Company announcement
No. 26/2019

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

Interim Report First Half 2019

"We have made substantial progress in the first half of 2019. We confirmed our filing strategy for arimoclomol in Niemann-Pick disease type C (NPC) with both the European and, more recently, the US regulators, and with this major step forward we will be introducing an Early Access Program for NPC during the fall. We are on track to file for our first marketing authorization during H1 2020 and continue our preparations for launch of arimoclomol in key markets. Arimoclomol is also in clinical development in three other indications of very high unmet need: Amyotrophic Lateral Sclerosis (ALS), sporadic Inclusion Body Myositis (sIBM), and Gaucher disease. We were particularly pleased with the great interest in our ALS phase 3 trial, which is now fully enrolled ahead of the expected timeline. This means that both potential registration trials in ALS and sIBM have now completed enrollment and we look forward to reporting headline results from the trials during the first half of 2021. Furthermore, we have completed enrollment in our phase 2 Gaucher disease trial and results are now expected in H1 2020" said Anders Hinsby, Chief Executive Officer of Orphazyme.

Anders Hinsby continued, "*As previously announced, I will hand over the position of Chief Executive Officer of Orphazyme to Kim Stratton on October 1, 2019. The change in leadership will allow the timely transformation of the company to be able to successfully make arimoclomol available to patients after an approval. It has been a great privilege to work closely with talented scientists, dedicated clinicians, and most of all the awe-inspiring patient organizations. I have always, along with our whole team at Orphazyme, been fueled by these close collaborations and the daunting task of matching the courage, commitment, and perseverance of our close collaborators.*"

Business Highlights First Half 2019 and Subsequent Events

- Pivotal trial primary endpoint with arimoclomol for NPC showed 74% reduction in disease progression after 12 months, subgroups showed statistically significant efficacy. Anticipated H1 2020 filing submission in Europe and USA, following positive meetings with regulatory authorities
- Phase 3 trial with arimoclomol for ALS on-going; completed enrollment in July 2019. Topline results from full analysis remain on track for H1 2021
- Phase 2/3 trial with arimoclomol for sIBM on-going; completed enrollment in April 2019. Results from full trial expected H1 2021 (interim analysis expected H1 2020)
- Enrollment of phase 2 Gaucher disease trial completed in August 2019; results expected in H1 2020
- Several leads for new molecular entities that constitute potentially new intellectual property opportunities
- Appointed Kim Stratton as Chief Executive Officer, succeeding Anders Hinsby as of October 1, 2019
- Introduced a new board incentive program, a new share-based incentive program, and a new phantom share program in July 2019
- Strengthened balance sheet in August 2019 with EUR 9 million financing from Kreos Capital

Financial Results First Half 2019

- For the first six months of 2019, Orphazyme reported a net loss of MDKK 164 or DKK 8.21 per share (basic and diluted) compared to a net loss of MDKK 108 or DKK 5.41 per share (basic and diluted) for the same period in 2018
- Research and development expenses for the period totaled MDKK 142 compared to MDKK 94 for the same period in 2018
- General and administrative expenses for the period totaled MDKK 23 compared to MDKK 15 for the same period in 2018
- As of June 30, 2019, Orphazyme held cash totaling MDKK 226 compared to MDKK 513 as of June 30, 2018 and MDKK 395 as of December 31, 2018

Outlook

Orphazyme maintains the 2019 operating loss outlook in the range of DKK 315-345 million and the anticipated cash position at year-end 2019 to be greater than DKK >110 million, as published in the Annual Report of 2018 on March 1, 2019.

Conference Call

Orphazyme will be hosting an investor call at which Chief Executive Officer, Anders Hinsby, and Chief Financial Officer, Anders Vadsholt, will be presenting the Interim Report First Half 2019. The presentation will be followed by a Q&A session.

The call will be held on: **Wednesday, August 28, 2019 at 11.00 AM CEST/5.00 AM EDT.**

Dial-in details:

- Denmark: +45 32 72 80 42
- United Kingdom: +44 (0) 844 571 88 92
- United States: +1 6315 107 495

Event Title: Orphazyme Interim Report First Half 2019

Confirmation code: **8655918**

The presentation will also be available via webcast: https://edge.media-server.com/mmc/p/v2haoix5

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

Condensed Consolidated Key Figures

TDKK	Jun 30, 2019	Jun 30, 2018	Dec 31, 2018
Statement of profit or loss and other comprehensive income			
Research and development expenses	(141,710)	(94,387)	(196,525)
General and administrative expenses	(23,345)	(15,060)	(35,127)
Operating loss	**(165,055)**	**(109,447)**	**(231,652)**
Net financial items	(1,348)	(448)	(3,448)
Loss before tax	**(166,403)**	**(109,895)**	**(235,100)**
Income tax benefit	2,495	2,090	5,500
Net loss for the period	**(163,908)**	**(107,805)**	**(229,600)**
Total comprehensive income	**(163,927)**	**(107,779)**	**(229,558)**
Loss per share, basic and diluted (DKK)	(8.21)	(5.41)	(11,50)
Statement of financial position			
Licenses and intangibles	10,500	9,497	10,744
Right-of-use asset (Note 2)	11,706	-	-
Property, plant, and equipment	2,643	1,866	1,940
Non-current assets	32,883	16,657	17,965
Cash	225,560	513,370	394,706
Other current assets	20,479	14,350	28,678
Total assets	278,922	544,377	441,349
Share capital	19,984	19,940	19,939
Equity	224,824	509,045	388,249
Current liabilities	44,435	35,332	52,995
Cash flow statement			
Net cash used in operating activities	(166,597)	(118,044)	(234,764)
Net cash used in investing activities	(1,225)	(349)	(2,346)
Net cash used in financing activities	(1,061)	-	-
Other			
Share price (DKK)	58.00	61.20	43.35
Total outstanding shares	19,984,799	19,939,564	19,939,564
Market capitalization (MDKK)[1]	1,159.1	1,220.3	864.4
Equity ratio[2]	80.6%	93.5%	88.0%
Equity per share (DKK)[3]	11.25	25.5	19.47
Average number of employees	66	39	46
Number of full-time employees (FTEs) at end of period	72	45	57

[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

MDKK	2019 guidance	2018 actual result
Operating loss	(315)-(345)	(232)
Cash position at year-end	>110	395

Operating result
We maintain our outlook that our 2019 operating loss will be in the range of DKK 315-345 million.

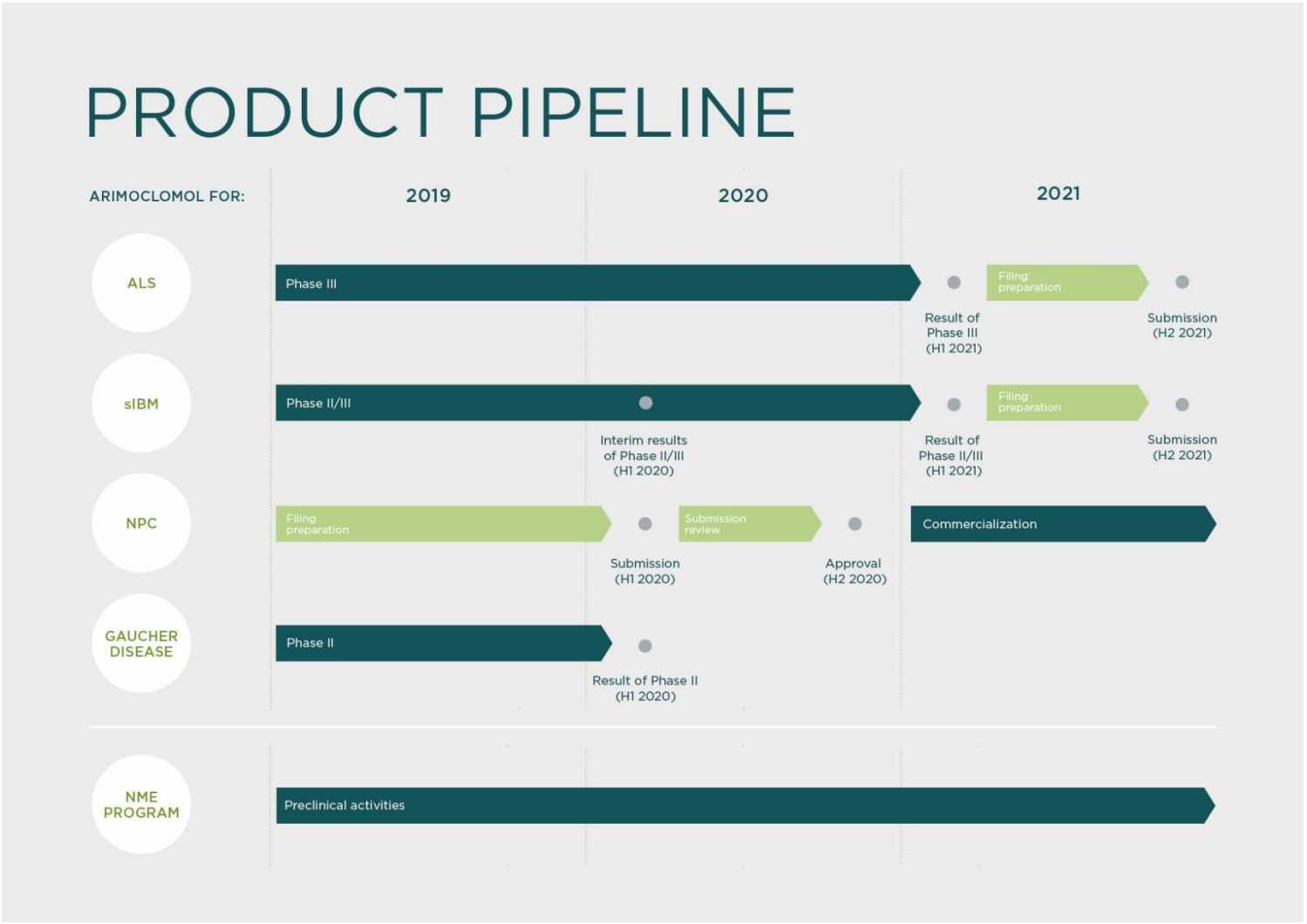
Cash position
We update the outlook on our anticipated cash position at year-end 2019 to be greater than DKK 110 million.

Risks and assumptions
As of June 30, 2019, Orphazyme held total cash of DKK 226 million and for the remainder of the year will continue to incur costs associated with the on-going clinical trials to assess the effect of arimoclomol in the treatment of sIBM, ALS, NPC, and Gaucher disease. In August 2019, the Group entered into a structured debt facility to secure funding of EUR 9 million, as described in Note 4 to the interim financial statements. Various funding alternatives are continuously being considered and therefore, management considers it appropriate to prepare the financial statements on a going concern basis.

2019 Objectives

Priority	√	Targeted milestone
ALS	√	• Complete enrollment in H2
sIBM	√	• Complete enrollment in H1
NPC	√	• Regulatory feedback mid-2019
Gaucher disease	-	• Phase II results in H2 2019 – *now expected H1 2020*
New molecular entities (NME) program		• Preclinical studies with NMEs in protein-misfolding diseases

Product Pipeline



Amyotrophic Lateral Sclerosis
Orphazyme initiated a Phase III trial in August 2018 to support the application for a marketing authorization for arimoclomol in Amyotrophic Lateral Sclerosis (ALS). The Phase III ALS trial, which has been agreed with the regulatory authorities, is an 18-month, randomized, placebo-controlled trial being conducted at 30 centers of excellence in North America and Europe. Enrollment in the trial was completed in July 2019, earlier than anticipated due to support from the global ALS community, patients and care givers. Headline results from the full analysis are expected in H1 2021. The trial design and patient baseline characteristics have been 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. The primary endpoint is determined as a combined assessment of function and survival. Patients completing the Phase III trial are offered participation in an open-label extension trial.

Sporadic Inclusion Body Myositis
In April 2019, Orphazyme completed enrolment in a Phase II/III trial investigating the safety and efficacy of arimoclomol in patients with sporadic Inclusion Body Myositis (sIBM). The trial was initiated in August 2017 in the USA and is intended to support registration of arimoclomol for the treatment of sIBM. The Phase II/III study is a 150-patient, 20-month, randomized, double blind placebo-controlled trial being conducted at 11 centers of excellence in the US and one in the UK. An interim analysis at 12 months is expected to take place in H1 2020, with study completion due by end 2020. Results from the completed study are expected in H1 2021. The 20-month primary analysis timepoint was chosen to maximize chances of success, allowing for greater separation between treatment groups at 20 months, while maintaining the possibility to terminate the trial for efficacy after a 12-month interim readout. Orphazyme has initiated an open-label extension of the sIBM trial which is available to patients who have completed the study.

Niemann-Pick disease Type C
Positive results from the NPC-002 Phase II/III pivotal trial of arimoclomol in Niemann-Pick disease Type C (NPC) were reported in January 2019. The primary endpoint showed a 74% reduction in disease progression after 12 months in patients on routine clinical care randomized to arimoclomol versus placebo (p-value = 0.0506). In a predefined subgroup of patients of 4 years and older, the treatment difference was statistically significant with a minimal progression at month 12 in the arimoclomol-treated group. Biomarker results demonstrated statistically significant biological response to treatment. In mid-2019, Orphazyme confirmed that the Company has initiated filing processes for arimoclomol in NPC in USA and Europe based on positive advice from both the US Food and Drug Administration (FDA) and the European Medicines Agency (EMA). Both agencies were supportive of the available data being adequate for review of a marketing application and, hence, Orphazyme is currently working diligently to enable a regulatory filing as expected in H1 2020, with potential approval in H2 2020.

Gaucher disease
A 6-month, placebo-controlled phase 2 clinical trial of arimoclomol in patients with Gaucher disease was initiated in India in June 2018. 39 patients were randomized 1:1:1:1 into four treatment arms – active treatment at three different doses and placebo. Following the placebo-controlled period, patients will be rolled over to a 6-month extension. The primary endpoint is the percentage change in serum chitotriosidase levels from baseline to 6 months. Enrollment of the phase 2 Gaucher disease trial was completed in August 2019 and results are now expected in H1 2020, which implies a delay to the previously communicated timeline (i.e. H2 2019).

New Molecular Entities
Orphazyme is developing a new series of heat-shock protein (HSP) amplifying drugs based on its expertise and know-how about the convergence of HSPs, protein aggregation, and cellular recycling systems and how these can be targeted for therapeutic benefit. Orphazyme has several leads that constitute potentially new intellectual property opportunities.

Financial Review

Income statement
The net result for the first six months of 2019 was a loss of DKK 163.9 million compared to a loss of DKK 107.8 million for the same period in 2018. The higher loss is primarily due to increased research and development activities as well as general and administrative expenses.

Research and development expenses
Research and development expenses totaled DKK 141.7 million for the first six months of 2019 compared to DKK 94.4 million for the same period in 2018. The increase is primarily due to the steady progression of enrollment in our two largest clinical trials, specifically ALS, which only launched in late H1-2018 and was nearing full enrollment at the end of H1-2019, and sIBM, which was fully enrolled by April 2019. In H1-2019 we also initiated the open label extension trials for ALS and sIBM, which notably added to the R&D expenses. In addition, the various R&D functions have been significantly increased from 40 FTEs on June 30, 2018 to 60 FTEs in June 30, 2019.

General and administrative expenses
General and administrative expenses totaled DKK 23.3 million for the first six months of 2019 compared to DKK 15.1 million for the same period in 2018. The increase is primarily due to the costs of establishing our U.S. subsidiary, which was incorporated in late H1-2018. We also increased our G&A FTE count in Denmark from 6 to 12.

Net financial items
Net financial items totaled an expense of DKK 1.3 million for the first six months of 2019 compared to an expense of DKK 0.4 million for the same period in 2018. The increase is mainly due to interest paid on our cash balance in the bank as well as the interest expense recognized on our office lease liability that is recognized on balance sheet as of January 1, 2019 following the adoption of the new lease accounting standard, IFRS 16.

Income tax benefit
Income tax benefit for both periods is comparable at DKK 2.5 million for the first six months of 2019 compared to DKK 2.1 million for the same period in 2018. 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.

Statement of financial position
Cash
As of June 30, 2019, Orphazyme held cash of DKK 225.6 million compared to DKK 394.7 million as of December 31, 2018. The decrease reflects the operating loss for the period.

Equity
As of June 30, 2019, equity amounted to DKK 224.8 million compared to DKK 388.2 million as of December 31, 2018. Similar to our cash balance, the decrease reflects operating loss.

Cash flows
Cash flow from operating activities
Net cash flow from operating activities amounted to an outflow of DKK 166.6 million in the six-month period ended June 30, 2019 compared to DKK 118 million in the six-month period ended June 30, 2018. Net cash flow from operating activities is attributable primarily to the initiation and progression of clinical development activities, as well as general and administrative expenses.

Cash flow from investing activities
Net cash flow from investing activities amounted to an outflow of DKK 1.2 million in the six-month period ended June 30, 2019 compared to DKK 0.3 million in the six-month period ended June 30, 2018. Investing activities comprise investment in equipment for research and development purposes as well as refurbishment of a new facility.

Cash flow from financing activities
Net cash flow from financing activities amounted to an outflow of DKK 1.1 million in the six-month period ended June 30, 2019 compared to no cash flow attributed to financing activities in the six-month period ended June 30, 2018. The increase is attributable mainly to the repayments of lease obligations following the adoption of the IFRS leases standard.

Statements of Profit or Loss and Other Comprehensive Income

	Six months ended Jun 30, 2019 TDKK	Six months ended Jun 30, 2018 TDKK
Research and development expenses	(141,710)	(94,387)
General and administrative expenses	(23,345)	(15,060)
Operating loss	**(165,055)**	**(109,447)**
Financial income	152	2
Financial expenses	(1,500)	(450)
Loss before tax	**(166,403)**	**(109,895)**
Income tax benefit	2,495	2,090
Net loss for the period	**(163,908)**	**(107,805)**
Other comprehensive income/(loss)	(19)	26
Total comprehensive loss	**(163,927)**	**(107,779)**
Loss per share, basic and diluted (Note 3)	(8.21)	(5.41)

See accompanying notes to these financial statements.

Statements of Financial Position

	Jun 30, 2019 TDKK	Dec 31, 2018 TDKK
ASSETS		
Non-current assets		
Licenses and intangibles	10,500	10,744
Right-of-use asset (Note 2)	11,706	-
Property, plant, and equipment	2,643	1,940
Corporation tax receivable	5,500	2,750
Prepayments and deposits	2,534	2,531
Total non-current assets	**32,883**	**17,965**
Current assets		
Corporation tax receivable	5,500	5,500
Prepayments and other receivables	14,979	23,178
Cash	225,560	394,706
Total current assets	**246,039**	**423,384**
TOTAL ASSETS	**278,922**	**441,349**
EQUITY & LIABILITIES		
Equity		
Share capital	19,984	19,939
Share premium	924,021	924,021
Other reserves	7,896	9,112
Accumulated deficit	(727,077)	(564,823)
Total equity	**224,824**	**388,249**
Non-current liabilities		
Lease liability (Note 2)	9,541	-
Other non-current liabilities	122	105
Total non-current liabilities	**9,663**	**105**
Current liabilities		
Trade payables and accruals	29,705	42,183
Other liabilities	14,730	10,812
Total current liabilities	**44,435**	**52,995**
TOTAL EQUITY AND LIABILITIES	**278,922**	**441,349**

See accompanying notes to these financial statements.

Statements of Changes in Shareholders' Equity

	Share capital TDKK	Share premium TDKK	Other reserves: Foreign currency translation reserve TDKK	Other reserves: Share-based compensation – acquisition of intangible assets TDKK	Accumulated deficit TDKK	Total TDKK
Balance as of December 31, 2017	**19,928**	**924,021**	**-**	**9,972**	**(338,219)**	**615,702**
Net loss for the period	-	-	-	-	(107,805)	(107,805)
Other comprehensive loss for the period	-	-	26	-	-	26
Total other comprehensive income/(loss)	-	-	*26*	-	*(107,805)*	*(107,779)*
Transactions with owners						
Capital increase, subscribed and paid	12	(12)	-	-	-	-
Capital increase, subscribed but not paid	-	-	-	-	-	-
Expenses, capital increase	-	-	-	-	1,122	1,122
Total transactions with owners	*12*	*(12)*	-	-	*1,122*	*1,122*
Balance as of June 30, 2018	**19,940**	**924,109**	**26**	**9,972**	**(444,902)**	**509,045**

	Share capital TDKK	Share premium TDKK	Other reserves: Foreign currency translation reserve TDKK	Other reserves: Share-based compensation – acquisition of intangible assets TDKK	Accumulated deficit TDKK	Total TDKK
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 income/(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**

See accompanying notes to these financial statements.

Statements of Cash Flow

	Six months ended Jun 30, 2019 TDKK	Six months ended Jun 30, 2018 TDKK
Operating activities		
Operating loss	(165,055)	(109,447)
Adjustments to reconcile loss before tax to cash flows from operating activities:		
Share-based payment expense	483	1,122
Depreciation and amortization	2,066	690
Change in prepayments, deposits and other receivables	8,197	1,829
Change in payables, accruals and other liabilities	(11,384)	(11,790)
Corporate taxes received / (paid)	(255)	-
Interest received /(paid)	(649)	(448)
Net cash used in operating activities	**(166,597)**	**(118,044)**
Investing activities		
Investments in intangibles	(112)	-
Investment in property, plant, and equipment	(1,113)	(349)
Net cash used in investing activities	**(1,225)**	**(349)**
Financing activities		
Repayment of lease liability	(1,080)	-
Proceeds from issuance of Matching Shares (LTIP)	19	-
Net cash provided by financing activities	**(1,061)**	**-**
Net change in cash and cash equivalents	**(168,883)**	**(118,393)**
Cash balance at beginning of period	**394,706**	631,735
Net foreign exchange differences	(263)	28
Cash balance at end of period	**225,560**	**513,370**

See accompanying notes to these financial statements.

Notes to the Financial Statements

NOTE 1 – CORPORATE INFORMATION

Orphazyme A/S (the "Company") is a limited liability company incorporated and domiciled in Denmark. The registered office is located in Copenhagen, Denmark. In April 2018, a fully-owned subsidiary, Orphazyme US, Inc., was incorporated in Massachusetts, USA (together with Orphazyme A/S, "Orphazyme" or the "Group"). Orphazyme US, Inc. will directly support the US market to 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, 2019 have been prepared in accordance with IAS 34 Interim Financial Reporting and additional Danish disclosure requirements for interim reports of listed companies.

The interim condensed consolidated financial statements do not include all the information and disclosures required in the annual financial statements and should be read in conjunction with Orphazyme A/S' latest annual financial statements as of December 31, 2018.

As at June 30, 2019, Orphazyme held total cash of DKK 226 million and for the remainder of the year will continue to incur costs associated with the on-going clinical trials to assess the effect of arimoclomol in the treatment of sIBM, ALS, NPC, and Gaucher disease. In August 2019, the Group entered into a structured debt facility to secure funding of EUR 9 million, as described in Note 4. Various funding alternatives are continuously being considered and therefore, management considers it appropriate to prepare the financial statements on a going concern basis.

Updates to the Group's accounting policies
The accounting policies adopted in the preparation of the interim condensed consolidated financial statements are consistent with those followed in the preparation of Orphazyme A/S' annual consolidated financial statements for the year ended December 31, 2018.

On January 1, 2019, Orphazyme adopted IFRS 16, *Leases*, using the modified retrospective method. Under this method, the cumulative effect of initially applying IFRS 16 is recognized at January 1, 2019. Upon adoption, Orphazyme recognized a right-of-use asset and a lease liability based on the present value of the remaining lease payments in the amount of TDKK 13,006 for the office lease previously classified as an operating lease. Since the application of IFRS 16, Orphazyme has recognized finance expense on the lease liability and depreciation expense on the right-of-use asset. As of January 1, 2019, Orphazyme has applied the following accounting policy:

Leases
For contracts which are, or contain, a lease, Orphazyme recognizes a right-of-use asset and a lease liability. The right-of-use asset is initially measured at cost, being the initial amount of the lease liability adjusted for any lease payments made at or before the commencement date. The right-of-use asset is subsequently depreciated using the straight-line method over the lease term. The right-of-use asset is periodically reduced by impairment losses, if any, and adjusted for certain remeasurements of the lease liability. The lease liability is initially measured at the present value of the lease payments outstanding at the commencement date, discounted using Orphazyme's incremental borrowing rate. The lease liability is measured using the effective interest method. It is remeasured when there is a change in future lease payments, typically due to a change in index or rate (e.g. inflation) on property leases, or if there is a reassessment of whether an extension or termination option will be exercised. A corresponding

adjustment is made to the right-of-use asset, or in the income statement when the right-of-use asset has been fully depreciated. Right-of-use assets and non-current liabilities are presented on separate line items in the Group's consolidated statement of financial position. The current portion of lease liabilities is presented under other liabilities. Lease contracts that have a lease term of 12 months or less and low value assets are not recognized on the balance sheet. These lease payments are expensed on a straight-line basis over the lease term.

NOTE 3 – LOSS PER SHARE

The following reflects the net loss attributable to shareholders and share data used in the basic and diluted earnings/(loss) per share computations for the six months ended June 30, 2019 and 2018:

	Six months ended Jun 30, 2019 TDKK	**Six months ended Jun 30, 2018 TDKK**
Loss for the period	(163,908)	(107,805)
Weighted-average shares outstanding	19,956,473	19,937,794
Loss per share	**(8.21)**	**(5.41)**

Basic loss per share amounts are calculated by dividing the net earnings/(loss) attributable to ordinary shareholders for the period by the weighted average number of ordinary shares outstanding during each period. 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. Equity instruments that could potentially dilute basic earnings per share in the future are those under share-based incentive programs.

NOTE 4 – SUBSEQUENT EVENTS

Management has evaluated its financial statements for potential subsequent events occurring after the balance sheet date of June 30, 2019 and prior to the date that these financial statements were issued and found the following events to disclose:

In July 2019, Orphazyme introduced a four-year phantom share-based incentive program for employees (the "2019 Phantom Shares Program") that will be settled in cash in January 2024. The 2019 Phantom Shares Program has the same terms and conditions as the phantom share-based incentive program introduced in June 2018 and described in the 2018 Annual Report.

In July 2019, Orphazyme introduced a Long-term Incentive Program ("2019 LTIP") for the Company's executive management and certain key employees. The 2019 LTIP is largely based on the LTIP introduced after the Company's IPO in November 2017 and described in the 2018 Annual Report.

Also in July 2019, Orphazyme introduced a new Board incentive program for the Company's members of the board of directors. The Board incentive program is based on restricted share units ("RSUs"), which entitle the participant to acquire or subscribe for shares in the Company corresponding to the number of RSUs held at a price per share corresponding to the volume weighted average share price of the Company's shares as quoted on Nasdaq Copenhagen A/S during the ten trading days preceding the date of

grant. The RSUs will have a vesting period from the date of grant until the date of the next annual general meeting of the Company following the date of grant. Vesting is not conditional on any financial or performance criteria, but is, however, conditional upon the continued membership by the participant in the Company's Board of Directors. Upon vesting, the RSUs may be exercised within a period of twelve months.

The above-mentioned incentive programs will be accounted for in accordance with the Group's share-based payment accounting policy as described in the 2018 Annual Report.

Loan Agreement

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

In addition, the lender may, at any time in its sole discretion in eight years, depending on certain events defined in the Loan Agreement, notify the Company that a Facilitation Fee is due and payable ("Notification"). The Facilitation Fee is an amount equal to the greater of (i) 10% of the aggregate amount of the amount borrowed and (ii) the percentage increase in the Company's share price between the 30-day volume-weighted average share price on the date of the Loan Agreement and the closing share price on the day immediately preceding the date of the notification applied to the aggregate amount of amounts borrowed.

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 condensed consolidated financial statements of Orphazyme A/S for the period January 1-June 30, 2019. These interim condensed consolidated financial statements have not been reviewed or audited by the Group's independent auditors.

The interim condensed consolidated financial statements for the period January 1-June 30, 2019 have been prepared in accordance with IAS 34 *Interim Financial Reporting*. The accounting policies adopted in the preparation are consistent with those accounting policies applied in the 2018 Annual Report of Orphazyme, except for the adoption of new, amended or revised IFRS standards and interpretations as published by the IASB that are endorsed by the European Union effective January 1, 2019. This includes IFRS 16, *Leases*, applied using the modified retrospective method. Furthermore, the financial report for the first six months of 2019 and Management's Review are prepared in accordance with 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 the Group's assets, liabilities, and financial position at June 30, 2019 and of the results of the Group's operations and cash flows for the period January 1-June 30, 2019. 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, 2019.

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

Anders Hinsby	Anders Vadsholt
Chief Executive Officer	*Chief Financial Officer*

Forward-looking statement

This company announcement may contain certain forward-looking statements. Although the Company believes its expectations are based on reasonable assumptions, all statements other than statements of historical fact included in this company announcement about future events are subject to (i) change without notice and (ii) factors beyond the Company's control. These statements may include, without limitation, any statements preceded by, followed by, or including words such as "target," "believe," "expect," "aim," "intend," "may," "anticipate," "estimate," "plan," "project," "will," "can have," "likely," "should," "would," "could", and other words and terms of similar meaning or the negative thereof. Forward-looking statements are subject to inherent risks and uncertainties beyond the Company's control that could cause the Company's actual results, performance, or achievements to be materially different from the expected results, performance, or achievements expressed or implied by such forward-looking statements. 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.