

## Company announcement

No. 36/2022

### Orphazyme A/S

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## Interim Report First Half 2022

**Copenhagen, Denmark, September 26, 2022** – Orphazyme A/S (ORPHA.CO) (the “Company”), today announces its Interim Report First Half 2022 for the period January 1 – June 30, 2022.

Anders Vadsholt, Chief Executive Officer and Chief Financial Officer said, *“During the first half of 2022, we executed the sale of substantially all of Orphazyme’s assets, including the continuing development of arimoclomol, to KemPharm Denmark A/S, a wholly owned subsidiary of KemPharm, Inc. Our employees demonstrated unwavering commitment throughout this difficult time, and I am delighted that many of them are pursuing their mission to bring new treatment options to patients with NPC at KemPharm. We are now focused on exploring ways to realize value from the Company’s remaining assets and may engage in activities and investments related to biopharmaceutical research and development, participate in partnerships or co-operate with other businesses.”*

### Corporate Updates First Half 2022

- In January 2022, Christophe Bourdon, Chief Executive Officer (CEO), resigned from his position as CEO of Orphazyme to take on the role of CEO at another company. Anders Vadsholt was appointed CEO of Orphazyme, effective March 1, 2022, in addition to his position as Chief Financial Officer (CFO).
- In February 2022, Orphazyme was notified of a negative trend vote by the Committee for Medicinal Products for Human Use (CHMP) of the European Medicine’s Agency (EMA) relating to its marketing authorization application (MAA) for arimoclomol in Niemann-Pick disease type C, or NPC, in Europe. The Company withdrew the MAA in March 2022, ahead of the CHMP’s final vote.
- On March 11, 2022, the Company’s board of directors (the “Board of Directors”) filed to initiate in-court restructuring proceedings under the Danish Insolvency Act with the aim of exploring whether a basis could be established for all or part of Orphazyme’s operations to continue, including a basis for injecting further capital, and/or a basis for a sale of all or parts of its assets.
- In March 2022, the Company voluntarily delisted American Depositary Shares (ADSs) representing Orphazyme’s ordinary shares from the Nasdaq Global Select Market in the United States, effective March 31, 2022.
- In May 2022, the Company signed an agreement to sell substantially all Orphazyme’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 amount of USD 12.8 million in cash and assumed liabilities estimated to equal approximately USD 5.2 million (the “Sale of Assets”).
- Stephanie Okey, Carolee Barlow and Martin Bonde stepped down from their positions as members of the Board of Directors as of May 23, 2022.
- The restructuring proposal was approved on May 30, 2022, by Orphazyme’s known creditors and affirmed by the Danish Maritime and Commercial High Court; the in-court restructuring proceedings were discontinued on May 30, 2022, and the Sale of Assets was completed on May 31, 2022.
- In June 2022, Bo Jesper Hansen was elected Chairman of the Board of Directors and John Sommer Schmidt and Anders Vadsholt were elected to the Board of Directors. Georges Gemayel, former Chairman of the Board of Directors and Andrew Mercieca, member of the Board of Directors did not stand for re-election at the Company’s Annual General Meeting.

## Financial Highlights First Half 2022

Unless otherwise stated, comments in this announcement refer to H1 performance from continuing operations. From January 1, 2022, the business operations and activities that were part of the Sale of Assets agreement with KemPharm have been reclassified as discontinued operations. 2021 figures have been restated according to IFRS.

- Operating expenses for continuing operations for the six months ended June 30, 2022 totaled DKK 27.3 million compared to DKK 57.3 million for the same period in 2021.
- Loss from continuing operations totaled DKK 24.4 million compared DKK 55.0 million for the same period in 2021.
- The net result from operations held for sale (discontinued operations) was a profit of DKK 87.4 million compared to a net loss of DKK 408.8 million for the same period in 2021, primarily due to the Sale of Assets to KemPharm.
- The total comprehensive profit/loss (both continuing and discontinued operations) for the first half 2022 was a net profit of DKK 62.5 million, or DKK 1.79 per share (basic and diluted), compared to a total comprehensive loss of DKK 463.7 million, or DKK (13.27) per share (basic and diluted) for the same period in 2021.
- As of June 30, 2022, Orphazyme held cash totaling DKK 49.5 million compared to DKK 102.3 million as of December 31, 2021.

## Subsequent Events

- As announced in April 2022, the Deposit Agreement among the Company, The Bank of New York Mellon, as depository, and owners and American Depositary Receipt (ADRs) holders was terminated, effective July 6, 2022.
- On August 11, 2022, the U.S. District Court for the Northern District of Illinois granted in part and denied in part defendants' motion to dismiss the amended complaint in the putative class action lawsuit, captioned *Busic v. Orphazyme A/S, et al.*, No. 21- cv-03640 (N.D. Ill.). The Court dismissed all claims under the Securities Act of 1933 relating to the Company's registration statement for its IPO of ADSs and dismissed as inactionable a number of challenged statements for the lead plaintiff's claims under the Securities Exchange Act of 1934. On August 26, 2022, the Court amended its August 11, 2022 order to also dismiss certain claims against certain individual defendants. The Court's dismissal of claims in its August 11 and 26, 2022 orders was without prejudice to the filing of a further amended complaint. On September 8, 2022, the lead plaintiff filed a second amended complaint. Defendants have until October 6, 2022 to respond to the second amended complaint. Management does not believe these claims have any merit and believe that the outcome will not materially affect the Company's financial position.

## Outlook

As announced September 25, 2022 (Company announcement No. 35/2022), Orphazyme has adjusted its outlook for 2022, as published on June 7, 2022. The change is driven by the separation of activities into continuing and discontinued operations. The discontinued activities are related to the Sale of Assets to KemPharm A/S. The continuing business activities are the limited ongoing operational business activities with only two employees. For the full-year 2022, we anticipate a loss from continuing operations in the range DKK 40 – 45 million and a positive result from discontinued operations relating to the Sale of Assets to KemPharm of DKK 87.4 million (previously operating profit in the range DKK 10 – 30 million). As previously communicated, we expect to end 2022 with more than DKK 30 million in cash and equivalents.

## Condensed Consolidated Key Figures

Following the Sale of Assets to KemPharm Denmark A/S, key figures and ratios are presented for continuing activities unless otherwise stated. Comparative periods have been restated accordingly for the statement of profit or loss and other comprehensive income and the cash flow statements.

DKK (000)	As of and for the six-months ended Jun 30, 2022	As of and for the six-months ended Jun 30, 2021	As of and for the year ended Dec 31, 2021
<b>Statement of profit or loss and other comprehensive income</b>			
Net revenue	-	-	-
Research and development expenses	-	-	-
General and administrative expenses	(27,296)	(57,344)	(83,472)
<b>Operating loss</b>	<b>(27,296)</b>	<b>(57,344)</b>	<b>(83,472)</b>
Net financial items	56	25	37
<b>Loss before tax</b>	<b>(27,239)</b>	<b>(57,319)</b>	<b>(83,436)</b>
Income tax benefit	2,793	2,318	4,941
<b>Loss from continuing operations</b>	<b>(24,446)</b>	<b>(55,002)</b>	<b>(78,495)</b>
Net result from operations held for sale ("discontinued operations")	87,429	(408,778)	(548,044)
<b>Net result for the period</b>	<b>62,983</b>	<b>(463,780)</b>	<b>(626,539)</b>
<b>Total comprehensive income (loss)</b>	<b>62,531</b>	<b>(463,704)</b>	<b>(626,841)</b>
Profit / (loss) per share, basic and diluted (DKK)	1.79	(13.27)	(17.94)
<b>Statement of financial position</b>			
Intangible assets	-	2,194	2,152
Right-of-use assets & Property, plant and equipment	-	13,928	8,419
Other non-current assets	5,500	8,214	3,714
Cash	49,532	334,184	102,255
Other current assets	38,557	69,511	56,689
Total assets	93,589	428,031	173,229
Share capital	35,312	34,952	34,952
Total equity	78,205	166,761	9,339
Non-current liabilities	-	37,610	34,798
Current liabilities	15,384	223,660	129,092
<b>Cash flow statement</b>			
Net cash (used in) provided by operating activities	(116,472)	2,591	(78,975)
Net cash used in investing activities	-	-	-
Net cash provided by (used in) financing activities	5,310	(1,184)	(1,184)
Net cash flow from operations held for sale	63,577	(399,463)	(552,710)
<b>Other</b>			
Share price (DKK)	1.81	46.84	17.16
Total outstanding shares	35,312,241	34,952,241	34,952,241
Market capitalization (DKK million) <sup>1</sup>	64.3	1,637.2	599.8
Equity ratio <sup>2</sup>	83.6%	39.0%	5%
Equity per share (DKK) <sup>3</sup>	2.21	4.77	0.27
Purchase of property, plant and equipment (DKK 000)	-	-	92
Average number of employees	34	164	130
Number of full-time employees (FTEs), end of period	2	180	62

(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's Review

**The first half of 2022 was a challenging time for Orphazyme, resulting in a significant shift in our focus and business operations by the end of the period. We were able to sell substantially all of our assets to enable the ongoing development of arimoclomol by a third party, and were pleased to have been able to transition, to KemPharm, many of the employees involved in arimoclomol's journey. We now have limited operations and are focused on exploring ways to realize value from the Company's remaining assets and may engage in activities and investments related to biopharmaceutical research and development, participate in partnerships or co-operate with other businesses.**

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

In February 2022, we were notified of a negative trend vote by Committee for Medicinal Products for Human Use (CHMP) of the European Medicine's Agency (EMA) relating to our MAA for arimoclomol in Niemann-Pick disease type C (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 filed to initiate in-court restructuring proceedings of Orphazyme under the Danish Insolvency Act, which commenced on March 11, 2022. In accordance with the in-court restructuring proceedings, 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 American Depositary Shares (ADSs) representing our ordinary shares from Nasdaq Global Select Market in the U.S., effective on March 31, 2022. In April 2022, we announced that the Deposit Agreement among the Company, The Bank of New York Mellon, as depositary, and owners and American Depositary Receipt (ADRs) holders was to be terminated, effective July 6, 2022.

In accordance with the Statutory Restructuring Plan, in April 2022 we also 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 our 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 our 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 our 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 our known creditors and affirmed by the Danish Maritime and Commercial High Court. Following the approval of the restructuring proposal, the in-court restructuring proceedings were discontinued with immediate effect on May 30, 2022, and the Sale of Assets was completed on May 31, 2022. Completion of the Sale of Assets provided full coverage to creditors with undisputed claims based on the claims filed during the restructuring. All (undisputed and unconditional) debts related to the time prior to restructuring have been or will be paid in full, including all obligations outstanding under our 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. 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.

At the Annual General Meeting in June 2022, the Board of Directors was further reduced to three members, and consists of Bo Jesper Hansen, Anders Vadsholt and John Sommer Schmidt. Georges Gemayel, former Chairman of the Board of Directors, and Andrew Mercieca, member of the Board of Directors did not stand for re-election.

Further, at the Annual General Meeting, it was decided to amend the purpose of the Company in light of the Sale of Assets, and the shareholders adopted resolutions permitting the Company to engage in activities and investments related to biopharmaceutical research and development, as well as to carry out any associated activities as deemed relevant by the Board of Directors. Furthermore, the Company may, within its line of business, participate in partnerships or co-operate with other businesses.

We entered the second half of 2022 with limited ongoing operations and DKK 49.5 million in cash as at June 30, 2022.

## Outlook

As announced September 25, 2022 (Company announcement No. 35/2022), Orphazyme has adjusted its outlook for 2022, as published on June 7, 2022. The change is driven by the separation of activities into continuing and discontinued operations. The discontinued activities relate to the Sale of Assets to KemPharm Denmark A/S. The continuing business activities relate to the limited ongoing operational business activities and the now two employees.

In millions of DKK	2022 guidance
Loss from continuing operations	(40 – 45)
Positive result from discontinued operations	87.4
Net result for the year <sup>1</sup>	42.5 - 47.5
Cash position at year-end	>30

(1) From continuing and discontinued operations

There are inherent risks and uncertainties in our outlook for 2022 given the recent closing of the KemPharm Sale of Assets transaction, transfer of operating activities from us to KemPharm and our future operational plans and prospects.

## Risk management

The following significant risks and uncertainties were identified as of June 30, 2022, which could have a significant impact on our outlook and financial prospects:

- **Securities litigation:**

- We are subject to securities litigation described below, which is expensive and could divert management's attention.
- On July 9, 2021, a putative class action lawsuit, captioned *Busic v. Orphazyme A/S, et al.*, No. 21- cv-03640 (N.D. Ill.), was filed against us and certain of our current and former directors and officers in the U.S. District Court for the Northern District of Illinois. On September 13, 2021, the Court appointed a lead plaintiff for the putative class. On November 19, 2021, the lead plaintiff filed an amended complaint asserting claims under the U.S. Securities Act of 1933 and the U.S.

Securities Exchange Act of 1934 for alleged misrepresentations and/or omissions in our registration statement for our IPO of ADSs in the United States and in subsequent public statements. The amended complaint seeks an unspecified amount of damages on behalf of a putative class comprised of all persons and entities other than the defendants that purchased or otherwise acquired the ADS in connection with the IPO in September 2020 and/or between September 29, 2020 and November 4, 2021. On August 11, 2022, the Court granted in part and denied in part defendants' motion to dismiss the amended complaint. The Court dismissed all claims under the Securities Act of 1933 relating to our registration statement for our IPO of ADSs and dismissed as inactionable a number of challenged statements for the lead plaintiff's claims under the Securities Exchange Act of 1934, including all of the challenged statements prior to June 2021. On August 26, 2022, the Court amended its August 11, 2022 order to also dismiss certain claims against certain individual defendants. The Court's dismissal of claims in its August 11 and 26, 2022 orders was without prejudice to the filing of a further amended complaint. On September 8, 2022, the lead plaintiff filed a second amended complaint with new allegations regarding an April 2021 statement by our CEO at the time. Defendants have until October 6, 2022 to respond to the second amended complaint.

- **Financial position and business prospects:**

- 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 employees. As such our future prospects are uncertain.
- We may choose to wind down our business, which could result in additional costs associated with any distribution which would further limit funds to shareholders.

- **Risks Related to the Sale of Assets:**

- 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.

- **Political and geopolitical conflict:**

- The conflict in Ukraine and any retaliatory measures taken by the United States and NATO could threaten global security and result in further regional conflict and otherwise have a lasting impact on regional and global economies, any or all of which could adversely affect our business.

- **Currency risk:**

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

Actions to mitigate these risks and uncertainties:

- **Securities litigation:**

- We believe that the claims raised in the *Busic v. Orphazyme A/S, et.al.* suit are without merit and we intend to continue to vigorously defend against the claims.

- **Financial position and business prospects:**

- As of June 30, 2022, we had cash of DKK 49.5 million. We have limited operating activities and prudently manage our expenses. We plan to explore ways to realize value from the Company's remaining assets and may engage in activities and investments related to biopharmaceutical research and development, participate in partnerships or co-operate with other businesses.

For more information on risks and uncertainties relating to Orphazyme's business please refer to our Statutory Annual Report 2021 and our Annual Report on Form 20-F that is filed with the United States Securities and Exchange Committee (SEC) and is available on our website.

The risks and uncertainties discussed above are those that the Company currently views as material and specific to the Company, but there can be no assurance that these are the only risks and uncertainties that the Company might face. Additional risks and uncertainties, including risks that are not known to the Company at present or that its management currently deems immaterial or non-specific to the Company, may also arise or become material or specific to the Company in the future, which could, if such risks were to materialize, have a material and adverse effect on the Company's business, financial condition, and/or results of operations.

With the uncertainties listed above, there can be no assurance that the Company will achieve or sustain profitability or positive cash flows from operations. These interim condensed consolidated financial statements do not include any adjustments that might result from the outcome of these uncertainties. Based on the mitigating actions described above, Management is of the view that a going concern assumption is appropriate as of June 30, 2022.

## Financial Review

### Introduction

Substantially all of Orphazyme's assets and business activities, including those relating to the development and approval of arimoclomol, the full claw back liability related to the French early access program and costs associated with certain employees, were transferred to KemPharm Denmark A/S (KemPharm), a wholly owned subsidiary of KemPharm, Inc., on June 1, 2022 in line with the terms of the Sale of Assets agreement. These activities have been reclassified as discontinued operations, cf note 4, in the presentation of these consolidated interim financial statements. Please note the figures below are presented on a continuing basis, unless otherwise described.

### Income statement

The net result for H1 2022 (from continuing and discontinued operations) was a profit of DKK 63.0 million compared to a net loss of DKK 463.8 million for the same period in 2021. Loss from continuing operations was DKK 24.4 million in H1 2022 compared to a net loss of DKK 55.0 million in H1 2021. The net result from discontinued operations was a profit of DKK 87.4 million in H1 2022 compared to a net loss of DKK 408.8 million for the same period in 2021. The difference primarily relates to the net effect of income received from the Sale of Assets to KemPharm and higher costs during the same period the prior year associated with preparations for commercial launch of arimoclomol in the U.S., accelerated close out of certain clinical trials and impairment costs following unfavorable results, initiation of a restructuring program and termination of onerous contracts.

### Net revenue

As substantially all of Orphazyme's business activities were sold to KemPharm, including any revenue from sales of arimoclomol, net revenue from continuing operations for H1 2022 was DKK 0 million (DKK 0 million for H1 2021). Net revenue in H1 2022 from discontinued operations was DKK 22.3 million compared to DKK 13.2 million in the same in 2021.

### Research and development expenses

Following the Sale of Assets to KemPharm, research and development (R&D) expenses have also been reclassified as discontinued operations. As such, R&D expenses from continuing operations totaled DKK 0 million in H1 2022 (DKK 0 million in H1 2021). R&D expenses from discontinued operations totaled DKK 30.0 million in H1 2022 compared to DKK 264.7 million in H1 2021. The difference primarily relates to higher costs in H1 2021 associated with manufacturing, accelerated close out of certain clinical trials following unfavorable results and associated impairment costs, initiation of a restructuring program and termination of onerous contracts.

### General and administrative expenses

General and administrative (G&A) expenses from continuing operations totaled DKK 27.3 million in H1 2022, a reduction of approx. DKK 30 million compared to the prior year (DKK 57.3 million in H1 2021). G&A expenses in H1 2022 include costs associated with employees and Board of Directors, service providers and external assistance, legal and technology expenses. G&A expenses from discontinued operations were DKK 39.8 million in H1 2022 compared to DKK 156.8 million in H1 2021. The difference primarily relates to higher costs in H1 2021 associated with the build-up of a commercial organization, including commercial launch preparation activities, as well as expenses related to support functions and an impairment charge for software and the employee restructuring program.

### Net financial items

Net financial income from continuing operations was DKK 56 thousand in H1 2022 compared to DKK 25 thousand in H1 2021. Net financial items from discontinued operations were a loss of DKK 10.9 million in H1 2022 compared to a loss of DKK 0.4 million in H1 2021. The difference primarily relates to fluctuations in foreign exchange rates year over year, particularly the USD / DKK exchange rate.



## **Income tax benefit**

Income tax benefit from continuing operations totaled DKK 2.8 million in H1 2022 compared to DKK 2.3 million in H1 2021. This includes income tax benefit related to tax credits for research and development expenses at the applicable tax rate under the Danish Corporate Income Tax Act. Income tax benefit from discontinued operations was DKK 0 million during H1 2022 compared to DKK 0 million during H1 2021.

## **Statement of financial position**

### *Cash*

As of June 30, 2022, Orphazyme held cash of DKK 49.5 million compared to DKK 102.3 million as of December 31, 2021.

### *Equity*

As of June 30, 2022, total equity amounted to DKK 78.2 million compared to DKK 9.3 million as of December 31, 2021. The increase relates to positive impact from the Sale of Assets transaction with KemPharm.

## **Cash flows**

### *Cash flow from operating activities*

Net cash flow from continuing operating activities amounted to an outflow of DKK 116.5 million in the six-month period ended June 30, 2022 compared to an inflow of DKK 2.6 million in the six month period ended June 30, 2021. Net cash flow from discontinued operating activities was an inflow of DKK 102.5 million during H1 2022, compared to an outflow of DKK 384.3 million in H1 2021. The difference in cash flow from discontinued operating activities during the period was due to the Sale of Assets to KemPharm in H1 2022 compared to higher costs in the prior year relating to clinical, manufacturing and pre-commercial activities relating to arimoclomol, among other operating costs.

### *Cash flow from investing activities*

Net cash flow from investing activities from continuing operations amounted to DKK 0 million in H1 2022 (DKK 0 million in H1 2021). Net cash flow from investing activities relating to discontinued operations was an inflow of DKK 1.5 million in H1 2022 compared to an outflow of DKK 0.8 million in H1 2021.

### *Cash flow from financing activities*

Net cash flow from financing activities from continuing operations amounted to an inflow of DKK 5.3 million in H1 2022 compared to an outflow of DKK 1.2 million in H1 2021. The difference relates to cash inflow relating to the U.S. At-the-market program share issue in H1 2022 compared to outflow in H1 2021 relating to bonus share issues due to the license agreement with KLSDC and UCL (note 10). Cash flow from financing activities relating to discontinued operations was an outflow of DKK 40.4 million in H1 2022 compared to an outflow of DKK 14.4 million in the same period the prior year. The difference primarily relates to repayment of the debt facility with Kreos Capital in H1 2022.

## Consolidated Statements of Profit or Loss and Other Comprehensive Income

	Six months ended Jun 30, 2022 DKK (000)	Six months ended Jun 30, 2021 DKK (000)
Net revenue (Note 5)	-	-
Research and development expenses (Note 6)	-	-
General and administrative expenses (Note 7)	(27,296)	(57,344)
<b>Operating expenses</b>	<b>(27,296)</b>	<b>(57,344)</b>
Financial income	56	25
Financial expenses	-	-
<b>Loss before tax</b>	<b>(27,239)</b>	<b>(57,319)</b>
Income tax benefit	2,793	2,318
<b>Loss from continuing operations</b>	<b>(24,446)</b>	<b>(55,002)</b>
Net result from discontinued operations (Note 4)	87,429	(408,778)
<b>Net result for the period</b>	<b>62,983</b>	<b>(463,780)</b>
Exchange differences from translation of foreign operations	(452)	76
<b>Total comprehensive income (loss)</b>	<b>62,531</b>	<b>(463,704)</b>
<b>Profit/(loss) per share, basic and diluted (Note 11)</b>	<b>1.79</b>	<b>(13.27)</b>
Continuing operations	(0.69)	(1.58)
Discontinued operations	2.48	(11.71)

The accompanying notes are an integral part of these interim consolidated financial statements

<b>Consolidated Statements of Financial Position</b>	<b>Jun 30, 2022 DKK (000)</b>	<b>Dec 31, 2021 DKK (000)</b>
<b>ASSETS</b>		
<b>Non-current assets</b>		
Intangible assets	-	2,152
Right-of-use assets	-	5,434
Property, plant, and equipment	-	2,985
Corporation tax receivable	5,500	2,750
Deferred tax assets (Note 9)	-	-
Prepayments and deposits	-	964
<b>Total non-current assets</b>	<b>5,500</b>	<b>14,285</b>
<b>Current assets</b>		
Corporation tax receivable	7,388	7,229
Trade receivables	23,765	29,268
Prepayments and other receivables	7,404	20,192
Cash	49,532	102,255
<b>Total current assets</b>	<b>88,089</b>	<b>158,944</b>
<b>TOTAL ASSETS</b>	<b>93,589</b>	<b>173,229</b>
<b>EQUITY &amp; LIABILITIES</b>		
<b>Equity</b>		
Share capital (Note 10)	35,312	34,952
Share premium (Note 10)	2,087,436	2,082,486
Other reserves	2,447	2,899
Accumulated deficit	(2,046,990)	(2,110,998)
<b>Total equity</b>	<b>78,205</b>	<b>9,339</b>
<b>Non-current liabilities</b>		
Borrowings	-	2,482
Lease liabilities	-	3,925
Discount and rebate liabilities	-	28,293
Other non-current liabilities	-	98
<b>Total non-current liabilities</b>	<b>-</b>	<b>34,798</b>
<b>Current liabilities</b>		
Borrowings	-	30,983
Lease liabilities	-	2,578
Trade payable and accruals	11,362	57,524
Corporation tax payable	334	584
Discount and rebate liabilities	-	7,900
Other liabilities	3,688	29,523
<b>Total current liabilities</b>	<b>15,384</b>	<b>129,092</b>
<b>TOTAL EQUITY AND LIABILITIES</b>	<b>93,589</b>	<b>173,229</b>

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

## Consolidated Statements of Changes in Shareholders' Equity

	Share capital DKK (000)	Share premium DKK (000)	Other reserves		Accumulated deficit DKK (000)	Total DKK (000)
			Foreign currency translation reserve DKK (000)	Share-based compensation – acquisition of intangible assets DKK (000)		
<b>Balance as of December 31, 2020</b>	<b>34,698</b>	<b>2,082,254</b>	<b>714</b>	<b>5,780</b>	<b>(1,502,921)</b>	<b>620,525</b>
Net loss for the period	-	-	-	-	(463,780)	(463,780)
Other comprehensive loss for the period	-	-	76	-	-	76
<i>Total other comprehensive loss</i>	-	-	76	-	(463,780)	(463,704)
<i>Transactions with owners</i>						
Capital increase, issuance of Matching Shares, net of costs (Note 10)	170	-	-	-	-	170
Capital increase, Bonus Shares (Note 10)	22	-	-	(1,645)	1,623	-
Cash settlement of Bonus Shares (Note 10)	-	-	-	(1,648)	-	(1,648)
Capital increase, issuance of sign-on bonus shares to former CEO (Note 10)	58	-	-	-	-	58
Capital increase, exercise of RSUs (Note 10)	4	232	-	-	-	236
Share-based payment costs (Note 8)	-	-	-	-	11,123	11,123
<i>Total transactions with owners</i>	<i>254</i>	<i>232</i>	<i>-</i>	<i>(3,293)</i>	<i>12,746</i>	<i>9,939</i>
<b>Balance as of June 30, 2021</b>	<b>34,952</b>	<b>2,082,486</b>	<b>790</b>	<b>2,487</b>	<b>(1,953,955)</b>	<b>166,761</b>

	Share capital DKK (000)	Share premium DKK (000)	Other reserves		Accumulated deficit DKK (000)	Total DKK (000)
			Foreign currency translation reserve DKK (000)	Share-based compensation – acquisition of intangible assets DKK (000)		
<b>Balance as of December 31, 2021</b>	<b>34,952</b>	<b>2,082,486</b>	<b>412</b>	<b>2,487</b>	<b>(2,110,998)</b>	<b>9,339</b>
Net loss /profit for the period	-	-	-	-	62,983	62,983
Other comprehensive loss for the period	-	-	(452)	-	-	(452)
<i>Total other comprehensive loss</i>	-	-	(452)	-	62,983	62,531
<i>Transactions with owners</i>						
Capital increase (Note 10)	360	4,950	-	-	-	5,310
Share-based payment costs (Note 8)	-	-	-	-	1,025	1,025
<i>Total transactions with owners</i>	<i>360</i>	<i>4,950</i>	<i>-</i>	<i>-</i>	<i>1,025</i>	<i>6,335</i>
<b>Balance as of June 30, 2022</b>	<b>35,312</b>	<b>2,087,436</b>	<b>(40)</b>	<b>2,487</b>	<b>(2,046,990)</b>	<b>78,205</b>

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

## Consolidated Statements of Cash Flow

	Six months ended Jun 30, 2022 DKK (000)	Six months ended Jun 30, 2021 DKK (000)
<b>Operating activities</b>		
Operating loss	(27,296)	(57,344)
<i>Adjustments to reconcile loss before tax to cash flows from operating activities:</i>		
Change in trade receivables, prepayments, deposits and other receivables	19,255	(11,377)
Change in provisions, trade payables, accruals and other liabilities	(108,288)	75,468
Corporation taxes received / (paid)	-	(4,181)
Interest received / (paid), net	(143)	25
<b>Net cash (used in) provided by operating activities</b>	<b>(116,472)</b>	<b>2,591</b>
<b>Investing activities</b>		
Purchase of intangible assets	-	-
Purchase of property, plant, and equipment	-	-
<b>Net cash used in investing activities</b>	<b>-</b>	<b>-</b>
<b>Financing activities</b>		
Proceeds from issuance of shares (Note 10)	5,310	464
Cash settlement of Bonus Shares (Note 10)	-	(1,648)
<b>Net cash provided by (used in) financing activities</b>	<b>5,310</b>	<b>(1,148)</b>
<b>Changes in cash and cash equivalents from continuing operations</b>	<b>(111,162)</b>	<b>1,407</b>
Net cash flow from operations held for sale (Note 4)	63,577	(399,463)
<b>Net change in cash and cash equivalents</b>	<b>(47,585)</b>	<b>(398,056)</b>
Cash balance at beginning of period	102,255	726,929
Effect of changes in exchange rates	(5,138)	5,311
<b>Cash balance at end of period</b>	<b>49,532</b>	<b>334,184</b>

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

## Notes to the Interim Consolidated Financial Statement

### NOTE 1 – CORPORATE INFORMATION

Orphazyme A/S (the “Company”) was, 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. 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 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 the Company filed a petition for an in-court restructuring of the Company, which commenced in March 2022. In May 2022, as part of the in-court restructuring proceedings, the Company 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 the Company’s remaining Danish employees. The in-court restructuring proceedings were discontinued on May 30, 2022. As of the date of publication of this interim report, the Company has limited ongoing operational business activities and only two employees.

Orphazyme is headquartered in Copenhagen, Denmark and is publicly traded on Nasdaq Copenhagen.

In April 2018, a wholly owned subsidiary of Orphazyme, Orphazyme U.S., Inc., was incorporated in Delaware, USA and in March 2020, another wholly owned subsidiary, Orphazyme Schweiz GmbH, was incorporated in Zug, Switzerland (together with Orphazyme A/S, “Orphazyme” or “the Group” as applicable). The Company is considering winding down these subsidiaries at an appropriate time.

In September 2020, the Company listed American Depositary Shares (ADSs) on the Nasdaq Global Select Market. In March 2022, the Company initiated voluntary delisting of the ADSs representing its shares from Nasdaq Global Select Market in the United States and the ADSs are not listed for trading on any other national securities exchange in the United States. The ADS facility was terminated on July 6, 2022.

These interim consolidated financial statements were approved and authorized for issuance by the Board of Directors on September 26, 2022.

#### **Basis of preparation**

The interim condensed consolidated financial statements for the six months ended June 30, 2022 have been prepared in accordance with IAS 34 *Interim Financial Reporting* as issued by the International Accounting Standards Board (IASB) and as adopted by the EU, and additional Danish disclosure requirements for interim reports of companies listed on the Nasdaq Copenhagen.

The interim condensed consolidated financial statements do not include all the information and disclosures required in annual financial statements and should be read in conjunction with the Company’s latest consolidated annual financial statements as of December 31, 2021. These interim condensed consolidated financial statements have been prepared in accordance with the going concern assumption (Note 3).

#### *COVID-19*

As of June 30, 2022 and as of the date of publication of the interim report, there is no material impact directly related to COVID-19 on the Group’s consolidated financial statements, including the judgements and estimates applied.

### **Updates to the Group's accounting policies**

The accounting policies used in the preparation of the interim condensed consolidated financial statements are consistent with those used in the preparation of Orphazyme's annual consolidated financial statements for the year ended December 31, 2021, with any additions included in the respective notes below. In addition, Orphazyme has implemented IFRS 5 as described below.

#### Discontinued operations and assets held for sale

Assets classified as held for sale comprise non-current assets and disposal groups, which are held for sale. Disposal groups are defined as a group of assets to be disposed of, by sale or otherwise, together as a group in a single transaction. Liabilities associated with assets held for sale are those liabilities directly associated with the assets that will be transferred in the transaction. Assets are classified as "held for sale" if the carrying amount will be recovered principally through a sale within 12 months in accordance with a formal plan rather than through continuing use.

Assets or disposal groups held for sale are measured at the lower of carrying amount at the date of the classification as "held for sale" and fair value less costs to sell. Assets are not depreciated or amortized from the date when they are classified as held for sale.

Impairment losses on the initial classification as "held for sale" and gains and losses on subsequent measurement at the lower of the carrying amount and fair value less costs to sell are recognized in the income statement in the items to which they relate. Gains and losses are disclosed in the notes.

Discontinued operations comprise a separate major line of business whose activities and cash flows can be clearly distinguished, operationally and for financial reporting purposes, from the other business areas and where the unit either has been disposed of or is held for sale and where the sale is expected to be carried out within twelve months in accordance with a formal plan.

The profit/loss of discontinued operations after tax, value adjustments of related assets and liabilities after tax and gains and losses on disposal are presented as a separate line item in the income statement, and comparative figures are restated accordingly.

Revenue, costs, value adjustments and tax relating to discontinued operations are disclosed in the notes. Assets and liabilities for discontinued operations are presented in separate lines in the statement of financial position without restatement of comparative figures and the major classes of assets and liabilities are disclosed in the notes.

Cash flows from operating, investing, and financing activities of the discontinued operations are disclosed in a note.

Additional disclosures are provided in note 4. All other notes to the Interim consolidated financial statements include amounts for continuing operations, unless indicated otherwise.

### **Significant accounting estimates and judgements**

The significant accounting estimates and judgements disclosed in note 1.4 of the consolidated financial statements for the year ended December 31, 2021 are still applicable.

#### **NOTE 3 – GOING CONCERN**

The expected net result for the year and the limited ongoing operational business activities makes us comfortable with preparing the interim consolidated financial statements on a going concern basis.

#### **NOTE 4 – DISCONTINUED OPERATIONS AND DISPOSAL GROUP HELD FOR SALE**

In May 2022, Orphazyme announced that it had signed an agreement to sell substantially all of the Company's assets and business activities, including those relating to the development

and approval of arimoclomol and the full claw back liability related to the French early access program, to KemPharm Denmark A/S (KemPharm), a wholly owned subsidiary of KemPharm, Inc.

From January 1, 2022, the business operations and activities that were part of the Sale of Assets agreement with KemPharm have been reclassified as discontinued operations. The net result related to the above-mentioned operations held for sale are therefore presented separately in the income statement and the statement of cash flows. The comparative figures have been restated accordingly.

Net result and net cash flow from operations held for sale and assets and liabilities in disposal group held for sale are specified below:

	<b>Six months ended Jun 30, 2022 DKK (000)</b>	<b>Six months ended Jun 30, 2021 DKK (000)</b>
Net revenue (Note 5)	22,324	13,152
Research and development expenses (Note 6)	(29,988)	(264,683)
General and administrative expenses (Note 7)	(39,833)	(156,823)
<b>Operating expenses</b>	<b>(69,822)</b>	<b>(421,505)</b>
Financial income	-	6,495
Financial expenses	(10,893)	(6,921)
<b>Loss before tax</b>	<b>(58,391)</b>	<b>(408,778)</b>
Income tax benefit	-	-
<b>Net result for the period</b>	<b>(58,391)</b>	<b>(408,778)</b>
Gain from disposal of assets and liabilities held for sale	145,820	-
<b>Net result from discontinued operations</b>	<b>87,429</b>	<b>(408,778)</b>
Cash flow from operating activities	102,505	(384,315)
Cash flow from investing activities	1,460	(806)
Cash flow from financing activities	(40,388)	(14,343)
<b>Net cash flow from operations held for sale</b>	<b>63,577</b>	<b>(399,463)</b>

#### NOTE 5 – NET REVENUE

Following the Sale of Assets to KemPharm, net revenue is fully presented as discontinued operations in note 4.

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. Any repayment liability has been transferred to KemPharm as part of the Sale of Assets transaction.

All sales and distributions of arimoclomol are included in the service agreement with Clinigen Health Limited, who keeps Orphazyme goods on a consignment stock until transferred to a third-party customer. The service agreement with Clinigen Health Limited and the consignment stock have been transferred to KemPharm as part of the Sale of Assets transaction.

For the six-month period ended June 30, 2022, Orphazyme recognized net revenue of DKK 22.3 million (compared to DKK 13.2 million for the six-month period ended June 30, 2021), which is net of certain estimated repayment amounts that may become due following potential approval of the drug product by the French authorities.

Trade receivables in the amount of DKK 23.8 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.

Cost of goods sold relating to products sold under the French nATU is recognized as research and development expenses.

#### NOTE 6 – RESEARCH AND DEVELOPMENT EXPENSES

Research and development expenses are fully presented as discontinued operation in note 4.

Research and development expenses of DKK 30.0 million for the six-month period ended June 30, 2022 (June 30, 2021: DKK 264.7 million) included 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. Further, research and development expenses include costs of goods sold relating to products sold under the French early access compassionate use program (note 5).

#### NOTE 7 – GENERAL AND ADMINISTRATIVE EXPENSES

General and administrative expenses of DKK 67.2 million for the six-month period ended June 30, 2022 (June 30, 2021: DKK 214.2 million) include salaries for administrative employees and Executive Management, remuneration to the Board of Directors, share-based compensation costs, audit fees, legal costs, and investor relations costs; as well as an impairment charge related to software. In addition, General and administrative expenses comprise costs as well as expenses related to restructuring activities, i.e. employee redundancies and termination of onerous contracts.

General and administrative expenses is comprised of general and administrative expenses from continuing operations of DKK 27.3 million for the six-month period ended June 30, 2022 (June 30, 2021: DKK 57.3 million) and of general and administrative expenses from discontinued operations of DKK 39.8 million for the six-month period ended June 30, 2022 (June 30, 2021: DKK 156.8 million). Discontinued operations are disclosed in note 4.

## NOTE 8 – SHARE-BASED COMPENSATION COSTS

Please refer to Note 2.6 of the Group’s consolidated financial statements included in the 2021 Annual Report for a description of the share-based compensation programs and the accounting policies and estimates applied. The activities in the respective programs are outlined below. The respective share-based compensation programs allow the vesting of awards through the employment release date. As such, there are no forfeitures to be disclosed or expense to be reversed due to the termination of employees as a result of our restructuring.

### **a) Long-term incentive programs (equity-settled)**

*2021 Long-term incentive program ("2021 LTIP"):*

Similar to the 2017 LTIP, the 2019 LTIP and the 2020 LTIP described in Note 2.6 of the consolidated financial statements for the year ended December 31, 2021, the 2021 LTIP offers all permanent employees of Orphazyme a certain number of Restricted Share Units (RSUs) contingent on continued employment and Performance Share Units (PSUs) depending on the development of the Company’s share price. Although the grant date of these awards was in April 2021, management has determined that the service commencement date of the participants was the earlier of January 1, 2021, or the employee’s employment date, and consequently the compensation expense is recognized from the service commencement date.

In October 2021, the Group initiated a modified 2021 LTIP also described in Note 2.6 of the consolidated financial statements for the year ended December 31, 2021.

The fair value of the 2021 LTIP awards (original and modified) was estimated separately for the RSUs and PSUs using a Black-Scholes model and a Monte-Carlo simulation, respectively, at the grant date of April 21 and October 7, 2021, respectively. The following inputs were used in the valuation models:

	Modified		Original	
	RSUs	PSUs	RSUs	PSUs
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 (years)	0.23-2.23	2.23	0.69-2.69	2.69
Share price (DKK)	25.70	25.70	59.05	59.05
Fair value of awards at measurement date (DKK 000)	24.72	12.56	58.04	20.02

The risk-free interest rate has been estimated based on Danish government bonds with similar maturities. Expected volatility has been determined based on Orphazyme’s own historic share price volatility. Based on the fair value of the 2021 LTIP, a credit of DKK 0.4 million was recognized for the six-month period ended June 30, 2022 compared to DKK 6.0 recognized for the six-month period ended June 30, 2021. The net income is because of reversal of expenses related to PSUs that has lapsed.

In addition, for the six-month period ended June 30, 2022, an aggregate expense of DKK 1.4 million was recognized for the 2020 LTIP and 2019 LTIP for the six-month period ended June 30, 2022 compared to DKK 5.1 million recognized for the six-month period ended June 30, 2021.

### **b) Phantom share-based incentive programs (cash-settled)**

As described in Note 2.6 of the consolidated financial statements for the year ended December 31, 2021, the Phantom programs are cash-settled and their fair value is re-

assessed at each reporting date.

The risk-free interest rate has been estimated based on Danish government bonds with similar maturities. Expected volatility has been determined based on Orphazyme's own historic volatility. Based on the fair value of the awards on June 30, 2022, a credit of DKK 0.0 million was recognized for the six-month period ended June 30, 2022 compared to a credit of DKK 0.0 million recognized for the six-month period ended June 30, 2021.

**c) Restricted share units (cash-settled)**

In March 2020, the RSUs granted to the Board of Directors under the 2019 RSU program fully vested. As of June 30, 2022, all but one board member has exercised the fully vested RSUs. The unexercised RSUs lapsed.

Also 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 RSUs granted under the 2019 program described in Note 2.6 of the consolidated financial statements for the year ended December 31, 2021. 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 Company'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. As of June 30, 2022, all but one board member has exercised the fully vested RSUs. The unexercised RSUs lapsed.

In May 2021, an aggregate of 30,450 RSUs were granted to the Board of Directors under the 2021 RSU program under similar terms and conditions as the 2019 RSU program described in Note 2.6 of the consolidated financial statements for the year ended December 31, 2021. As of June 30, 2022, all but 12,071 RSUs have lapsed following resignation of the eligible board members. The remaining unexercised were fully vested but unexercised.

The risk-free interest rate has been estimated based on Danish government bonds with similar maturities. Expected volatility has been determined based on Orphazyme's own historic volatility. Based on the fair value of the RSUs granted under the 2021 RSU Program on June 30, 2022, a credit of DKK 0.0 million was recognized for the six-month period ended June 30, 2022 compared to an expense of DKK 0.3 million recognized for the six-month period ended June 30, 2021. In addition, a credit of DKK 0.0 million was recognized for the 2020 RSUs for the six-month period ended June 30, 2022 compared to DKK 0.3 million recognized for the six-month period ended June 30, 2021.

**d) Bonus shares**

As part of the license agreement with KLSDC and UCL described in Note 3.1 of the consolidated financial statements for the year ended December 31, 2021, 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. Subsequently, it was agreed, that as an alternative to the issuance of shares, a cash equivalent of the value of the Bonus Shares may be paid instead. No aggregate costs incurred by KLSDC and UCL in January 2022.

**NOTE 9 – DEFERRED TAX ASSETS**

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 period ended June 30, 2022, the Company has unrecognized net tax loss carry-forwards in the Danish entity in the amount of DKK 2,127 million which is equal to a deferred tax asset of DKK 468 million as of June 30, 2022.

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.

## NOTE 10 – EQUITY

The following table summarizes the Company's share activity:

	<u>Ordinary Shares</u>
<b>December 31, 2020</b>	<b>34,697,703</b>
Issuance of Bonus Shares as part of license agreement	22,553
Issuance of Matching Shares	170,131
Issuance of sign-on bonus shares to former CEO, Kim Stratton	58,000
Issuance of shares due to exercise of restricted share-units	3,854
<b>June 30, 2021</b>	<b>34,952,241</b>
<b>December 31, 2021</b>	<b>34,952,241</b>
Issuance of shares related to at the market offering in US	360,000
<b>June 30, 2022</b>	<b>35,312,241</b>

In January 2021, the Company issued 22,553 bonus shares to KLSDC and UCL under the terms of the license agreement, discussed in note 8 above.

In January 2021, the Company issued 170,131 Matching Shares to participants in the 2020 LTIP, discussed in note 8 above.

In February 2021, the Company issued 58,000 new shares to former CEO, Kim Stratton following the service agreement, discussed in note 8 above.

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, discussed in note 8 above.

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 USD 835,668.00.

Following the above activity, the total nominal share capital of the Company as of June 30, 2022 was DKK 35,312,241, representing 35,312,241 ordinary shares each with a nominal value of DKK 1.

Following the above activity, the total nominal share capital of the Company as of June 30, 2021, was DKK 34,952,241, representing 34,952,241 ordinary shares each with a nominal value of DKK 1.

## NOTE 11 – PROFIT/LOSS PER SHARE

The following reflects the net loss attributable to shareholders and share data used in the basic and diluted loss per share computations for the six months ended June 30, 2022 and 2021:

	<b>Six months ended June 30, 2022 DKK (000)</b>	<b>Six months ended June 30, 2021 DKK (000)</b>
Profit/loss for the period	62,983	(463,780)
Loss from continuing operations	(24,446)	(55,002)
Net result from discontinued operations	87,429	(408,778)
Weighted-average shares outstanding	35,250,241	34,903,711
<b>Profit/loss per share</b>	<b>1.79</b>	<b>(13.27)</b>
Continuing operations	(0.69)	(1.58)
Discontinued operations	2.48	(11.71)

Basic profit/loss per share is calculated by dividing the net profit/loss attributable to ordinary shareholders for the period by the weighted-average number of ordinary shares outstanding during each period. Diluted profit/loss per share is calculated by dividing the net loss by the weighted-average number of ordinary shares outstanding during the period increased by the dilutive effect of the assumed issuance of any outstanding share-based awards. Due to the fact that the share-based programs are considered to be "out-of-the money" as at June 30, 2022 and due to the fact that Orphazyme has incurred losses for the comparative period presented, the potential shares issuable related to outstanding equity awards have been excluded from the calculation of diluted loss per share. Therefore, basic and diluted loss per share are the same for each period presented.

#### NOTE 12 – COMMITMENTS AND CONTINGENCIES

##### ***Putative class action lawsuit***

On July 9, 2021, a putative class action lawsuit, captioned *Busic v. Orphazyme A/S, et al.*, No. 21- cv-03640 (N.D. Ill.), was filed against the Company and certain of its current and former directors and officers in the U.S. District Court for the Northern District of Illinois. On September 13, 2021, the Court appointed a lead plaintiff for the putative class. On November 19, 2021, the lead plaintiff filed an amended complaint asserting claims under the U.S. Securities Act of 1933 and the U.S. Securities Exchange Act of 1934 for alleged misrepresentations and/or omissions in the Company's registration statement for its IPO of ADSs in the United States and in subsequent public statements. The amended complaint seeks an unspecified amount of damages on behalf of a putative class comprised of all persons and entities other than the defendants that purchased or otherwise acquired the ADSs in connection with the IPO in September 2020 and/or between September 29, 2020 and November 4, 2021. On August 11, 2022, the Court granted in part and denied in part defendants' motion to dismiss the amended complaint. The Court dismissed all claims under the Securities Act of 1933 relating to the Company's registration statement for its IPO of ADSs and dismissed as inactionable a number of challenged statements for the lead plaintiff's claims under the Securities Exchange Act of 1934, including all of the challenged statements prior to June 2021. On August 26, 2022, the Court amended its August 11, 2022 order to also dismiss certain claims against certain individual defendants. The Court's dismissal of claims in its August 11 and 26, 2022 orders was without prejudice to the filing of a further amended complaint. On September 8, 2022, the lead plaintiff filed a second amended complaint with new allegations regarding an April 2021 statement by the Company's CEO at the time. Defendants have until October 6, 2022 to respond to the second amended complaint. Management does not believe these claims have any merit and believe that the outcome will not materially affect the Company's financial position.

##### ***KLSDC and UCL Bonus shares requested***

As part of the license agreement with Kansas Life Sciences Development Company, Inc ("KLSDC") and UCL Business PLC ("UCL"), 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. As of January 31, 2022, no costs were incurred by KLSDC and UCL in connection with such license agreement following discontinuation of the clinical study. As of March 7, 2022, KLSDC and UCL requested Bonus Shares representing a value of USD 284,919 from the Company. The Company has not agreed to deliver such Bonus Shares and has rejected such request. The Management believes that the outcome of this disagreement will not materially affect the Company's financial position.

#### NOTE 13 – RELATED PARTIES

Orphazyme's related parties are Orphazyme A/S' subsidiaries, Orphazyme U.S. Inc. and Orphazyme Schweiz GmbH, Board of Directors, Executive Management, and close

members of the family of these persons.

During the six-month period ended June 30, 2022, remuneration to Executive Management and the Board of Directors was made in line with the Company's remuneration policy. There were no material related party transactions during the first half of 2022.

#### NOTE 14 – SUBSEQUENT EVENTS

No events have occurred in the period from the balance sheet date until the date of release of this interim report that would materially affect an evaluation of the interim report.

## **Statement by the Board of Directors and the Executive Management**

The Board of Directors and the Executive Management have today reviewed and approved the interim financial report of Orphazyme A/S for the period January 1 - June 30, 2022. The interim financial report has not been reviewed or audited by the Company's independent auditors.

The interim financial report for the period January 1 - June 30, 2022 has been prepared in accordance with IAS 34 *Interim Financial Reporting*. The accounting policies used in the interim financial report are consistent with those accounting policies used in Orphazyme's 2021 Annual Report with the additions described and additional Danish disclosure requirements for interim reports of listed companies.

In our opinion, the interim condensed consolidated financial statements give a true and fair view of Orphazyme's assets, liabilities, and financial position at June 30, 2022 and of the results of its operations and cash flows for the period January 1 - June 30, 2022. Furthermore, in our opinion, Management's Review gives a true and fair account of the development and performance of the Group's activities.

Copenhagen, September 26, 2022.

### **Board of Directors**

Bo Jesper Hansen  
*Chairman of the Board*

Anders Vadsholt

John Sommer Schmidt

### **Executive Management**

Anders Vadsholt  
*Chief Executive Officer and Chief Financial Officer*

**Disclaimer**

This company announcement may contain certain forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995, including in respect of the company's anticipated operating plans and performance, financial position and ability to operate as a going concern. 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, including actions by regulatory agencies, effects of the global COVID-19 pandemic, and the impact of the Company's Sale of Assets. 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. In light of these risks and uncertainties, and other risks and uncertainties that are described in the Risk Factors section and other sections of the Company's Annual Report on Form 20-F, filed with the Securities and Exchange Commission (SEC) on March 2, 2021, and other filings that the Company makes with the SEC from time to time (which are available at <http://www.sec.gov>), the events and circumstances discussed in such forward-looking statements may not occur, and the Company's actual results could differ materially and adversely from those anticipated or implied thereby. Although the Company's forward-looking statements reflect the good faith judgment of its management, these statements are based only on facts and factors currently known by the Company. As a result, you are cautioned not to rely on these forward-looking statements. 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.