ORPHAZYME

ANNUAL REPORT 2017

Innovative therapies for protein-misfolding diseases

Orphazyme A/S
CVR no.: 32266355

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ORPHAZYME AT A GLANCE

OUR VISION

To profoundly impact the lives of patients with orphan diseases and their families

WHO WE ARE

Orphazyme is a Danish biopharmaceutical company listed on Nasdaq Copenhagen (ORPHA.CO). We are 34 employees in Copenhagen, Denmark, focused on developing novel treatment options for severe and debilitating protein-aggregation and lysosomal storage diseases.

Our lead candidate, arimoclomol, is being investigated in four orphan disease, with anticipated completion of three potential registration trials by end 2020. The first potential marketing authorization for the treatment of NPC could be obtained in 2020.

PIPELINE

Arimoclomol is being developed as a treatment for four severe orphan protein-misfolding diseases: sporadic Inclusion Body Myositis (sIBM), Amyotrophic Lateral Sclerosis (ALS), Niemann-Pick disease Type C (NPC), and Gaucher disease. Arimoclomol has been studied in seven Phase I and three Phase II trials and is currently being studied in two Phase II/III trials in NPC and sIBM. In January 2018, arimoclomol was granted Rare Pediatric Disease Designation by the US Food and Drug Administration (FDA) for the treatment of NPC.

STRATEGY

Orphazyme's strategy is to develop treatments for rare, or orphan, diseases with protein misfolding where it can apply its specialized know-how in heat-shock proteins (HSPs).

Important elements of Orphazyme's strategy are:

- To successfully conduct and complete the planned and on-going trials of arimoclomol for the treatment of sIBM, ALS, NPC, and Gaucher disease
- To develop a commercialization strategy with our own commercialization structure and operations in key markets, while entering into partnerships for markets outside the USA and Europe
- To use our expertise and proprietary know-how in the field of HSPs and protein misfolding to select and develop new leads for high unmet need diseases – including the current development of a suite of new molecular entities (NMEs)

2
LATE-STAGE CLINICAL TRIALS
(NPC, sIBM)

1
RARE PEDIATRIC DISEASE DESIGNATION
(NPC)

3
ORPHAN DESIGNATIONS IN EU & USA
(ALS, NPC, sIBM)

1
FAST-TRACK DESIGNATION
(NPC)

LETTER FROM THE CEO

OUR JOURNEY

Since Orphazyme's foundation in 2009, we have been fully committed to developing new therapies for patients affected by rare diseases and with limited treatment options. Founded on a platform of pioneering research, we have been on a great journey of translating research into a promising late-stage pipeline. In 2017, we successfully conducted an Initial Public Offering (IPO), raising the capital that will enable us to conduct four clinical trials with our lead product candidate, arimoclomol. We are now well on our way to reaching our goal of launching arimoclomol for NPC as early as 2020, with launches in three other indications in the following years. 2018 will be an important year for Orphazyme, with the clinical NPC trial results expected in Q3, potentially forming the basis for a marketing authorization application in the USA and Europe.

In 2017, we completed patient enrollment for our Phase II/III trial in NPC, 'AIDNPC', and initiated a Phase II/III trial in sIBM. Both trials may, with a positive outcome, lead to marketing authorization in these debilitating indications where treatment options are desperately needed. We ended the year with Orphazyme's IPO in November.

GETTING LISTED

On November 16, 2017, we rang the bell at Nasdaq Copenhagen's headquarters, celebrating the completion of our IPO. Our entire team worked tirelessly towards this goal during 2017 and on that day in November, we celebrated the beginning of a new chapter of our journey.

The successful completion of the IPO represents a major milestone for Orphazyme. Raising DKK 600 million, we now have the financial means to conduct our clinical development. Moreover, we have the resources to strengthen our organization by further attracting top talent. We strive not to lose momentum, because every day matters to the patients with these serious diseases. With a skilled and dedicated team, we are well on our way – staying on track.

STRONG PIPELINE

We are developing our lead program in four orphan indications within protein-aggregation and lysosomal storage diseases. Currently, arimoclomol is being tested in two late-stage trials for NPC and sIBM, respectively, and we are planning to initiate two additional clinical trials in ALS and Gaucher disease during 2018.

In 2017, we completed the enrollment of patients in the Phase II/III NPC trial, obtained an Orphan Drug Designation in sIBM from the FDA, and initiated a Phase II/III trial in sIBM. The first clinical trial with arimoclomol in sIBM was led by a dedicated team of clinical researchers from the University of Kansas and University College London, providing exciting data that led them, supported by Orphazyme, to apply for and receive an FDA grant to conduct a Phase II/III trial. In 2017, we assumed the sponsorship of the Phase II/III trial while maintaining a strong collaboration with the world-leading academic experts who have been instrumental in bringing arimoclomol to this stage in sIBM.

2017 was also the year in which we presented favorable trial results from a clinical Phase II trial in a subpopulation of ALS patients with a particularly aggravated disease progression: SOD1-ALS. The data from this trial further highlight the potential of arimoclomol in this truly devastating and fatal disease. The data was published in the journal Neurology® in January 2018[1].

Early 2018, arimoclomol was moreover granted Rare Pediatric Disease Designation by the FDA for the treatment of NPC. With this grant, Orphazyme is eligible to receive a Priority Review Voucher upon marketing approval, which can be redeemed to provide Priority Review of a subsequent marketing application for a different product.

(1) Benatar et al. Neurology® Journal (2018)

Also, in line with our previously-communicated commercialization strategy, we have recently begun to take steps to prepare for the future commercialization of arimoclomol by welcoming Paul Merrigan to our team as Chief Commercial Officer. Paul will be laying the pre-commercial foundation to not only prepare our organization but the markets for the coming of arimoclomol, aligning our development activities with the needs and expectations of our key stakeholders, including the patient communities, healthcare professionals, and payers we plan to serve.

LOOKING FORWARD

This year, we are looking forward to reporting the trial results from our Phase II/III trial in NPC, which are anticipated in Q3 2018. By the end of 2020, our ambition is to have completed three potential registration trials with arimoclomol, with the first possible marketing authorization as early as 2020. Orphazyme's path to achieving these goals is clear and we are fully focused on performing the best possible clinical trials in collaboration with experts and the patients affected by the diseases. Our purpose remains unchanged: To develop new therapies for patients affected by rare diseases.

I want to express my gratitude to the Orphazyme employees for their determined effort, to the patient organizations working tirelessly to improve the lives of patients and their families, and to our long-standing academic collaborators who contribute greatly to the development of arimoclomol. Finally, thank you to our shareholders, old as new, for making this journey possible. 2018 will be a transformative year and we are looking forward to taking you with us on this life-changing journey.

Anders Hinsby
Chief Executive Officer

KEY FIGURES

TDKK	2017	2016	2015[1]	2014/15	2013/14[2]
Statement of profit and loss and other comprehensive income					
Research and development costs	(99,048)	(55,817)	(25,478)	(31,604)	-
General and administrative expenses	(31,994)	(7,703)	(4,044)	(5,494)	-
Operating loss	**(131,042)**	**(63,520)**	**(29,522)**	**(37,098)**	**(37,717)**
Net financial items	(662)	85	40	(1,369)	(1)
Loss before tax	**(131,704)**	**(63,435)**	**(29,482)**	**(38,467)**	**(37,718)**
Income tax benefit	5,500	5,500	2,750	5,875	6,250
Net loss for the period	**(126,204)**	**(57,935)**	**(26,732)**	**(32,592)**	**(31,468)**
Total comprehensive loss	**(126,204)**	**(57,935)**	**(26,732)**	**(32,592)**	**(31,468)**
Loss per share, basic (DKK)	(10.46)	(5.89)	(2.75)	(3.60)	(3.63)
Statement of financial position					
Licenses	9,853	-	-	-	-
Property, plant, and equipment	1,851	987	1,487	1,748	1,694
Investment in property, plant, and equipment	1,491	238	25	558	114
Non-current assets	14,864	4,047	4,448	7,807	1,873
Cash and cash equivalents	631,735	14,349	68,014	78,161	25,732
Other current assets	16,218	13,545	12,490	9,379	7,959
Total assets	662,817	31,941	84,952	95,347	35,564
Share capital	19,928	3,361	3,346	3,218	2,175
Equity	615,702	17,509	74,143	89,380	31,727
Current liabilities	47,115	14,431	10,809	5,967	4,017
Cash flow statement					
Cash flow from operating activities	(95,426)	(54,724)	(21,372)	(36,438)	-
Cash flow from investing activities	(1,491)	(238)	(25)	(558)	-
Cash flow from financing activities	714,303	1,300	11,250	89,425	-
Other					
Share price (DKK)[3]	76.00	-	-	-	-
Total outstanding shares	19,928,184	3,360,541	3,345,755	3,218,031	2,175,208
Market capitalization (MDKK)[4]	1,514.5	-	-	-	-
Equity ratio[5]	92.9%	54.8%	87.3%	93.7%	89.2%
Equity per share (DKK)[6]	30.90	5.21	22.16	27.77	14.59
Average number of employees	26	17	13	10	9
Number of employees at the end of the year	34	21	15	14	8

(1) 2015 covers the period July 1-December 31, 2015 as the Company changed its financial year to follow the calendar year
(2) The financial period 2013/14 is prepared in accordance with the Danish Financial Statements Act's requirements for class B enterprises and presented as reported in the annual report for 2013/14
(3) There is no official share price for the reporting periods prior to 2017 since the Company only went public in 2017
(4) Market capitalization is calculated as the share price multiplied with the total outstanding shares as of the balance sheet date
(5) Equity ratio is calculated as the equity divided by the total assets as of the balance sheet date
(6) Equity per share is calculated as the total equity divided by the total outstanding shares as of the balance sheet date

2017 ACHIEVEMENTS

FINANCIAL PERFORMANCE

- The financial result is in line with the guided outlook for 2017. The net result for 2017 was a loss of DKK 126.2 million compared to DKK 57.9 million in 2016. The increase is primarily due to increased research and development activities and costs associated with the IPO

- Research & development costs totaled DKK 99.0 million in 2017 compared to DKK 55.8 million in 2016. The increase was mainly due to the on-going NPC Phase II/III trial, preparation costs for the Phase II trial in Gaucher disease, a Phase II/III trial in ALS, IND transfer costs for the sIBM study, and arimoclomol manufacturing costs

- General and administrative expenses increased from DKK 7.7 million in 2016 to DKK 32.0 million in 2017, mainly due to IPO-related costs and the expanded management and additional staff

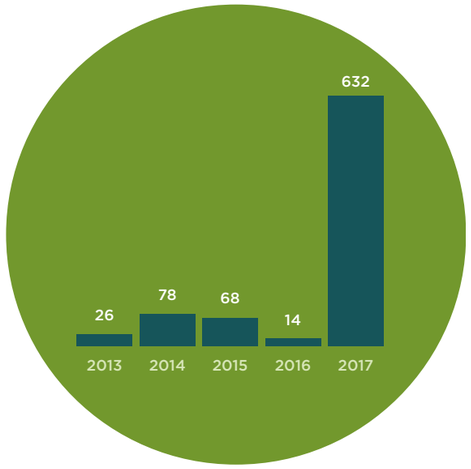
- The net financials totaled an expense of DKK 0.7 million in 2017 compared to an income of DKK 0.1 million in 2016, primarily reflecting exchange rate gains

- As of December 31, 2017, Orphazyme had cash and cash equivalents of DKK 631.7 million compared to DKK 14.3 million as of December 31, 2016. The increase reflects the net proceeds from the IPO of DKK 557.4 million and capital increases of DKK 156.8 million prior to the IPO

CASH POSITION YEAR-END 2017

DKK 631.7M

CASH AND CASH EQUIVALENTS



BUSINESS PROGRESS

Priority	✓	Targeted milestone
sIBM	✓	• Initiate Phase II/III trial
	✓	• Assume sponsorship of Phase II/III trial
ALS	✓	• Conduct 'end of Phase II' meeting with FDA
	✓	• Prepare design for Phase II/III trial
NPC	✓	• Complete patient enrollment in Phase II/III trial
	✓	• Rare Pediatric Disease Designation from FDA
Financing	✓	• Venture financing to expand clinical development program
	✓	• Prepare and complete Initial Public Offering

2018 OUTLOOK

MDKK	2018 guidance	2017 actual result	2017 guidance**
Operating loss	(245) - (275)	(126)	(125) - (135)
Cash position at year-end*	>350	632	-

*Cash, cash equivalents, and marketable securities
**Prospectus, November 2017.

OPERATING RESULT

The financial guidance for 2017 was met with an operating loss of DKK 126.2 million. We anticipate that our 2018 operating loss will be in the range of DKK 245–275 million. The outlook range reflects the inherent uncertainty related to the timing of patient enrollment and similar operational uncertainties. The increase is driven by the advancement of arimoclomol into new indications and an increase in employees to support our clinical and operational activities. The two most relevant cost drivers affecting the outlook are the progress in patient enrollment in the sIBM trial and the planned ALS trial.

CASH POSITION

At year-end 2018, we anticipate a cash position of DKK >350 million compared to DKK 631.7 million as of December 31, 2017.

RISKS AND ASSUMPTIONS

In addition to factors already mentioned, the estimates above are subject to change due to numerous reasons, including but not limited to the timing and variation of development activities.

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: 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 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 described on page 10, Product Pipeline, as to the activities planned for 2018.

Disclaimer
This annual report contains forward-looking statements. The words "believe", "expect", "anticipate", "intend", "plan", and similar expressions identify forward-looking statements. Actual results or performance may differ materially from any future results or performance expressed or implied by such statements. The important factors that could cause our actual results or performance to differ materially include, among others, risks associated with product discovery and development, uncertainties related to the outcome and conduct of clinical trials, including unforeseen safety issues, uncertainties related to product manufacturing, and other factors. For a further discussion of these risks, please refer to the section Risk Management on page 25 in this annual report. Orphazyme A/S does not undertake any obligation to update or revise forward-looking statements in this annual report nor to confirm such statements in relation to actual results, unless required by law.

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 II/III trial in Q3
NPC	• Phase II/III trial 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

OBJECTIVES

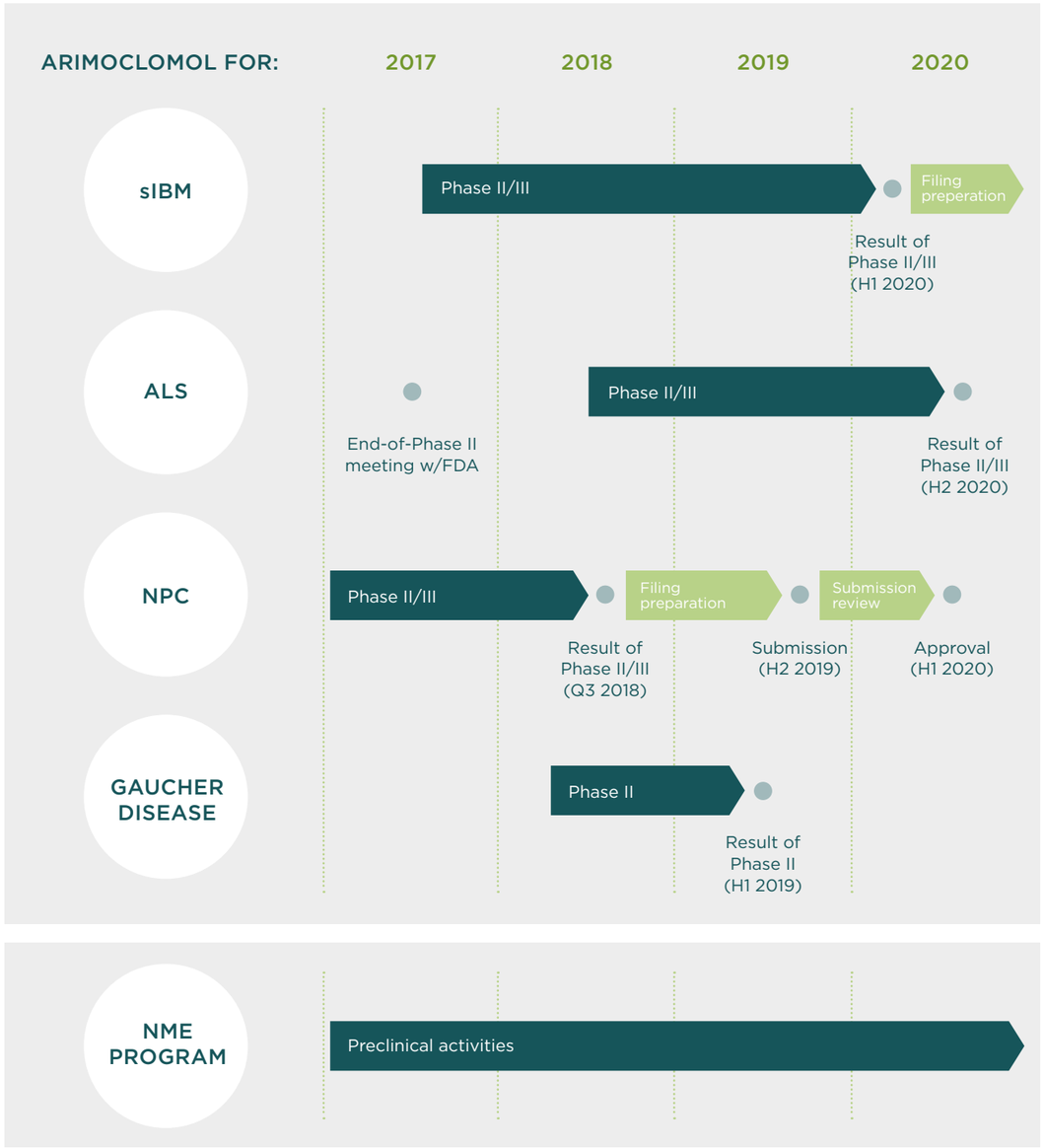
Our long-term objective is to develop new treatment options for orphan diseases with none or inadequate therapies available. In 2018, we will move towards that goal by furthering the development of arimoclomol.

COMPLETE ENROLLMENT OF PHASE II/III SIBM TRIAL BY YEAR-END

ALS PHASE II/III
We expect to be initiating a Phase II/III trial in ALS in Q3 2018.

Q3 2018
TOPLINE
RESULTS EXPECTED FORPHASE II/III TRIAL IN NPC

PRODUCT PIPELINE



Orphazyme's current clinical programs investigate arimoclomol as a treatment for four protein-misfolding indications: The protein-aggregation diseases sporadic Inclusion Body Myositis (sIBM) and Amyotrophic Lateral Sclerosis (ALS), and the lysosomal storage diseases Niemann-Pick disease Type C (NPC) and Gaucher disease. The new molecular entity (NME) program focuses on the development of new molecules as a treatment for relevant protein-misfolding diseases. In the following, we will elaborate on our pipeline.

PROTEIN-AGGREGATION DISEASES

The protein-aggregation diseases sIBM and ALS are caused by the misfolding of proteins inside the cells and the formation of protein aggregates, impairing normal protein recycling. Protein aggregation can cause cell stress and eventually cell death. sIBM is characterized by the loss of muscle, primarily affecting the function of the limbs of the patients, but also other muscle functions, such as swallowing. In ALS, the primary pathology is found in the nerve cells controlling the muscles, which in turn leads to loss of muscle control and ability to move, and eventually the loss of vital functions such as breathing.

LYSOSOMAL STORAGE DISEASES

Lysosomal storage diseases (LSDs) are inherited metabolic disorders caused by a deficiency in the recycling machinery in the cells of the body. These deficiencies are often caused by mutations leading to protein misfolding. Lysosomes are membrane-bound compartments of enzymes, located in the body's cells, used to break down and recycle fats, proteins, and other large molecules into the respective building blocks. Loss of lysosomal enzyme activity due to their misfolding and dysfunction prohibits the lysosomes from performing their normal function and results in accumulation of metabolites in the cells. Lysosomal storage diseases are comprised of more than 50 different diseases that may affect different parts of the body, e.g. the brain, central nervous system, skeleton, skin, and heart, and new disorders continue to be discovered. Orphazyme is currently conducting clinical research with arimoclomol for NPC and expect to start a Phase II trial in Gaucher disease in Q2 2018.

NEW MOLECULAR ENTITIES PROGRAM

Orphazyme is developing a new series of heat-shock protein (HSP) amplifying drugs based on its expertise and know-how about the convergences 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.

sIBM & ALS

PROTEIN-AGGREGATION DISEASES

NPC & GAUCHER DISEASE

LYSOSOMAL STORAGE DISEASES

SPHINGOLIPIDOSES

Sphingolipidoses are a group of lipid (fat) storage diseases where the metabolism of the class of sphingolipids inside the cells is disturbed. This results in build-up of these lipids in large depots inside the cells that disturb cell function or lead to cell death. Sphingolipids are produced in many tissues, but especially nerve tissues, and sphingolipid diseases therefore very often affect the nervous system. Important diseases in this group include NPC, Gaucher disease, Fabry disease, Krabbe disease, Tay–Sachs disease, and metachromatic leukodystrophy.

sIBM

THE DISEASE

Sporadic Inclusion Body Myositis (sIBM) is an acquired, rare, and slowly-progressing protein-aggregation disease. It is the most common muscle-wasting disorder in the elderly population and is characterized by progressive degeneration and weakness and atrophy of muscles, especially of the arms and legs. sIBM patients experience impaired hand function as well as difficulty standing up and walking. Some patients also have problems with swallowing (dysphagia) due to weakness of the throat muscles. In most cases, the disease progresses relentlessly over 10-15 years until the affected patient has lost mobility entirely. The cause of sIBM is not fully elucidated, but degenerative factors, i.e. the build-up of tangled and misfolded proteins (inclusion bodies), play a major role.

TREATMENT OPTIONS

As of today, there are no approved drugs for the treatment of sIBM. The standard treatment option for sIBM consists only of supportive therapy (physical, speech, and occupational therapy), and there is no evidence that any form of treatment slows progression.

PREVALENCE

The size of the patient population in the USA and Europe is not fully elucidated, but a 2017 publication estimated a patient population of approximately 24.8 and 45.6 per million or at least 17,000-31,000 individuals in the USA and the major European countries[1].

TRIAL STATUS

A Phase II clinical trial in 24 sIBM patients conducted in London and Kansas found that arimoclomol was well-tolerated and showed marked efficacy trends. A Phase II/III arimoclomol trial for sIBM was initiated in August 2017 in the USA and Europe. The trial is intended to support a registration of arimoclomol for the treatment of sIBM. Results from the trial are expected in H1 2020.

ALS

THE DISEASE

The rare neuromuscular disease Amyotrophic Lateral Sclerosis (ALS), also called Lou Gehrig's disease, is rapidly progressive and invariably fatal, usually within two to five years. The disease attacks the neurons responsible for controlling muscles leading to paralysis of all skeletal muscles as well as involuntary muscles that involve breathing, speaking, and swallowing. The cause of damage to the neurons includes protein misfolding and aggregation.

Familial and sporadic ALS

Around 10% of ALS cases are associated with pathogenic mutations (familial ALS) while the rest have no identified genetic component (sporadic ALS). Among the familial ALS cases, 20% harbor mutations in a gene coding for the superoxide dismutase (SOD1) enzyme. ALS associated with mutations in the SOD1 gene is often very aggressive with a life expectancy <2 years from diagnosis. Arimoclomol has so far been tested in two Phase II ALS trials, one dose-ranging trial in sporadic ALS, and one trial in familial SOD1.

TREATMENT OPTIONS

As of today, ALS patients have very limited treatment options; Rilutek (riluzole) which reduces the levels of the neurotransmitter glutamate, potentially slowing down the progression of ALS; and Radicava (edaravone) which can act against the reactive oxygen species that may damage the nerve cells in ALS patients. Thus, a high unmet medical need remains for new therapies for ALS patients.

PREVALENCE

The incidence of ALS is estimated at between 1-3 per 100,000 individuals per year globally. The patient population in the USA and Europe is estimated to be approximately 50,000 patients. In Japan, it is estimated that there are between 8,000-14,000 patients with ALS.

TRIAL STATUS

A placebo-controlled Phase II trial in 38 patients with SOD1 ALS was reported, confirming that 200mg arimoclomol three times daily up to 12 months is well-tolerated. Although not powered for therapeutic effect, the consistency of results across the range of efficacy outcome measures suggested a therapeutic benefit of arimoclomol on survival, function (ALSFRS-R), vital capacity, as well as the combined assessment of function and survival (CAFS). Orphazyme intends to conduct a Phase II/III trial to support the application for a marketing authorization in ALS. The trial is planned to be initiated in Q3 2018.

(1) Benatar et al. Neurology® Journal (2018)

NPC

THE DISEASE

Niemann-Pick disease Type C (NPC) is a rare, inherited, progressive, and often fatal neurodegenerative disease. NPC is a lysosomal storage disorder caused by genetic mutations that most often lead to misfolded variants of the NPC proteins. Misfolded NPC protein does not function properly and is subject to rapid degradation. As a consequence, lipids, that would normally be cleared, build up in the lysosomes of cells throughout the body. Accumulation of lipids in the tissues and organs, including the brain, leads to loss of cell function and organ damage. Neurologic involvement is common and results in progressive motor and cognitive impairment. The NPC disease is caused by mutations on one of two genes, NPC1 or NPC2. Approximately 95% of individuals with the disease have mutations in NPC1.

TREATMENT OPTIONS

The majority of current treatment options are palliative and are only directed towards the specific symptoms apparent in each individual (e.g. prescription of anti-seizure medications to prevent seizures).

Only one drug, Zavesca (miglustat), is currently marketed for NPC and only in certain jurisdictions. The product may reduce progression of disease in some patients, but there is still a very large unmet need for new therapy in NPC.

PREVALENCE

NPC often presents in childhood but can present at any age. The incidence of the disease is estimated to be 1 in 120,000 births and it is estimated that the NPC patient population is between 1,000-2,000 in the USA and Europe.

TRIAL STATUS

Arimoclomol is currently being investigated in a clinical Phase II/III trial as a potential treatment for NPC. Topline results are expected to be reported in Q3 2018.

GAUCHER DISEASE

THE DISEASE

Gaucher disease is an inherited metabolic disorder caused by mutations in a protein called glucocerebrosidase, which leads to the accumulation of certain sugar-containing lipids. The usual symptoms of Gaucher disease include an abnormally enlarged liver and/or spleen (hepatosplenomegaly), low levels of circulating red blood cells (anemia), blood cells promoting clotting (thrombocytopenia), and skeletal abnormalities. Disease of the nervous system is observed in a significant subpopulation of Gaucher disease (neuropathic Gaucher).

TREATMENT OPTIONS

Two types of treatment are currently available for patients with Gaucher disease: Enzyme-replacement therapy and substrate reduction therapy. None of these are useful in the treatment of neuropathic Gaucher disease.

PREVALENCE

The total number of Gaucher patients in the USA and Europe is conservatively estimated at 10,000-15,000 individuals. Of the total market, Orphazyme focuses on the 10-30% with neuropathic Gaucher disease, where no treatments are available today.

TRIAL STATUS

A Phase II clinical trial of arimoclomol in Gaucher disease is planned to commence in Q2 2018.

TECHNOLOGY

Our technology is based on amplifying human cells' defense against protein aggregation and misfolding. This defense consists of a system of HSPs, which work by rescuing proteins from misfolding and aggregation. Orphazyme's lead compound, arimoclomol, works by increasing the body's own production of heat-shock proteins in cells experiencing stress or toxicity.

ARIMOCLOMOL

Arimoclomol works by increasing the production of HSPs inside the cells and thereby enhancing the natural biological mechanisms that reduce protein misfolding and aggregation.

Protein-misfolding diseases

Proteins are our cells' molecular machines, making certain that each cell in the body fulfils its function. To do their job, they must attain the correct shape. In many diseases protein misfolding may cause toxicity either as a consequence of protein aggregation or the loss of protein function. Protein aggregates, a hallmark of diseases such as sIBM and ALS, leads to toxicity as aggregates perturb the cell functions. Loss of function, a classical feature of the LSDs such as NPC and Gaucher disease, leads to an accumulation of toxic substances, which in turn leads to development of disease.

HSPs

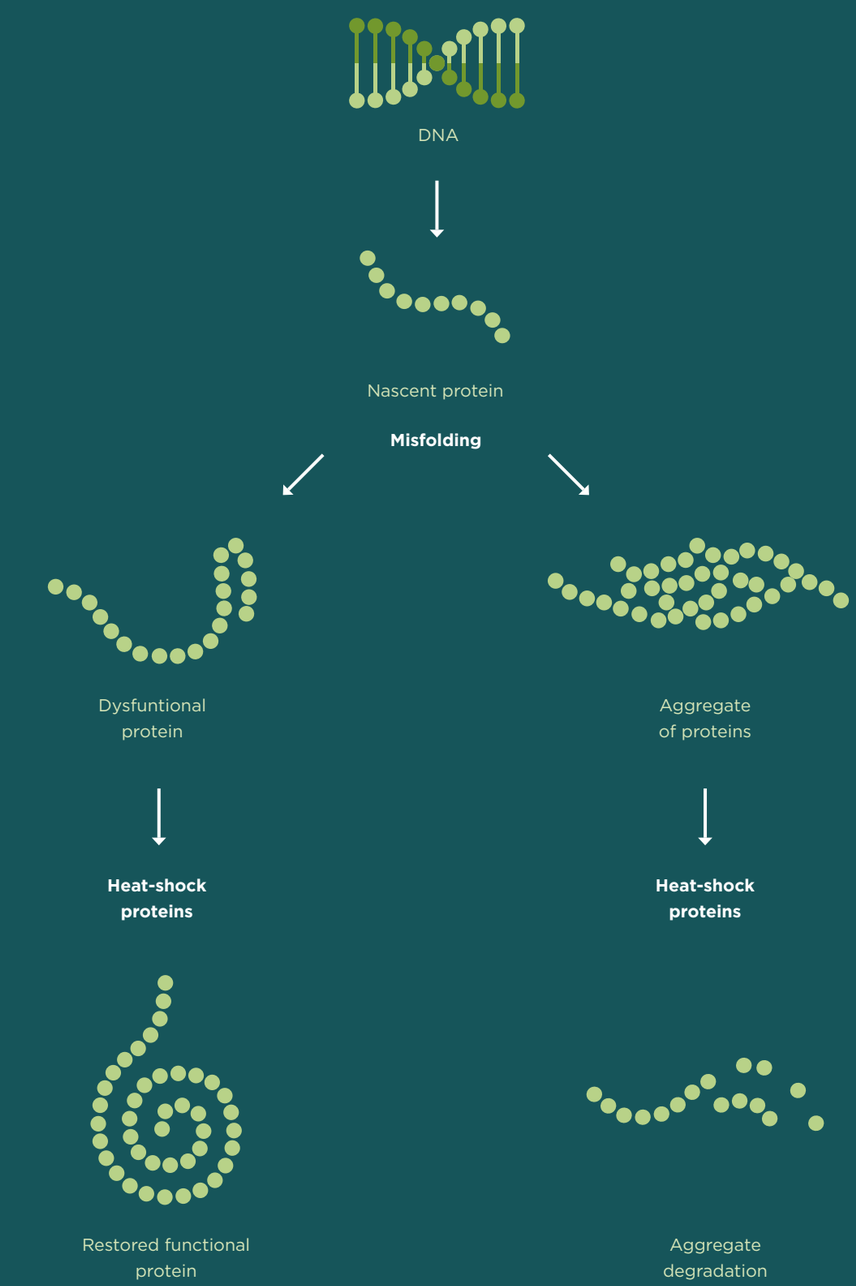
The HSPs constitute a natural system that makes other proteins work correctly and guard against the toxicity arising from misfolded proteins and dysfunctional cellular recycling systems. In particular, HSPs are molecular chaperones that promote the survival of stressed cells by re-folding misfolded proteins into their correct conformation, or by directing 'terminally'-misfolded proteins to be broken down. They also protect cells by inhibiting lysosomal membrane permeabilization; stabilizing lysosomes (cellular structures where waste products are broken down), allowing cells to clear away waste and return to their healthy status.

There are several different types of HSPs which work in conjunction – a cardinal member is HSP70, which Orphazyme uses as the key parameter to measure activity of its drug candidates. HSP70 has been shown to protect against the formation of protein aggregates which are the defining characteristic of a number of protein-aggregation diseases including sIBM and ALS. In addition, HSP70 has been identified as a co-factor for lysosomal sphingolipid breakdown, a necessary step in the metabolism of stored lipids which cause toxicity if accumulated in the lysosome. In both NPC and Gaucher disease, as well as other LSDs, mutations lead to misfolding and loss of enzyme functions involved in the breakdown and recycling of critical cellular components within the cells recycling centers, the lysosomes. By amplifying the production of HSPs, this pathological cascade can be addressed by rescuing the function of the recycling enzymes and helping them perform better in the lysosomes.

Mechanism of action

Arimoclomol stimulates the production of HSPs. The production of HSPs is regulated by a transcription factor, heat-shock factor 1 ("HSF1"). A transcription factor is a protein that regulates production of other proteins in the cell. In the case of HSF1, the proteins being regulated are HSPs. Activation of HSF1 starts the production of the major stress-inducible HSP70-chaperone along with other HSP-chaperones, which help reshape the cells' misfolded proteins and take care of the recycling systems. Under normal cellular conditions, HSF1 is inactive. However, the transcription factor can be activated by an initial cellular stress, such as protein misfolding, and becomes fully activated under a sustained stress signal. Arimoclomol amplifies and prolongs the activated, HSP-producing state of HSF1. This leads to an amplification in the production of cell protective HSPs, but only in physiologically stressed cells.

OUR TECHNOLOGY



FINANCIAL REVIEW

INCOME STATEMENT

The net result for the financial year 2017 was a loss of DKK 126.2 million compared to DKK 57.9 in 2016. The increase is primarily due to increased research and development activities as well as costs associated with the Company's IPO.

RESEARCH AND DEVELOPMENT COSTS

Research and development costs totaled DKK 99.0 million in 2017 compared to DKK 55.8 million in 2016. The increase was mainly due to the on-going NPC Phase II/III trial initiated in mid-2016, the costs for the preparations of a Phase II trial in India for Gaucher disease, a Phase II/III trial for ALS, and costs for taking over sponsorship of the sIBM Phase II/III trial. Furthermore, the costs in 2017 are impacted by costs of manufacturing arimoclomol for the clinical trials.

GENERAL AND ADMINISTRATIVE EXPENSES

General and administrative expenses were DKK 32.0 million in 2017 compared to DKK 7.7 million in 2016. The increase is mainly due to costs for lawyers, auditors, and consultants relating to the IPO, which was successfully completed and the shares had the first trading day on November 16, 2017. Furthermore, the costs are also impacted by the hiring of new members of management and additional staff.

IPO-related costs amounted to DKK 56.1 million of which DKK 14.0 million affecs the income statement under general and administrative expenses and DKK 42.6 million affects the equity and comprised costs for external advisors – mainly fees to bookrunners, lawyers, auditors, and other advisors. Costs, which can be considered directly related to the IPO, is allocated to the equity based on the percentage of new shares issued compared to the total number of shares and the remaining amount is allocated to the income statement. However, costs such as bank fees and advertising for the IPO has been allocated 100% to equity.

NET FINANCIAL ITEMS

Net financials totaled an expense of DKK 0.7 million in 2017 compared to an income of DKK 0.1 million in 2016. Financial income primarily reflects exchange rate gains. Financial expenses primarily cover interest expenses on bank accounts, exchange rate losses, and bank fees.

INCOME TAX BENEFIT

Income tax benefit totaled DKK 5.5 million in 2017 compared to DKK 5.5 million in 2016. Income tax benefits for the two periods include a tax credit for research and development expenses at the applicable tax rate under the Danish Corporate Income Tax Act.

STATEMENT OF FINANCIAL POSITION

Cash and cash equivalents

As of December 31, 2017, Orphazyme had cash and cash equivalents of DKK 631.7 million compared to DKK 14.3 million as of December 31, 2016. The increase reflects the net proceeds from the IPO of DKK 557.4 million and capital increases of DKK 156.8 million prior to the IPO.

Equity

As of December 31, 2017, equity amounted to DKK 615.7 million compared with DKK 17.5 million as of December 31, 2016. The increase reflects the gross proceeds from the IPO of DKK 600.0 million and capital increases of DKK 156.8 million prior to the IPO.

CASH FLOWS

Cash flow from operating activities

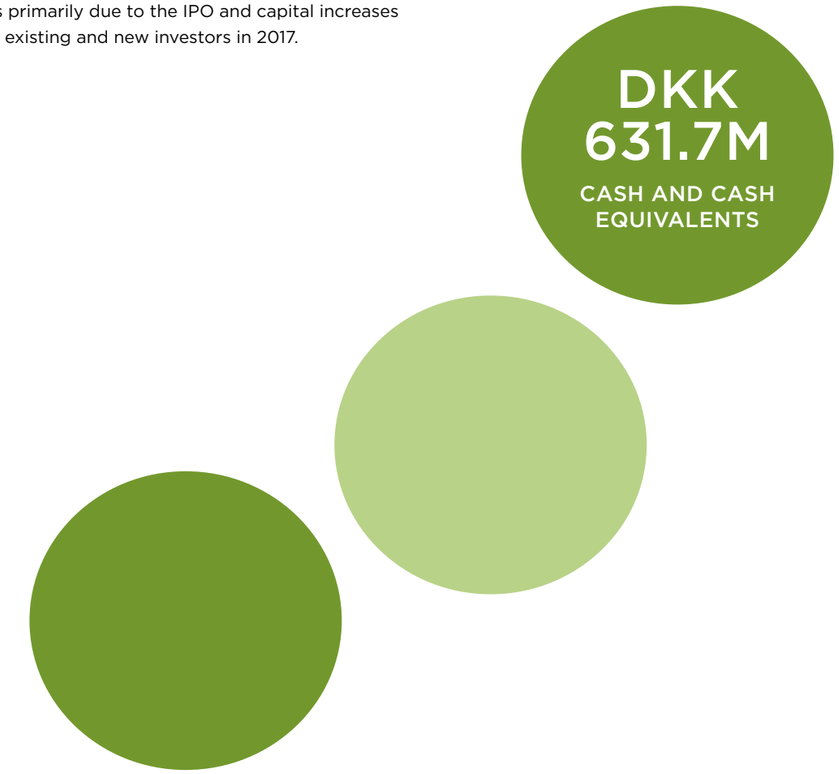
Net cash flow from operating activities amounted to an outflow of DKK 95.4 million in the twelve-month period ended December 31, 2017 compared to DKK 54.7 million in the twelve-month period ended December 31, 2016. Net cash flow from operating activities is attributable primarily to the initiation and progression of clinical development activities, as well as general and administrative expenses.

Cash flow from investing activities

Net cash outflow from investing activities amounted to an outflow of DKK 1.5 million in the twelve-month period ended December 31, 2017 compared to DKK 0.2 million in the twelve-month period ended December 31, 2016. 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 inflow of DKK 714.3 million in the twelve-month period ended December 31, 2017 compared to DKK 1.3 million in the twelve-month period ended December 31, 2016. The increase is primarily due to the IPO and capital increases from both existing and new investors in 2017.



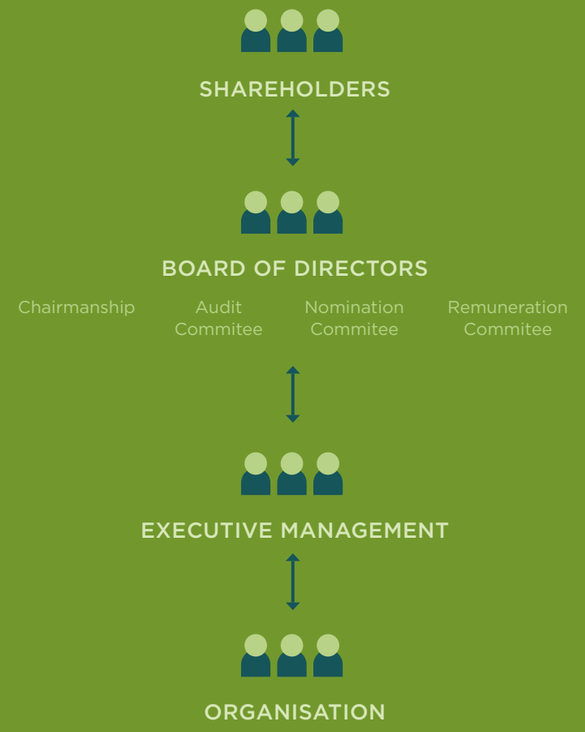
CORPORATE GOVERNANCE

In order to maintain the trust of the Company's stakeholders, Orphazyme is committed to ensure transparent and good corporate governance. As a company listed on Nasdaq Copenhagen A/S, Orphazyme is subject to the Recommendations on Corporate Governance from May 2013 (as amended November 2014).

The Recommendations on Corporate Governance are best practice guidelines for the management of companies admitted to trading on a regulated market.

Orphazyme intends to comply with the Recommendations on Corporate Governance in all material respects, however, due to the current size of the Company and the nature of its present operations, the company has opted to deviate from the recommendations in the following areas:

- Orphazyme has decided only to publish half-yearly financial reports;
- the current Articles of Association do not stipulate a retirement age for Orphazyme's Board of Directors;
- share options may be included in the remuneration of the Board of Directors; and
- share-based instruments that would be granted to board members shall have a maturity of one year from the date of allocation.



Orphazyme's corporate governance statement includes a summary of the company's governance structure, Orphazyme's position on the Recommendations on Corporate Governance as well as a complete list of the company's comments to each recommendation.

The corporate governance statement is available on www.orphazyme.com.

Corporate Governance Statement

BOARD OF DIRECTORS

Name	Position	Independent[1]	Year of first appointment	Expiration of term
Georges Gemayel	Chairman	Independent	2012	2018
Bo Jesper Hansen	Deputy Chairman	Independent	2010	2018
Martin Bonde	Member	Independent	2010	2018
Sten Verland	Member	Independent	2010	2018
Rémi Droller	Member	Independent	2015	2018
Martijn Kleijwegt	Member	Independent	2017	2018
Anders Hedegaard	Member	Independent	2017	2018
Cathrine Moukheibir	Member	Independent	2017	2018

[1] The Company has based its assessment of independence on the basis of criteria set out in the current Coporate Governance Recommendations (as defined below).

BOARD OF DIRECTORS

The Board of Directors is responsible for the overall and strategic management and proper organization of Orphazyme's business and operations and it supervises the Company's activities, management, and organization. The Board of Directors appoints and dismisses the members of the Executive Management, who are responsible for the day-to-day management of the Company.

The Board of Directors normally holds at least five regular meetings annually, including a strategy review, plus ad-hoc meetings as required. Extraordinary board meetings are convened by the Chairman when necessary or when requested by a member of the Board of Directors, a member of the Executive Management, or by the Company's auditor. The Board of Directors forms a quorum when more than half of its members are represented, including the Chairman or the Deputy Chairman. Resolutions of the Board of Directors are passed by a simple majority of the votes present at the meeting. In the event of equal votes, the Chairman or, in his absence, the Deputy Chairman shall have the casting vote. The Board of Directors conducts an annual evaluation of the effectiveness, performance, achievements, and competencies of the Board of Directors and of the individual members as well as the collaboration with the Executive Management.

The members of the Board of Directors elected by the general meeting are elected for a term of one year. Members of the Board of Directors may be re-elected.

The Company believes that the members of the Board of Directors possess the professional skills and experience required to serve as board members of the Company.

BOARD COMMITTEES

To support the Board of Directors in its duties, the Board of Directors has established and appointed an Audit Committee, a Nomination Committee, and a Remuneration Committee. These committees are charged with reviewing issues pertaining to their respective fields that are due to be considered at board meetings. Written charters specifying the tasks and responsibilities for each of the committees are available on the Company's website www.orphazyme.com.

Committee Charter

AUDIT COMMITTEE

The Audit Committee reviews accounting and audit matters that by decision of the Board of Directors or the Audit Committee require a more thorough evaluation, and assess the internal controls and risk management systems of Orphazyme. Its duties also include supervision of the Company's auditors and review of the audit process. In accordance with the Recommendations on Corporate Governance of the Danish Committee on Corporate Governance issued in May 2013 and updated in November 2014 (the "Corporate Governance Recommendations"), the Company has decided that the Chairman of the Board of Directors may not also be the Chairman of the Audit Committee and that a majority of the members of the Audit Committee are required to meet the independence requirements set out in the Corporate Governance Recommendations. In addition, at least one member shall have accounting or audit qualifications and between them, the members shall possess such expertise and experience as

to provide an updated insight into, and experience in, the financial, accounting, and audit aspects of companies with shares admitted to trading and official listing on a regulated market.

The Audit Committee shall consist of no less than three members appointed by and among the Board of Directors, including the Chairman of the Audit Committee, and consists of Catherine Moukheibir as Chairman, Martijn Kleijwegt, and Sten Verland. All of the members of the Audit Committee meet the independence requirement set out in the Corporate Governance Recommendations. The CEO and/or the CFO and the Company's external auditor shall participate in meetings of the Audit Committee if so requested by the Audit Committee and the external auditor shall attend at least one meeting per year or the relevant part hereof where the Executive Management is not present.

NOMINATION COMMITTEE

The Nomination Committee shall assist the Board of Directors with ensuring that appropriate plans and processes are in place for the nomination of candidates to the Board of Directors, the Executive Management, and the board committees. Moreover, the Nomination Committee shall evaluate the composition of the Board of Directors and the Executive Management. This includes making recommendations for nomination or appointment of members of (a) the Board of Directors, (b) the Executive Management, and (c) the board committees established by the Board of Directors.

The Nomination Committee consists of no less than three members appointed by and among the Board of Directors, including Georges Gemayel as Chairman, Martin Bonde, and Sten Verland. All of the members of the Nomination Committee meet the independence requirements set out in the Corporate Governance Recommendations.

REMUNERATION COMMITTEE

The Remuneration Committee ensures that the Company maintains a Remuneration Policy for the members of the Board of Directors and the Executive Management, which includes the overall guidelines on incentive pay for the Board of Directors and Executive Management in accordance with Section 139 of the Danish Companies Act, and to evaluate and make recommendations for the remuneration of the members of the Board of Directors and the Executive Management.

The Remuneration Committee shall consist of no less than three members appointed by and among the Board of Directors, including Bo Jesper Hansen as Chairman, Rémi Droller, and Anders Hedegaard. All of the members of the Remuneration Committee meet the independence requirements set out in the Corporate Governance Recommendations.

DESCRIPTION OF INTERNAL CONTROL AND FINANCIAL REPORTING PROCEDURES

The Board of Directors, the Audit Committee, and the Executive Management are ultimately responsible for Orphazyme's risk management and internal controls in relation to its financial reporting and approve Orphazyme's general policies in that regard. The Audit Committee assists the Board of Directors in overseeing the reporting process and the most important risks involved in this respect. The Executive Management is responsible for the effectiveness of the internal controls and risk management and for the implementation of such controls aimed at mitigating the risk associated with the financial reporting. Orphazyme has internal control and financial reporting procedures aimed at enabling it to monitor its performance, operations, funding, and risk.

CORPORATE SOCIAL RESPONSIBILITY

Orphazyme has currently not developed and implemented a formal Corporate Social Responsibility (CSR) policy. For 2018, we will increase our focus on CSR, seeking to become compliant to the extent possible in accordance with the given regulations.

As a part of our focus and commitment to developing treatments for patients suffering from orphan diseases, though, we naturally find it important to conduct responsible business. Below we will pin out our approach to CSR.

As a biopharmaceutical company, we are aware of our ethical responsibilities towards society, patients, stakeholders, and employees and we believe that our corporate behavior should always be of the highest ethical standard. When conducting our business, we strive to act ethically and comply with international regulations, guidelines, and standards for clinical trials and drug development as well as production, testing, marketing, and sale of pharmaceutical drugs. We believe that an ethical behavior is a significant factor for Orphazyme's status as an attractive workplace for both current and future employees.

Working closely with our stakeholders and partners, Orphazyme is socially and environmentally responsible and strives to comply with all relevant laws, regulations, guidelines, and policies.

> **We believe that an ethical behavior is a significant factor for our status as an attractive workplace for both current and future employees.**

ORGANIZATION

Orphazyme supports human and labor rights, but has, due to a limited number of suppliers and employees working in a highly regulated industry, not found it necessary to implement a separate human and labor rights policy.

Orphazyme views diversity as an integrated part of a socially responsible company. In accordance with applicable law, we have adopted a Diversity Policy, which sets out our goals for increasing the diversity in the Board of Directors and at other management levels. We encourage diversity, including age, ethnicity, nationality, religion, education, and skills. The Company seeks to be an attractive workplace for both women and men, and endeavors to ensure that women and men have equal opportunities for furthering their careers and for attaining and occupying management positions. Currently, our staff consists of 65.5% females and 34.6% males, including 70% female and 30% male employees on director level or above.

The Board of Directors currently comprises one woman and seven men. There has not been a new election to the Board of Directors. The Board of Directors' target is to include at least two female board members by the end of 2021.

Please refer to page 22, Human Resources, for more information on our employee composition. Orphazyme's Diversity Policy is available on our website www.orphazyme.com.

Diversity Policy

ENVIRONMENT

Orphazyme focuses on compliance with environmental laws and other applicable requirements with an emphasis on minimizing pollution and the environmental impact of its business. We also strive to conduct our business in a sustainable manner and our employees are encouraged to work in an environmentally friendly way.

HUMAN RESOURCES

At Orphazyme, we are determined to recruit highly skilled employees that will benefit the development of our clinical pipeline

Orphazyme is located in Copenhagen, Denmark at COBIS, Ole Maaløes Vej 3, DK-2200 Copenhagen N.

The office in Copenhagen serves as headquarter and focuses on coordination and execution of the drug development process and on the conduct of preclinical and clinical trials and administration. As of December 31, 2017, the organization comprised:

- Senior Management (4 persons comprising the Executive Management and Key Employees)
- Administration (2 persons)
- Clinical development (12 persons)
- CMC/QA (as defined below) (3 persons)
- Regulatory (2 persons)
- Research (11 persons)

Administration is responsible for supporting the Senior Management, Human Resources, Finance, and Investor Relations. Clinical development comprises clinical trial and central laboratory oversight. Chemistry, Manufacturing, and Controls (CMC) and Quality Assurance (QA) comprise internal and external audits. Regulatory comprises interactions with regulators and related strategic oversight. Research comprises internal and external efforts primarily to support on-going clinical development and to a lesser extent identify new lead compounds with potential to enter clinical development.

At Orphazyme, we are dependent on adequate knowledge resources and as such, we are determined to recruit highly skilled employees that will benefit the development of our clinical pipeline.

KEY EMPLOYEE RATIOS

	2017		2016	
	Male	**Female**	**Male**	**Female**
Orphazyme A/S	34.4%	65.6%	50.0%	50.0%
Executive Management and Key Employees	100%	0.0%	100%	0.0%
Director level and above	30.0%	70.0%	50.0%	50.0%
Below director level	22.2%	77.8%	33.3%	66.7%

OTHER KEY EMPLOYEE RATIOS

	2017	2016
Employees at year-end	34	22
Research & Development	28	16
Administration	2	1
Number of nationalities	6	5

PARTNERSHIPS

Orphazyme strives to develop its expertise within its therapeutic areas of interest through close collaborations with academic experts and patient organizations. Through these partnerships, Orphazyme supports the advancement of molecular and clinical understandings and performs preclinical evaluations in biological models of relevant diseases. Orphazyme's academic partners include academic professors and clinicians from institutions such as the University of Oxford, University Hospital of Udine, University College London, University of Miami, University of Cambridge, University of Kansas, and University of Helsinki. Please refer to page 24 for key agreements.

UNIVERSITY OF MIAMI

Option Agreement

In May 2017, Orphazyme entered into an option agreement with the University of Miami. Pursuant to the option agreement, Orphazyme is during an initial period granted a first option to negotiate a world-wide royalty bear- ing, exclusive license to data, know- how and patent rights generated by the University of Miami in a Phase II clinical trial of arimoclomol to treat ALS with the SOD1 mutation to use or apply the study data. Orphazyme has also been granted internal development use rights to the data, know-how, and patent rights. Please refer to Orphazyme's Prospectus of November 2017 for further details

UNIVERSITY OF KANSAS AND UNIVERSITY COLLEGE LONDON

License Agreement

In October 2017, Orphazyme entered into a license agreement with University of Kansas and UCL Business PLC (a wholly-owned subsidiary of University College London). The license agreement grants Orphazyme the world-wide, royalty-bearing exclusive license to develop and commercialize products under all data generated in the course of the on-going Phase II/III clinical trial of arimoclomol for the treatment of sIBM. Orphazyme's license includes any inventions and know-how included in such data. The trial was initiated in August 2017 with the University of Kansas as sponsor, but the license agreement provides that the Investigational New Drug (IND) and trial sponsorship shall be transferred to Orphazyme on Orphazyme's request. Under the terms of the license agreement, Orphazyme shall pay an aggregate royalty of 2% of net sales of products sold for the treatment of sIBM. Orphazyme is required to use commercially diligent efforts to develop and commercialize such products. The license agreement also provides that Orphazyme in consideration of the license shall issue bonus shares in favor of the University of Kansas and UCL Business PLC, for up to an aggregated value of USD 2.5 million (around DKK 15.8 million) in total depending on the size of the grants awarded to the universities under the trial (with a price per share calculated based on the average closing price of the shares on Nasdaq Copenhagen for the 30 days immediately prior to the date of issuance). The shares shall be issued or delivered on a yearly basis subject to certain reporting requirements

CYTRX

Asset Purchase Agreement

In May 2011, Orphazyme entered into an Asset Purchase Agreement with the US biopharmaceutical company CytRx. Pursuant to this agreement, CytRx irrevocably sold and transferred certain preclinical and clinical data, intellectual property rights, and other assets, including contractual rights and obligations relating to a portfolio of chemical compounds, including arimoclomol, to Orphazyme. Under the terms of the Asset Purchase Agreement, Orphazyme made an up-front cash payment of USD 150,000 in 2011 and further agreed to make future payments to CytRx contingent upon the achievement of specified clinical/regulatory and sales milestones as well as royalty payments based on a specified percentage of any eventual net sales of products containing one of the compounds purchased. Please refer to Orphazyme's Prospectus of November 2017 for further details

RISK MANAGEMENT

OUR VISION

Our vision is to profoundly impact the lives of patients with orphan diseases and their families.

OUR STRATEGY

Development of treatments for orphan diseases with protein misfolding where we can apply our specialized know-how in HSPs.

Our vision is to profoundly impact the lives of patients with orphan diseases and their families. Our strategy to accomplish this vision is to develop treatments for orphan diseases with protein misfolding where we can apply our specialized know-how in HSPs. We are implementing this strategy through three key objectives as listed on the following pages. Failure to achieve these key objectives may have significant and adverse effects on our business, financial condition, results, and future prospects.

RISK THREATENING

Key objective	Risks that threaten the achievement of our key objectives	Our actions to mitigate the risks
1. To successfully conduct and complete the on-going and planned trials of arimoclomol for the treatment of sIBM, ALS, NPC, and Gaucher disease.	Designing and conducting clinical trials is complex, costly, and time-consuming and the results are unpredictable. There is a risk that no matter how well-designed and diligent our preparation has been, the clinical trial results will not demonstrate sufficient evidence of safety and efficacy to ensure the requisite regulatory approvals are granted. If we are not able to successfully conduct and complete on-going and planned clinical trials, we will not be able to commercialize any pharmaceutical products.	We make every effort to design and plan our clinical trials in the most diligent manner. We partner with professional organizations to conduct our clinical trials in order to maximize quality, safety, and efficacy. We maintain frequent interactions with the regulatory bodies in the USA, Europe, and India to advance our programs towards approval in the most expedient manner.
2. To develop our commercialization strategy and build commercial structure and operations.	We face competition from other life science companies developing treatments for similar diseases, and as the potential market for our pharmaceutical products is not so vast, competing products may gain wider acceptance within the market and could render our products obsolete or limit our ability to generate revenues. An important dimension to our commercialization is the ability to obtain and maintain orphan designation/status, which will provide us with marketing exclusivity. Finally, a lack of in-house commercialization capabilities, including sales and marketing expertise, can threaten our product roll-out.	We are refining our strategy and planning our commercialization roll-out in anticipation of positive clinical results and subsequent regulatory approvals. As the clinical development program for arimoclomol progresses, we intend to refine and finalize our commercialization strategy and build our commercial structure and operations. We intend to build our own sales force in key markets, being the USA and Europe. Outside of these key markets, we currently intend to partner with local or regional distributors or license partners.
3. To use our expertise, including proprietary know-how, to select and develop new molecular entities (NMEs) for other protein-misfolding diseases.	To a large extent, our success depends on our ability to obtain and maintain patents and other intellectual property rights for our products. Our IP is the basis for our current products and any potential new leads, and thus any threats to our IP rights could be detrimental to our future pipeline of product candidates.	We are developing a suite of new molecular entities (NMEs) with improved characteristics. We have attracted highly talented resources to continue to develop and explore new leads. In addition, we are consistently monitoring our IP in order to, not only protect our rights and minimize legal claims, but also strengthen our rights and current technology platform. We believe that our patent portfolio has a wide scope of protection and geographical coverage.

In addition to risks threatening the achievement of our key objectives, we are exposed to pervasive risks that threaten our business.

PERVASIVE RISKS

Pervasive risk	How the risk threatens our business	Our actions to mitigate the risks
Lack of funding.	In order to execute our strategy, we may need to raise additional capital and additional funding may not be available on favorable terms. If we are not able to obtain timely financing, it may cause a delay in our clinical trials, commercialization activities, or other critical activities.	Following our IPO in November, we expect to be able to fund our operating plans through at least until 2020. If needed, we are prepared to raise additional funds, obtain debt financing, or seek partnerships or other financing arrangements in order to have adequate funds at our disposal in order to complete clinical trials in all indications simultaneous with pursuing filing and registration activities and preparation of commercialization activities.
Data privacy concerns and cybersecurity breaches.	We possess sensitive personal data, including information from clinical trials and other health data. We are subject to data protection laws, privacy requirements, and other regulatory restrictions in which we operate. Any failure, on our part, to comply with these requirements could result in penalties, fines, or suspension of our approvals or registrations. Also, cybersecurity attacks on our servers, databases, or information systems could compromise the privacy of our data or cause interruption to our operations.	We are in process of adopting the new procedures in order to comply with the EU General Data Protection Regulation that enters into force in May 2018. Our IT-security level is being scrutinized and new procedures implemented in order to reduce the risk of cybercrime.
Non-compliance with legislation and industry standards	In all stages and phases of our operations we are subject to regulatory and legislative obligations in order to conduct business. Regulatory and legislative requirements are subject to change and if we do not remain abreast of the regulations and actively work to comply, we are at risk of either losing or not obtaining required approvals in order to implement our business strategy.	Our organization consists of two full-time regulatory resources who interact with regulators and actively monitor the current regulatory environment. As of the publication date of this report, a new regulatory resource has been employed as well as a dedicated QA resource to prepare the organization for the preparation to file activities.

Orphazyme's vision is to profoundly impact the lives of patients with orphan diseases and their families.

SHAREHOLDERS AND SHARE INFORMATION

OWNERSHIP

Since November 16, 2017, Orphazyme is listed on Nasdaq Copenhagen under the ticker symbol ORPHA.CO. In compliance with the rules and regulations stipulated for companies listed on this exchange, we conduct our communication in accordance with the Danish Financial Supervisory Authority and Nasdaq Copenhagen.

As of December 31, 2017, the number of registered shareholders totaled 3,182 shareholders holding a total of 15,424,472 shares, which represented 74.43% of the total share capital of 19,928,184.

All shares have the same rights, including in respect of eligibility to receive dividends and participate in share buybacks. Orphazyme has not declared or made any dividend payments for the last two financial years. Currently, the Company intends to use all available financial resources as well as revenue, if any, for purposes of the Company's current and future business. As of the date hereof, the Company does not expect to make dividend payments within the foreseeable future.

Below is a table containing the details of the major shareholders as at December 31, 2017.

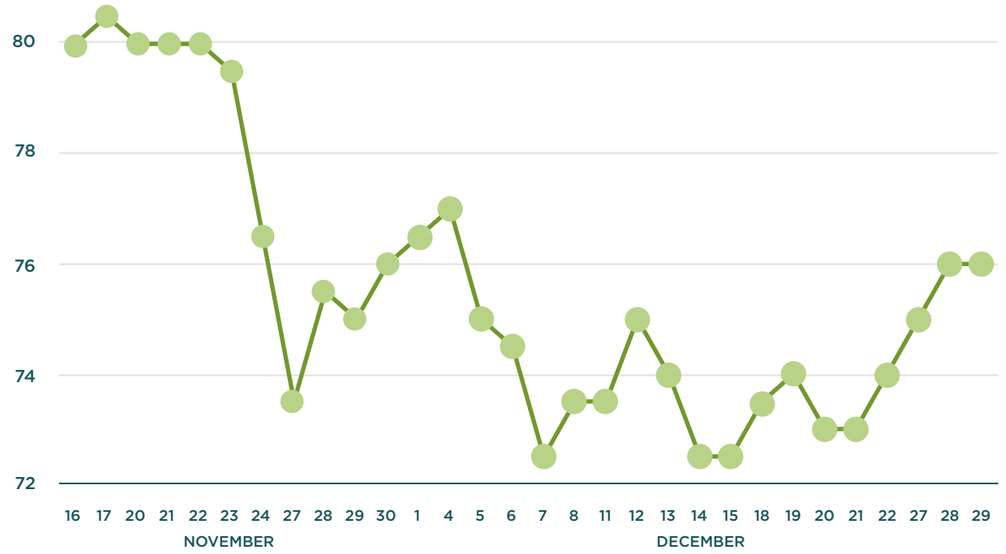
Concurrent to Orphazyme's admission on Nasdaq Copenhagen, the Company has entered into lock-up agreements with a number of stakeholders. The lock-up agreements entail that the parties must not sell, directly or indirectly, any Orphazyme shares or securities convertible into shares. The agreements are valid from the admission date until Orphazyme's publication of the results for the on-going NPC Phase II/III trial, currently expected in Q3 2018, although at the earliest 180 days and at the latest 360 days from admission.

MAJOR SHAREHOLDERS, DECEMBER 31, 2017

Major shareholder	Company address	Share capital % at December 31, 2017
Novo Holdings A/S	Tuborg Havnevej 19, 2900 Hellerup, Denmark	19.2%
LSP V Coöperatieve U.A.	Wilhelmina Tower 7th Floor, Delflandlaan 1, 1062 EA Amsterdam, Netherlands	13.6%[1]
Sunstone Life Science Ventures Fund II K/S	Lautrupsgade 7, 2100 Copenhagen, Denmark	9.1%
Coöperatieve Aescap Venture I U.A.	Science Park 406, building Matrix V, Amsterdam, 1098 XH, Netherlands	8.9%

[1] Includes a direct shareholding of 1.4% and an indirect shareholding of 12.2% of the total share capital and voting rights held through Orpha Pooling B.V. (a joint venture between LSP V Coöperatieve U.A. and ALS Invest 2 B.V.)

SHARE PERFORMANCE 2017*



*November 16-December 31, 2017

Lock-up agreements as entered into with the following parties:

- The Board of Directors
- Executive Management
- Key Employees

In accordance with Orphazyme's Articles of Association article 12, all communication from the Company to shareholders, including notices convening general meetings, may take place electronically by posting on Orphazyme's website or by email. Registered shareholders may register their electronic contact details via our investor portal, which can be accessed via www.orphazyme.com.

SHARE PERFORMANCE

Please note that the share performance chart and geographical split are based on information starting at the listing date, November 16, 2017, until year-end, December 31, 2017.

For more information on Orphazyme's share capital, please refer to note 11.

INVESTOR RELATIONS

Orphazyme's Investor Relations' primary goal is to ensure a timely communication of anything interesting or relevant to our stakeholders. This, we do by communicating both company announcements (containing potentially share-sensitive information) and investor news (pertaining to interesting news that is not share-sensitive).

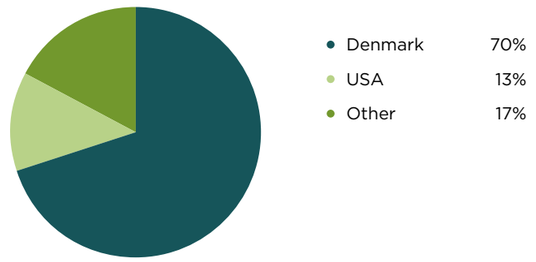
We strive to keep our shareholders and investors informed at all times, by providing annual and half-year reports, hosting analyst and investor meetings, upholding an informative and transparent website, containing all relevant reports, announcements, policies, etc. Orphazyme is followed by three analysts: Carnegie, Danske Equities, and Oddo.

Please visit our website for further details: www.orphazyme.com.

Carnegie, Danske Equities, and Oddo

If you have any questions for Investor Relations, please feel free to contact Orphazyme's CFO, Anders Vadsholt: afv@orphazyme.com.

GEOGRAPHICAL SHAREHOLDER DISTRIBUTION



NEWS

2017 COMPANY ANNOUNCEMENTS

24 OCTOBER 24
Orphazyme announces intention to launch an Initial Public Offering

6 NOVEMBER 6
Orphazyme publishes offering circular and the indicative offer price range for its IPO

14 NOVEMBER 14
Early close of offering of shares in Orphazyme

16 NOVEMBER 16
- Election of new members of the Board of Directors
- Orphazyme announces the result of its IPO, including an offer price of DKK 80 per share
- Financial calendar
- Stabilization period begins
- New share-based incentive program
- Reporting of transactions made by persons discharging managerial responsibilities and persons closely associated with them in Orphazymes' shares
- Major shareholder announcement

20 NOVEMBER 20
- No termination or withdrawal of Orphazyme's IPO
- Completion of the Offering and registration of share capital increase

21 NOVEMBER 21
Reporting of transactions made by persons discharging managerial responsibilities in Orphazyme A/S

24 NOVEMBER 24
Stabilization measures taken

5 DECEMBER 5
Stabilization measures taken

14 DECEMBER 14
Stabilization measures taken and end of stabilization period

18 DECEMBER 18
Major shareholder announcement

2017 INVESTOR NEWS

MARCH 8*
Orphazyme raises EUR 14 million to further expand its clinical development program

MAY 15*
Orphazyme completes enrollment of patients for Phase II/III clinical trial in Niemann-Pick disease Type C

NOVEMBER 7*
Orphazyme receives Orphan Drug Designation to arimoclomol for Inclusion Body Myositis from the US FDA

DECEMBER 15
Orphazyme assumes sponsorship of Phase II/III sIBM trial

*Published prior to IPO completed on November 16, 2018.

CORPORATE INFORMATION

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

Nordea
Vesterbrogade 8
0900 Copenhagen C

LEGAL COUNSEL
Gorrissen Federspiel, Advokatpartnerselskab
Axeltorv 2
DK-1609 Copenhagen V

INDEPENDENT AUDITORS
EY
Osvald Helmuths Vej 4
DK-2000 Frederiksberg

Annual report
Copies of this annual report in English are available upon request.

Annual General Meeting
The Annual General Meeting will be held on April 12, 2018 at 5.00 PM CET at:
Gorrissen Federspiel, Axeltorv 2,
DK-1609 Copenhagen V

FINANCIAL CALENDAR 2018
Annual General Meeting
Thursday, April 12, 2018

Interim report for the period ending June 30, 2018
Tuesday, August 28, 2018

BOARD OF DIRECTORS

Georges Gemayel holds a Master and a PhD in Pharmacology from Paris-Sud University and a Docteur d'Exercice en Pharmacie from the St. Joseph University.

Georges Gemayel is currently Chairman of the Board of Directors of Enterome SA and OxThera AB and a member of the Board of Directors of Momenta Pharmaceuticals Inc. (publ) and Supernus Pharmaceuticals Inc. (publ).

Member since: 2012 (Chairman since 2014)
Born in: 1960
Nationality: American
Committees: Nomination Committee (Chairman)

Bo Jesper Hansen holds an MD and a PhD in Medicine from the University of Copenhagen.

Bo Jesper Hansen is currently Chairman of the Board of Directors of Laborie Inc. and Innoventa Medica ApS and a member of the Board of Directors of Azanta A/S and Ascelia Pharmaceuticals AB.

Member since: 2010 (Deputy Chairman since 2017)
Born in: 1958
Nationality: Danish
Committees: Remuneration Committee (Chairman)

Martin Bonde holds a Graduate Diploma in Business Administration from Copenhagen Business School, a Master of Science, and a PhD in Chemical Engineering from the Technical University of Denmark.

Martin Bonde is currently Chief Executive Officer of Vaccibody AS and Bohrs Towers IVS as well as a member of the Board of Directors and the Executive Management of Biotopix ApS. Martin Bonde is furthermore Chairman of the Board of Directors of the trade organization DANSK BIOTEK.

M**ember since:** 2010
Born in: 1963
Nationality: Danish
Committees: Nomination Committee

Anders Hedegaard holds a Master of Science in Chemical Engineering and Biochemistry from the Technical University of Denmark.

Anders Hedegaard is currently Chief Executive Officer of GN Store Nord A/S and GN Hearing A/S and a member of the Board of Directors of the Confederation of Danish Enterprise, Hearing Instrument Manufacturers Software Association A/S, and HIMSA II A/S.

Member since: 2017
Born in: 1960
Nationality: Danish
Committees: Remuneration Committee

Rémi Droller holds a Master in Molecular Biology from Université Pierre et Marie Curie and a Master in Finance and Management of Innovation from Masternova.

Rémi Droller is currently Managing Partner of Kurma Partners SA and Chairman of the Board of Directors in Dyncaure SAS and Im-Check SAS. Member of the Board of Directors of OxThera AB, AM Pharma BV, STAT Dx S.L., and Pharvaris BV.

Member since: 2015
Born in: 1975
Nationality: French
Committees: Remuneration Committee

Martijn Kleijwegt holds a Master's degree from the University of Amsterdam.

Martijn Kleijwegt is currently Founder and Manging Partner at LSP Management Group BV. and a member of the Board of Directors at Kiadis Pharma N.V. (publ), OxThera AB, Eloxx Pharmaceuticals Ltd., and Pharvaris BV.

Member since: 2017
Born in: 1955
Nationality: Dutch
Committees: Audit Committee

Sten Verland holds a Master in Biology and Mathematics and a PhD in Immunology from the University of Copenhagen.

Sten Verland is currently Co-Founder of Sunstone Capital A/S, Senior Partner at Sunstone Life Science Ventures A/S, in the Executive Management of Verland Capital ApS, Verland Holding ApS, Verland Holding II ApS, Genobiotix ApS, and in companies in or associated with the Sunstone Group. Member of the Board of Directors of Anergis SA, Vaximm AG, F2G Ltd., MinervaX ApS, OxThera AB, Danish Venture Capital and Private Equity Asscociation (DVCA), and in companies in or associated with the Sunstone Group.

Member since: 2010
Born in: 1957
Nationality: Danish
Committees: Nomination Committee, Audit Committee

Catherine Moukheibir holds a Master in Economics and an MBA, both from Yale University.

Catherine Moukheibir is currently Chairman of the Board of Directors of MedDay Pharmaceuticals SA and a member of the Board of Directors of Zealand Pharma A/S (publ), Genkyotex SA (publ), Ablynx NV (publ), and Cerenis Therapeutics SA (publ).

Member since: 2017
Born in: 1959
Nationality: American, Lebanese, and British
Committees: Audit Committee (Chairman)

EXECUTIVE MANAGEMENT

PhD in Medicine, University of Copenhagen. Previously at BankInvest Biomedical Venture and Assistant Professor in Systems Biology.

Born in: 1973
Nationality: Danish

MBA in Finance, Melbourne University. 20+ years' experience from biotech and corporate finance. Previously at Topotarget, BankInvest Biomedical Venture, 7TM Pharma, and Carnegie.

Joined in: 2016
Born in: 1969
Nationality: Danish

KEY EMPLOYEES

PhD in Medicine, University of Copenhagen. Orphazyme's scientific rationale published in Nature and ScienceTM with +200 references. Inventor of +60 patents in heat-shock response therapy.

Born in: 1977
Nationality: Danish

MD, University of Zürich. Board-certified neurologist with 12+ years' experience in neuroscience development. Previously at Roche, Bristol-Myers Squibb, and Novartis.

Joined in: 2016
Born in: 1967
Nationality: Swiss

Executive MBA, Boston University. 30+ years' experience from the biotech and biopharm**aceutical in**dustry. Previously at Catalyst Pharmaceuticals, Aegerion Pharmaceuticals, Genzyme, Genentech, Marion Laboratories, and Pfizer.

Joined in: 2018
Born in: 1960
Nationality: American

2017 ORPHAZYME

FINANCIAL STATEMENTS

STATEMENT OF PROFIT OR LOSS AND OTHER COMPREHENSIVE INCOME

Note		Twelve months ended December 31, 2017 TDKK	Twelve months ended December 31, 2016 TDKK
4,5,9, 10	Research and development expenses	(99,048)	(55,817)
5,10, 11	General and administrative expenses	(31,994)	(7,703)
	Operating loss	**(131,042)**	**(63,520)**
7	Financial income	26	182
8	Financial expenses	(688)	(97)
	Loss before tax	**(131,704)**	**(63,435)**
12	Income tax benefit	5,500	5,500
	Net loss for the period	**(126,204)**	**(57,935)**
	Other comprehensive income/(loss)	-	-
	Total comprehensive loss	**(126,204)**	**(57,935)**
13	Loss per share, basic and diluted	(10.46)	(5.89)

Distribution of the year's result
The Board of Directors proposes that the net loss for 2017 TDKK 126,204 (2016: net loss of TDKK 57,935) be carried forward to next year by transfer to accumulated deficit.

See accompanying notes to these financial statements.

STATEMENT OF FINANCIAL POSITION

Note		December 31, 2017 TDKK	December 31, 2016 TDKK
	ASSETS		
	Non-current assets		
9	Licenses	9,853	-
10	Property, plant, and equipment	1,851	987
12	Corporation tax receivable	2,750	2,750
	Leasehold deposits	410	310
	Total non-current assets	**14,864**	**4,047**
	Current assets		
12	Corporation tax receivable	5,500	5,500
	Other receivables	5,871	3,421
	Prepayments	4,847	4,624
	Cash and cash equivalents	631,735	14,349
	Total current assets	**647,953**	**27,894**
	TOTAL ASSETS	**662,817**	**31,941**
	EQUITY AND LIABILITIES		
	Equity		
11	Share capital	19,928	3,361
11	Share premium	924,143	226,285
9	Share-based compensation – acquisition of intangible rights	9,972	-
	Accumulated deficit	(338,341)	(212,137)
	Total equity	**615,702**	**17,509**
	Current liabilities		
	Trade payables	13,436	4,718
	Other payables	33,679	9,714
	Total current liabilities	47,115	**14,432**
	TOTAL EQUITY AND LIABILITIES	**662,817**	**31,941**

1	Accounting policies
15	Contractual obligations and contingencies

See accompanying notes to these financial statements.

STATEMENT OF CHANGES IN SHAREHOLDERS' EQUITY

	Note	Share capital TDKK	Share premium TDKK	Share-based compensation – aquisition of intangible assets TDKK	Accumulated deficit TDKK	Total TDKK
Balance as of December 31, 2015		**3,346**	**224,999**	**-**	**(154,202)**	**74,143**
Net loss for the period		-	-	-	(57,935)	(57,935)
Other comprehensive loss for the period		-	-	-	-	-
Total other comprehensive income/(loss)		-	-	-	(57,935)	(57,935)
Transactions with owners		-	-	-	-	-
Capital increase		15	1,316	-	-	1,331
Expenses, capital increase		-	(30)	-	-	(30)
Share-based payment expense	6	-	-	-	-	-
Total transactions with owners		15	1,286	-	-	1,301
Balance as of December 31, 2016		**3,361**	**226,285**	**-**	**(212,137)**	**17,509**
Net loss for the period		-	-	-	(126,204)	(126,204)
Other comprehensive loss for the period		-	-	-	-	-
Total other comprehensive income/(loss)		-	-	-	(126,204)	(126,204)
Transactions with owners		-	-	-	-	-
Contribution of a license agreement	9	-	-	9,972	-	9,972
Share issue in connection with conversion of former preference shares into ordinary shares	11	6,488	(6,488)	-	-	-
Proceeds from IPO	11	7,500	592,500	-	-	600,000
Costs related to IPO		-	(42,605)	-	-	(42,605)
Capital increase	11	1,741	155,010	-	-	156,751
Exercise of warrants for cash	6	838	323	-	-	1,161
Costs related to non-IPO related capital increases		-	(1,004)	-	-	(1,004)
Share-based payment expense	6	-	122	-	-	122
Total transactions with owners		16,567	697,858	9,972	-	724,397
Balance as of December 31, 2017		**19,928**	**924,143**	**9,972**	**(338,341)**	**615,702**

See accompanying notes to these financial statements

STATEMENT OF CASH FLOWS

	Note	Twelve months ended December 31, 2017 TDKK	Twelve months ended December 31, 2016 TDKK
Operating activities			
Operating loss		(131,042)	(63,520)
Adjustments to reconcile loss before tax to cash flows from operating activities			
Share-based payment expense	6	122	-
Depreciation and amortization	9,10	627	706
Gain/(loss) on sale and disposal of assets		119	33
Change in other receivables		(2,555)	(2,876)
Change in prepayments		(223)	1,347
Change in trade payables		8,718	2,271
Change in other payables		23,970	1,352
Corporate taxes received	12	5,500	5,875
Interest received/(paid)		(662)	85
Net cash used in operating activities		**(95,426)**	**(54,727)**
Investing activities			
Investment in property, plant, and equipment	10	(1,491)	(238)
Net cash used in investing activities		**(1,491)**	**(238)**
Financing activities			
Proceeds from IPO	11	600,000	-
Costs related to IPO		(42,605)	-
Capital contributions from shareholders	11	157,912	1,330
Costs related to capital contributions		(1,004)	(30)
Net cash provided by financing activities		**714,303**	**1,300**
Net change in cash and cash equivalents		**617,386**	**(53,665)**
Cash and cash equivalents at the beginning of the period		14,349	68,014
Cash and cash equivalents at the end of the period		**631,735**	**14,349**

See accompanying notes to these financial statements

NOTES TO THE FINANCIAL STATEMENTS

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 2017 and 2016, the company had no subsidiaries, and consequently, the financial statements are an individual financial statement. After the balance sheet date, the company has established a 100%-controlled subsidiary in the USA. On November 16, 2017, the Company completed an IPO process and was listed on Nasdaq Copenhagen in the Mid-Cap segment. The financial statements for the year ended December 31, 2017 were authorized for approval at the Annual General Meeting to be held on April 12, 2018, with a resolution of the Board of Directors on March 15, 2018.

NOTE 1 – ACCOUNTING POLICIES

Basis of preparation
The financial statements of the Company have been prepared in accordance with International Financial Reporting Standards, or IFRS, as adopted by the EU and additional requirements under the Danish Financial Statements Act.

The financial statements have been prepared on a historical cost basis except for share-based payment. The financial statements are presented in Danish Kroner, or DKK, which is the functional currency of the Company based on facts and circumstances and the technical requirements of IFRS.

Segment information
For management purposes, the Company is managed and operated as one business unit that is reflected in the organizational structure and internal reporting. No separate lines of business or separate business entities have been identified with respect to any product candidate or geographical market and no segment information is currently disclosed in the Company's internal reporting. Accordingly, it has been concluded that it is not relevant to include segment disclosures in the financial statements as the Company's business activities are not organized into business units, products, or geographical areas.

Foreign currency transactions, and balances
On initial recognition, transactions denominated in foreign currencies are translated at the foreign exchange spot rate at the transaction date. Differences arising between the foreign exchange spot rates at the transaction date and the date of payment are recognized in the income statement as financial income or financial expenses.

Receivables and payables and other monetary items denominated in foreign currencies are translated at the foreign exchange spot rates at the balance sheet date. The difference between the foreign exchange spot rates at the balance sheet date and the date at which the balance was recognized are recognized in the income statement as financial income or financial expenses.

Share-based payment
Employees and Management of the Company receive remuneration in the form of equity-settled awards whereby services are rendered as consideration for warrants. The cumulative expense recognized for equity-settled transactions at each reporting date until the vesting date reflects the extent to which the vesting period has expired and the company's best estimate of the number of equity 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 fair value of these equity-settled 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 equity awards that may occur over the service period. Fair value of warrants and options granted prior to the IPO has been determined using the Black-Scholes model. Fair value of warrants and options granted after the IPO has been determined using the Monte-Carlo model.

The cost of share-based payments is 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. For grants which vest subject to obtaining a specified share price, a compensation expense is recognized regardless of whether the share price condition is met if all other vesting conditions are met. 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. Share-based payments related to acquisition of intangible rights are discussed below.

Employee benefits
Employee benefits are primarily made up of salaries, share-based payments, and pension. The cost of these benefits is recognized as an expense as services are delivered. The Group's contributions to the employee pension plan have not been material. All employee pension plans are defined contribution plans and not defined benefit plans.

Leases
Leases that do not transfer substantially all the risks and rewards incident to the ownership to the Company are classified as operating leases. Payments relating to operating leases and any other leases are recognized in the income statement over the term of the lease. The Company's aggregate liabilities relating to operating leases and other leases are disclosed under contingencies, etc.

Public grants
The public grants received by the Company provides compensation for a part of certain project-specific research and development expenses, including wages and salaries. Public grants are recognized where there is reasonable assurance that the grant will be received and all attached conditions will be complied with. Grants relating to expense items are recognized in the statement of profit or loss and set off against the related research and development expenses on a systematic basis over the periods that the related expenses for which it is intended to compensate, are expensed. The terms of the grants do not obligate the Company to repay any of portion of the grant.

Financial instrument valuation hierarchy
Financial instruments recognized at fair value are allocated to one of the following valuation hierarchy levels:

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

No financial instrument is measured at fair value as of the balance sheet dates presented in the financial statements.

Intangible assets
Intangible assets such as license rights to develop and commercialize products that are acquired separately are measured on initial recognition at cost. For acquisition of intangible rights involving equity-settled share-based payment transactions, Orphazyme either measures the fair value of the rights received, and the corresponding increase in equity (presented as "Share-based compensation – acquisition of intangible rights " within equity), directly, at the fair value of the rights received, unless that fair value cannot be estimated reliably. If Orphazyme cannot estimate reliably the fair value of the rights received, Orphazyme measures their value, and the corresponding increase in equity, indirectly, by reference to the fair value of the equity instruments granted.

Following initial recognition, intangible assets are carried at cost less any accumulated amortization and accumulated impairment losses.

Development costs are capitalized when they fulfill the criteria set out in IAS 38 and are expected to represent significant amounts for the development initiatives as a whole. Development costs are otherwise expensed under research and development costs. No development costs has been capitalized as the management assesses that they do not fulfill the criteria set out in IAS 38.

The useful lives of intangible assets are assessed as either finite or indefinite.

Intangible assets with finite lives such as license rights to develop and commercialize products are amortized over the useful economic life and assessed for impairment whenever there is an indication that the intangible asset may be impaired.

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

The amortization period for intangible licensed rights is based on the duration of the underlying license agreement and has been estimated to be approximately 14 years.

Property, plant, and equipment
Property, plant, and equipment includes fixtures, fittings, leasehold improvements. and other plant and equipment, and are measured at cost less accumulated depreciation and impairment losses. Cost includes the acquisition price and costs directly related to the acquisition until the time at which the asset is ready for use. Depreciation is calculated on a straight-line basis over the expected useful lives of the underlying assets of five years. The residual values of equipment are not material. The useful life of and method of depreciation of equipment are reviewed by management at least each year-end or more often based on changes in facts and circumstances. Changes in useful lives or residual values are adjusted prospectively as changes in accounting estimates.

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

Corporation tax receivable
Corporation tax receivable is recognized in the balance sheet as the tax benefit computed on the taxable loss for the year, adjusted for any changes to the prior year benefit due to changes in the taxable loss of prior years and for any taxes already paid or refunded. Management periodically evaluates positions taken in the tax returns with respect to situations in which applicable tax regulations are subject to interpretation or "uncertainty" and establishes provisions where appropriate. To date, there have been no provisions established for uncertain tax positions.

Deferred tax is measured using the balance sheet liability method on all temporary differences between the carrying amount and the tax value of assets and liabilities, with the exception of temporary differences occurring at the time of acquisition and liabilities neither affecting the result of operation nor the taxable income. The Company has no deferred tax balances as of December 31, 2017 or 2016.

For further details please refer to note 2 and note 11.

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

- When the sales tax incurred on a purchase of assets or services is not recoverable from the taxation authority, in which case, the sales tax is recognized as part of the cost of acquisition of the asset or as part of the expense item, as applicable
- When receivables and payables are stated with the amount of sales tax included

The net amount of sales tax recoverable from, or payable to, the taxation authority is included as part of receivables or payables in the statement of financial position.

Leasehold deposits
Deposits for property leased by the Company are measured at amortized cost.

Prepayments
Prepayments include prepaid costs that will be incurred in subsequent financial reporting periods on a current basis.

Other receivables
Other receivables include prepaid costs that will be incurred on a noncurrent basis.

Financial assets
Initial recognition and measurement
Financial assets that meet certain criteria are classified at initial recognition as either financial assets at fair value through profit or loss, available for sale financial assets, held to maturity investments or receivables.

The Company's financial assets include other receivables and cash and cash equivalents. The Company does not hold assets that have been classified at fair value through profit or loss, available for sale or held to maturity. Generally, the Company's financial assets are available to support current operations; however, amounts expected to be realized within the next twelve months are classified within the statement of financial position as current assets.

The Company has no derivative financial assets nor has there been a change in classification of a financial asset after initial recognition and measurements as discussed herein. Financial assets are not acquired for trading or speculative purposes. The Company has not placed any assets as security for loans at either December 31, 2017 or 2016.

The Company's financial assets are recognized initially at fair value plus, in the case of financial assets not carried at fair value through profit and loss, transaction costs that are attributable to the acquisition of the financial asset, if any.

Subsequent measurement
Historically, the Company's receivables are due within a short period and therefore the impact of using the effective interest rate method on the Company's financial statements has been immaterial. The same applies to cash and cash equivalents that comprise cash at banks available on demand.

Financial asset impairment
The Company assesses at the end of each reporting period whether there has been objective evidence that a financial asset or Company of financial assets may be impaired. Impairment losses are incurred if there is objective evidence of impairment and the evidence indicates that estimated future cash flows will be negatively impacted. For financial assets held at amortized costs, the amount of impairment loss to be recognized in the financial statements is measured as the difference between the carrying value of the financial asset and the present value of the expected cash flows of the financial asset using the original effective interest rate. The Company did not experience an impairment of a financial asset for either the twelve months ended at December 31, 2016 or the twelve months ended December 31, 2017.

Cash and cash equivalents
Cash includes cash on hand and in banks, as well as short-term marketable securities that are subject to an insignificant risk of changes in value.

Financial liabilities
Historically, the Company's financial liabilities have included bank debt, trade payables, and other payables.

Bank debt
The Company has no bank debt as of December 31, 2017 and as of December 31, 2016. Bank debt is measured at amortized cost.

Trade payables
Trade payables relate to the Company's purchase of products and services from various vendors in the normal course of business. Bank debt is measured at amortized cost.

Other payables
Other payables are measured at net realizable value.

Statement of Profit or Loss and Other Comprehensive Income

Revenue
The Company does not have any revenue in any of the reporting periods.

Research and development costs
Research and development costs include salaries including share-based compensation and costs arising from research activities, clinical development, legal expenses related to the protection, defense and enforcement of the Company's intellectual property, and rent associated with facilities used for research purposes. Given the uncertainty regarding the recoverability of clinical development costs, the Company has expensed all such expenses in the statement of profit and loss and comprehensive loss for the periods presented.

General and administrative expenses
General and administrative expenses include salaries for administrative staff and management, costs of share-based payment, rent associated with facilities not used for research purposes, investor relations, and costs incurred in connection with the Initial Public Offering (which has not been set off against equity).

Financial income and expense
Financial income and expense include interest income and expense, gains and losses due to changes in foreign exchange rates, interest expenses on convertible debt, allowances, and surcharges related to the advance payment of tax scheme, and other miscellaneous items of financial income and expense.

Income tax benefit
Income tax benefit includes the current benefit due from the current period's taxable loss and deferred tax adjustments. The benefit is comprised primarily of refundable tax credits for costs incurred in connection with research and development activities under the Danish Tax Credit Regime.

Statement of cash flows
The statement of cash flows is presented using the indirect method and shows cash flows resulting from operating activities, investing activities, financing activities, and the Company's cash and cash equivalents at the beginning and end of the year.

Cash flows used in operating activities primarily comprise the net loss for the year adjusted for non-cash items, such as foreign exchange gains and losses, depreciation, changes in working capital and cash received for interest and taxes.

Cash flows from investing activities are comprised primarily of investment in property, plant, and equipment.

Cash flows from financing activities are comprised of repayment of bank debt, proceeds from share issuance net of transaction costs, including the proceeds from the IPO.

NOTE 2 - SIGNIFICANT ACCOUNTING JUDGMENTS, ESTIMATES, AND ASSUMPTIONS

The preparation of the financial statements requires management to make judgments, estimates, and assumptions that affect the reported amounts of income, expenses, assets, and liabilities as well as the accompanying disclosures. Uncertainty about these assumptions and estimates could result in outcomes that require a material adjustment to the carrying amount of the asset or liability affected in future periods.

Judgments made in applying accounting policies
In the process of applying the Company's accounting policies, management has made the following judgments and estimates that have the most significant effect on the amounts recognized in the financial statements. Refer to the following notes for more details:

- Estimation of accruals and prepaid costs for clinical research trials
- Estimation of share-based compensation (note 6)
- Estimation of fair value of licenses (note 9)
- Judgment in respect of IPO and IPO-related costs
- Judgment in respect of recognition of deferred taxes related to taxable losses to be carried forward (note 12)

Estimates and assumptions
The key assumptions concerning the future and other key sources of estimation uncertainty at the reporting date that have a significant risk of causing a material adjustment to the carrying amounts of assets and liabilities within the next financial year, are listed below. The Company based its assumptions and estimates on information available when the financial statements were prepared.

Estimation of accruals and prepaid costs for clinical trial development costs
The Company incurs substantial costs associated with clinical trials related to the AIDNPC program. The objective of the program is to develop a pharmaceutical drug for treatment of Niemann-Pick disease Type C (NPC). NPC is a lysosomal storage disease affecting around 1 in 150,000 newborns and is caused by mutations in the NPC1 or NPC2 genes.

Accounting for clinical trials relating to activities performed by CROs and other external vendors requires management to exercise significant estimates in regards to the timing and accounting for these costs. The diverse nature of 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 adds 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 upfront in connection with clinical activities. In estimating the relevant periods etc., the Company evaluates the start-up, treatment and wrap-up periods, compensation arrangements, and services rendered attributable to each clinical trial. Estimated costs are regularly tested against payment plans and trial completion assumptions.

The Company has recognized accruals related to clinical trial development costs of TDKK 6,952 and TDKK 0 for the twelve months ended December 31, 2017 and the twelve months ended December 31, 2016, respectively.

The Company has recognized prepaid costs related to clinical trial development costs of TDKK 1,305 and TDKK 3,794 for the twelve months December 31, 2017 and the twelve months ended December 31, 2016, respectively.

Estimation of share-based payment
Estimating fair value for the company's post-IPO long-term incentive programs requires determination of the most appropriate valuation model, which depends on the terms and conditions of the respective grant. This estimate also requires determination of the most appropriate inputs to the valuation model including the expected life of the LTIP, volatility dividend pay-out ratio, and risk-free interest rate and making assumptions about them. For the measurement of the fair value of post-IPO long-term incentive programs at the grant date, the Group uses a Monte-Carlo simulation model for the warrant program 2015/21.

The assumptions and models used for estimating fair value for share-based payment transactions are discussed further in note 6.

Estimation of fair value of licenses
Licenses contains an agreement entered into with University of Kansas and University College London, in which the Company will get access to data and know-how generated in the course of research in connection with the sIBM trial. Payment for license is done by issuing new shares to the contract partners for a value corresponding to the costs incurred during a calendar year. The value of the license is estimated by management based on the expected costs over the contract period.

Judgment in respect of IPO and IPO-related costs
IPO and IPO-related costs are costs that have been incurred in connection with the Company's IPO and mainly covers fees to bookrunner's, banks, and advisors. Certain costs relating to preparing the prospectus, searching for new investors, issuing comfort letters, and other directly related activities to listing the Company's shares has been charged to equity based on the percentage of the newly issued shares. The remaining costs has been charged to the income statement. Costs relating to support activities in connection with the IPO such as legal work in connection with incentive programs, conversion of the Company to an A/S, financial assistance, and the like has been charged directly to the income statement.

Judgement related to deferred taxes related to taxable losses to be carried forward
The Company recognizes deferred income tax assets if it is probable that sufficient taxable income will be available in the future against which the temporary differences and unused tax losses can be utilized. Management has considered future taxable income in assessing whether deferred income tax assets should be recognized and has concluded that the deferred income tax assets related to taxable losses to be carried forward do not meet the criteria for being recognized as assets in the statement of financial position.

The Company has net tax loss carry-forwards that are not recognized of MDKK 235 and MDKK 135 for the twelve months December 31, 2017 and the twelve months ended December 31, 2016, respectively.

The Company's tax losses can be carried forward infinitely subject to the general rules on limited deductibility due to ownership changes.

Reference is made to note 12.

NOTE 3 – STANDARDS ISSUED BUT NOT YET EFFECTIVE

The IASB has issued a number of new standards that become effective on or after January 1, 2018. Management's current expectation is that the new standards will be adopted by the company at the effective date. Depending on the stage of development of the company as of this point in time, the following new standards could have an impact of the financial statements:

IFRS 9 – Financial instruments
This standard addresses the accounting for financial assets and liabilities including their recognition, classification and measurement, and hedge accounting. The Company adopts IFRS 9 from the mandatory effective date of January 1, 2018 and will make use of the relief from restating comparative figures. The adoption of IFRS 9 is not expected to have an impact on the financial figures.

IFRS 15 – Revenue from contracts with customers
This standard addresses the accounting for revenue recognition. The Company adopts IFRS 15 from the mandatory effective date of January 1, 2018 and will make use of the relief from restating comparative figures. The adoption of IFRS 15 is not expected to have an impact on the financial figures as the company currently do not have revenue from contracts with customers.

IFRS 16 - Leases
IFRS 16 was issued in January 2016 and it replaces IAS 17 Leases, IFRIC 4 Determining whether an Arrangement contains a Lease, SIC-15 Operating Leases-Incentives and SIC-27 Evaluating the Substance of Transactions Involving the Legal Form of a Lease. IFRS 16 sets out the principles for the recognition, measurement, presentation, and disclosure of leases and requires lessees to account for all leases under a single on-balance sheet model similar to the accounting for finance leases under IAS 17. The standard includes two recognition exemptions for lessees – leases of 'low-value' assets (e.g., personal computers) and short-term leases (i.e., leases with a lease term of 12 months or less). At the commencement date of a lease, the Company will as lessee recognize a liability to make lease payments (i.e., the lease liability) and an asset representing the right to use the underlying asset during the lease term (i.e., the right-of-use asset). The Company will be required to separately recognize the interest expense on the lease liability and the depreciation expense on the right-of-use asset.

Upon implementation on January 1, 2019, assuming that no new leases are entered into and no amendments to existing leases are made, the Company is expected to recognize a liability to make lease payments (i.e. the lease liability) of approximately TDKK 0 and an asset representing the right to use the underlying asset during the lease term (i.e. the right to use asset) of approximately TDKK 0. The expected accumulated effect on equity and total assets at January 1, 2019 approximates TDKK 0 and TDKK 0, respectively. Reference is made to note 15 in respect of operating leasing obligations as at December 31, 2017.

The Company will be also required to remeasure the lease liability upon the occurrence of certain events (e.g., a change in the lease term, a change in future lease payments resulting from a change in an index or rate used to determine those payments). The Company will generally recognize the amount of the remeasurement of the lease liability as an adjustment to the right-of-use asset.

The Company expects to implement IFRS 16 applying a simplified transition method, whereby comparative figures will not be restated. Furthermore, the company expects to use the other available reliefs, e.g. in regards to low-value assets and leases with a maturity of less than 12 months to the widest possible extent.

NOTE 4 – GOVERNMENT GRANT

	Twelve months ended December 31, 2017 TDKK	Twelve months ended December 31, 2016 TDKK
Receivable/(prepayment) at January 1	1,347	-
Received during the year	(5,307)	(1,439)
Recognized in the statement of profit or loss	3,941	2,786
Receivable/(prepayment) at December 31	**(19)**	**1,347**

Government grants comprise research funding from the Danish government and the EU. Government grants are recognized in the period where the expenses funded by the grants have been incurred. Government grants are recognized as a reduction in research and development expenses as the grants are considered to be cost refunds.

Receivables are included in the statement of financial position under Other Receivables and prepaid grants are included under Other Payables.

None of the government grants received are subject to repayment clauses.

NOTE 5 – STAFF COSTS

The amounts disclosed in the table on the following page are the amounts recognized as an expense during the reporting periods. Executive Management consists of the Company's Chief Executive Officer and the Chief Financial Officer for the twelve months ended December 31, 2017 and 2016. Executive Management is also identical to the registered management of the Company. See note 16 for compensation paid to the members of the Board of Directors.

The Executive Management is eligible to receive an annual performance-based cash bonus subject to certain predefined corporate and individual goals as determined by the Board of Directors on an annual basis. These predefined goals may include financial and/or operational targets, e.g. related to financing, working capital needs, organizational development as well as preclinical and clinical development. A cash bonus received under the STIP (short-term incentive programs) may not exceed 100% of the annual fixed salary of the participants. For the financial year 2017, the company expensed TDKK 1,507 on cash bonuses and TDKK 351 on IPO bonuses for the Executive Management (2016: TDKK 385).

In connection with the completion of the IPO, all employees of Orphazyme (including the Executive Management and Key Employees) received a cash bonus corresponding to 10% of their respective annual base salaries (excluding pension contributions). The aggregate value of the cash bonus was TDKK 2,276.

NOTE 5 – STAFF COSTS (CONTINUED)

	Twelve months ended December 31, 2017 TDKK	Twelve months ended December 31, 2016 TDKK
Staff costs		
Wages/salaries	21,186	12,957
Cash bonus	3,233	781
Share-based payment (note 6)	44	-
Pensions	968	60
Other social security costs	172	147
Other staff costs	338	372
Total staff costs	**25,941**	**14,317**
Executive Management remuneration		
Wages/salaries	3,204	2,140
Cash bonus	1,858	385
Share-based payment (note 6)	78	-
Pensions	-	-
Other social security costs	7	6
Other staff costs	0	-
Total Executive Management remuneration	**5,147**	**2,531**
Total staff and Executive Management costs	**31,088**	**16,848**
Staff and Executive Management costs are recognized as follows in the financial statements:		
Research and development expenses	25,648	14,417
General and administrative expenses	5,440	2,4131
Total staff and Executive Management costs	**31,088**	**16,848**
Average number of employees	26	17

NOTE 6 – SHARE-BASED INCENTIVE PROGRAMS

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 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

- the grant date fair value of the old program under the original vesting terms, plus
- 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, 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 upon the completion of the IPO in November 2017.

The table below summarizes the activity related to the warrants for the twelve months ended December 31, 2017 and the 12 months ended December 31, 2016:

	Executive Management	Employees	Board of Directors	Consultants	Total Warrants	Warrants exercisable
Outstanding at December 31, 2015	**211,879**	**76,176**	**124,122**	**9,700**	**421,877**	**266,621**
Granted	-	-	-	-	-	-
Exercised	-	-	-	-	-	-
Expired	-	-	-	-	-	-
Outstanding at December 31, 2016	**211,879**	**76,176**	**124,122**	**9,700**	**421,877**	**324,078**
Granted	333,964	313,815	182,813	-	830,592	-
Exercised	(333,964)	(313,815)	(182,813)	(7,500)	(838,092)	-
Expired	-	-	-	-	-	-
Forfeited	(211,879)	(76,176)	(124,122)	(2,200)	(414,377)	-
Outstanding at December 31, 2017	**-**	**-**	**-**	**-**	**-**	**-**

All warrants were exercised in November 2017 upon completion of the IPO. The average share price was DKK 80.

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 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). The Board of Directors may decide to offer other current or new employees of Orphazyme to participate 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.

The table below summarizes the activity related to the LTIP for the twelve months ended December 31, 2017 and the 12 months ended December 31, 2016:

	Executive Management	Key Employees	Board of Directors	Consultants	Total Warrants	Warrants exercisable
Outstanding at December 31, 2015	**-**	**-**	**-**	**-**	**-**	**-**
Granted	-	-	-	-	-	-
Exercised	-	-	-	-	-	-
Expired	-	-	-	-	-	-
Outstanding at December 31, 2016	**-**	**-**	**-**	**-**	**-**	**-**
Granted	9,000	5,875	-	-	14,875	-
Exercised	-	-	-	-	-	-
Expired	-	-	-	-	-	-
Forfeited	-	-	-	-	-	-
Outstanding at December 31, 2017	**9,000**	**5,875**	**-**	**-**	**14,875**	**465**

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.

Fair value of the LTIP has been estimated at approximately TDKK 3,895 using a Monte-Carlo simulation model, taking into account the terms and conditions on which the LTIPs were granted.

The weighted average remaining contractual life for LTIPs outstanding as at December 31, 2017 was 3.88 years (2016: 0 years).

The exercise prices for LTIPS outstanding at the end of the year was DKK 1 (2016: DKK 0).

The following tables list the inputs to the models used for the three plans for the years ended December 31, 2017:

	Twelve months ended December 31, 2017 TDKK
Weighted average fair values at the measurement date	
Dividend yield (%)	-
Expected volatility (%)	44,6
Risk-free interest rate (%)	(0.43)
Expected life of share options/SARs (years)	3.875
Weighted average share price (DKK)	1.00
Model applied	Monte-Carlo

A four-year volatility estimate of 44.6%, an expected dividend yield of 0%. This implies an expected number of Performance Shares to be allocated of 12,711. Expected volatility has been determined on the basis of the historic volatility for comparable listed companies.

In 2017, TDKK 122 was recognized a compensation expense.

NOTE 7 – FINANCIAL INCOME

	Twelve months ended December 31, 2017 TDKK	Twelve months ended December 31, 2016 TDKK
Exchange gain	26	180
Other interest income	-	2
Total financial income	26	182

NOTE 8 – FINANCIAL EXPENSES

	Twelve months ended December 31, 2017 TDKK	Twelve months ended December 31, 2016 TDKK
Interest expense	647	71
Other financial expenses	41	26
Exchange losses	-	-
Total financial expenses	688	97

NOTE 9 – LICENSES

Contractual obligations	Licenses TDKK
Cost at December 31, 2015	-
Additions	-
Cost at December 31, 2016	-
Additions	9,972
Cost at December 31, 2017	9,972
Accum. amortization at December 31, 2015	-
Amortization expense	-
Accum. amortization at December 31, 2016	-
Amortization expense	119
Accum. amortization at December 31, 2017	119
Net book value at	
December 31, 2016	-
December 31, 2017	9,853

The Company entered into a license agreement in 2017 with KU Center for Technology Commercialization, Inc., University of Kansas, Kansas Life Sciences Development Company, Inc., and UCL Business PLC. The price for the license to develop and commercialize products under all data generated in the course of the on-going Phase II/III clinical trial on arimoclomol for the treatment of sIBM depends on actual incurred costs by Kansas Life Sciences Development Company, Inc. and UCL Business PLC and is capped at TUSD 2,500 (DKK 15.8 million). The payment will be done by issuing bonus shares to the two parties measured at the end of each year. As at December 31, 2017 the costs incurred by these parties amounted to TDKK 902, and the bonus shares with a corresponding value were issued in January 2018. Reference is made to note 17 – subsequent events.

Under the terms of the license agreement, Orphazyme shall furthermore pay an aggregate royalty of 2% of net sales of products sold for the treatment of sIBM. Orphazyme expects to generate income from such products sold for the treatment of sIBM which will exceed any royalty payments due. Orphazyme has no liabilities prior to the occurrence of a potential future sale of products sold for the treatment of sIBM and accordingly, neither such liabilities have been recognized nor contingent considerations as part of the rights acquired.

Amortization expense is included within operating loss as follows:

	Twelve months ended December 31, 2017 TDKK	Twelve months ended December 31, 2016 TDKK
Research and development expenses	119	-
General and administrative expenses	-	-
Total depreciation expense	119	-

NOTE 10 – PROPERTY, PLANT, AND EQUIPMENT

	Fixtures and fittings, other plant, and equipment TDKK	Leasehold improvements TDKK	
Cost at December 31, 2015	2,349	540	2,889
Additions	218	20	238
Disposals	-	(38)	(38)
Cost at December 31, 2016	2,567	522	3,089
Additions	1,165	326	1,491
Disposals	-	(502)	(502)
Cost at December 31, 2017	3,732	346	4,078
Accum. depreciation at December 31, 2015	1,137	265	1,402
Depreciation expense	472	100	572
Impairment	-	134	134
Disposals	-	(6)	(6)
Accum. depreciation at December 31, 2016	1,609	493	2,102
Depreciation expense	574	53	627
Impairment	-	-	-
Disposals	-	(502)	(502)
Accum. depreciation at December 31, 2017	2,183	44	2,227
Net book value at			
December 31, 2016	958	29	987
December 31, 2017	1,549	302	1,851

At the end of 2016, the Company started the process of moving to new premises and this process was completed in 2017. In connection with this move, the book value of the leasehold improvement has been impaired and written down to its expected net realizable value as of December 31, 2016. The write down has been recognized under research and development expenses in the statement of profit or loss and other comprehensive income. There has been no impairment of property, plant, and equipment in the twelve months ended December 31, 2017.

Depreciation expense is included within operating loss as follows:

	Twelve months ended December 31, 2017 TDKK	Twelve months ended December 31, 2016 TDKK
Research and development expenses	620	472
General and administrative expenses	7	-
Total depreciation expense	**627**	472

NOTE 11 – EQUITY

The following table summarizes the Company's share activity:

	Class A ordinary shares	Class B preferred shares	Class C preferred shares	Common shares
December 31, 2015	125,000	2,050,208	1,170,547	-
Capital increase	-		14,786	-
December 31, 2016	125,000	2,050,208	1,185,333	-
Capital increase	-	-	1,741,669	
Conversion of shares prior to IPO	(125,000)	(2,050,208)	(2,927,002)	5,102,210
Non-cash share issue in connection with conversion	-	-	-	6,487,882
Warrant exercise	-	-	-	838,092
IPO	-	-	-	7,500,000
December 31, 2017	-	-	-	19,928,184

On November 2, 2017, in preparation of the IPO, the capital structure of Orphazyme was adjusted by way of a merger of the three previous share classes into one combined with an issue of bonus Shares in order to account for the now abolished preference shares (the "2017 Capital Structure Adjustment"). In connection with the 2017 Capital Structure Adjustment, the class B and C preference shares of the Company were converted into Shares on a 1:1 ratio. In order to account for the preferential rights attached to the preference shares, a directed issue of 6,487,882 bonus Shares using free reserves of the Company was carried out at par value in favor of the preference shareholders

On January 6, 2015, the company issued 1,704,554 indemnification warrants to the investors subscribing shares at this date. The warrants entitle the holders to subscribe C class shares at DKK 1 per share if certain liabilities of the company prove higher than warranted by the company in the Investment agreement. The warrants have been considered as an adjustment mechanism to the subscription price and not as separate derivative liabilities. The warrants are accounted for as equity instruments. The warrants may only be exercised in the event of the occurrence of one or more Warranty Claims as defined in the Investment Agreement dated December 30, 2014. Exercise of the warrants is further subject to the submission to the Company by the Owner of written notice of one or more Warranty Claims on or before January 6, 2016, or, in the event of a Warranty Claim which relates to tax issues, before January 6, 2018. The number of warrants exercisable is determined by the amount of the Claim. Neither as at December 31, 2017 nor after the balance sheet date such Warrant Claim related to tax issues have been raised and therefore the indemnification warrants lapsed without compensation in connection with Initial Public Offering.

On December 9, 2016, share capital was increased by 14,786 shares through issuance of shares for TDKK 1,331 in cash. In connection with the capital increase, the Company incurred expenses totaling TDKK 30.

In first quarter 2017, the Company has finished 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, the Company 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.

On November 16, 2017, the Company successfully completed the IPO by issuing 7,500,000 new ordinary shares for gross proceeds of TDKK 600,000. In connection with the capital increase, the Company incurred expenses totaling tDKK 56,061, of which TDKK 42,605 was recognized directly in equity.

In connection with the IPO on November 16, 2017 a total of 838,092 warrants were exercised for net proceeds of TDKK 1,161.

The Company has not declared or made any dividend payments for the last two financial years. Currently, the Company intends to use all available financial resources as well as revenue, if any, for purposes of the Company's current and future business. As of the date hereof, the Company does not expect to make dividend payments within the foreseeable future.

NOTE 11 – EQUITY (CONTINUED)

Subsequent to December 31, 2017, the company 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 (see note 10). Following this share capital increase, the total nominal share capital will be DKK 19,939,564, divided into 19,939,564 shares each with a nominal value of DKK 1.

NOTE 12 – INCOME TAX AND DEFERRED TAX

	Twelve months ended December 31, 2017 TDKK	Twelve months ended December 31, 2016 TDKK
Current tax on benefit on net loss	28,975	13,962
Adjustment to prior years	-	-
Tax credit research and development expenses	5,500	5,500
Change in unrecognized deferred tax before tax credit	(27,592)	(13,951)
Permanent deviations	(1,383)	(11)
Total income tax benefit for the period	**5,500**	**5,500**

Reconciliation of effective tax rate to Danish statutory tax rate

	December 31, 2017 TDKK	December 31, 2016 TDKK
Net loss before tax	(131,704)	(63,465)
Corporate income tax rate in Denmark	22%	22%
Computed income tax benefit	**28,975**	**13,962**
Tax effect of:		
Other non-deductible expenses, including IPO-related costs and share-based compensation	(1,383)	(11)
Deferred tax asset not recognized	(22,092)	(8,451)
Total income tax benefit for the period	**5,500**	**5,500**

Deferred tax in the statement of financial position

	December 31, 2017 TDKK	December 31, 2016 TDKK
Tax deductible losses	51,644	29,716
Other temporary differences	277	(113)
	51,921	29,603
Deferred tax asset not recognized	(51,921)	(29,603)
Carrying amount included on statement of financial position	**-**	**-**

The Company had net tax loss carry-forwards in Denmark for income tax purposes of MDKK 235 and MDKK 135 as of December 31, 2017 and 2016.

Income tax benefit for the year includes a tax credit for research and development at the applicable tax rate under the Danish Corporate Income Tax Act.

The tax loss carry forwards have no expiry date. The Company's ability to use tax loss carry forwards in any one year is limited to 100% of the first MDKK 7.5 of taxable income plus 60% of taxable income above MDKK 7.5.

The Company recognizes deferred tax assets, including the tax base of tax loss carry forwards, if management assesses that these tax assets can be offset against positive taxable income within a foreseeable future. Significant management judgment is required to determine the amount of deferred tax assets that can be recognized, based upon the likely timing and the level of future taxable profits together with future tax planning strategies. This judgment is made periodically after considering current facts and circumstances, budgets and business plans as well as the risks and uncertainty associated with the Company's ability successfully commercialize and defend its intellectual property.

Significant judgment
The development of therapeutic products within the biopharmaceutical industry is subject to significant risks and uncertainties and there is no assurance a therapeutic product will be successfully developed. As the result of this uncertainty and since the Company has reported significant losses since inception, has no commercial products or revenues and does not expect to generate revenues or profits for the foreseeable future, management has concluded that deferred tax assets should not be recognized as of December 31, 2017 or at any other prior date. The tax assets are currently not deemed to meet the criteria for recognition as management is not able to provide convincing positive evidence that taxable profits will be available in the future to utilize the benefit from the tax assets.
As of December 31, 2017 there are no tax audits in process nor has management been notified of any pending tax audit. As of December 31, 2017, the tax years that remain open for audit by the Danish tax authorities include 2013 through 2016.

NOTE 13 – 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 twelve months ended December 31, 2017 and 2016:

	Twelve months ended December 31, 2017 TDKK	Twelve months ended December 31, 2016 TDKK
Loss for the period	(126,204)	(57,935)
Weighted-average shares outstanding	12,064,113	9,834,500
Loss per share	**(10.46)**	**(5.89)**

Basic loss per share amounts are calculated by dividing the net earnings/(loss) for the period by the weighted average number of shares outstanding during each period.

On November 2, 2017, in preparation of the IPO, the capital structure of Orphazyme was adjusted by way of a merger of the three previous share classes into one combined with an issue of bonus Shares in order to account for the now abolished preference shares. The total number of bonus shares issued was 6,487,882, which has in accordance with the provisions of IAS 33 Earnings per Share been included in the number of weighted average shares outstanding for both the twelve months ended December 31, 2017 and 2016.

Due to the fact that all warrants not exercised in connection with the IPO has been terminated, basic and diluted loss per share are the same for each period presented.

Subsequent to December 31, 2017, the company has issued 11,380 shares, which will have an impact on the calculation of earnings per share from 2018 and forward.

NOTE 14 – CAPITAL MANAGEMENT

For the purpose of the Company's capital management, capital includes issued capital, share premium and all other equity reserves attributable to the equity holders of the Company. The primary objective of the Company's capital management is to maximize shareholder value while limiting the financial risk. The Board of Directors' policy is to maintain a strong capital base so as to maintain investor, creditor and market confidence, and a continuous advancement of the Company's intellectual property, product pipeline, and business. Cash, cash equivalents, and financial assets are monitored on a regular basis by management and the Board of Directors in assessing current and long term capital needs. As of December 31, 2017, the Company held cash and cash equivalents totaling MDKK 631.7 that will be sufficient to provide adequate funding to allow the Company to meet its planned operating activities, including increased levels of research and development activities, in the normal course of business for the next twelve months. The Company currently has no significant planned capital expenditures.

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

Foreign Currency
The Company maintains operations in Denmark and uses the DKK as its functional currency. The Company conducts cross border transactions where the functional currency is not always used. Accordingly, future changes in the exchange rates of the DKK, the EUR, the USD and/or the GBP will expose the Company to currency gains or losses that will impact the reported amounts of assets, liabilities, income and expenses and the impact could be material. For the year ended December 31, 2017 and 2016 the impact on the Company's statement of loss for possible changes in the EUR, USD, and GBP exchange rates against the Company's functional currency of DKK would be as follows:

Currency	Possible change	Twelve months ended December 31, 2017 TDKK	Twelve months ended December 31, 2016 TDKK
EUR	+/2%	(215) / +215	+177 / (177)
USD	+/10%	(936) / +936	(21) / +21
GBP	+/10%	(358) / +358	(327) / +327

Interest Rate Risk
The Company has no interest bearing debt. Due to the current interest level in Denmark the Company incurs negative interest on bank deposits. For the year ended December 31, 2017 and 2016 the impact on the Company's statement of loss for possible changes in the interest rate on DKK and EUR deposits would be as follows:

Cash deposits	Possible change	Twelve months ended December 31, 2017 TDKK	Twelve months ended December 31, 2016 TDKK
DKK	+/1%	(6,313) / +6,313	+53 / (53)
EUR	+/1%	(1) / +1	(89) / +89
GBP	+/1%	0 / 0	(1) / +1
USD	+/1%	(3) / +3	0 / 0

Credit Risk
The Company's credit risk is associated with cash held in banks. The Company does not trade financial assets for speculative purposes and invests with the objective of preserving capital.

The Company's cash and cash equivalents are held primarily at two banks in Denmark with Moody's long-term credit ratings exceeding of A1.

NOTE 15 – CONTRACTUAL OBLIGATIONS AND CONTINGENCIES

Contractual obligations
The Company has the following non-cancelable contractual obligations related to its lease and other rent liabilities:

December 31, 2017	Less than 1 year	1-3 years	3-5 years	More than 5 years	Total
Operating lease obligations	902	-	-	-	902
R&D contractual obligations	47,306	293	-	-	47,599
Total	**48,208**	**293**	**-**	**-**	**48,501**

December 31, 2016	Less than 1 year	1-3 years	3-5 years	More than 5 years	Total
Operating lease obligations	418	-	-	-	418
R&D contractual obligations	48,500	43,891	-	-	92,391
Total	**48,918**	**43,891**	**-**	**-**	**92,809**

Total expense under non-cancelable contractual obligations was TDKK 1,671 and TDKK 848 for the twelve months December 31, 2017 and 2016.

The Company furthermore has contracts with CRO's, where the CRO will carry out clinical trials for the Company. There is a contract in place between the parties but the amounts are not fixed as it depends on reaching milestones, number of enrolled patients, etc.

Contingencies
Contingencies are assets and liabilities that arise from past events but whose existence will only be confirmed by the occurrence or non occurrence of future events that in some situations are beyond the Company's control.

Under the terms of the license agreement described in note 10, Orphazyme shall pay an aggregate royalty of 2% of net sales of products sold for the treatment of sIBM. Orphazyme is required to use commercially diligent efforts to develop and commercialize such products. Orphazyme expects to generate income from such products sold for the treatment of sIBM which will exceed any royalty payments due. Orphazyme has no liabilities prior to the occurrence of a potential future sale of products sold for the treatment of sIBM and accordingly, neither such liabilities have been recognized nor contingent considerations as part of the rights acquired.

As of December 31, 2017 there are no contingent assets or liabilities besides the above and as of December 31, 2016 there are no contingent assets or liabilities.

NOTE 16 – RELATED PARTY DISCLOSURES

The Company is not ultimately controlled by any of the investors. See note 5 for additional related party trans-actions, related to the remuneration paid to key management. In addition to Novo A/S, LSP V Coöperatieve U.A., Sunstone Life Science Ventures Fund II K/S, and Coopera-tive Aescap Venture I U.A. all owns more than 5%.

There have been no transactions between related parties in the 12 months ended December 31, 2017 and the twelve months ended December 31, 2016 besides capital increases as described in note 11.

Terms and conditions of transactions with related parties
Amounts due to related parties are uncollateralized and interest free. There have been no guarantees provided or received for any related party receivables or payables. For the twelve months ended December 31, 2017 and the twelve months ended December 31, 2016 the Company has not recorded any impairment of receivables relating to amounts owed by related parties. There are no related party receivables at any of the balance sheet dates.

Transactions with key management
The Company has not granted any loans, guarantees, or other commitments to or on behalf of any of the members of the Board of Directors or Executive Management.

Other than the remuneration described in note 5, no other significant transactions have taken place with Executive Management personnel during the period presented herein.

	Number of shares December 31, 2017	Number of shares December 31, 2016	Number of warrants December 31, 2017	Number of warrants December 31, 2016	LTIP 2017	LTIP 2016
Anders Hinsby	204,596	18,750	-	112,701	5,000	-
Anders Vadsholt	127,806	-	-	-	4,000	-

Compensation paid to members of the Board of Directors
Compensation paid to members of the Board of Directors are classified as administrative expense within the statement of loss. The following table lists compensation paid to members of the Board of Directors:

	Twelve months ended December 31, 2017 TDKK	Twelve months ended December 31, 2016 TDKK
Director fees	1,211	524
Warrants	-	-
Other fees	-	-
Total compensation paid to the Board of Directors	1,211	524

NOTE 16 – RELATED PARTY DISCLOSURES (CONTINUED)

	Number of shares December 31, 2017	Number of shares December 31, 2016	Number of warrants December 31, 2017	Number of warrants December 31, 2016
Georges Gemayel	87,758	-	-	51,842
Bo Jesper Hansen	79,945	-	-	22,390
Martijn Kleijwegt	-	-	-	-
Martin Bonde	46,009	-	-	32,609
Martin Rahbek Kornum	41,476	18,750	-	17,281
Nanna Lüneborg	-	-	-	-
Patrick J.H. Krol	-	-	-	-
Rémi Droller	-	-	-	-
Sten Verland	-	-	-	-
Anders Hedegaard	6,250	-	-	-
Catherine Moukheibir	7,980	-	-	-

Transactions with shareholders and affiliates
There have been no transactions with shareholders or affiliates of shareholders during the twelve months ending December 31, 2017 or the twelve months ended December 31, 2016, except for the capital increases disclosed in note 11.

NOTE 17 – SUBSEQUENT EVENTS

Management has evaluated its financial statements for potential subsequent events occurring after the balance sheet date of December 31, 2017 but prior to the date that these financial statements were issued.

On January 19, 2018 the Company announced that arimoclomol has been granted rare pediatric disease designation by US Food and Drug Administration (FDA) for the treatment of Niemann-Pick Disease Type C (NPC).

On January 29, 2018, the company 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 (see note 9). Following this share capital increase, the total nominal share capital will be DKK 19,939,564, divided into 19,939,564 shares each with a nominal value of DKK 1.

On February 5, 2018, Orphazyme hired Paul Merrigan as Chief Commercial Officer in line with the Company's commercial strategy as set out in the Prospectus relating to the IPO in November 2017. In this connection, a 100%-controlled subsidiary has been established in Boston, USA.

Other than the event disclosed above, there were no other events that were required to be reported or disclosed that are not already included within these financial statements.

Fees for other assistance in 2017 are primarily for services provided in relation to the process leading up to the IPO on Nasdaq Copenhagen.

NOTE 18 – FEES PAID TO AUDITORS APPOINTED AT THE ANNUAL GENERAL MEETING

	Twelve months ended December 31, 2017	Twelve months ended December 31, 2016
	TDKK	TDKK
Fee for statutory audit	250	200
Fee for statutory audit, previous years	142	-
Assurance engagements	30	-
Tax consultancy	-	-
Other assistance	2,735	40
Total fee paid to auditor	3,157	240

STATEMENTS AND SIGNATURES

STATEMENT BY THE BOARD OF DIRECTORS AND EXECUTIVE MANAGEMENT

The Board of Directors and Executive Management have today considered and approved the Annual Report of Orphazyme A/S for the financial year January 1-December 31, 2017.

The financial statements have been prepared in accordance with International Financial Reporting Standards (IFRS) as adopted by the EU and additional requirements in the Danish Financial Statements Act.

In our opinion, the financial statements provide a fair presentation of the Company's assets, liabilities, and financial position at December 31, 2017 and of the results of the company's operations and cash flows for the financial year January 1–December 31, 2017.

In our opinion, the Management Review provides a fair presentation of the development in the Company's operations and financial circumstances, of the results for the year, and of the overall financial position of the Company as well as a description of the most significant risks and elements of uncertainty facing the Company.

We recommend that the Annual Report be adopted at the Annual General Meeting on April 12, 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, PhD
Chief Executive Officer, Co-Founder

Anders Vadsholt, MBA
Chief Financial Officer

INDEPENDENT AUDITOR'S REPORT

TO THE SHAREHOLDERS OF ORPHAZYME A/S

OPINION

We have audited the financial statements of Orphazyme A/S for the financial year January 1–December 31, 2017, which comprise income statement, statement of comprehensive income, balance sheet, statement of changes in equity, cash flow statement and notes, including accounting policies. The financial statements are prepared in accordance with International Financial Reporting Standards as adopted by the EU and additional requirements of the Danish Financial Statements Act.

In our opinion, the financial statements give a true and fair view of the financial position of the Company at December 31, 2017 and of the results of the Company's operations and cash flows for the financial year January 1–December 31, 2017 in accordance with International Financial Reporting Standards as adopted by the EU and additional requirements of the Danish Financial Statements Act.

Our opinion is consistent with our long-form audit report to the Audit Committee and the Board of Directors.

BASIS FOR OPINION

We conducted our audit in accordance with International Standards on Auditing (ISAs) and additional requirements applicable in Denmark. Our responsibilities under those standards and requirements are further described in the "Auditor's responsibilities for the audit financial statements" section of our report. We believe that the audit evidence we have obtained is sufficient and appropriate to provide a basis for our opinion.

Independence

We are independent of the Company in accordance with the International Ethics Standards Board for Accountants' Code of Ethics for Professional Accountants (IESBA Code) and additional requirements applicable in Denmark, and we have fulfilled our other ethical responsibilities in accordance with these rules and requirements.

To the best of our knowledge, we have not provided any prohibited non-audit services as described in article 5(1) of Regulation (EU) no. 537/2014.

Appointment of auditor

On November 16, 2017, Orphazyme A/S completed its Initial Public Offering and was admitted to trading and official listing on Nasdaq Copenhagen A/S. We were initially appointed as auditor of Orphazyme A/S on December 4, 2015 for the financial year 2015. We have been reappointed annually by resolution of the general meeting for a total consecutive period of 3 years up until and including the financial year 2017.

KEY AUDIT MATTERS

Key audit matters are those matters that, in our professional judgement, were of most significance in our audit of the financial statements for the financial year 2017. These matters were addressed during our audit of the financial statements as a whole and in forming our opinion thereon. We do not provide a separate opinion on these matters. For each matter below, our description of how our audit addressed the matter is provided in that context.

We have fulfilled our responsibilities described in the "Auditor's responsibilities for the audit of the financial statements" section, including in relation to the key audit matters below. Accordingly, our audit included the design and performance of procedures to respond to our assessment of the risks of material misstatement of the financial statements. The results of our audit procedures, including the procedures performed to address the matters below, provide the basis for our audit opinion on the financial statements.

KEY AUDIT MATTERS

Complex contractual arrangements

As a company in the Biopharmaceutical industry, Orphazyme has entered into several complex contractual arrangements, such as in-licensing agreements with universities related to intangible rights and knowhow, clinical research contracts with Clinical Research Organizations (CROs) and share-based compensation arrangements (warrants and long-term-incentive programmes (LTIPs)) with employees etc.

Some in-licensing arrangements include complex mechanisms for sharing profit in the event of future commercialization of biopharmaceutical products. Furthermore, the CRO contracts contains complex mechanisms of considerations for services received from the CROs. The accounting for share-based compensation requires application of complex valuation models, such as the Black-Scholes model and the Monte-Carlo simulation model.

Due to the complexity of these arrangements, management has to exercise significant judgment in interpreting such arrangements. Furthermore, recognition of the transactions requires management to make significant estimates.

We focused on these matters because the arrangements and the related accounting treatment of in-licensing of intangible rights, accruals accounting for CRO costs and recognition of share-based compensation is complex and because the accounting requires significant judgement and estimation by Management.

Refer to note 2, 6, and 9 to the financial statements.

HOW OUR AUDIT ADDRESSED THE KEY AUDIT MATTER

We have evaluated relevant processes including Management's review controls to ensure that in-licensing agreements, CRO contracts and share-based compensation programmes are recognized and measured in accordance with applicable accounting standards on an ongoing basis.

We obtained Management's calculations of fair value of acquired rights, CRO accruals and share-based compensation models, and corroborated inputs and key assumptions to underlying terms and conditions in the agreements and relevant internal and external sources – and assessed the accuracy of the intangible assets recognised, accruals for CRO services received and share-based compensation incurred in the periods. Furthermore, estimation models applied by management, including CRO accrual models, Black-Scholes models and Monte-Carlo simulation models have been tested for mathematical accuracy.

We assessed whether the disclosures in relation to intangible rights, CRO accruals and share-based compensation were appropriate and met the requirements of accounting standards.

STATEMENT ON THE MANAGEMENT'S REVIEW

Management is responsible for the Management's review.

Our opinion on the financial statements does not cover the Management's review, and we do not express any form of assurance conclusion thereon.

In connection with our audit of the financial statements, our responsibility is to read the Management's review and, in doing so, consider whether the Management's review is materially inconsistent with the financial statements or our knowledge obtained during the audit, or otherwise appears to be materially misstated.

Moreover, it is our responsibility to consider whether the Management's review provides the information required under the Danish Financial Statements Act.

Based on the work we have performed, we conclude that the Management's review is in accordance with the financial statements and has been prepared in accordance with the requirements of the Danish Financial Statements Act. We did not identify any material misstatement of the Management's review.

MANAGEMENT'S RESPONSIBILITIES FOR THE FINANCIAL STATEMENTS

Management is responsible for the preparation of financial statements that give a true and fair view in accordance with International Financial Reporting Standards as adopted by the EU and additional requirements of the Danish Financial Statements Act and for such internal control as Management determines is necessary to enable the preparation of financial statements that are free from material misstatement, whether due to fraud or error.

In preparing the financial statements, Management is responsible for assessing the Company's ability to continue as a going concern, disclosing, as applicable, matters related to going concern and using the going concern basis of accounting in preparing the financial statements unless Management either intends to liquidate the Company or to cease operations, or has no realistic alternative but to do so.

AUDITOR'S RESPONSIBILITIES FOR THE AUDIT OF THE FINANCIAL STATEMENTS

Our objectives are to obtain reasonable assurance as to whether the financial statements as a whole are free from material misstatement, whether due to fraud or error, and to issue an auditor's report that includes our opinion. Reasonable assurance is a high level of assurance, but is not a guarantee that an audit conducted in accordance with ISAs and additional requirements applicable in Denmark will always detect a material misstatement when it exists. Misstatements can arise from fraud or error and are considered material if, individually or in the aggregate, they could reasonably be expected to influence the economic decisions of users taken on the basis of the financial statements.

As part of an audit conducted in accordance with ISAs and additional requirements applicable in Denmark, we exercise professional judgement and maintain professional scepticism throughout the audit. We also:

- Identify and assess the risks of material misstatement of the financial statements, whether due to fraud or error, design and perform audit procedures responsive to those risks and obtain audit evidence that is sufficient and appropriate to provide a basis for our opinion. The risk of not detecting a material misstatement resulting from fraud is higher than for one resulting from error, as fraud may involve collusion, forgery, intentional omissions, misrepresentations or the override of internal control.

- Obtain an understanding of internal control relevant to the audit in order to design audit procedures that are appropriate in the circumstances, but not for the purpose of expressing an opinion on the effectiveness of the Company's internal control.

- Evaluate the appropriateness of accounting policies used and the reasonableness of accounting estimates and related disclosures made by Management.

- Conclude on the appropriateness of Management's use of the going concern basis of accounting in preparing the financial statements and, based on the audit evidence obtained, whether a material uncertainty exists related to events or conditions that may cast significant doubt on the Company's ability to continue as a going concern. If we conclude that a material uncertainty exists, we are required to draw attention in our auditor's report to the related disclosures in the financial statements or, if such disclosures are inadequate, to modify our opinion. Our conclusions are based on the audit evidence obtained up to the date of our auditor's report. However, future events or conditions may cause the Company to cease to continue as a going concern.

- Evaluate the overall presentation, structure and contents of the financial statements, including the note disclosures, and whether the financial statements represent the underlying transactions and events in a manner that gives a true and fair view.

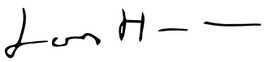
We communicate with those charged with governance regarding, among other matters, the planned scope and timing of the audit and significant audit findings, including any significant deficiencies in internal control that we identify during our audit.

We also provide those charged with governance with a statement that we have complied with relevant ethical requirements regarding independence, and to communicate with them all relationships and other matters that may reasonably be thought to bear on our independence, and where applicable, related safeguards.

From the matters communicated with those charged with governance, we determine those matters that were of most significance in the audit of the financial statements of the current period and are therefore the key audit matters. We describe these matters in our auditor's report unless law or regulation precludes public disclosure about the matter or when, in extremely rare circumstances, we determine that a matter should not be communicated in our report because the adverse consequences of doing so would reasonably be expected to outweigh the public interest benefits of such communication.

Copenhagen, March 15, 2018
ERNST & YOUNG
Godkendt Revisionspartnerselskab
CVR no. 30 70 02 28

Christian Schwenn Johansen
State Authorized Public Accountant
mne33234



Lars Hansen
State Authorized Public Accountant
mne24828

Orphazyme A/S
Ole Maaløes Vej 3
DK-2200 Copenhagen N
Approval at Annual General Meeting (AGM): April 12, 2018
Chairman of AGM: Rikke Schiøtt Petersen, Gorrissen Federspiel Advokatpartnerselskab