks at this time.

† Our future CSR activities will be commensurate with the size and limited operations of the Company. We will continue to strive to uphold our values and responsibilities, where possible, as we execute our strategy.

2021 CSR reporting areas

Human Rights

Risk

- Limited: Orphazyme conducts business in a highly regulated industry.

Actions

- Continued to respect internationally declared human rights and did not employ child labor.
- Trained employees in relation to whistleblower policy.
- Continued to support Board diversity, electing one additional female member to the board of directors during 2021.

Policies in place

- Diversity policy to increase diversity among members of the Board of Directors and other management levels.
- Whistleblower policy to enable reporting of potential violations.

Results

- No diversity related incidents reported.
- No human rights violations reported.
- Balanced employee composition: 58% female, 42% male.
- Leadership (director level and above) 53% / 47% gender balance.

- Reached board of directors diversity target. As at year end 2021 there were 2 female board members, equating to 40% of the board. As of the date of this annual report, the Board of Directors is comprised of three members, none of which are women.

Future Plans†

- Continue to support and respect internationally declared human rights and will not employ child labor.

Anti-Corruption & Bribery

Risk

- Limited: Orphazyme conducts business in a highly regulated industry. 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.

Actions

- Anti-corruption and bribery training in onboarding of all employees
- Legal & Compliance training refreshers, including anti-corruption and bribery
- Expanded our commitment to anti-bribery & corruption and implemented a global policy on good promotional practice, global policy on interactions with healthcare professionals and healthcare organizations and global policy on fee-for-service engagement of healthcare professionals, healthcare organizations and patient organizations..

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 2021.

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 (direct risk): We have a modest number of employees and use external suppliers for certain activities such as clinical trials and product manufacturing. The Company conducts its business in a highly regulated industry and follows applicable rules on hazardous substances. As such, use of hazardous substances is connected with a very low and controlled risk.

Actions

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

Policies in place

- Considering the business of the Company, Orphazyme's general potential impact on the environment and climate and the impact of the climate on Orphazyme's business is viewed as minimal. As such, specific environment and climate policies have not been developed at this time.

Results

- Continued to keep records of all accidents.
- No records of spill of hazardous substances.

- Continued to focus on efficient management of office materials.

Future Plans †

- Orphazyme is no longer active in research and development activities associated with the use of hazardous substances and such risks have accordingly been minimized. Further, with only limited operating activities and employees, the general potential impact on the environment and climate and the impact of the climate on Orphazyme's business is viewed as minimal and no specific environment and climate policies are planned at this time.

Social / Employees

Risk

- Limited: Orphazyme conducts business in a highly regulated industry. We believe a diverse, skilled, and healthy workforce is crucial to the success of Orphazyme and our ability to serve patients and the rare disease community. We value diversity in gender, age, ethnicity, nationality, religion, education, sexual orientation, work history, perspectives, opinions, and skills at all levels of our business.

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 mandatory and ongoing education relating to workplace safety.
- 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.
- Recruitment process focused on balancing representation in our teams.
- Fostered an inclusive workplace committed to freedom from discrimination, harassment, and bullying.
- Continued our employee initiatives including all staff townhall meetings, social events, hybrid flexible working, and fostering an open and trusting work environment. Characterized by psychological safety.
- Provided frequent updates to keep staff healthy and safe during COVID pandemic.
- 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.
- Maintained a diverse workforce consisting of 58% women and 42% men.
- Leadership (director level and above) 53% / 47% gender balance.
- Workplace assessment survey: 92% satisfaction with physical work environment; 82% satisfaction with psychosocial work environment.
- Culture survey confirmed that Orphazyme has a clear and meaningful purpose.
- Established a resilient culture centered on trust and collaboration.

Future Plans†

- As of the date of this annual report, the Company has limited ongoing business activities and only two employees. 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 diverse and inclusive workplace, as we execute our strategy.

Business Ethics

Risk

- Limited: Orphazyme is committed to conducting business in a manner that ensures that no Orphazyme representatives are influenced by undue personal interests and that the Company's directors, officers, and employees adhere to specific guidelines in the conduct of the Company's business.

Actions

- Continued to train existing employees in business ethics and conflict of interest through our risk and compliance system.
- Continued to include business ethics and conflict of interest in onboarding training of all new employees.

Policies in place

- Conflict of interest policy.
- Code of business conduct & ethics.

Results

- High awareness on ethical business conduct by Orphazyme representatives.
- No instances of unethical behaviors escalated to senior management.

Future Plans[†]

- Continue our commitment to conducting our business ethically, fairly, and with integrity.

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 now has limited operational business activities, particularly in the area of clinical trials where the importance of data privacy and regulations are paramount, we will evaluate our practices to ensure they align with the new statutory requirements set forth in Section 99d of the Danish Financial Statements Act.

Diversity metrics

In July 2021, we implemented a restructuring plan, reducing our workforce by two-thirds while retaining core expertise to enable us to achieve our goal of gaining approval for arimoclomol in NPC.

As of December 31, 2021, we 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.

Overall, most employees at year-end 2021 were in the areas of finance, legal, IT and administration (25%), clinical (22%) and commercial/pre-launch (20%).

Since the end of 2021, there have been some voluntary employee departures and in March 2022 Orphazyme entered in-court restructuring proceedings, resulting in a reduction in its workforce of approximately 50%.

As of the date of this annual report, there are two employees, including Executive Management.

The Board of Directors also saw some changes during 2021, with the departure of three male members and one female member. At year-end 2021, the Board of Directors was comprised of five members, of which two (40%) were women. In February 2022, Andrew Mercieca was appointed to the Board of directors, taking the Board to six members, of which two (33%) are women. The Board of Directors was further changed in May 2022, with the departure of Stephanie Okey, Carrolee Barlow and Martin Bonde. As of the date of this annual report, the Board of Directors is comprised of three members, none of which are women.

Covid-19

The COVID-19 pandemic continued to restrict the world's activities during 2021. We supported our employees with work-from-home and hybrid-working, IT support, COVID-19 testing, and various other initiatives aimed at keeping our business operating successfully and our team healthy and engaged. There was no material impact directly related to COVID-19 on the Group's consolidated financial statements, including the judgements and estimates applied.

Board of Directors and Executive Management

Board of Directors

Georges Gemayel, Chairman of the Board

- Member since: 2012 (Chairman, 2014)
- Born in: 1960
- Nationality: American
- Committees: Audit; Remuneration; Nomination

Special competencies: Dr Gemayel has significant management and executive experience from the global pharmaceutical industry. He holds a Master's and PhD in Pharmacology from Paris-Sud University and a Docteur d'Exercice en Pharmacie, St. Joseph University.

Current positions: Interim CEO and Chairman of the Board of Gemini Therapeutics, Chairman of the Board of Dynacure, Enterome SA, and OxThera AB, and a member of the Board of Directors of Supernus Pharmaceuticals Inc. (publ).

Bo Jesper Hansen, Deputy Chairman of the Board

- Member since: 2010 (Deputy Chairman, 2017)
- 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: Chairman of the Board of Laborie Inc., Deputy Chairman of SOBI AB, member of the Board of directors Innoventa Medica ApS, and Reapplix 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.

Andrew Mercieca

- Member since: 2022
- Born in: 1966
- Nationality: British
- Committees: Audit (Chair), Remuneration

Special competencies: Andrew has extensive experience in International Finance and across Life Science and Technology sectors in corporate, private and non-profit organizations. He has an exceptional track record of driving business performance, changing and optimizing finance functions, leveraging the use of technology, building strong teams, and ensuring agile and effective risk management. He holds a BA in Accounting and Finance from the University of Portsmouth and is a fellow of the Institute of Chartered Accountants in England & Wales.

Current positions: Director of Octant Limited, providing Financial Consultancy services.

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.

Anders has 25+ years' experience from biotech and corporate finance. Previously he was at Topotarget, BankInvest Biomedical Venture and Carnegie Investment Bank.

Anders is currently owner of Alpha Healthcare Investments ApS.

Anders holds a BSc in Corporate Law from the University of Aalborg, an MBA in Finance and Strategy from the University of Melbourne, and an MSc in Corporate Law and Economics from Copenhagen Business School.

Corporate information

Commercial bankers

Danske Bank
Holmens Kanal 2-12
DK-1092 Copenhagen K

Nordea
Vesterbrogade 8
DK-1620 Copenhagen

BNY Mellon
240 Greenwich St New York, NY
10286 USA

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*.

2021 Consolidated Financial Statements

CONSOLIDATED STATEMENTS OF PROFIT OR LOSS AND OTHER COMPREHENSIVE INCOME

For the years ended December 31,

DKK 000, except per share and share data	Note	2021	2020	2019
Net revenue	2.1	**36,193**	—	—
Research and development expenses	2.2, 2.3	(329,980)	(361,284)	(285,413)
General and administrative expenses	2.4	(339,516)	(247,250)	(50,541)
Operating loss		**(633,303)**	**(608,534)**	**(335,954)**
Financial income	2.7	12,432	2,444	316
Financial expenses	2.7	(10,609)	(29,071)	(7,359)
Loss before tax		**(631,480)**	**(635,161)**	**(342,997)**
Income tax benefit	2.8	4,941	1,915	5,500
Net loss for the year		**(626,539)**	**(633,246)**	**(337,497)**
Items that will be reclassified subsequently to the Statement of Profit or Loss:				
Exchange difference from translation of foreign operations		(302)	605	67
Total comprehensive loss		**(626,841)**	**(632,641)**	**(337,430)**
Weighted-average shares outstanding		34,924,702	28,366,469	20,024,692
Loss per share, basic and diluted (DKK)	4.3	(17.94)	(22.32)	(16.87)

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

CONSOLIDATED STATEMENTS OF FINANCIAL POSITION

As of December 31,

DKK 000 ASSETS	Note	2021	2020
Non-current assets			
Intangible assets	3.1	2,152	12,454
Right-of-use assets	3.2	5,434	14,859
Property, plant, and equipment	3.3	2,985	4,687
Corporation tax receivable	2.8	2,750	2,750
Deferred tax assets	2.8	—	2,065
Prepayments and deposits	3.4	964	2,014
Total non-currents assets		**14,285**	**38,829**
Current assets			
Corporation tax receivable	2.8	7,229	5,500
Trade receivables	3.5	29,268	—
Prepayments and other receivables	3.4	20,192	51,235
Inventory	3.6	—	—
Cash	3.8	102,255	726,929
Total current assets		**158,944**	**783,664**
Total assets		**173,229**	**822,493**

EQUITY AND LIABILITIES	Note	2021	2020
Equity			
Share capital	4.2	34,952	34,698
Share premium		2,082,486	2,082,254
Other reserves		2,899	6,494
Accumulated deficit		(2,110,998)	(1,502,921)
Total equity		**9,339**	**620,525**
Non-current liabilities			
Borrowings	3.7	2,482	23,830
Lease liabilities	3.2	3,925	9,877
Discount and rebate liabilities	3.7	28,293	—
Other non-current liabilities	3.7	98	1,634
Total non-current liabilities		**34,798**	**35,341**
Current liabilities			
Provisions		—	—
Borrowings	3.7	30,983	33,349
Lease liabilities	3.2	2,578	3,657
Trade payables and accruals	3.7	57,524	72,135
Tax payables	2.8	584	4,159
Discount and rebate liabilities	3.7	7,900	—
Other liabilities	3.7	29,523	53,327
Total current liabilities		**129,092**	**166,627**
Total equity and liabilities		**173,229**	**822,493**

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

CONSOLIDATED STATEMENTS OF CHANGES IN SHAREHOLDERS' EQUITY

DKK 000

	Notes	Share capital	Share premium	Other reserves: Foreign currency translation reserve	Other reserves: Share-based compensation – acquisition of intangible assets	Accumulated deficit	Total
Balance as of December 31, 2018		**19,939**	**924,021**	**42**	**9,070**	**(564,823)**	**388,249**
Net loss for the year						(337,497)	(337,497)
Other comprehensive income (loss)				67		—	67
Total other comprehensive income (loss)		**—**	**—**	**67**	**—**	**(337,497)**	**(337,430)**
Transactions with owners:							
Capital increase in connection with issuance of bonus shares	3.1	26			(1,197)	1,171	—
Issuance of Matching Shares, net of costs	2.6	19					19
Share-based compensation expense	2.6					2,131	2,131
Total transactions with owners		**45**	**—**	**—**	**(1,197)**	**3,302**	**2,150**
Balance as of December 31, 2019		**19,984**	**924,021**	**109**	**7,873**	**(899,018)**	**52,969**
Net loss for the year						(633,246)	(633,246)
Other comprehensive income				605			605
Total other comprehensive income (loss)		**—**	**—**	**605**	**—**	**(633,246)**	**(632,641)**
Transactions with owners:							
Capital increase in connection with issuance of bonus shares	3.1	21			(2,094)	2,073	—
Capital increase in connection with exercise of RSUs	4.2	13	717				730
Capital increase related to directed issue and private placement	4.2	7,033	738,458				745,491
Transaction costs related to directed issue and private placement			(51,243)				(51,243)
Capital increase in connection with US listing	4.2	7,616	526,918				534,534
Transaction costs related to the US listing			(56,616)				(56,616)
Issuance of Matching Shares, net of costs	2.6	31					31
Share-based compensation expense	2.6					27,270	27,270
Total transactions with owners		**14,714**	**1,158,233**	**—**	**(2,094)**	**29,343**	**1,200,196**
Balance as of December 31, 2020		**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**

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

CONSOLIDATED STATEMENTS OF CASH FLOWS

For the years ended December 31,

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

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

Notes to Financial Statements

SECTION 1 Basis of preparation and significant accounting policies

1.1 CORPORATE INFORMATION

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

The Company is headquartered in Copenhagen, Denmark and is publicly traded on Nasdaq Copenhagen. In September 2020, the Company listed American Depositary Shares (ADSs) on the Nasdaq Global Select Market. In March 2022, Orphazyme A/S initiated voluntary delisting of the ADSs representing its shares from Nasdaq Global Select Market in the US. It is expected that deregistration of the ADSs will become effective approximately 90 days after the filing of the Form 25 with the SEC, which occurred on March 21, 2022.

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

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

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

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

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

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

Information on COVID-19

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

1.2 BASIS OF PREPARATION

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

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 loss for the year and actual cash receipts and payments are included.

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

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

Segment information

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

1.4 SIGNIFICANT ACCOUNTING ESTIMATES AND JUDGEMENTS

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

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

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

1.5 NEW IFRS STANDARDS APPLICABLE TO THE GROUP

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

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

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

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

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

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

On 28 May 2020, the IASB issued Covid-19-Related Rent Concessions - amendment to IFRS 16 Leases. The amendments provide relief to lessees from applying IFRS 16 guidance on lease modification accounting for rent concessions arising as a direct consequence of the Covid-19 pandemic. As a practical expedient, a lessee may elect not to assess whether a Covid-19 related rent concession from a lessor is a lease

modification. A lessee that makes this election accounts for any change in lease payments resulting from the Covid-19 related rent concession the same way it would account for the change under IFRS 16 if the change were not a lease modification.

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

New IFRS Standards not yet effective

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

1.6 SIGNIFICANT EVENTS AFTER THE REPORTING PERIOD

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

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

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

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

During the restructuring proceedings, mass redundancies took place and employees were released from their duties to reduce payroll costs. The MAA filed with EMA was withdrawn in March 2022. In addition, the Company initiated voluntary delisting of the ADSs representing its ordinary shares from Nasdaq Global Select Market in the US, which became effective in March 2022. It is expected that deregistration of the ADSs will become effective approximately 90 days after the filing of the Form 25 (Notification of Removal From Listing and/or Registration under Section 12(b) of the Securities Exchange Act of 1934) with the SEC, which occurred on March 21, 2022.

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

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

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

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

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

SECTION 2 Result of the Year

2.1 NET REVENUE

§ ACCOUNTING POLICIES

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

allocating the price to the performance obligations; and 5) recognizing revenue when the performance obligations have been fulfilled.

Net revenue comprises revenue from the sale of arimoclomol for the treatment of NPC under the remunerated early access compassionate use program ("nATU") in France. An early access compassionate use program is a program giving specific patients access to a drug, which is not yet approved for commercial sale. Only drugs targeting serious or rare indications and for which there is currently no appropriate treatment are considered for early access compassionate use programs. Further, to be considered for the early access compassionate use program, the drug must have proven efficacy and safety and must either be undergoing price negotiations or seeking marketing approval.

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

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

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

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

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

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

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

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

2.2 RESEARCH AND DEVELOPMENT EXPENSES

§ ACCOUNTING POLICIES

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

Development expenses comprise costs incurred during the different phases of clinical drug development starting in phase 1, when the drug is administered to humans for the first time, through phases 2 and 3, and subsequent activities to obtain marketing authorizations, which will permit Orphazyme to eventually market and sell the drug products.

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

Clinical trial costs are a significant component of research and development expenses. The 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	2021	2020	2019
External costs	229,942	261,136	218,143
Employee costs (Note 2.5)	86,329	96,108	64,167
Depreciation, amortization and impairment (Notes 3.1, 3.2, 3.3)	13,709	4,040	3,103
Total research and development expenses	**329,980**	**361,284**	**285,413**

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

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

Estimate of research and development expenses associated with clinical trials

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

2.3 GOVERNMENT GRANTS

§ ACCOUNTING POLICIES

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

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

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

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

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

2.4 GENERAL AND ADMINISTRATIVE EXPENSES

§ ACCOUNTING POLICIES

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

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

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

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

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

2.5 EMPLOYEE COSTS

§ ACCOUNTING POLICIES

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

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

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

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

2.6 SHARE-BASED COMPENSATION COSTS

§ ACCOUNTING POLICIES

Equity-settled awards

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

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

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

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

Cash-settled awards

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

	2021	**2020**
Program		
2020 LTIP	2.0	3.0
2019 LTIP	1.7	2.7

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

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

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

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

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

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

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

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

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

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

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

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

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

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

c) Restricted Share Units (cash-settled)

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

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

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

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

In September 2020, a new RSU incentive program was announced (2020-2 RSU program), which comprised 22,993 RSUs in total, including an on-boarding grant to a new board member in accordance with the Group's remuneration policy. The 2020-2 RSU program runs in parallel with the 2020 RSU program and board members can only exercise RSUs under one of the programs.

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

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

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

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

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

Program	December 31, 2021 2021 RSUs	December 31, 2020 2020 RSUs
Fair value at valuation date (DKK 000)	16	1,913
Dividend yield (%)	—	—
Expected volatility (%)	146,9%	45.9%
Risk-free interest rate (%)	(0.63%)	(0.57%)
Expected life of awards (years)	0,75	0.50
Weighted average share price (DKK)	17.16	67.10

d) Sign-on bonus shares to former CEO

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

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

e) Sign-on bonus shares to new CEO

As part of the new CEO service agreement, Christophe Bourdon was granted 34,941 RSUs in connection with the on-boarding, which will have a total vesting period of three years (beginning on January 1, 2021) and with one third of the granted RSUs vesting on each January 1 in the following three financial years. Vesting is not conditional upon achieving any financial or non-financial targets. However, in case of termination of employment and designation as a Good Leaver, the right to receive vested RSUs will be prorated and calculated through the date of release of the Participant's work obligations. The vested RSUs can only be exercised after the expiration of the total vesting period. However, the delivery period may be extended to the

next open trading window in certain circumstances. The RSUs were valued at grant date, April 2021, using a Black Scholes option valuation model similar to the original 2021 LTIP. The valuation of the award at grant date was DKK 2.0 million. The share-based compensation expense was classified as administrative and with recognition from January 2021.

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

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

Please see Note 3.1.

Summary of share-based compensation

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

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

2.7 FINANCIAL INCOME AND FINANCIAL EXPENSES

§ ACCOUNTING POLICIES

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

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

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

2.8 INCOME TAXES

§ ACCOUNTING POLICIES

Income tax benefit includes the current benefit due from the current period's taxable loss and deferred tax adjustments. The benefit is comprised primarily of refundable tax credits for costs incurred in connection with research and development activities under the Danish Tax Credit Regime. Income tax expense relates to tax imposed on income recognized in our subsidiaries in the United States and Switzerland as a result of our transfer pricing agreements.

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

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

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

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

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

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

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

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

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

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

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

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

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

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

SECTION 3 Assets and liabilities

3.1 INTANGIBLE ASSETS

§ ACCOUNTING POLICIES

Intangible assets comprise software development costs and license rights to develop and commercialize products and are acquired separately and measured on initial recognition at cost. Software assets consist of implementation costs to get cloud computing arrangements ready for use, as long as they meet the requirements

of IAS 38, Intangible Assets. These cloud computing arrangements begin to be amortized when they are ready for intended use and are amortized over seven years.

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

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

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

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

Estimate of the fair value of licenses

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

Estimate related to recoverable amounts

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

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

CytRx Asset Purchase Agreement

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

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

The Asset Purchase Agreement further includes sales milestones and royalty payments to be made by Orphazyme based on a specified percentage of any eventual net sales of products containing one of the compounds purchased. In addition, under the terms of the Asset Purchase Agreement, the Company was assigned and became party to a royalty agreement with ALS Charitable Remainder Trust pursuant to which the Company is obliged to pay a 1% royalty to the ALS Charitable Remainder Trust on global net sales of products to treat ALS. Orphazyme has no liabilities prior to the occurrence of future sales of products and accordingly neither such liabilities nor contingent consideration have been recognized as part of the license agreement. Remaining life for this intangible asset is not possible to determine until approval of the underlying asset has been received from regulatory authorities.

License Agreement with KLSDC and UCL

In 2017, the Company entered into a license agreement with KU Center for Technology Commercialization Inc., University of Kansas, Kansas Life Sciences Development Company, Inc., ("KLSDC") and UCL Business PLC ("UCL") granting Orphazyme the right to develop and commercialize products under all data generated in the course of the on-going Phase 2/3 clinical trial on arimoclomol for the treatment of IBM worldwide. The total consideration for the license is to be paid out in bonus shares to KLSDC and UCL up to an aggregate value of USD 2.5 million (DKK 15.8 million), depending on the amount of grants awarded to KLSDC and UCL for use in the trial. At the time the license agreement was executed, Management estimated the aggregate amount of the funding to be received by KLSDC and UCL to be USD 1.6 million (DKK 10 million), which has been recognized as an intangible asset (License) with a corresponding increase in equity reserves (Share-based compensation - acquisition of intangible assets).

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

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

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

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

License Agreement with the University of Miami

In September 2019, the Company entered into an exclusive license agreement with the University of Miami. Pursuant to the exclusive license agreement, the Company was granted a global royalty-bearing,

exclusive license to all data, know-how, inventions and technology generated by the University of Miami and certain other institutions in a Phase 2 clinical trial of arimoclomol in ALS with the A4V SOD1 mutation to research, develop, make, use or sell certain pharmaceutical products or processes containing arimoclomol.

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

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

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

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

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

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

3.2 LEASES

§ ACCOUNTING POLICIES

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

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

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

Right-of-use assets

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

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

Lease liabilities

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

Short-term leases and leases of low-value assets

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

Lease modifications

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

Estimate relating to the incremental borrowing rate

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

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

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

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

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

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

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

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

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

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

3.3 PROPERTY, PLANT, AND EQUIPMENT

§ ACCOUNTING POLICIES

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

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

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

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

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

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

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

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

3.4 PREPAYMENTS, DEPOSITS, AND OTHER RECEIVABLES

§ ACCOUNTING POLICIES

Prepayments

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

Deposits

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

Other receivables

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

Sales tax

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

- *when the sales tax incurred on a purchase of assets or services is not recoverable from the taxation authority, in which case, the sales tax is recognized as part of the cost of acquisition of the asset or as part of the expense item, as applicable*

- *when receivables and payables are stated with the amount of sales tax included*

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

Estimate of prepayments related to clinical trial development costs

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

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

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

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

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

Current prepayments and other receivables are specified below:

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

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

3.5 TRADE RECEIVABLES

§ ACCOUNTING POLICIES

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

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

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

3.6 PRE-LAUNCH INVENTORY

§ ACCOUNTING POLICIES

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

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

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

Pre-launch inventory intended for commercial sale

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

3.7 FINANCIAL ASSETS AND LIABILITIES

§ ACCOUNTING POLICIES

Financial assets

Initial recognition and measurement

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

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

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

Subsequent measurement

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

Financial asset impairment

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

Financial liabilities

Borrowings

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

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

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

Trade payables and accruals

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

Other liabilities

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

Discount and rebate liabilities

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

Estimate of accruals related to clinical trial development costs

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

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

The Group's financial assets include mainly cash (Note 3.8). The Group has no derivative financial assets nor has there been a change in classification of a financial asset after initial recognition and measurements

as discussed herein. Financial assets are not acquired for trading or speculative purposes, nor has the Group placed any assets as security for loans at either December 31, 2021 or 2020.

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

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

Kreos Debt Facility

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

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

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

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

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

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

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

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

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

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

Maturities of financial liabilities

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

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

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

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

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

			Non-cash changes				
DKK 000	December 31, 2020	Cash flows	Disposals	Adjustments and modifications	Accumulated interest	Exchange rate adjustments	December 31, 2021
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**

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

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

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

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

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

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

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

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

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

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

3.8 CASH

§ ACCOUNTING POLICIES

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

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

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

3.9 COMMITMENTS AND CONTINGENCIES

Pledges and securities for loans.

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

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

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

SECTION 4 Other disclosures

4.1 CAPITAL MANAGEMENT

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

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

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

4.2 EQUITY

The following table summarizes the Company's share activity:

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

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

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

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

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

The transaction consisted of a directed issue and private placement of up to 3,961,264 new shares of a nominal value of DKK 1 each (the "New Shares") and private placement of up to 3,071,673 existing shares of a nominal value of DKK 1 each (the "Existing Shares" and together with the New Shares, the "Offer Shares") at an offer price of DKK 106 per Offer Share, as determined by the Board of Directors of the Company through a

book-building process (the "Offering"). The New Shares will be issued without pre-emption rights for existing shareholders.

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

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

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

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

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

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

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

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

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

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

- Issue of new shares at market price without pre-emption rights by up to a nominal amount of DKK 6,989,767 in the period until 25 March 2026 (Article 3.1 of the Articles of Association)

- Issue of new shares against cash payment at a subscription price, which may be below the market price, to members of the Board of Directors, executives and/or employees of the Company without pre-emption rights by up to a nominal amount of DKK 1,300,000 in the period until 2 November 2022 (Article 3.2 of the Articles of Association). The authorization in article 3.2 has been partly exercised following which a nominal value of DKK 294,331 of the authorization has been issued.

- Directed issues of bonus shares, and/or directed issues of new shares effected by cash payment, to Kansas Life Sciences Development Inc. and UCL Business PLC (or entities designated by them), respectively, without pre-emption rights by up to a nominal amount of DKK 15,750,000 in the period until 2 November 2022 (Article 3.3 of the Articles of Association). The capital increase shall take place at par value (i.e. below market price). The value of such new shares to be issued can in any case not exceed a maximum of USD 2.5 million with a fixed exchange rate of DKK 6.30 per 1 USD based on the average closing price of the Company's shares on Nasdaq Copenhagen A/S for the 30 days immediately prior to the date

of issuance. The authorization in article 3.3 has been partly exercised following which a nominal value of DKK 80,643 of the authorization has been issued.

- Issue of new shares at a subscription price which may be below the market price with pre-emption rights by up to a nominal amount of DKK 25,000,000 in the period until 25 January 2025 (Article 3.4 of the Articles of Association).

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

The authorisations granted to the Board of Directors at December 31, 2021 pursuant to Articles 3.2 and 3.5 of the Articles of Association could in the aggregate only be exercised to increase the share capital by a maximum nominal amount of DKK 2,000,000.

4.3 LOSS PER SHARE

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

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

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

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

4.4 FINANCIAL RISKS

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

Liquidity Risk

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

Foreign Currency Risk

The Group's foreign currency risk is assessed to be high. The Group conducts cross border transactions where the functional currency of the respective group entity is not always used. Accordingly, future changes in the exchange rates of the DKK against the EUR, the USD, the CHF and/or the GBP will expose the Group to

currency gains or losses that will impact the reported amounts of assets, liabilities, income and expenses and the impact could be material.

Interest Rate Risk

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

Credit Risk

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

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

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

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

4.5 REMUNERATION OF BOARD OF DIRECTORS AND EXECUTIVE MANAGEMENT

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

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

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

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

cash bonus. The Executive Management is also eligible to receive an extraordinary bonus at the discretion of the Board of Directors.

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

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

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

Remuneration paid to members of the Board of Directors is made up of board and committee fees, a travel allowance, ad-hoc fees for additional services provided as described in Note 4.6, and share-based compensation related to the Restricted Share Units (RSUs) as described in Note 2.6. Board remuneration is recognized as general and administrative expenses in the Statement of Profit or Loss. The following table lists Board of Directors remuneration for the years ended December 31:

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

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

Andrew Mercieca			
Board and committee fees	24	—	—
Total	**24**	**—**	**—**
Total remuneration to the Board of Directors	**3,834**	**4,315**	**3,033**

(1) George Gemayel received ad-hoc fee in connection with a consultancy agreement for support during the interim period until Christophe Bourdon joined as CEO in April 2021

(2) Rémi Droller resigned effective as of June 30, 2021. At the same time all historical board fee was reversed.

4.6 RELATED PARTIES

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

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

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

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

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

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

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

4.7 FEES TO AUDITORS

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

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

Audit services

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

Audit-Related services

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

Other assistance

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

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

Pre-approval policies

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

2021 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	2021	2020
Net revenue	2.1	**36,193**	**-**
Research and development expenses	2.2	(331,046)	(362,413)
General and administrative expenses	2.3	(340,316)	(258,933)
Operating loss		**(671,362)**	**(621,346)**
Financial income	2.5	11,983	2,427
Financial expenses	2.5	(10,438)	(28,985)
Loss before tax		**(633,624)**	**(647,905)**
Income tax benefit	2.6	5,500	5,500
Net loss for the year		**(628,124)**	**(642,405)**
Items that will be reclassified subsequently to the Statement of Profit or Loss:			
Exchange difference from translation of foreign operations		(57)	(54)
Total comprehensive loss		**(628,181)**	**(642,459)**

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

STATEMENTS OF FINANCIAL POSITION

DKK 000 ASSETS	Note	2021	2020
Non-current assets			
Intangible assets	2.7	2,152	10,681
Right-of-use assets	2.8	3,726	12,862
Property, plant, and equipment	2.9	2,205	3,770
Corporation tax receivable		2,750	2,750
Investment in subsidiaries	2.10	-	139
Prepayments and deposits	2.11	593	1,666
Total non-currents assets		**11,426**	**31,805**
Current assets			
Corporation tax receivable	2.6	5,500	5,500
Trade receivables		29,268	-
Prepayments and other receivables	2.11	25,056	262,173
Inventory		-	-
Cash	2.13	85,693	487,322
Total current assets		**145,517**	**754,995**
Total assets		**156,943**	**786,800**

EQUITY AND LIABILITIES	Note	2021	2020
Equity			
Share capital		34,952	34,698
Share premium		2,082,487	2,082,255
Other reserves		2,324	5,674
Accumulated deficit		(2,120,884)	(1,511,221)
Total equity		**(1,121)**	**611,406**
Non-current liabilities			
Borrowings	2.12	2,482	23,830
Lease liabilities	2.8	2,427	8,071
Discount and rebate liabilities	2.12	28,293	-
--Other non-current liabilities	2.12	98	1,634
Total non-current liabilities		**33,300**	**33,535**
Current liabilities			
Borrowings	2.12	30,983	33,349
Lease liabilities	2.8	2,122	3,276
Trade payables and accruals	2.12	49,445	49,074
Discount and rebate liabilities	2.12	7,900	-
Other liabilities	2.12	34,314	56,160
Total current liabilities		**124,764**	**141,859**
Total equity and liabilities		**156,943**	**786,800**

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

STATEMENTS OF CHANGES IN SHAREHOLDERS' EQUITY

DKK 000	Notes	Share capital	Share premium	Other reserves: Foreign currency translation reserve	Other reserves: Share-based compensation – acquisition of intangible assets	Accumulated deficit	Total
Balance as of December 31, 2019		**19,984**	**924,021**	**(51)**	**7,873**	**(898,159)**	**53,668**
Net loss for the year						(642,405)	(642,405)
Other comprehensive income (loss)				(54)			(54)
Total other comprehensive income (loss)							**(642,459)**
Transactions with owners:							
Issuance of bonus shares to KLSDC and UCL		21			(2,094)	2,073	-
Issuance of shares due to exercise of RSUs		13	717				730
Issuance of LTIP matching shares		31					
Issuance of new shares in private placement		7,033	738,458				
Transaction costs			(51,243)				
Issuance of new shares in U.S listing		7,616	526,918				
Transaction costs			(56,616)				
Share-based payments costs for the period						27,270	27,270
Total transactions with owners, 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)							**(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)**

STATEMENTS OF CASH FLOWS

DKK 000	Note	2021	2020
Net loss		(628,124)	(642,405)
Reversal of non-cash items:			
Equity-settled share-based compensation expense	2.4	12,493	20,823
Depreciation and amortization	2.7, 2.8, 2.9	14,870	4,742
Financial income	2.5	(11,983)	(2,427)
Financial expenses	2.5	10,438	28,985
Income tax benefit	2.6	(5,500)	(5,500)
Change in working capital:			
Change in prepayments, deposits, and other receivables	2.11	2,344	(32,552)
Change in trade payables, accruals, and other liabilities	2.12	12,638	49,502
Change in intercompany receivables	2.11	153,123	(211,878)
Change in intercompany payables	2.12	53,998	7,907
Exchange rate adjustments on intercompany balances		9,055	(4,044)
Cash generated by operating activities before financial items and tax		**(376,648)**	**(786,845)**
Interest paid		(6,098)	(10,639)
Interest received		0	28
Income taxes received		5,500	5,500
Cash flow from operating activities		**(377,246)**	**(791,957)**
Investment in intangible assets	2.7	(85)	(896)
Investment in property, plant and equipment	2.9	-	(1,259)
Proceeds from sale of property, plant and equipment		1,040	-
Cash flow from investing activities		**955**	**(2,155)**
Proceeds from issue of shares and exercise of RSUs		(1,184)	1,280,786
Transaction costs		-	(107,859)
Repayment of borrowing		(25,657)	(10,535)
Repayment of leasing liabilities		(3,090)	(2,913)
Cash flow from financing activities		**(29,931)**	**1,159,478**
Changes in cash and cash equivalents		**(406,222)**	**365,366**
Cash and cash equivalents at the beginning of the period		487,322	123,250
Exchange rate adjustment on cash and cash equivalents		1,857	(1,294)
Cash and cash equivalents at the end of the period		**82,957**	**487,322**

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.

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.

SECTION 2 Notes

2.1 NET REVENUE

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

2.2 RESEARCH AND DEVELOPMENT EXPENSES

DKK 000	2021	2020
External costs	229,252	256,445
Employee costs	79,388	93,261
Intercompany expenses	8,697	8,752
Depreciation and amortization	13,709	3,955
Total research and development expenses	**331,046**	**362,413**

2.3 GENERAL AND ADMINISTRATIVE EXPENSES

DKK 000	2021	2020
External costs	136,148	54,251
Employee costs	64,294	64,313
Intercompany expenses	138,713	139,582
Depreciation and amortization	1,161	787
Total research and development expenses	**340,316**	**258,933**

2.4 EMPLOYEE COSTS

DKK 000 **Employee costs**	2021	2020
Salaries	99,110	97,444
Cash bonus	10,886	21,636
Share-based compensation (Note 2.6)	12,050	19,976
Pension	8,306	8,914
Other social security contributions	1,900	2,020
Other staff costs	7,596	3,267
Total employee costs excluding board remuneration	**139,848**	**153,257**
Board remuneration (Note 4.5)	3,391	3,470
Board share-based compensation (Note 2.6 and Note 4.5)	443	847
Total employee costs	**143,683**	**157,574**
Recognized as follows in the statement of Profit or Loss		
Research and development expenses	79,388	93,261
General and administrative expenses	64,294	64,313
Total employee costs	**143,683**	**157,574**
Average number of full-time employees	99	94
Year-end number of full-time employees	49	102

2.5 FINANCIAL INCOME AND FINANCIAL EXPENSES

DKK 000	2021	2020
Interest income on cash balances		28
Foreign currency exchange gains	11,438	1,649
Gain on embedded call option	546	750
Total financial income	**11,983**	**2,427**
Interest expense on Loan Agreement	7,350	9,921
Interest expense on lease liabilities	490	499
Interest expense on cash balances	(330)	3,608
Foreign currency exchange loss	1,364	14,805
Bank fees and other charges	80	152
Total financial expenses	**10,438**	**28,985**

2.6 INCOME TAXES

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

DKK 000	2021	2020
Current tax benefit on net loss	139,397	142,539
Adjustments prior years	-	(1,065)
Tax credit research and development expenses	5,500	5,500
Change in unrecognized deferred tax before tax credit	(150,544)	(144,256)
Permanent differences	11,147	2,782
Total income tax benefit for the year	**5,500**	**5,500**

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

DKK 000	2021	2020
Net loss before tax	(633,624)	(647,905)
Corporate income tax rate in Denmark	22 %	22%
Computed income tax benefit	**139,397**	**142,539**
Tax effect of:		
Adjustments prior years	-	(1,065)
Other non-deductible expenses, including US listing-related costs and share-based compensation	11,147	2,782
Effect of different tax rate	-	
Deferred tax asset not recognized after tax credit	(145,044)	(138,756)
Total income tax benefit for the year	**5,500**	**5,500**

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

DKK 000	2021	2020
Tax deductible losses	319,811	192,837
Deferred tax on intangible assets	132,310	112,192
Other temporary differences	1,882	2,019
	454,003	307,048
Deferred tax asset not recognized	454,003	307,048
Carrying amount included in the Statement of Financial Position		
	-	-

2.7 INTANGIBLE ASSETS

DKK 000	Software	Licenses	Total
Cost at December 31, 2019	—	12,083	12,083
Additions	896	—	896
Cost at December 31, 2020	896	12,083	12,979
Additions	85		85
Cost at December 31, 2021	**981**	**12,083**	**13,064**
Accumulated amortization at December 31, 2019	—	1,544	1,544
Amortization expense	42	712	754
Accumulated amortization at December 31, 2020	42	2,256	2,298
Amortization expense	391	119	510
Impairment expense	0	8,105	8,105
Accumulated amortization at December 31, 2021	**433**	**10,480**	**10,912**
Net carrying value at			
December 31, 2020	854	9,827	10,681
December 31, 2021	549	1,603	2,152

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 buildings	Operating equipment	Total
At December 31, 2019	**10,095**	**3,808**	**13,903**
Additions	1,712	-	1,712
Depreciation expense	(2,351)	(401)	(2,752)
Modifications		-	
At December 31, 2020	**9,456**	**3,406**	**12,862**
Additions	-	-	-
Disposals	(1,177)	-	(1,177)
Depreciation expense	(2,387)	(167)	(2,554)
Impairment expense	-	(3,239)	(3,239)
Modifications	(2,166)	-	(2,166)
Exchange rate adjustments			
At December 31, 2021	**3,726**	**-**	**3,726**

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

DKK 000	2021	2020
At January 1	11,346	12,689
Additions	-	1,712
Accretion of interest	490	499
Disposals	(1,212)	
Payments	(3,579)	(3,554)
Exchange rate adjustments		
Modifications	(2,496)	-
At December 31	**4,549**	**11,346**
Current	2,122	3,275
Non-current	2,427	8,071

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

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

DKK 000	2021	2020
Depreciation and impairment expense of right-of-use assets (R&D)	5,386	2,441
Depreciation and impairment expense of right-of-use assets (G&A)	407	311
Interest expense on lease liabilities	490	499
Gain on lease modification and disposals	(330)	—
Total amount recognized in the Statement of Profit or Loss	**5,953**	**3,574**

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, 2019	5,532	2,066	7,598
Additions	1,259		1,259
Disposals	—	—	—
Cost at December 31, 2020	6,791	2,066	8,857
Additions			
Disposals	(597)	—	(597)
Cost at December 31, 2021	**6,194**	**2,066**	**8,260**
Accumulated depreciation at December 31, 2019	3,614	300	3,914
Depreciation expense	919	317	1,236
Exchange rate adjustments		0	
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)	0	(411)
Exchange rate adjustments		0	
Accumulated depreciation at December 31, 2021	**5,121**	**934**	**6,055**
Net carrying value at			
December 31, 2020	2,258	1,449	3,707
December 31, 2021	1,073	1,132	2,205

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

DKK 000	2021	2020
Research and development expenses	1,031	802
General and administrative expenses	285	434
Total depreciation expense	**1,316**	**1,236**

2.10 INVESTMENT IN GROUP COMPANIES

DKK 000	2021	2020
Cost at January 1	1,346	1,207
Additions	2,596	139
Cost end of year December 31	**3,942**	**1,346**
Adjustment January 1	(1,207)	(1,207)
Impairment	(2,735)	-
End of year December 31	**(3,942)**	**(1,207)**
Carrying amount of investment	**-**	**139**

DKK 000	Registered office	Ownership interest (%)	Share capital	Equity	Net result
Orphazyme US, Inc.	Delaware, USA	100 %	USD 1 (USD 000)	1,630	(1,776)
Orphazyme GmhH (CH)	Zug, Switzerland	100 %	CHF 20,000 (CHF 000)	592	301

2.11 PREPAYMENTS, DEPOSITS, AND OTHER RECEIVABLES

DKK 000	2021	2020
Deposits with vendors	-	296
Prepayments to vendors	-	280
Leasehold deposit	593	1,090
Total non-current prepayments and deposits	**593**	**1,666**

DKK 000	2021	2020
Prepayments to vendors	12,436	37,342
Grant income receivable	-	81
VAT receivable, net	2,903	10,331
Foreign VAT receivable	1,627	1,304
Receivables from group entities	5,300	211,878
Other current receivables	2,790	1,237
Total current prepayments and other receivables	**25,056**	**262,173**

2.12 FINANCIAL ASSETS AND LIABILITIES

DKK 000	2021	2020
Borrowings	33,465	57,180
Lease liabilities (Note 3.2)	4,549	11,346
Trade payables	41,395	18,376
Accruals	8,050	30,698
Total liabilities measured at amortized cost	**87,459**	**117,600**

DKK 000	2021	2020
Remuneration to the Board of Directors	293	2,840
Payables to group entities	8,451	7,907
Payroll and employee-related costs	25,570	45,413
Total current other liabilities	**34,314**	**56,160**

			Non-cash changes				
DKK 000	December 31, 2020	Cash flows	Additions	Adjustments and modifications	Accumulated interest	Exchange rate adjustments	December 31, 2021
Borrowings	57,180	(30,904)	-	-	7,350	(160)	33,466
Lease liabilities	11,346	(3,579)	(2,496)	(1,212)	490	-	4,549
Total liabilities from financing activities	**68,526**	**(34,483)**	**(2,496)**	**(1,212)**	**7,840**	**(160)**	**38,015**

DKK 000	December 31, 2020	Cash flows	Accruals	December 31, 2021
Discount and rebate liabilities	—	—	36.193	36.193
Total liabilities from accrued discount and rebates	**—**	**—**	**36.193**	**36.193**

2.13 CASH

DKK 000	2021	2020
DKK	53,291	483,872
USD	27,440	1,765
EUR	4,070	644
CHF	-	-
GBP	892	1,051
Total cash	**85,693**	**487,332**

2.14 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	2021	2020
Transaction with subsidiaries		
Service fee costs	147,410	148,334
Balances with subsidiaries		
Current receivables	5,300	211,878
Current payables	8,451	7,907

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

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, 2021.

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, 2021 and the results of the Group's and Parent Company's operations and cash flows for the financial year January 1–December 31, 2021.

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 June 29, 2022

Copenhagen, June 7, 2022

BOARD OF DIRECTORS

Georges Gemayel
Chairman of the Board

Bo Jesper Hansen
Deputy Chairman of the Board

Andrew Mercieca

EXECUTIVE MANAGEMENT

Anders Vadsholt
Chief Executive Officer and
Chief Financial Officer

Independent Auditors' Report

Report of Independent Registered Public Accounting Firm

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 2021, 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 2021 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 2021 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.

Appointment of auditor

We were initially appointed as auditor of Orphazyme A/S on 4 December 2015 for the financial year for 1 July to 31 December 2015. We have been reappointed annually by resolution of the general meeting for a total consecutive period of 7 years up until the financial year 2021.

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 2021. 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 the 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 clawback liability related to the Company's sales under the early access compassionate use programs in France

As disclosed in Note 2.1 to the consolidated financial statements, revenue from the sale of arimoclomol for the treatment of NPC under the remunerated early access compassionate use program ("nATU") in France is measured net of estimated clawback liability to the French Authorities.

The estimates for clawback liability to the French Authorities are recognised as a reduction to gross product sales in the period in which the underlying sales are recognised. As of December 31, 2021, the liabilities for clawback amounts to DKK 36.2 million, as disclosed in Note 3.7 in the consolidated financial statements.

Estimating the clawback liability to the French Authorities is complex due to the judgmental nature of management's estimates, which involves assumptions of the final price to be agreed with the health authorities in France once the drug product is approved in France or otherwise agreement of price in the event of no approval in France, as not all conditions are known at the time of sale.

Management's assumptions are based on available relevant market information regarding average treatment cost of the most comparable drugs possible in the rare disease area in Europe. The Company is operating within rare disease therapeutic area where there is unmet treatment need and hence a limited number of comparable commercialized drugs products. The limited available relevant market information for directly comparable commercialized drugs within rare deceases increases the uncertainty in managements estimate.

How our audit addressed the key audit matter

Our procedures included, among others, checking clerical accuracy of management's calculation of liabilities for clawback. We assessed the assumptions applied by management and compared them with the conditions laid out in the Early Access Programs in France. We reperformed procedures on management's compilation of available relevant market information regarding average treatment cost of the most comparable drugs possible in the rare disease area in Europe.

Further, we examined subsequent settlement obligations to assess completeness and accuracy of the recorded liabilities, including obtaining the sales contract with KemPharm, and validating the transfer of clawback liability to KemPharm at the carrying amount subsequently to the balance sheet date.

In addition, we have assessed the adequacy of the Company's disclosures on clawback liabilities related to the matter described above.

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 or when, in extremely rare circumstances, we determine that a matter should not be communicated in our report because the adverse consequences of doing so would reasonably be expected to outweigh the public interest benefits of such communication.

Report on compliance with the ESEF Regulation

As part of our audit of the financial statements of Orphazyme A/S we performed procedures to express an opinion on whether the annual report for the financial year 1 January – 31 December 2021 with the file name 54930025OZD2GGSQ7L42-2021-12-31-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.

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 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;
- 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 for the financial year 1 January – 31 December 2021 with the file name 54930025OZD2GGSQ7L42-2021-12-31-en is prepared, in all material respects, in compliance with the ESEF Regulation.

Copenhagen, 7 June 2022

EY Godkendt Revisionspartnerselskab

CVR no. 30 70 02 28

Christian Schwenn Johansen State Authorised Public Accountant mne33234	Anders Roe Eriksen State Authorised Public Accountant mne46667

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