Company announcement
No. 45/2020

Orphazyme A/S
Ole Maaløes Vej 3
DK-2200 Copenhagen N
CVR No.: 32266355

Interim Report First Half 2020

Copenhagen, Denmark, August 28, 2020 – Orphazyme A/S (ORPHA.CO), a late-stage biopharmaceutical company pioneering the Heat-Shock Protein response for the treatment of neurodegenerative orphan diseases, today announces its Interim Report First Half 2020 for the period January 1–June 30, 2020.

Pipeline Highlights First Half 2020

- Reported positive data from 12-month open-label phase 2/3 extension clinical trial in Niemann-Pick disease Type C (NPC) showing sustained effect of arimoclomol in reducing disease progression over two years
- Announced a U.S. Early Access Program for NPC
- Initiated rolling submission of New Drug Application (NDA) for arimoclomol with the U.S. Food and Drug Administration (FDA) in NPC, which was subsequently completed in July 2020
- Reported top-line data from a phase 2 clinical trial in Gaucher disease Type 1 and Type 3 demonstrating marked improvements in key clinical markers following 6 months of treatment with arimoclomol
- Received U.S. Fast-Track Designation for arimoclomol in Amyotrophic Lateral Sclerosis (ALS)

Financial and Business Highlights First Half 2020

- For the first six months of 2020, Orphazyme reported a net loss of DKK 251.4 million or DKK 9.88 per share (basic and diluted) compared to a net loss of DKK 163.9 million or DKK 8.20 per share (basic and diluted) for the same period in 2019
- Research and development expenses for the period totaled DKK 167.0 million compared to DKK 141.7 million for the same period in 2019, as our pipeline continued to advance through late-stage trials
- General and administrative expenses for the period totaled DKK 78.6 million compared to DKK 23.3 million for the same period in 2019 due to build-up of our commercial organization as well as related support functions in anticipation of the potential approval of arimoclomol in NPC
- Completed offering of 7,032,937 shares in a directed issue and private placement, raising approximately DKK 745 million in gross proceeds (approximately USD 110 million)
- As of June 30, 2020, Orphazyme held cash totaling DKK 610.4 million compared to DKK 225.6 million as of June 30, 2019 and DKK 123.6 million as of December 31, 2019

Subsequent Events

- Completed rolling submission of NDA for arimoclomol with U.S. FDA in NPC
- Announced confidential submission of draft registration statement for a potential registered public offering of American Depositary Shares in the United States

- Announced collaboration with The Michael J. Fox Foundation on Parkinson's disease research, joining its Research Tools Consortium which brings together experts from the medical community and industry to identify and develop new tools to address unmet research needs in Parkinson's disease

"During the first half of the year, we made significant progress in our efforts to bring our first product candidate to market and in July completed the submission of a rolling New Drug Application (NDA) to the FDA for arimoclomol to NPC. We are pleased to be able to already provide patients in the U.S. access to this much needed treatment for NPC through an Early Access Program, which we initiated during Q1. We continue to work at pace in building a highly specialized commercial organization in anticipation of potential approval in NPC in the U.S. and in preparing to submit a marketing application for NPC to regulatory authorities in Europe during the second half of this year", said Kim Stratton, Chief Executive Officer of Orphazyme. *"Our belief that arimoclomol has pipeline-in-a-product potential was also further underlined during the first half of this year with the phase 2 data readout supporting further studies in Gaucher disease and the award by the U.S. regulators of Fast-Track Designation for arimoclomol for the treatment of ALS. Also importantly, the first half of this year saw a boost to our efforts in building a strong international biopharmaceutical company that can maximize the value of arimoclomol, with a successful private placement raising gross proceeds of DKK 745 million (~USD 110 million). We believe the next twelve months hold significant potential for Orphazyme as we near the anticipated commercial launch of arimoclomol, if approved, in its first indication and as we look forward to data readouts from pivotal stage clinical trials testing arimoclomol in ALS and sporadic Inclusion Body Myositis (sIBM) during H1 2021."*

Outlook
The company maintains its 2020 outlook as announced on February 28, 2020. Net operating expenses are anticipated to be in the range of DKK 500–550 million and our cash position at year-end 2020 is anticipated to be greater than DKK 300 million.

Conference Call
Orphazyme will host an investor call at which Chief Executive Officer, Kim Stratton, and Chief Financial Officer, Anders Vadsholt, will present the Interim Report First Half 2020. The presentation will be followed by a Q&A session.

The call will be held on **Friday, August 28, 2020 at 2.00 PM CET/8.00 AM ET.**

Dial-in details:

- Denmark: +45 3272 8042
- United States: +1 6315 107 495
- Standard International: +44 (0) 2071 928000
- United Kingdom: +44 (0) 8445718892
- France: +33 (0) 176700794
- Netherlands: +31 (0) 207143545
- Sweden: +46 (0) 850692180

Event Title: Orphazyme Interim Report First Half 2020

Confirmation code: **5555608**

The presentation will also be available via webcast: *https://edge.media-server.com/mmc/p/guyn7ubp*

After the call, the presentation will be available via the webcast link above.

Condensed Consolidated Key Figures

DKK (000)	As of and for the six-months ended Jun 30, 2020	As of and for the six-months ended Jun 30, 2019	As of and for the year ended Dec 31, 2019
Statement of profit or loss and other comprehensive income			
Research and development expenses	(166,980)	(141,710)	(285,413)
General and administrative expenses	(78,575)	(23,345)	(50,541)
Operating loss	**(245,555)**	**(165,055)**	**(335,954)**
Net financial items	(7,841)	(1,348)	(7,043)
Loss before tax	**(253,396)**	**(166,403)**	**(342,997)**
Income tax benefit	1,981	2,495	5,500
Net loss for the period	**(251,415)**	**(163,908)**	**(337,497)**
Total comprehensive loss	**(251,550)**	**(163,927)**	**(337,430)**
Loss per share, basic and diluted (DKK)	(9.88)	(8.20)	(16,87)
Statement of financial position			
Intangible assets	10,773	10,500	10,539
Right-of-use assets	15,542	11,706	13,903
Property, plant, and equipment	4,247	2,643	3,685
Corporation tax receivable	5,500	5,500	2,750
Prepayments and deposits	2,108	2,534	1,652
Non-current assets	38,170	32,883	32,529
Cash	610,448	225,560	123,588
Other current assets	27,742	20,479	24,637
Total assets	676,360	278,922	180,754
Share capital	27,045	19,984	19,984
Equity	506,135	224,824	52,969
Non-current liabilities	48,419	9,663	61,797
Current liabilities	121,806	44,435	65,988
Cash flow statement			
Net cash used in operating activities	(204,169)	(166,597)	(326,818)
Net cash used in investing activities	(1,760)	(1,225)	(3,285)
Net cash provided by financing activities	692,944	(1,061)	58,939
Other			
Share price (DKK)	89.30	58.00	72.40
Total outstanding shares	27,044,929	19,984,799	19,984,799
Market capitalization (DKK million)[1]	2,415.1	1,159.1	1,446.9
Equity ratio[2]	74.8%	80.6%	29%
Equity per share (DKK)[3]	18.71	11.25	2.65
Average number of employees	106	66	74
Number of full-time employees (FTEs) at end of period	114	72	86

[1] Market capitalization 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.

Outlook

In millions of DKK	2020 guidance	2019 actual operating loss
Operating loss	(500) – (550)	(336)
Cash position at year-end	>300	124

Orphazyme is maintaining its anticipated 2020 outlook as published on February 28, 2020.

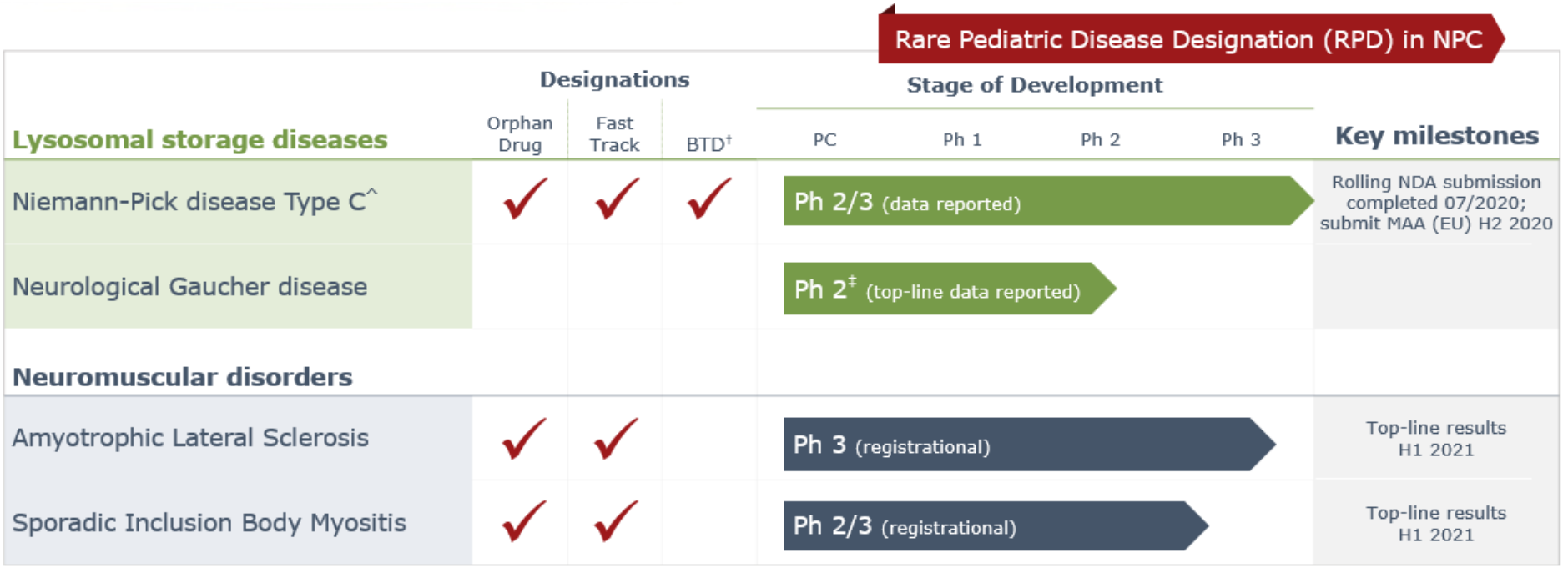
Risks and assumptions
As of June 30, 2020, Orphazyme held total cash of DKK 610.4 million, which is anticipated to cover our planned clinical development and pre-launch commercial activities well into 2021. For the remainder of the year, we will continue to incur costs associated with preparing for potential launch of arimoclomol in NPC, as well as ongoing clinical development activities. Our outlook assumes increasing spend on launch preparation activities in the second half of 2020. We continuously monitor our cash position to identify risks relating to our liquidity needs and, in the future, may seek to raise new capital. If we are not successful in gaining access to new funding, or such funding is not on satisfactory terms, our future plans and our ability to execute our strategy could be adversely affected and we may need to take measures to limit or implement planned activities in several stages.

COVID-19
At this time, there is no material impact on the Company's consolidated financial statements, including the judgements and estimates applied. Our business, operations, and clinical development plans could be adversely impacted by the effects of COVID-19. Our clinical studies are continuing, however with expected increased total costs for the clinical studies arising from the implications of COVID-19. The COVID-19 pandemic may also have an effect on other aspects of our business, including: Our third-party manufacturers, CROs, and other third parties; the productivity of our staff; ability to attract, integrate, manage and retain qualified personnel or key employees; our global supply chains and relationships with vendors and other parties; significant disruption of global financial markets; and reduced ability to secure additional funding. We will continue to monitor the COVID-19 pandemic and its potential impact on our business and financials.

Priorities H2 2020 and beyond

Priority	Targeted milestone	Estimated timing
NPC registration and launch	• Submit New Drug Application U.S. • Submit Marketing Authorization Application in EU	• Completed rolling NDA in July 2020 • H2 2020
Expand beyond NPC	• Phase 2/3 trial top-line results for sIBM • Phase 3 trial top-line results for ALS	• H1 2021 • H1 2021

Product Pipeline



^Early access program in US underway; † Breakthrough Therapy Designation; ‡ Type 1 and Type 3 Gaucher disease

Niemann-Pick disease Type C (NPC)
Orphazyme reported positive results from a phase 2/3 clinical trial of arimoclomol in NPC in January 2019 and additional positive results from the open-label extension clinical trial in January 2020. In the trial, including the extension, arimoclomol was observed to be well-tolerated and had a clinically meaningful effect on slowing the disease progression in NPC. Based on the data from the phase 2/3 clinical trial, Orphazyme completed a rolling submission of an NDA in the United States in July 2020 and plans to submit a Marketing Authorization Application (MAA) in Europe in the second half of 2020 for arimoclomol as a treatment for NPC.

Gaucher disease
Orphazyme initiated a randomized, double-blinded, dose-ranging phase 2 clinical trial of arimoclomol in patients with Gaucher disease Type 1 and 3 naïve to therapy in June 2018. Positive top-line results were reported in June 2020, in which arimoclomol was observed to be well-tolerated and demonstrated a relative reduction in serum chitotriosidase activity from baseline to six months, the primary endpoint, across all dosages compared to placebo, although statistical significance was not achieved. However, a statistically significant and clinically meaningful dose-dependent reduction in liver size ranging from -15% to -20% relative to placebo (dose trend analysis $p<0.05$) was observed, in addition to a clinically meaningful dose-dependent reduction in spleen size ranging from -5 to -21% relative to placebo. Based on these results, Orphazyme intends to advance into pivotal-stage clinical development for arimoclomol in neurological Gaucher disease. An extension of the phase 2 trial is currently ongoing across the three weight-adjusted dose levels and Orphazyme will continue to evaluate clinical outcomes, monitor safety, and further explore relevant biomarkers.

Amyotrophic Lateral Sclerosis (ALS)
Orphazyme initiated a phase 3 trial in August 2018 to support the application for a marketing authorization in ALS. The phase 3 ALS trial is an 18-month, placebo-controlled trial including 245 patients randomized 2:1 to arimoclomol 400mg three times per day or placebo. The primary endpoint is a combined assessment of function and survival (CAFS). Top-line results are expected in H1 2021. The trial design and trial patient baseline characteristics were defined based on systemic analysis of data from the largest publicly available repository of ALS clinical trial data (PROACT) in conjunction with arimoclomol ALS trial data.

Sporadic Inclusion Body Myositis (sIBM)
A multicenter, randomized 1:1, double blind, placebo-controlled phase 2/3 clinical trial investigating efficacy and safety of arimoclomol 400 mg three times per day compared to placebo in patients with sIBM was initiated in August 2017 in the U.S. and UK. The trial, which is intended to support the registration of arimoclomol for the treatment of sIBM, is a 20-month trial with the Inclusion Body Myositis-Functional Rating Scale (IBMFRS) as a primary endpoint. Top-line results from the phase 2/3 trial are expected in H1 2021.

New Molecular Entities (NMEs)
Orphazyme is actively developing a proprietary suite of next generation Heat-Shock Protein (HSP) amplifiers and lysosome biology-targeting compounds and intends to select protein-misfolding diseases for these NMEs based on genetic and mechanistic insights. For new indications and molecule development, Orphazyme plans to continue to closely collaborate with academic experts and patient organizations and intends to leverage its learnings to inform a selection of additional indications involving related biological mechanisms.

Financial Review

Income statement
The net result for the first six months of 2020 was a loss of DKK 251.5 million compared to a loss of DKK 163.9 million for the same period in 2019. The increased net loss was primarily due to higher research and development expenses as our pipeline continued to advance, as well as commercial launch activities included under general and administrative expenses.

Research and development expenses
Research and development (R&D) expenses totaled DKK 167.0 million for the first six months of 2020 compared to DKK 141.7 million for the same period in 2019. The increase of DKK 25.3 million was mainly attributable to an increase of DKK 14.2 million for the initiation of three clinical pharmacology registration trials in H1 2020 and an increase of DKK 11.1 million in R&D employee costs due to the increase in full-time research and development employees from 60 on June 30, 2019 to 77 FTEs on June 30, 2020.

General and administrative expenses
General and administrative expenses totaled DKK 78.6 million for the first six months of 2020 compared to DKK 23.3 million for the same period in 2019. The increase of DKK 55.3 million was primarily due to the build-up of our commercial organization as well as expenses related to our support functions.

Pre-launch expenses represented DKK 40.4 million of the DKK 55.3 million increase, which was mainly due to the escalation of commercial launch preparation activities, including the strengthening of our U.S.-based and Switzerland-based commercial team of 12 additional full-time employees; and an increase in medical affairs activities, particularly for NPC, as we further engaged with the scientific community through our communication and education programs.

Administrative expenses represented the remaining DKK 14.9 million of the DKK 55.3 million increase, which was mainly due to audit, legal, investor relations, other external assistance, and share-based payment expenses; and the hiring of 10 additional administrative, finance, and legal full-time employees to support our growing organization.

Net financial items
Net financial items totaled an expense of DKK 7.8 million for the first six months of 2020 compared to an expense of DKK 1.3 million for the same period in 2019. The increase of DKK 6.5 million was mainly related to the Loan Agreement with Kreos, including interest expense of DKK 5 million and an increase of DKK 0.7 million related to the change in fair value of the facilitation fee accounted for as an embedded call option. The remaining DKK 0.8 million increase results from interest paid on cash balances in the bank due to negative interest rates.

Income tax benefit
Income tax benefit totaled DKK 2.0 million for the first six months of 2020 compared to DKK 2.5 million for the same period in 2019. Income tax benefits 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 2020 has been reduced by an income tax expense in our subsidiaries in the U.S. and Switzerland.

Statement of financial position

Cash
As of June 30, 2020, Orphazyme held cash of DKK 610.4 million compared to DKK 123.6 million as of December 31, 2019. The increase was a result of the cash inflow from the financing completed in February, net of transaction costs.

Equity
As of June 30, 2020, equity amounted to DKK 506.1 million compared to DKK 53.0 million as of December 31, 2019. Similar to our cash balance, the decrease reflects operating loss since December 31, 2019.

Cash flows

Cash flow from operating activities
Net cash flow from operating activities amounted to an outflow of DKK 204.2 million in the six-month period ended June 30, 2020 compared to DKK 166.6 million in the six-month period ended June 30, 2019. Net cash flow from operating activities was attributable primarily to the progression of clinical development activities, as well as commercial launch preparation activities.

Cash flow from investing activities
Net cash flow from investing activities amounted to an outflow of DKK 1.8 million in the six-month period ended June 30, 2020 compared to DKK 1.2 million in the six-month period ended June 30, 2019. The increase in investing activities mainly comprises the capitalization of our new ERP system.

Cash flow from financing activities
Net cash flow from financing activities amounted to an inflow of DKK 692.9 million in the six-month period ended June 30, 2020 compared to an outflow of DKK 1.1 million in the six-month period ended June 30, 2019. The increase reflects the net cash inflow from the financing completed in February.

Consolidated Statements of Profit or Loss and Other Comprehensive Income

	Six months ended Jun 30, 2020 DKK (000)	Six months ended Jun 30, 2019 DKK (000)
Research and development expenses (Note 3)	(166,980)	(141,710)
General and administrative expenses (Note 4)	(78,575)	(23,345)
Operating loss	**(245,555)**	**(165,055)**
Financial income	126	152
Financial expenses	(7,967)	(1,500)
Loss before tax	**(253,396)**	**(166,403)**
Income tax benefit	1,981	2,495
Net loss for the period	**(251,415)**	**(163,908)**
Exchange differences from translation of foreign operations	(135)	(19)
Total comprehensive loss	**(251,550)**	**(163,927)**
Loss per share, basic and diluted (Note 8)	(9.88)	(8.20)

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

Consolidated Statements of Financial Position

	Jun 30, 2020 DKK (000)	Dec 31, 2019 DKK (000)
ASSETS		
Non-current assets		
Intangible assets	10,773	10,539
Right-of-use assets	15,542	13,903
Property, plant, and equipment	4,247	3,685
Corporation tax receivable	5,500	2,750
Prepayments and deposits	2,108	1,652
Total non-current assets	**38,170**	**32,529**
Current assets		
Corporation tax receivable	-	5,500
Prepayments and other receivables	27,742	19,137
Cash	610,448	123,588
Total current assets	**638,190**	**148,225**
TOTAL ASSETS	**676,360**	**180,754**
EQUITY & LIABILITIES		
Equity		
Share capital	27,045	19,984
Share premium	1,611,630	924,021
Other reserves	5,753	7,982
Accumulated deficit	(1,138,293)	(899,018)
Total equity	**506,135**	**52,969**
Non-current liabilities		
Borrowings	36,827	51,606
Lease liabilities	10,976	9,813
Other non-current liabilities	616	378
Total non-current liabilities	**48,419**	**61,797**
Current liabilities		
Current borrowings	29,954	12,813
Lease liabilities	3,183	2,876
Trade payable and accruals	58,769	32,390
Tax payable	665	-
Other liabilities	29,235	17,909
Total current liabilities	**121,806**	**65,988**
TOTAL EQUITY AND LIABILITIES	**676,360**	**180,754**

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

Consolidated Statements of Changes in Shareholders' Equity

	Share capital DKK (000)	Share premium DKK (000)	Other reserves: Foreign currency translation reserve DKK (000)	Other reserves: Share-based compensation – acquisition of intangible assets DKK (000)	Accumulated deficit DKK (000)	Total DKK (000)
Balance as of December 31, 2018	**19,939**	**924,021**	**42**	**9,070**	**(564,823)**	**388,249**
Net loss for the period	-	-	-	-	(163,908)	(163,908)
Other comprehensive loss for the period	-	-	(19)	-	-	(19)
Total other comprehensive loss	-	-	*(19)*	-	*(163,908)*	*(163,927)*
Transactions with owners						
Capital increase, Bonus Shares	26	-	-	(1,197)	1,171	-
Capital increase, LTIP Matching Shares	19	-	-	-	-	19
Share-based payment costs	-	-	-	-	483	483
Total transactions with owners	*45*	-	-	*(1,197)*	*1,654*	*502*
Balance as of June 30, 2019	**19,984**	**924,021**	**23**	**7,873**	**(727,077)**	**224,824**

	Share capital DKK (000)	Share premium DKK (000)	Other reserves: Foreign currency translation reserve DKK (000)	Other reserves: Share-based compensation – acquisition of intangible assets DKK (000)	Accumulated deficit DKK (000)	Total DKK (000)
Balance as of December 31, 2019	**19,984**	**924,021**	**109**	**7,873**	**(899,018)**	**52,969**
Net loss for the period	**-**	**-**	**-**	**-**	(251,415)	(251,415)
Other comprehensive loss for the period	**-**	**-**	(135)	**-**	**-**	(135)
Total other comprehensive loss	-	-	*(135)*	-	*(251,415)*	*(251,550)*
Transactions with owners						
Capital increase, Bonus Shares (Note 7)	21	-	-	(2,094)	2,073	-
Capital increase, exercise of RSUs (Note 7)	7	394	-	-	-	401
Capital increase, private placement (Note 7)	7,033	738,458	-	-	-	745,491
Transaction costs (Note 7)	-	(51,243)	-	-	-	(51,243)
Share-based payment costs (Note 5)	-	-	-	-	10,067	10,067
Total transactions with owners	*7,061*	*687,609*	-	*(2,094)*	*12,140*	*704,716*
Balance as of June 30, 2020	**27,045**	**1,611,630**	**(26)**	**5,779**	**(1,138,293)**	**506,135**

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

Consolidated Statements of Cash Flow

	Six months ended Jun 30, 2020 DKK (000)	Six months ended Jun 30, 2019 DKK (000)
Operating activities		
Operating loss	(245,555)	(165,055)
Adjustments to reconcile loss before tax to cash flows from operating activities:		
Equity-settled share-based payment expense (Note 5)	10,067	483
Depreciation and amortization	2,250	2,066
Change in prepayments, deposits and other receivables	(9,061)	8,197
Change in trade payables, accruals and other liabilities	37,400	(11,384)
Corporate taxes received / (paid)	5,500	(255)
Interest paid	(4,770)	(649)
Net cash used in operating activities	**(204,169)**	**(166,597)**
Investing activities		
Purchase of intangible assets	(590)	(112)
Purchase of property, plant, and equipment	(1,170)	(1,113)
Net cash used in investing activities	**(1,760)**	**(1,225)**
Financing activities		
Proceeds from issuance of shares (Note 7)	745,892	19
Transaction costs	(51,243)	-
Repayment of lease obligations	(1,705)	(1,080)
Net cash provided by (used in) financing activities	**692,944**	**(1,061)**
Net change in cash and cash equivalents	**487,015**	**(168,883)**
Cash balance at beginning of period	123,588	**394,706**
Effect of changes in exchange rates on cash	(155)	(263)
Cash balance at end of period	**610,448**	**225,560**

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

Notes to the Financial Statements

NOTE 1 – CORPORATE INFORMATION

Orphazyme A/S (the "Company") is a late-stage biopharmaceutical company harnessing the amplification of Heat Shock Proteins, or HSPs, in order to develop and commercialize novel therapeutics for the treatment of neurodegenerative orphan diseases. The Company is a limited liability company publicly traded on Nasdaq Copenhagen with headquarters in Copenhagen, Denmark. In April 2018, a wholly-owned subsidiary, Orphazyme U.S., Inc., was incorporated in Massachusetts, USA and in March 2020, a wholly-owned subsidiary, Orphazyme Schweiz GmbH, was incorporated in Zug, Switzerland (together with Orphazyme A/S and Orphazyme U.S., Inc., "Orphazyme" or "the Group"). By establishing local subsidiaries, the Company aims to directly support the U.S. and European markets and establish closer relationships with the medical, patient, and financial communities as Orphazyme expands its development programs and global reach.

NOTE 2 – BASIS OF PREPARATION AND UPDATES TO THE GROUP'S ACCOUNTING POLICIES

Basis of preparation
The interim condensed consolidated financial statements for the six months ended June 30, 2020 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 Orphazyme A/S' latest consolidated annual financial statements as of December 31, 2019.

Evaluation of Going Concern Basis

The Company periodically monitors its funding position in order to identify risks relating to future liquidity needs and to ensure that it has access to sufficient funds to continue its activities as planned or alternatively take corrective actions to allow the Company to continue as a going concern. Management continuously evaluates various funding options for the Company, public or private debt or equity financings and believes it is probable that new funding will be obtained to enable the Company to continue its activities associated with the ongoing clinical trials and the escalation of activities to prepare for commercial launch as planned. If, contrary to management's expectations, the Company is not successful in getting access to new funding, the Company may down-size or delay planned activities to allow the Company to fund operations to at least June 30, 2021. Based on these factors, management considers it appropriate to prepare these financial statements on a going concern basis.

COVID-19

At this time, there is no material impact on the Company's consolidated financial statements, including the judgements and estimates applied. Our business, operations and clinical development plans could be adversely impacted by the effects of COVID-19. Our clinical studies are continuing, however with expected increased total costs for the clinical studies arising from the implications of COVID-19. The COVID-19 pandemic may also have an effect on other aspects of our business, including: our third-party manufacturers, CROs and other third parties; the productivity of our staff; ability to attract, integrate, manage and retain qualified personnel or key employees; our global supply chains and relationships with vendors and other parties; significant disruption of global financial markets; and reduced ability to secure additional funding. We will continue to monitor the COVID-19 pandemic and its potential impact on our business and financials.

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 A/S' annual consolidated financial statements for the year ended December 31, 2019. A number of new or amended standards became applicable for the current reporting period; however, these did not have an impact on the interim condensed consolidated financial statements of the Group.

NOTE 3 – RESEARCH AND DEVELOPMENT EXPENSES

Research and development expenses of DKK 167 million (June 30, 2019: DKK 141.7 million) include employee costs for research and development personnel (i.e. salaries, bonuses, employer contributions to pension schemes, share-based compensation) and external costs for further development of our pipeline, including ongoing clinical trials and clinical pharmacology registration trials.

NOTE 4 – GENERAL AND ADMINISTRATIVE EXPENSES

General and administrative expenses include pre-launch costs of DKK 44.1 million (June 30, 2019: DKK 3.7 million) associated 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 and medical affairs activities to further engage with the scientific community through communication and education programs. Furthermore, included in general and administrative expenses is DKK 34.5 million (June 30, 2019: DKK 19.6 million) mainly due to 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.

NOTE 5 – SHARE-BASED COMPENSATION COSTS

Please refer to Note 2.5 of the Group's consolidated financial statements included in the 2019 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:

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

2020 Long-term incentive program granted after the balance sheet date:

Similar to the 2017 LTIP and 2019 LTIP described in Note 2.5 of the consolidated financial statements for the year ended December 31, 2019, the 2020 LTIP offers the Executive Management as well as certain key employees of Orphazyme who have subscribed for a certain number of Investment Shares, an equal number of Matching Shares and a certain number of Performance Shares depending on the development of the Company's share price. Although the grant date of these awards is subsequent to the balance sheet date, management has determined that the service commencement date of the participants was during the period January 1 and June 30, 2020 and consequently compensation expense is recognized from the respective service commencement date of the participants. An estimate of the fair value of the awards was made at the balance sheet date for the purpose of recognizing the services received from the service commencement date through June 30, 2020. Once the grant date is established for all the awards, the estimate of the fair value as of June 30, 2020 will be revised so that the amounts recognized for services received in respect of the grant of awards are ultimately based on the grant date fair value of the equity instruments awarded.

The provisional fair value of the 2020 LTIP awards was estimated using a Monte-Carlo simulation model at the balance sheet date. The following inputs were used in the simulation:

	June 30, 2020
Dividend yield (%)	-
Expected volatility (%)	55.9%
Risk-free interest rate (%)	(0.56%)
Expected life (years)	3.5
Share price (DKK)	89.30
Provisional fair value at measurement date (DKK 000)	38,556

The risk-free interest rate 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. Based on the provisional fair value of the 2020 LTIP, an expense of DKK 8.2 million was recognized for the six-month period ending June 30, 2020. In addition, an aggregate expense of DKK 1.9 million for the 2017 LTIP and the 2019 LTIP was recognized for the six-month period ending June 30, 2020 compared to DKK 0.5 million recognized for the six-month period ended June 30, 2019.

In July 2020, the Matching Shares from the 2019 LTIP fully vested and were issued to the participants in exchange for the nominal value of DKK 1 per share. This resulted in cash received of DKK 31,250 and a capital increase of the same number of shares.

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

As described in Note 2.5 of the consolidated financial statements for the year ended December 31, 2019, the Phantom programs are cash-settled and their fair value is re-assessed at each reporting date. Management used a Monte-Carlo simulation model with the following inputs to estimate the fair value as of June 30, 2020:

	June 30, 2020	June 30, 2020	June 30, 2020
Program	2019-1	2018-2	2018-1
Dividend yield (%)	-	-	-
Expected volatility (%)	56%	58.5%	58.5%
Risk-free interest rate (%)	(0.55%)	(0.56%)	(0.56%)
Expected life (years)	3.64	2.58	2.58
Share price (DKK)	89.30	89.30	89.30
Fair value at measurement date (DKK 000)	437	72	215

	Dec 31, 2019	Dec 31, 2019	Dec 31, 2019
Program	2019-1	2018-2	2018-1
Dividend yield (%)	-	-	-
Expected volatility (%)	57.4%	57%	57%
Risk-free interest rate (%)	(0.50%)	(0.63%)	(0.63%)
Expected life (years)	4	3.08	3.08
Share price (DKK)	72.40	72.40	72.40
Fair value at measurement date (DKK 000)	347	53	152

The risk-free interest rate 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. Based on the fair value of the awards on June 30, 2020, an expense of DKK 0.2 million was recognized for the six-month period ended June 30, 2020 compared to DKK 0.1 million recognized for the six-month period ended June 30, 2019.

c) Restricted share units (cash-settled)

In March 2020, the 2019 RSUs granted to the board of directors in 2019 fully vested. During April, three board members exercised their RSUs, one exercised in July, subsequent to the reporting date, and the remaining participants have not exercised. See Note 7 for a description of the share issuances. The remaining RSUs expire in March 2021 if not exercised or paid out in cash.

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 2019 RSUs described in Note 2.5 of the consolidated financial statements for the year ended December 31, 2019. Management used a Black-Scholes model with the following inputs to estimate the fair value of the 2020 RSUs as of June 30, 2020:

	June 30, 2020
Dividend yield (%)	-
Expected volatility (%)	55.3%
Risk-free interest rate (%)	(0.48%)
Expected life (years)	1
Share price (DKK)	89.30
Fair value at measurement date (DKK 000)	420

The risk-free interest rate 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. Based on the fair value of the 2020 RSUs on June 30, 2020, an expense of DKK 0.1 million was recognized for the six-month period ended June 30, 2020. In addition, an amount of DKK 0.1 million was recognized for the 2019 RSUs during the same period. For the six-month period ended June 30, 2019 there was no expense recognized for the 2019 RSUs, as they were only granted in July 2019.

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, 2019, 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 at December 31, 2019 the aggregate costs incurred by KLSDC and UCL amounted to USD 0.3 million (DKK 2.2 million), and a total of 20,650 Bonus Shares ("2020 Bonus Shares") were issued to KLSDC and UCL in January 2020, based on the average 30-day closing price of Orphazyme's shares. 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.

NOTE 6 – FINANCIAL LIABILITIES

As disclosed in Note 3.5 of the consolidated financial statements for the year ended December 31, 2019, the structured debt facility with Kreos ("Loan Agreement") entered into in August 2019 includes a Facilitation Fee that is due and payable by Orphazyme at the sole discretion of the lender. 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 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 payment request notification by the lender 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. The call option is measured at fair value at level 2 in the fair value hierarchy 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 and the term of the option.

The table below shows the inputs used in the valuation of the call option and the estimated fair value on June 30, 2020 and December 31, 2019:

	June 30, 2020	**Dec 31, 2019**
Dividend yield (%)	-	-
Expected volatility (%)	58.5%	57%
Risk-free interest rate (%)	(0.56%)	(0.63%)
Expected life (years)	2.67	3.2
Share price (DKK)	89.30	72.40
Fair value of call option (DKK 000)	2,263	1,595

The change in fair value of the call option is recognized as a finance expense in the statement of profit or loss. Based on the fair value of the call option on June 30, 2020, an expense of DKK 0.7 million was recognized for the period. As the Loan Agreement was executed in August 2019, there was no expense recognized for the six-month period June 30, 2019.

NOTE 7 – 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	26,060
Issuance of Matching Shares as part of 2017 LTIP	19,175
June 30, 2019	**19,984,799**
(No share activity in H2 2019)	-
December 31, 2019	**19,984,799**
Issuance of Bonus Shares as part of license agreement	20,650
Issuance of shares in connection with a private placement offering	7,032,937
Issuance of shares due to exercise of 2019 RSUs by some participants	6,543
June 30, 2020	**27,044,929**

As discussed in Note 5 above, in January 2020 the Company issued 20,650 shares as part of consideration payable to KLSDC and UCL relating to the license agreement.

On February 7, 2020 Orphazyme 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 net proceeds of the directed issue and private placement is expected to support the U.S. and European filings for approval of arimoclomol for the treatment of Niemann-Pick disease Type C (NPC), as well as preparations for commercial launch.

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 a 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 were issued without pre-emption rights for existing shareholders.

The offering of Existing Shares was facilitated by a share loan to the Company from related parties Novo Holdings A/S and Orpha Pooling B.V. (the "Lending Shareholders") 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 were only facilitating the loan of the Lending Shares for purposes of the Company's offering of Existing Shares in the Offering.

As discussed in Note 5 above, in April 2020 certain members of the board of directors exercised their 2019 RSUs and the Company issued 6,543 shares in exchange for DKK 0.4 million.

Following the above activity, the total nominal share capital of the Company as of June 30, 2020 was DKK 27,044,929, representing 27,044,929 ordinary shares each with a nominal value of DKK 1.

Shares issued after the balance sheet date:

Subsequent to June 30, 2020, a board member exercised his 2019 RSUs and the Company issued 3,451 ordinary shares in exchange for DKK 0.4 million.

As mentioned above in Note 5, the Matching Shares from the 2019 LTI Program fully vested in July and the Company issued 31,250 ordinary shares to the participants in exchange for the nominal value of DKK 1 per share, or DKK 31,250.

NOTE 8 – 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, 2020 and 2019:

	Six months ended June 30, 2020 DKK (000)	Six months ended June 30, 2019 DKK (000)
Loss for the period	(251,415)	(163,908)
Weighted-average shares outstanding	25,447,748	19,977,123*
Loss per share, basic and diluted	**(9.88)**	**(8.20)***

*Recalculated retrospectively as a result of Bonus Shares issued in January 2020.

Basic loss per share is calculated by dividing the net loss attributable to ordinary shareholders for the period by the weighted-average number of ordinary shares outstanding during each period. Diluted 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 Orphazyme has incurred losses for each period presented, the potential shares issuable related to outstanding equity awards have been excluded from the calculation of diluted loss per share as the effect of such shares is anti-dilutive. Therefore, basic and diluted loss per share are the same for each period presented.

As disclosed in Note 4.3 of the consolidated financial statements for the year ended December 31, 2019, in January 2020, Bonus Shares were issued to KLSDC and UCL under the terms of the license agreement entered into in October 2017. Basic and diluted loss per share for the comparative period presented has been adjusted retrospectively to include these Bonus Shares in the number of weighted average shares outstanding for the six-months ended June 20, 2019.

NOTE 9 – SUBSEQUENT EVENTS

In July 2020, Orphazyme introduced the 2020 LTI Program as further described in Note 5 above.

Also in July 2020, the Matching Shares from the 2019 LTI Program were issued to the participants and a board member exercised his 2019 RSUs, as further described in Note 7 above.

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, 2020. 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, 2020 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 2019 Annual Report 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, 2020 and of the results of its operations and cash flows for the period January 1-June 30, 2020. Furthermore, in our opinion, Management's Review gives a true and fair account of the development and performance of the Group's activities.

Copenhagen, August 28, 2020.

Board of Directors

Georges Gemayel
Chairman of the Board

Bo Jesper Hansen
Deputy Chairman of the Board

Anders Hedegaard

Catherine Moukheibir

Martijn Kleijwegt

Martin Bonde

Rémi Droller

Sten Verland

Executive Management

Kim Stratton	Anders Vadsholt
Chief Executive Officer	*Chief Financial Officer*

Disclaimer

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