Company announcement
No. 11/2018

Interim Report First Half 2018

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Orphazyme A/S
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The first part of 2018 has taken Orphazyme further along the path of bringing arimoclomol to rare disease patients in great need of new treatment options. The clinical development programs remain on track, with two major clinical milestones met by the initiation of the Gaucher Phase II trial and the ALS Phase III trial. Another major milestone is coming up in the very near term, with the reporting of top-line results from the Phase II/III trial of arimoclomol for the treatment of Niemann-Pick disease Type C (NPC).

Business Highlights First Half 2018

- Arimoclomol was granted Rare Pediatric Disease Designation for the treatment of NPC, entailing eligibility for a Priority Review Voucher
- A US subsidiary was established in Massachusetts, USA, to establish closer relationships with the medical, patient, and financial communities, headed by Chief Commercial Officer, Paul Merrigan
- First patient dosed in arimoclomol Phase II trial in Gaucher disease. Results expected in H1 2019

Subsequent Events

- First patient dosed in Phase III trial of arimoclomol for the treatment of Amyotrophic Lateral Sclerosis (ALS)

"*The first half of 2018 has been very productive, and I am pleased that we have met our milestones with the progress of our clinical programs. We will keep working hard to maintain the forward momentum and remain fully dedicated to developing therapies for orphan diseases with high unmet needs. We are now looking forward to presenting the top-line results from our NPC trial, which are expected within weeks*", says Anders Hinsby, Chief Executive Officer of Orphazyme.

Financial Results First Half 2018

- For the first six months of 2018, Orphazyme reported a net loss of MDKK 108, or DKK 5.41 per share (basic and diluted) compared to a net loss of MDKK 52, or DKK 5.09 per share (basic and diluted) for the same period in 2017
- Research and development costs for the period totaled MDKK 94 compared to MDKK 47 for the same period in 2017
- General and administrative expenses for the period totaled MDKK 15 compared to MDKK 8 for the same period in 2017
- As of June 30, 2018, Orphazyme had cash and cash equivalents totaling MDKK 513 compared to MDKK 34 as of June 30, 2017 and MDKK 632 as of December 31, 2018

Outlook

Orphazyme maintains the 2018 financial guidance published in the Annual Report 2017 on March 15, 2018.

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 2018. The presentation will be followed by a Q&A session.

The call will be held on: **Tuesday, August 28, 2018 at 11.00 AM CET.**

Dial-in details:

- Denmark: +45 35 15 81 21
- United Kingdom: +44 (0) 330 336 9411
- United States: +1 929-477-0324

Event Title: Orphazyme Interim Report First Half 2018

Confirmation code: **2088051**

The presentation will also be available via webcast: https://edge.media-server.com/m6/p/5hejv92c

After the call, the presentation will be available by using the following dial-in details:

- Denmark: +45 70 14 50 87
- United Kingdom: +44 (0) 207 660 0134
- United States: +1 719-457-0820

Confirmation code: 2088051

Condensed Consolidated Key Figures

TDKK	Jun 30, 2018	Jun 30, 2017	Dec 31, 2017
Statement of profit or loss and other comprehensive income			
Research and development costs	(94,387)	(46,870)	(99,048)
General and administrative expenses	(15,060)	(7,972)	(31,994)
Operating loss	**(109,447)**	**(54,842)**	**(131,042)**
Net financial items	(448)	(122)	(662)
Loss before tax	**(109,895)**	**(54,964)**	**(131,704)**
Income tax benefit	2,090	2,841	5,500
Net loss for the period	**(107,805)**	**(52,123)**	**(126,204)**
Total comprehensive income	**(107,779)**	**(52,123)**	**(126,204)**
Loss per share, basis (DKK)	5.41	5.09	10.46
Statement of financial position			
Licenses	9,497	-	9,853
Property, plant, and equipment	1,866	1,007	1,851
Investment in property, plant, and equipment	349	311	1,491
Non-current assets	16,657	6,955	14,864
Cash and cash equivalents	513,370	33,589	631,735
Other current assets	14,350	105,168	16,218
Total assets	544,377	145,712	662,817
Share capital	19,940	5,103	19,928
Equity	509,045	121,297	615,702
Current liabilities	35,332	24,415	47,115
Cash flow statement			
Cash flow from operating activities	(118,044)	(45,307)	(95,426)
Cash flow from investing activities	(349)	(311)	(1,491)
Cash flow from financing activities	-	64,858	714,303
Other			
Share price (DKK)[1]	61.20	-	76.00
Total outstanding shares	19,939,564	5,102,210	19,928,184
Market capitalization (MDKK)[2]	1,220.3	-	1,514.5
Equity ratio[3]	93.5%	83.2%	92.9%
Equity per share (DKK)[4]	25.5	23.77	30.90
Average number of employees	39	24	26
Number of employees at end of period	45	28	34

[1] There is no official share price for the reporting periods prior to 2017 since the Company went public in November 2017.

[2] Market capitalization is calculated as the share price multiplied with the total outstanding shares as of the balance sheet date.

[3] Equity ratio is calculated as the equity divided by the total assets as of the balance sheet date.

[4] Equity per share is calculated as the total equity divided by the total outstanding shares as of the balance sheet date.

Outlook

MDKK	2018 guidance	2017 actual result
Operating loss	(245)-(275)	(126)
Cash position at year-end*	>350	632

*Cash and cash equivalents

Operating result
We anticipate that our 2018 operating loss will be in the range of DKK 245-275 million.

Cash position
At year-end 2018, we anticipate a cash position of DKK >350 million.

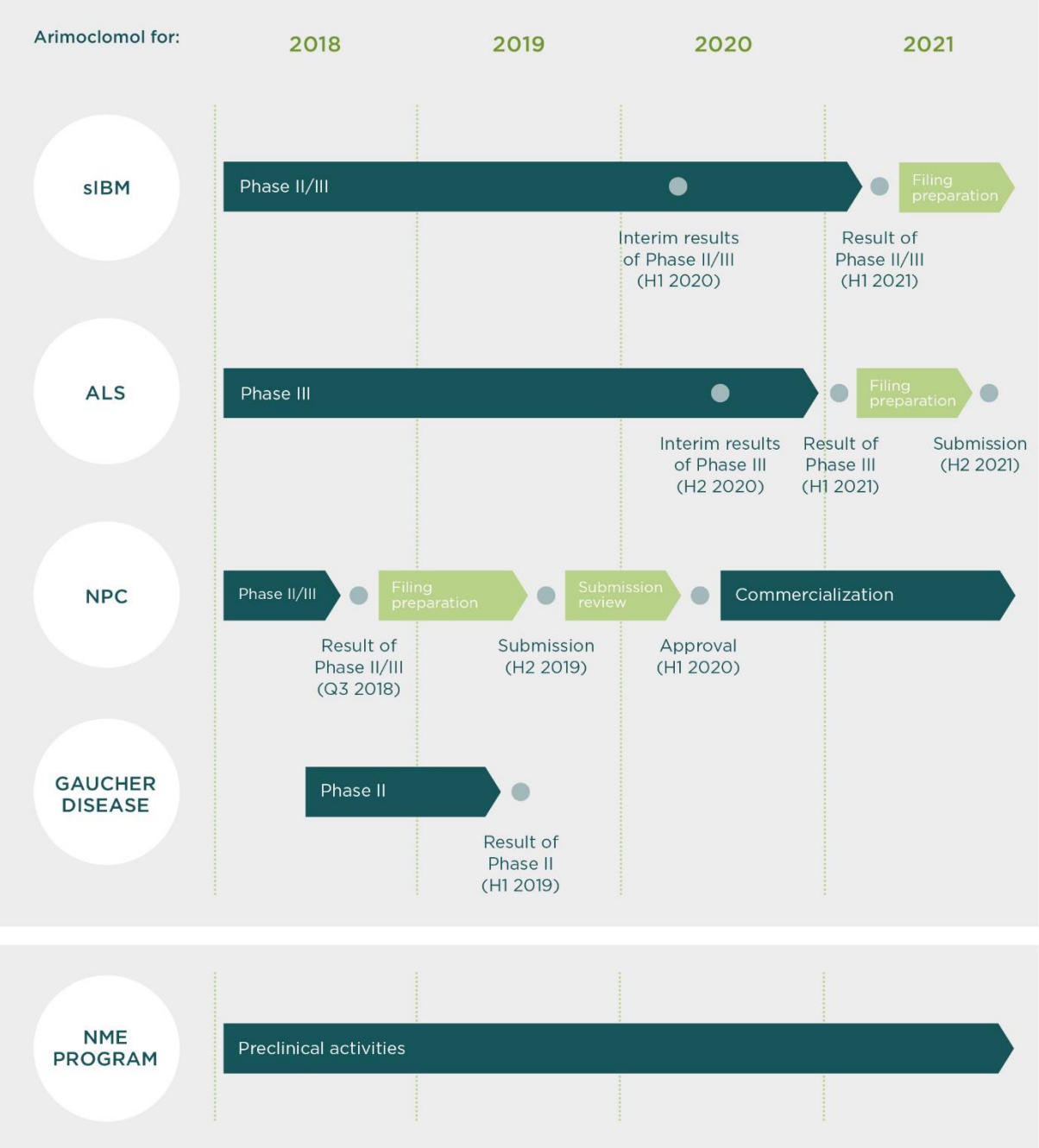
Risks and assumptions
For the financial year ending December 31, 2018, Orphazyme expects to incur substantial costs associated with clinical trials. The objective of the development programs is to develop a pharmaceutical drug for the treatment of the following diseases: Sporadic Inclusion Body Myositis (sIBM), ALS, NPC, and Gaucher disease.

The forecasting of costs associated with clinical trials relating to activities performed by Clinical Research Organizations (CROs) and other external vendors requires management to exercise significant estimates in regard to the timing and accounting for these costs. The diverse nature of the services being provided by CROs and other arrangements, the different compensation arrangements that exists for each type of service, and the limitations in respect of information related to certain clinical activities, add complexity to the estimation of accruals for services rendered by CROs and other vendors in connection with clinical trials. Furthermore, certain CROs and vendors are paid partly upfront in connection with clinical activities. The outlook for the financial year ending December 31, 2018 takes into consideration the trial designs for the respective product candidates, as to the activities planned for 2018.

2018 Objectives

Priority	√	Targeted milestone
sIBM		• Enroll patients in both USA and Europe • Complete Phase II/III trial enrollment by year-end
ALS	√	• Initiate Phase III trial in Q3
NPC		• Phase II/III topline results in Q3
Gaucher disease	√	• Initiate Phase II trial in Q2 • Complete trial enrollment before year-end
NME program		• Preclinical studies with new molecular entities

Product Pipeline



Sporadic Inclusion Body Myositis

A Phase II/III arimoclomol trial for sporadic Inclusion Body Myositis (sIBM) was initiated in August 2017 in the USA. The trial is intended to support a registration of arimoclomol for the treatment of sIBM. All clinical sites in both the USA and EU are expected to be enrolling patients by Q4 2018. The trial is a 20-month trial with a 12-month interim analysis. Interim results are expected in H1 2020. If positive, the trial may be deemed completed at this point in time for efficacy. In case the trial continues, final study results are expected in H1 2021. The 20-month primary analysis timepoint was made 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.

Amyotrophic Lateral Sclerosis

Orphazyme initiated a Phase III trial in August 2018 to support the application for a marketing authorization in Amyotrophic Lateral Sclerosis (ALS). As previously communicated, the Phase III ALS trial design has been agreed upon with the regulatory authorities: 18-month, placebo-controlled trial including 212 patients. Interim analysis at 70% completion in H2 2020 and full analysis 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. The primary endpoint is determined as a combined assessment of function and survival.

Niemann-Pick disease Type C

Data from the observational trial, NPC-001, showed a disease progression rate, which confirms the assumptions used to design the Phase II/III trial and candidate biomarkers were analyzed, confirming their potential use as disease biomarkers. Phase II/III trial enrollment was completed, and the last patient switched to the open-label extension study, in Q2 2018 and we are currently in the process of preparing the data set for analysis. This process includes data collection, checking the data set for completeness and errors to ensure that we get all relevant data compiled, and communicating with the health authorities to make sure that we are aligned in expectations before unblinding the data base and performing the analysis that will provide the top-line results. Top-line results are still expected to be reported in Q3 2018.

Gaucher disease

A Phase II clinical trial of arimoclomol for Gaucher disease was initiated in June 2018. The trial takes place at clinical sites in India and will include approximately 40 patients. Patients will be randomized 1:1:1:1 into four treatment arms – active treatment at three different doses and placebo. The patients will receive arimoclomol or placebo-controlled treatment for six months. Following the placebo-controlled period, the placebo group will be re-randomized into one of the three active treatment groups for a six-month extension. Results are expected in H1 2019.

New Molecular Entities

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

Financial Review

Income statement
The net result for the first six months of 2018 was a loss of DKK 107.8 million compared to a loss of DKK 52.1 million for the same period in 2017. The increased loss is primarily due to increased research and development activities as well as general and administrative expenses.

Research and development costs
Research and development costs totaled DKK 94.4 million for the first six months of 2018 compared to DKK 46.9 million for the same period in 2017. The increase is primarily due to the launch of the Phase II/III sIBM study, as well as preparation costs and the launch of the Phase II trial in Gaucher and preparation costs for the Phase III trial in ALS. Furthermore, the R&D organization has been increased in order to handle four clinical trials, compared to only one clinical trial in the same period in 2017. Lastly, arimoclomol manufacturing costs increased from 2017 to 2018.

General and administrative expenses
General and administrative expenses totaled DKK 15.1 million for the first six months of 2018 compared to DKK 8.0 million for the same period in 2017. The increase is primarily due to establishing a subsidiary in the US, including expanding management with an additional member. Furthermore, transitioning into a publicly-traded company has resulted in a growing organization with significant increases in costs.

Net financial items
Net financial items totaled an expense of DKK 0.4 million for the first six months of 2018 compared to a loss of DKK 0.1 million for the same period in 2017.

Income tax benefit
Income tax benefit totaled DKK 2.1 million for the first six months of 2018 compared to DKK 2.8 million for the same period in 2017. 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 and cash equivalents
As of June 30, 2018, Orphazyme had cash and cash equivalents of DKK 513.4 million compared to DKK 631.7 million as of December 31, 2017. The decrease reflects the operating loss as well as payment of bank fees from the IPO.

Equity
As of June 30, 2018, equity amounted to DKK 509.0 million compared to DKK 615.7 million as of December 31, 2017. The decrease reflects operating loss.

Cash flows
Cash flow from operating activities
Net cash flow from operating activities amounted to an outflow of DKK 118.0 million in the six-month period ended June 30, 2018 compared to DKK 45.3 million in the six-month period ended June 30, 2017. 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 0.3 million in the six-month period ended June 30, 2018 compared to DKK 0.3 million in the six-month period ended June 30, 2017. 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 0 million in the six-month period ended June 30, 2018 compared to DKK 64.9 million in the six-month period ended June 30, 2017.

Statement of Profit or Loss and Other Comprehensive Income

	Six months ended Jun 30, 2018 TDKK	Six months ended Jun 30, 2017 TDKK
Research and development expenses	(94,387)	(46,870)
General and administrative expenses	(15,060)	(7,972)
Operating loss	**(109,447)**	**(54,842)**
Financial income	2	-
Financial expenses	(450)	(122)
Loss before tax	**(109,895)**	**(54,964)**
Income tax benefit	2,090	2,841
Net loss for the period	**(107,805)**	**(52,123)**
Other comprehensive income/(loss)	26	-
Total comprehensive loss	**(107,779)**	**(52,123)**
Loss per share, basic and diluted (Note 5)	5.41	5.09

See accompanying notes to these financial statements.

Statement of Financial Position

	Jun 30, 2018 TDKK	Dec 31, 2017 TDKK
ASSETS		
Non-current assets		
Licenses	9,497	9,853
Property, plant, and equipment	1,866	1,851
Corporation tax receivable	4,840	2,750
Leasehold deposits	454	410
Total non-current assets	**16,657**	**14,864**
Current assets		
Corporation tax receivable	5,500	5,500
Other receivables	3,290	5,871
Prepayments	5,560	4,847
Cash and cash equivalents	513,370	631,735
Total current assets	**527,720**	**647,953**
TOTAL ASSETS	**544,377**	**662,817**
EQUITY & LIABILITIES		
Equity		
Share capital (Note 4)	19,940	19,928
Share premium	924,131	924,143
Share-based compensation – acquisition of intangible rights	9,972	9,972
Foreign currency translation reserve	26	-
Accumulated deficit	(445,024)	(338,341)
Total equity	**509,045**	**615,702**
Current liabilities		
Trade payables	27,024	13,436
Other payables	8,308	33,679
Total current liabilities	**35,332**	**47,115**
TOTAL EQUITY AND LIABILITIES	**544,377**	**662,817**

See accompanying notes to these financial statements.

Statement of Changes in Shareholders' Equity

	Share capital TDKK	Share premium TDKK	Foreign currency translation reserve TDKK	Share-based compensation – acquisition of intangible assets TDKK	Accumulated deficit TDKK	Total TDKK
Balance as of Dec 31, 2016	**3,361**	**226,285**	**-**	**-**	**(212,137)**	**17,509**
Net loss for the period	-	-	-	-	(52,123)	(52,123)
Other comprehensive loss for the period	-	-	-	-	-	-
Total other comprehensive income/(loss)	-	-	-	-	*(52,123)*	*(52,123)*
Transactions with owners						
Capital increase, subscribed and paid	1,742	63,689	-	-	-	65,431
Capital increase, subscribed but not paid	-	91,319	-	-	-	91,319
Expenses, capital increase	-	(839)	-	-	-	(839)
Total transactions with owners	*1,742*	*154,169*	-	-	-	*155,911*
Balance as of Jun 30, 2017	**5,103**	**380,454**	**-**	**-**	**(264,260)**	**121,297**

	Share capital TDKK	Share premium TDKK	Foreign currency translation reserve TDKK	Share-based compensation – acquisition of intangible assets TDKK	Accumulated deficit TDKK	Total TDKK
Balance as of Dec, 2017	**19,928**	**924,143**	**-**	**9,972**	**(338,341)**	**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 (Note 4)	**12**	**(12)**	**-**	**-**	**-**	**-**
Share-based payment costs (Note 3)	**-**	**-**	**-**	**-**	**1,122**	**1,122**
Total transactions with owners	-	-	-	-	*1,122*	*1,122*
Balance as of Jun 30, 2018	**19,940**	**924,131**	**26**	**9,972**	**(445,024)**	**509,045**

See accompanying notes to these financial statements.

Statement of Cash Flows

	Six months ended Jun 30, 2018 TDKK	Six months ended Jun 30, 2017 TDKK
Operating activities		
Operating loss	(109,447)	(54,842)
Adjustments to reconcile loss before tax to cash flows from operating activities:		
Share-based payment expense (Note 3)	1,122	-
Depreciation and amortization	690	291
Change in other receivables	2,539	(1,730)
Change in prepayments	(710)	1,379
Change in trade payables	13,582	2,896
Change in other payables	(25,372)	6,821
Corporate taxes received	-	-
Interest received/(paid)	(448)	(122)
Net cash used in operating activities	**(118,044)**	**(45,307)**
Investing activities		
Investment in property, plant, and equipment	(349)	(311)
Net cash used in investing activities	**(349)**	**(311)**
Financing activities		
Capital contributions from shareholders	-	65,431
Costs related to capital contributions	-	(573)
Net cash provided by financing activities	**-**	**64,858**
Net change in cash and cash equivalents	**(118,393)**	**19,240**
Cash and cash equivalents at beginning of period	631,735	14,349
Net foreign exchange differences	28	
Cash and cash equivalents at end of period	**513,370**	**33,589**

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. On November 16, 2017, the Company successfully completed its Initial Public Offering (IPO) on Nasdaq Copenhagen by issuing 7,500,000 new ordinary shares for gross proceeds of TDKK 600,000.

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, 2018 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, 2017.

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 financial statements for the year ended December 31, 2017.

On January 1, 2018, the Group adopted IFRS 9, "Financial Instruments", which did not have a significant impact on the interim condensed consolidated financial statements. In addition, there have been amendments to IFRS 2, *Share-Based Payment*, which the Group adopted, but did not have any impact on the Group's consolidated financial statements. The Group has not early adopted any other standard, interpretation, or amendment that has been issued but is not yet effective.

Due to the incorporation of Orphazyme US, Inc. in April 2018, the Group has adopted a consolidation policy and due to the establishment of a phantom share-based incentive program in June 2018, the Group has updated its share-based payment accounting policy, as follows:

Consolidated financial statements
The consolidated financial statements include Orphazyme A/S (the "Parent Company") and Orphazyme US, Inc., a fully-owned subsidiary over which the Parent Company has control. A parent company controls an entity when the parent company (i) is exposed to, or has rights to, variable returns from its involvement with the entity, (ii) has power over the entity (i.e. existing rights that give it the current ability to direct the activities of the entity), and (iii) has the ability to use its power to affect the returns of the entity.

The Parent Company re-assesses whether or not it controls an entity if facts and circumstances indicate that there are changes to one or more of the three elements of control. Consolidation of an entity begins when the Parent Company obtains control and ceases when the Parent Company has lost control of the entity.

Orphazyme US, Inc. has adopted the accounting policies of the Parent Company and therefore the Group's consolidated financial statements have been prepared by combining similar accounting items on a line-by-line

basis. On consolidation, intercompany income and expenses, intercompany receivables, and payables, and unrealized gains and losses on transactions between the consolidated companies are eliminated.

The functional currency of Orphazyme US, Inc. is the US dollar (USD) and the presentation currency of the Group is Danish kroner (DKK). On consolidation, the assets and liabilities of Orphazyme US, Inc are translated from USD to DKK at the exchange rate in effect at the balance sheet date and the statement of profit or loss and other comprehensive income is translated from USD to DKK at the date of the underlying transaction or average exchange rate of the period if there are no significant fluctuations in exchange rate throughout the period. The exchange rate differences arising on translation for consolidation are recognized in other comprehensive income (loss).

Share-based payment
Employees and Management of the Group receive remuneration in the form of both equity-settled and cash-settled awards. The cumulative expense recognized at each reporting date until the vesting date reflects the extent to which the vesting period has expired and the Group's best estimate of the number of instruments that will ultimately vest. The expense or credit in the statement of profit or loss for a period represents the movement in cumulative expense recognized as at the beginning and end of that period.

The warrants granted prior to the IPO and the long-term incentive program ("LTIP") are equity-settled awards. The fair value of these awards is determined at the date of grant, resulting in a fixed fair value at grant date that is not adjusted for future changes in the fair value of the awards that may occur over the service period. Fair value of warrants granted prior to the IPO has been determined using the Black-Scholes model. Fair value of the LTIP awards granted after the IPO has been determined using the Monte-Carlo model. Further details of the valuation models are presented in Note 3.

The phantom share-based incentive program established in June 2018 is cash-settled. A liability is recognized for the fair value of these awards, which is measured initially and at each reporting date up to and including the settlement date, with changes recognized at each reporting date. The fair value is expensed over the period until vesting date with recognition of a corresponding liability. The fair value is determined using the Monte-Carlo model, further details of which are presented in Note 3.

The cost of share-based payments in recognized as an expense together with a corresponding increase in equity over the period in which the performance and/or service conditions are fulfilled. In the event that equity instruments are granted conditionally upon an equal number of equity instruments granted in prior periods not being exercised, they are treated as a new grant for the current period and a modification of the equity instruments granted in the prior period.

The fair value of equity-settled awards with service conditions and non-market performance conditions is reported as compensation expense pro rata over the service period to the extent such awards are estimated to vest. The fair value of the cash-settled awards, which vest subject to obtaining a specified share price (i.e. market condition), a compensation expense if recognized regardless of whether the share price condition is met if all other vesting conditions are met. For these awards, fair value is determined taking into account the probability of meeting the share price target. No cost is recognized for awards that do not ultimately vest.

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

NOTE 3 – SHARE-BASED INCENTIVE PROGRAMS

Pre-IPO warrants
Up until December 31, 2016, the company issued warrants to employees, consultants providing similar services, and key management. The warrants could be settled by subscribing for A class shares of the company at an exercise price of DKK 44 per share.

Management applied a Black-Scholes option valuation model to determine fair value of the warrants. Fair value of the warrants granted in 2014/2015 and 2015 amounts to TDKK 3 and TDKK 12 respectively.

The most significant assumption applied is the underlying share price. Fair value of one A class share has been determined on the basis of the share of fair value of Orphazyme attributable to A class shares. Fair value of Orphazyme has been determined as the implied fair value, which can be derived from the subscription price in the most recent capital increase round prior to granting the warrants. Fair value per A class share has been determined to be in the range DKK 1.87 – DKK 3.72.

In the second half of 2017 and prior to the IPO, the company, without cancelling or modifying former warrant programs, issued 551,573 warrants under a new warrant program under which a mechanism was put in place ensuring that the respective warrant holders can only exercise warrants from either former programs or the new program. Orphazyme therefore had multiple warrant programs that ran 'in parallel'. The exercise price of new warrants was DKK 1.

The expense recognized by the company for warrant programs running in parallel and where management believes that both programs will vest, is determined based on (a) the grant date fair value of the old program under the original vesting terms, plus (b) the incremental fair value of the new warrant program, as at its grant date (being its fair value of the new programs less the fair value of the old programs at that date), over the vesting terms of the new program.

Due to the liquidation preference to B class and C class shares, the exercise price for the warrants were significantly above the fair value of one A class share at the respective issuance dates, and management determined that the incremental value was insignificant, and no expense has been recognized.

In addition, prior to the IPO in the second half of 2017, Orphazyme issued 279,019 warrants of which 130,541 warrants were exercisable subject to completion of an IPO. The other warrants vested gradually over 4 years subject to continued employment or upon an IPO or a change in control event. The fair value of the warrants granted amounts to TDKK 52 and the management determined the incremental value was insignificant and no expense has been recognized. Consequently, all these warrants vested, lapsed or were exercised upon the completion of the IPO in November 2017. The average share price upon exercise was DKK 80.

The table below summarizes the activity related to the warrants for the six months ended June 30, 2017:

	Executive Management	Employees	Board of Directors	Consultants	Total Warrants	Warrants Exercisable
Outstanding at Jan 1, 2017	211,879	76,176	124,122	9,700	421,877	324,078
Granted	211,879	215,573	124,122	-	551,574	-
Exercised	-	-	-	-	-	-
Expired	-	-	-	-	-	-
Outstanding at Jun 30, 2017	**423,758**	**291,749**	**248,244**	**9,700**	**973,451**	**440,354**

Post-IPO long-term incentive program

In connection with the completion of the IPO, the Executive Management and Key Employees were offered to subscribe for Offer Shares ("Investment Shares") at the Offer Price for a maximum amount corresponding to approximately 15% (CMO) and 20% (CEO, CFO, and CSO) of their respective current annual base salaries.

Under the post-IPO long-term incentive program (LTIP), the Executive Management as well as certain Key Employees of Orphazyme have subscribed to 14,875 ordinary shares ("Investment Shares") in connection with the IPO and admission to trading and official listing of Orphazyme on Nasdaq Copenhagen A/S ("Nasdaq Copenhagen") at the offer price (DKK 80). In April 2018, a Key Employee subscribed to 4,300 Investment Shares at the then-current market price of DKK 67.5. The Board of Directors may decide to offer other current or new employees of Orphazyme participation in the LTIP.

The participants may be allocated a number of shares in Orphazyme ("Performance Shares") at a price per Performance Share of DKK 1 at the end of a vesting period of four years from Orphazyme's first day of trading and official listing on Nasdaq Copenhagen. The number of Performance Shares shall be proportional to a potential increase in the price of Orphazyme's shares at the time of vesting compared to the offer price. The potential increase in the price of Orphazyme's shares will be calculated as the volume-weighted average share price as quoted on Nasdaq Copenhagen during the 10 trading days preceding the vesting date. The maximum allocation of Performance Shares will be six (CEO) and four (other participants) times the number of Investment Shares subscribed for in connection with the IPO. Performance Shares will be allocated on a linear scale with maximum allocation triggered by an 80% increase in share price, whereas no Performance Shares will be allocated, if the price of Orphazyme's shares has increased 20% or less at the time of vesting. Among other things, vesting is also subject to the participants having maintained ownership of their Investment Shares and continued employment at the time of vesting. Based on the number of Investment Shares subscribed for in connection with the IPO, a total maximum of 69,500 Performance Shares may be granted at the time of vesting.

Further, the participants may also be allocated a number of shares in Orphazyme ("Matching Shares") at a price per Matching Share of DKK 1 in connection with the first anniversary of Orphazyme's admission to trading and official listing on Nasdaq Copenhagen. The number of Matching Shares shall be equal to the number of Investment Shares subscribed for in connection with the IPO and vesting will be subject to the participants having maintained ownership of their Investment Shares and continued employment at the time of vesting. Based on the number of Investment Shares subscribed for in connection with the IPO, a total maximum of 14,875 Matching Shares may be granted at the time of vesting.

The LTIP was not in place for the six months ended June 30, 2017. The table below summarizes the activity related to the LTIP for the six months ended June 30, 2018:

	Executive Management	Key Employees	Board of Directors	Consultants	Total Awards	Awards Exercisable
Outstanding at January 1, 2018	9,000	5,875	-	-	14,875	
Granted	-	4,300	-	-	4,300	-
Exercised	-	-	-	-	-	-
Expired	-	-	-	-	-	-
Forfeited	-	-	-	-	-	-
Outstanding at June 30, 2018	**9,000**	**10,175**	**-**	**-**	**19,175**	**-**

The fair value of the LTIP upon the introduction of the plan was estimated at approximately TDKK 3,895 and the fair value of the LTIP awards granted in April 2018 was estimated at approximately TDKK 714. Fair value was estimated using a Monte-Carlo simulation model at the respective grant dates, considering the terms and conditions on which the awards were granted.

The weighted average remaining contractual life for LTIP awards outstanding as at June 30, 2018 was 3.38 years. The exercise price for each LTIP award outstanding as of June 30, 2018 was DKK 1.

Phantom share-based incentive program

In June 2018, Orphazyme introduced a four-year phantom share-based incentive program (the "Program") for all employees other than the Executive Management. The Program is based on phantom shares and entitles the participants to a potential cash bonus if there has been an increase of at least 20% in Orphazyme's share price compared to the entry price at the introduction of the Program. The Program will not have any dilutive effect on the shareholders of Orphazyme as the phantom shares do not constitute or qualify for actual shares in Orphazyme.

The overall objectives of the Program are (i) to retain qualified employees, (ii) to create long-term incentive for the participants of the Program, and (iii) to align the interests of the employees with those of Orphazyme's shareholders. Each employee participating in the Program earns the right to a certain number of phantom shares per month, depending on the employee's position. Subject to any adjustments to the Program made by the Board of Directors due to, for example, changes in Orphazyme's share capital structure or other significant events, each employee will be eligible to receive up to a total of 144 or 288 phantom shares under the Program. By the end of each calendar year 2018-2021, the participants will have earned phantom shares each year free of charge.

The entry price per phantom share is DKK 61 and has been calculated on the basis of the volume-weighted average closing price of Orphazyme's share on Nasdaq Copenhagen during a period of 10 trading days prior to the introduction of the Program. The phantom shares will automatically be settled in cash at the end of January 2023 by subtracting the entry price per share from the market price per share and multiplying the change by the total number of granted phantom shares, but only if Orphazyme's market price per share at that date exceeds the entry price per share by at least 20%. The market price per share will be based on the volume-weighted average closing price of Orphazyme's shares on Nasdaq Copenhagen during a period of 10 trading days prior to the settlement of the phantom shares in January 2023.

The employee's cash bonuses are capped and cannot exceed a gross amount of DKK 37,500 or DKK 75,000 per employee, depending on the number of phantom shares allocated to the respective employee under the Program. Based on the number of participants in the Program as of June 30, 2018, the Program will comprise up to 7,660 phantom shares in total.

As the Program is cash-settled, the fair value of the phantom shares granted as part of the Program is estimated at each reporting date. As of June 30, 2018, the fair value of the phantom shares was approximately TDKK 148.

As of June 30, 2018, all phantom shares granted under the Program were granted to employees of Orphazyme. No phantom shares were forfeited or expired, and none of the phantom shares were eligible for exercise.

The following table presents the inputs to the Monte-Carlo model used to estimate the fair value of the LTIP awards granted in April 2018 and the phantom shares remeasured as of June 30, 2018:

	Six months ended Jun 30, 2018	
	LTIP awards	Phantom shares
Weighted average fair values at the measurement date (TDKK)	714	148
Dividend yield (%)	-	-
Expected volatility (%)	41.8	47.9
Risk-free interest rate (%)	(0.28)	(0.27)
Expected life of awards (years)	3.38	4.58
Weighted average share price (DKK)	67.5	61.06
Model applied	**Monte-Carlo**	**Monte-Carlo**

Expected volatility has been determined on the basis of the historic volatility of comparable listed companies.

For the six-month period ended June 30, 2018, TDKK 1,158 (2017: 0) was recognized as compensation expense related to share-based incentive programs.

NOTE 4 – EQUITY

In the first quarter of 2017, Orphazyme A/S completed a capital increase by issuing 534,007 C class shares to existing shareholders for net proceeds received of TDKK 48,061. In connection with the capital increase, Orphazyme A/S incurred expenses totaling TDKK 73.

On March 8, 2017, the Company completed a TDKK 108,690 financing round by issuing 1,207,662 new C class shares to LSP V Coöperatieve U.A. and ALS Investment Fund.

Orphazyme A/S has not declared or made any dividend payments since its inception and intends to use all available financial resources as well as revenue, if any, for purposes of its current and future business. As of the date hereof, Orphazyme A/S does not expect to make dividend payments within the foreseeable future.

On January 29, 2018, Orphazyme A/S issued 11,380 bonus shares using free reserves of the company to KU Center for Technology Commercialization, Inc., University of Kansas, Kansas Life Sciences Development Company, Inc., and UCL Business PLC under the terms of the license agreement entered into in October 2017. Following this share capital increase, the total nominal share capital was DKK 19,939,564, divided into 19,939,564 shares each with a nominal value of DKK 1.

NOTE 5 – 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, 2018 and 2017:

	Six months ended Jun 30, 2018 TDKK	**Six months ended Jun 30, 2017 TDKK**
Loss for the period	(107,779)	(52,123)
Weighted-average shares outstanding	19,937,794	10,248,928
Loss per share	**5.41**	**5.09**

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 and are disclosed in Note 3.

NOTE 6 – SUBSEQUENT EVENTS

Management has evaluated its financial statements for potential subsequent events occurring after the balance sheet date of June 30, 2018, but prior to the date that these financial statements were issued and found no events to be disclosed.

Statement by the Executive Management and the Board of Directors on the Interim Condensed Consolidated Financial Statements for the period January 1–June 30, 2018

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, 2018. 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, 2018 have been prepared in accordance with IAS 34 "Interim Financial Reporting" as adopted by the European Union, 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 the Group's assets, liabilities, and financial position at June 30, 2018 and of the results of the Group's operations and cash flows for the period January 1-June 30, 2018. Furthermore, in our opinion, Management's Review gives a true and fair account of the development and performance of the Group's activities and of the Group's results for the period and the financial position as of June 30, 2018.

Copenhagen, August 28, 2018.

Board of Directors

Georges Gemayel
Chairman of the Board

Bo Jesper Hansen
Deputy Chairman of the Board

Martin Bonde

Anders Hedegaard

Rémi Droller

Martijn Kleijwegt

Sten Verland

Catherine Moukheibir

Executive Management

Anders Hinsby
Chief Executive Officer

Anders Vadsholt
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.