ORPHAZYME

ANNUAL REPORT 2018

Innovative therapies for protein-misfolding diseases

Lleyton Coombes (age 12, NPC) and his mother Joanne Coombes at NPUK event in 2016.
The Coombes family are well-known in the NPUK community due to their inspirational awareness and fund-raising efforts with their group 'Pride in Lleyton Appeal #makingmemories'.

Orphazyme A/S
CVR no.: 32266355

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AT A GLANCE

OUR VISION

To impact the lives of patients with orphan diseases

Copenhagen, DK
and Boston, USA

65 employees
as of today

ORPHAZYME
is a late-stage biopharmaceutical company listed on Nasdaq Copenhagen, focused on developing novel treatment options for rare diseases within the protein-misfolding space.

We are 62 employees in Copenhagen, Denmark, and 3 in Boston, MA, USA.

OUR LEAD PRODUCT
candidate arimoclomol, is in development for Amyotrophic Lateral Sclerosis (ALS), sporadic Inclusion Body Myositis (sIBM), Niemann-Pick disease Type C (NPC), and Gaucher disease.

For more information about Orphazyme, go to orphazyme.com

ARIMOCLOMOL
is orally available to patients

LATE-STAGE PIPELINE
with 3 Phase II/III trials

ALS & sIBM

NPC & GAUCHER DISEASE

2018 ACHIEVEMENTS

JANUARY
Receives Rare Pediatric Disease Designation for arimoclomol for Niemann-Pick disease Type C (NPC)

APRIL
Establishment of US subsidiary in Massachusetts

JUNE
First patient enrolled in Gaucher disease Phase II trial

AUGUST
First patient enrolled in ALS Phase III trial

SEPTEMBER
Reports encouraging arimoclomol clinical trial top-line data in Niemann-Pick disease Type C (NPC)

Priority	✓ Targeted milestone	– Complete Phase II/III trial enrollment by year-end
ALS	✓ Initiate Phase III trial in Q3	
sIBM	✓ Enroll patients in both US and Europe	
NPC	✓ Phase II/III trial top-line results in Q3	
Gaucher	✓ Initiate Phase II trial in Q2	– Complete trial enrollment before year-end
NME program	✓ Preclinical trials with new molecular entities (NME)	

LETTER FROM THE CHAIRMAN

PREPARING FOR LAUNCH

Having been a member of the Board of Directors since 2012, I have seen Orphazyme blossom into what it is today. From very humble beginnings, based on a solid scientific foundation, Orphazyme is today a successful, publicly-traded biopharmaceutical company with a potential to bring transformational therapies to patients in need in several diseases.

Looking back at 2018, Orphazyme has delivered on its promises to its shareholders and has kept the momentum of the development pipeline. The Company continues to simultaneously conduct clinical studies in four indications with high unmet medical needs. In addition, Orphazyme continues to build on its long-term potential through its very innovative preclinical pipeline.

With the positive results from the Phase II/III trial in Niemann-Pick disease Type C (NPC), Orphazyme is hoping to bring the much-needed therapy to the NPC community as soon as possible. In order to secure the patients' access to arimoclomol, Orphazyme is planning to launch a commercial organization as soon as the regulatory authorities in the US and Europe grant their approval to commercialize arimoclomol for the treatment of NPC. This organization will moreover create the foundation to build the commercial capabilities to launch the future indications of arimoclomol.

On behalf of the Board of Directors, I would like to thank the entire Management Team and every single employee in Orphazyme for their hard work and dedication. I am confident that 2019 will be as fruitful as 2018 and look forward to seeing the year unfold.

Georges Gemayel
Chairman of the
Board of Directors

LETTER FROM THE CEO

REACHING MILESTONES

In 2018, we successfully completed our Phase II/III clinical trial in Niemann-Pick disease Type C (NPC), and we are now closer than ever to our goal of providing NPC patients with a much needed new treatment option. We enter our 10th anniversary year, 2019, with optimism and resolve, and look forward to entering into dialogue with the regulatory authorities in both the US and Europe on how to best and most expediently bring arimoclomol to NPC patients.

2018 was a very important year for Orphazyme, with the initiation of two clinical trials and the successful completion of our Phase II/III clinical trial in Niemann-Pick disease Type C (NPC). For almost 10 years, we have been steadfast in our mission to develop a treatment option for patients suffering from this severely debilitating disease, and we are now closer than ever to achieving this goal. We are looking forward to 2019, during which we will start the preparations to bring arimoclomol to approval for NPC.

Orphazyme's listing on Nasdaq Copenhagen at the end of 2017 made it possible for us to pursue our ambition of becoming a leading biopharmaceutical company with a dedication to rare diseases. The proceeds from the Initial Public Offering allowed us to conduct clinical trials in three rare, debilitating diseases in addition to completing the Phase II/III trial in NPC. During 2018, we have added experienced and talented professionals to our organization, enabling us to drive our projects through clinical development towards regulatory approval and launch. Entering 2019, we are well-equipped to complete our journey and bring new medicine to NPC patients in great need of effective therapies.

NPC

NPC is a rare disease that affects both the physical and mental health of patients, who may become wheel-chair bound and experience severe deterioration of cognition, memory, and speech. The disease is fatal, with most patients not surviving into adulthood. Since the Company's inception nearly a decade ago, we have been working towards providing a treatment option for patients suffering from this terrible disease in close collaboration with patient organizations, expert scientists, and clinicians. We were therefore very pleased to report that arimoclomol put a break on disease progression. After 12 months, patients receiving arimoclomol had a 74% reduction in disease progression compared to placebo control (p-value 0.0506) measured by the 5-domain NPC Clinical Severity Scale. Moreover, the full data set supported a positive effect of arimoclomol by showing statistically significant benefit in large predefined subgroups as well as a treatment effect on biomarkers. In 2019, we look forward to entering into dialogue with the regulatory authorities in both the US and Europe on how to best and most expediently bring arimoclomol to NPC patients.

2018 MILESTONES

In addition to presenting results from our NPC trial, we were able to deliver on several other important milestones in 2018, including receipt of a Rare Pediatric Disease Designation from the US Food and Drug Administration (FDA) for the NPC/arimoclomol program, the initiation of two clinical trials – a Phase III trial in Amyotrophic Lateral Sclerosis (ALS) and a Phase II trial in Gaucher disease –, and the roll out of our clinical trial in sporadic Inclusion Body Myositis (sIBM) from one to 12 clinical trial sites.

THE YEAR AHEAD

Looking ahead, we expect a busy 2019, where we will start the preparations to bring arimoclomol to approval for NPC. Ten years in the making, this milestone is closer than ever: To provide NPC patients with a much needed treatment option. We therefore enter our 10th anniversary year with optimism and resolve about being able to deliver on our commitment to patients and caregivers, and enthusiastic about the future of our company.

In this industry there are no quiet days, and our team of talented and committed professionals continuously work hard to move us forward. I am grateful for their contributions as well as for the support of our shareholders. Thank you for taking this journey with us.

Anders Hinsby
Chief Executive Officer

KEY FIGURES

TDKK	2018	2017	2016	2015[1]	2014/15
Statement of profit and loss and other comprehensive income					
Research and development costs	(196,525)	(99,048)	(55,817)	(25,478)	(31,604)
General and administrative expenses	(35,127)	(31,994)	(7,703)	(4,044)	(5,494)
Operating loss	**(231,652)**	**(131,042)**	**(63,520)**	**(29,522)**	**(37,098)**
Net financial items	(3,448)	(662)	85	40	(1,369)
Loss before tax	**(235,100)**	**(131,704)**	**(63,435)**	**(29,482)**	**(38,467)**
Income tax benefit	5,500	5,500	5,500	2,750	5,875
Net loss for the period	**(229,600)**	**(126,204)**	**(57,935)**	**(26,732)**	**(32,592)**
Total comprehensive loss	**(229,558)**	**(126,204)**	**(57,935)**	**(26,732)**	**(32,592)**
Loss per share, basic (DKK)	(11.50)	(10.43)	(5.89)	(2.75)	(3.60)
Statement of financial position					
Licenses	10,744	9,853	-	-	-
Property, plant, and equipment	1,940	1,851	1,225	1,512	2,306
Total non-current assets	17,965	14,864	4,047	4,448	7,807
Cash	394,706	631,735	14,349	68,014	78,161
Other current assets	28,678	16,218	13,545	12,490	9,379
Total assets	441,349	662,817	31,941	84,952	95,347
Share capital	19,939	19,928	3,361	3,346	3,218
Total equity	388,249	615,702	17,509	74,143	89,380
Total current liabilities	52,995	47,115	14,432	10,809	5,967

[1] 2015 covers the period 1 July 2015 to 31 December 2015 as the Company changed its financial year to correspond to the calendar year

KEY FIGURES (CONTINUED)

TDKK	2018	2017	2016[1]	2015	2014/15[2]
Cash flow statement					
Cash flow from operating activities	(234,764)	(95,426)	(54,724)	(21,372)	(36,438)
Cash flow from investing activities	(2,346)	(1,491)	(238)	(25)	(558)
Cash flow from financing activities	-	714,303	1,300	11,250	89,425
Other					
Share price (DKK)[3]	43.35	76.00	-	-	-
Total outstanding shares	19,939,564	19,928,184	3,360,541	3,345,755	3,218,031
Market capitalization (MDKK)[4]	864.4	1,514.5	-	-	-
Equity ratio[5]	88.0%	92.9%	54.8%	87.3%	93.7%
Equity per share (DKK)[6]	19.47	30.90	5.21	22.16	27.77
Average number of employees	46	26	17	13	10
Number of employees at the end of the year	57	34	21	15	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

2019 OUTLOOK

MDKK	2019 guidance	2018 actual result	2018 guidance
Operating loss	(315) - (345)	(232)	(245) - (275)
Cash position at year-end	>50	395	>350

OPERATING RESULT

The operating loss of DKK 231.6 million was below the expected operating loss range of DKK 245–275 million. The difference to the outlook for 2018 was mostly due to slower patient enrollment in our clinical trials. We anticipate that our 2019 operating loss will be in the range of DKK 315–345 million. This range reflects the inherent uncertainty related to the timing of patient enrollment and related operational uncertainties. The cost increase is driven by our preparation to submit a New Drug Application/Marketing Authorization Application for arimoclomol for NPC, advancement of arimoclomol for sIBM and ALS, production of arimoclomol, and an increase in employees to support our expanding clinical and operational activities.

CASH POSITION

At year-end 2019, we anticipate a cash position of DKK >50 million compared to DKK 394.7 million as of December 31, 2018.

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, 2019, Orphazyme expects to incur substantial costs associated with clinical trials and filing activities. The objective of the development programs is to develop a pharmaceutical drug for the treatment of the following diseases: ALS, sIBM, NPC, and Gaucher disease.

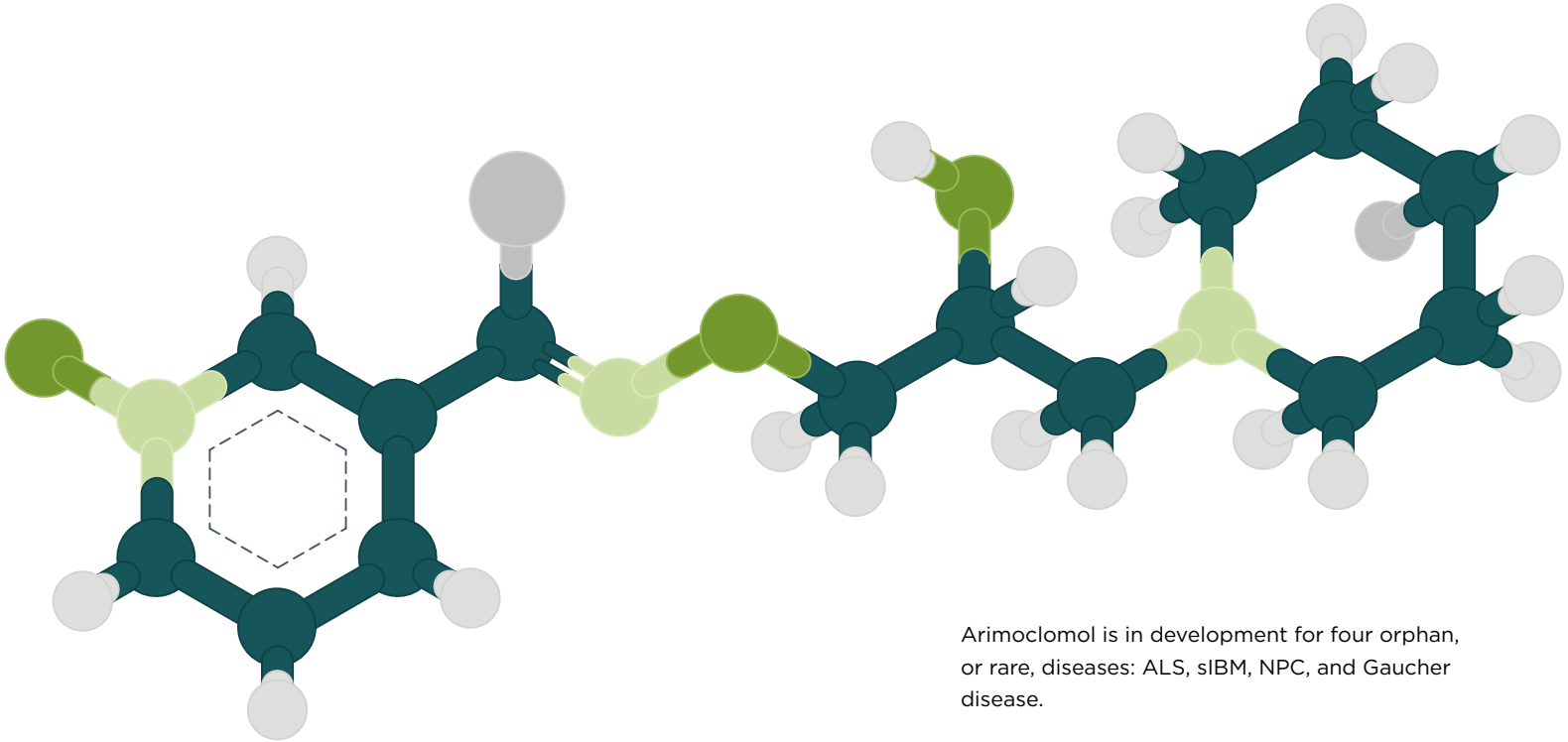
The forecasting of costs associated with clinical trials relating to activities performed by Clinical Research Organizations (CROs) and other external vendors requires management to exercise significant estimates with regard to the timing and accounting for these costs. The diverse nature of the services being provided by CROs and other arrangements, the different compensation arrangements that exist 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. The filing activities for arimoclomol for NPC will be influenced by the regulatory feedback from the US Food & Drug Administration (FDA) and the European Medicines Agency (EMA). The outlook for the financial year ending December 31, 2019 takes into consideration activities planned for 2019, including the trial designs for the respective indications and the filing activities as described on page 13, Product Pipeline.

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 pages 23-24 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.

2019 OBJECTIVES

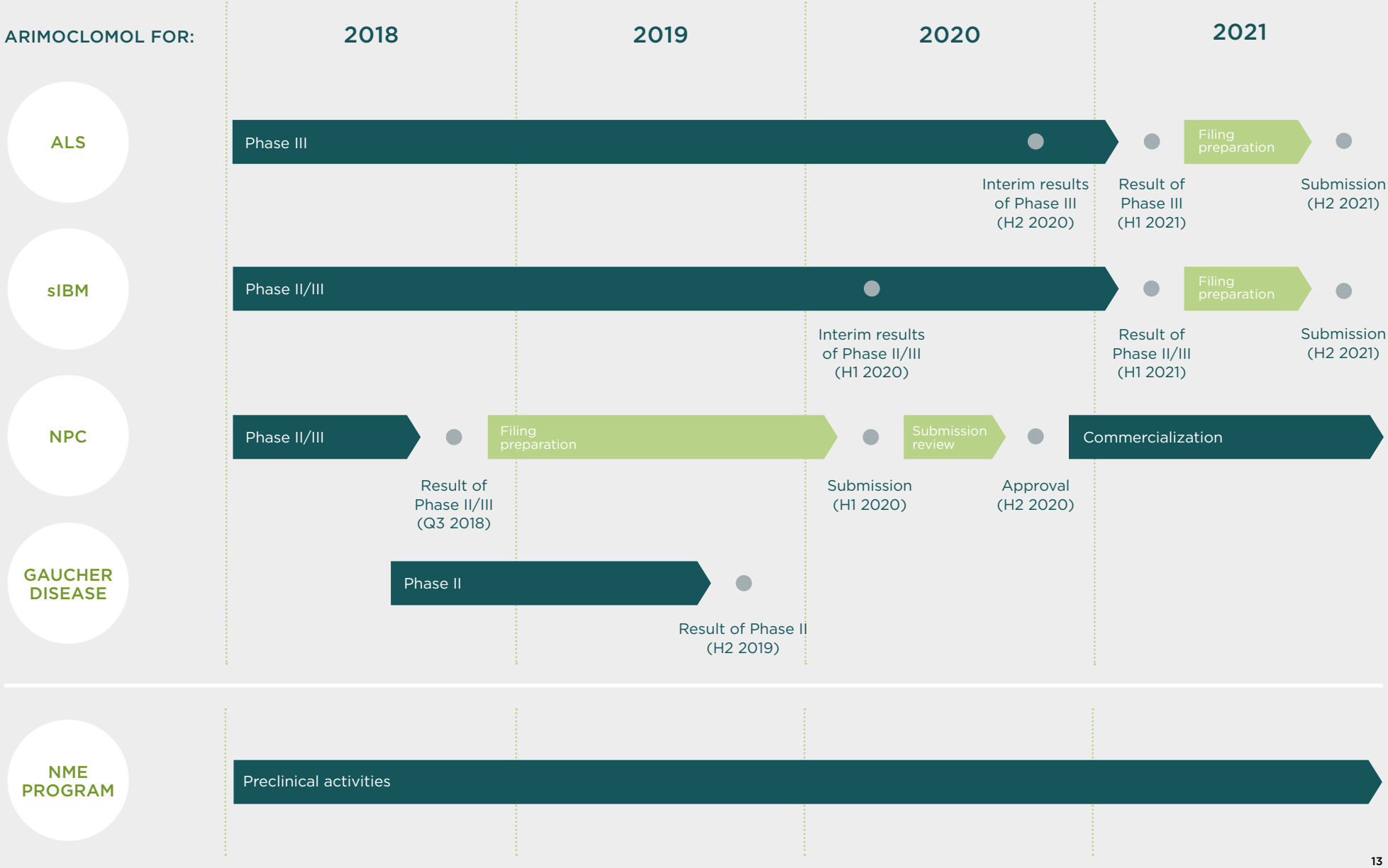
Priority	Targeted milestone
ALS	• Complete enrollment in H2
sIBM	• Complete enrollment in H1
NPC	• Regulatory feedback mid-2019
Gaucher disease	• Phase II results in H2
New molecular entities (NME) program	• Preclinical studies with NMEs in protein-misfolding diseases



Arimoclomol is in development for four orphan, or rare, diseases: ALS, sIBM, NPC, and Gaucher disease.

Lead compound arimoclomol is in development for rare diseases with high unmet needs

PRODUCT PIPELINE



Orphazyme's current clinical programs investigate arimoclomol as a treatment for four protein-misfolding indications: The protein-aggregation diseases Amyotrophic Lateral Sclerosis (ALS) and sporadic Inclusion Body Myositis (sIBM), 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.

ARIMOCLOMOL & HEAT-SHOCK PROTEINS

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

PROTEIN-AGGREGATION DISEASES

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

LYSOSOMAL STORAGE DISEASES

Lysosomal storage diseases (LSDs), such as NPC and Gaucher disease, 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 located in the body's cells and are used to break down and recycle fats, proteins, and other large molecules. Loss of lysosomal enzyme activity due to misfolding and dysfunction prohibits the lysosomes from performing their normal function.

SPHINGOLIPIDOSES

Within the category of LSDs, is a group of lipid (fat) storage diseases called sphingolipidoses. These diseases are characterized by a disturbance of the metabolism of the sphingolipids inside the cells, resulting in build-up that disturbs cell function or leads to cell death. Sphingolipids are produced in many tissues, but especially nerve tissues, and sphingolipid diseases therefore very often affect the nervous system. Diseases in this group include NPC, Gaucher disease, Fabry disease, Krabbe disease, Tay–Sachs disease, and metachromatic leukodystrophy.

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 today, Orphazyme has several leads that constitute potentially new intellectual property opportunities.

ALS & sIBM

PROTEIN-AGGREGATION DISEASES

NPC & GAUCHER DISEASE

LYSOSOMAL STORAGE DISEASES

ALS

THE DISEASE

The rare neuromuscular disease Amyotrophic Lateral Sclerosis (ALS), also called Lou Gehrig's disease, is rapidly progressive and fatal, usually within two to five years.

The disease attacks the neurons responsible for controlling muscles leading to paralysis of all skeletal muscles, eventually also affecting breathing, speaking, and swallowing.

The cause of damage to the neurons includes protein misfolding and aggregation.

Familial and sporadic ALS

Approximately 10% of ALS cases are associated with familial ALS, while the rest have no identified genetic component (sporadic ALS). Amongst familial ALS cases, 20% harbor mutations in a SOD1 enzyme. SOD1 ALS is often very aggressive with a life expectancy of less than 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 ALS caused by SOD1 mutations.

TREATMENT OPTIONS

There is currently very limited treatment options available to ALS patients, namely; 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. 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 US 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

In August 2018, Orphazyme initiated the enrollment for a Phase III trial in the US and Europe to support the application for a marketing authorization in ALS. Interim analysis at 70% completion is expected in H2 2020 and the full analysis in H1 2021. The trial design and trial patient baseline characteristics were defined based on systematic analysis of data from the largest publicly available repository of ALS clinical trial data (PROACT) in conjunction with arimoclomol ALS trial data. A machine learning approach was used to model disease progression and thereby inform trial design and statistics. The primary endpoint is determined as a combined assessment of function and survival.

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TRIAL SITES CURRENTLY OPEN

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, 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 known, but degenerative factors, i.e. the build-up of tangled and misfolded proteins (inclusion bodies), play a major role.

TREATMENT OPTIONS

There is currently no approved drug 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 US and Europe is not fully known, 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 US and the major European countries[1].

TRIAL STATUS

A Phase II/III arimoclomol trial for sIBM was initiated in August 2017 in the US and Europe. The trial is intended to support a registration of arimoclomol for the treatment of sIBM. Interim analysis expected in H1 2020, with final trial results expected in H1 2021. The extension from 12 to 20 months for the primary analysis was chosen to maximize the chances of success, allowing for greater separation (and thereby increase the clinical meaningfulness of the treatment benefit) between treatment groups at 20 months.

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TRIAL SITES CURRENTLY OPEN

[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.

NPC is caused by mutations on one of two genes, NPC1 or NPC2. Approximately 95% of individuals with the disease have mutations in NPC1.

The Johura family. Father Mohammed and daughter Amana (NPC) at NPUK event in 2018.

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 appears in childhood but can appears 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 US and Europe.

TRIAL STATUS

Arimoclomol is currently being tested in a clinical Phase II/III trial as a potential treatment for NPC. Positive results from the full data set were reported in January 2019, showing a reduction in disease progression of 74% after 12 months compared to placebo control. The open-label extension of the trial is still on-going and is intended to assess the long-term safety and effectiveness of treatment with arimoclomol beyond the 12-month randomized trial.
Orphazyme is to engage with the regulatory authorities in the US and Europe to discuss the best path forward to approval. We expect submission of filing in H1 2020, with a potential approval in H2 2020.

GAUCHER DISEASE

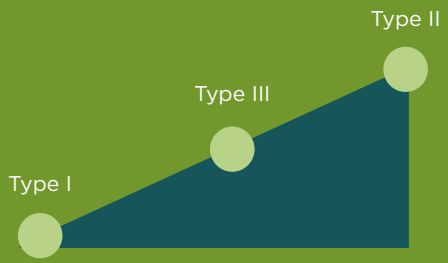
THE DISEASE

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).

There are three types of Gaucher disease, type II being the most severe.



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 US 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 was initiated in Q2 2018. Results are expected in H2 2019.

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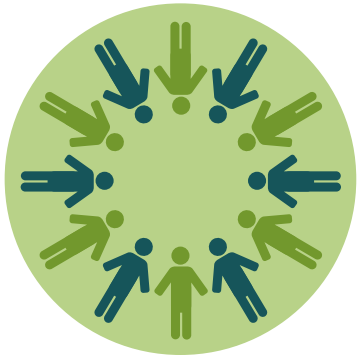
TRIAL SITES ACROSS INDIA

PATIENT FOCUS



WORKING WITH PATIENT ORGANIZATIONS

At the heart of Orphazyme's work to bring innovative therapies to patients with rare and serious diseases are our partnerships with community leaders and non-profit organizations. People living with rare disorders and their families are the authorities on these diseases and we rely on their expertise to inform our actions. In 2018, we took several steps to strengthen our partnerships with patient communities:

- Hired an Associate Director of Patient Advocacy Relations to formalize an advocacy program built on the foundation of Orphazyme's longstanding commitment to collaboration with patient communities
- Made strategic financial investments to encourage the success of rare disease non-profit organizations
- Attended 11 meetings hosted by patient organizations to engage with and learn from individuals and families living with the diseases we aim to treat
- Met personally with nearly 30 organizations over the course of the year and delivered numerous presentations to patient audiences to ensure information about Orphazyme's clinical programs is available and understood
- Formally endorsed consensus guidelines developed to enhance appropriate interactions between industry and rare disease patient advocacy organizations

Evy Reviers
CEO, ALS Liga Belgium and Board Chair, EUpALS (European Organization for Professionals and Patients with ALS)

"As the daughter of an ALS patient, I am daily confronted with the needs of people suffering from this incurable and deadly neuromuscular disease. This is my motivation to lead the team of ALS Liga Belgium. Together, we stimulate ALS research, offer direct support to ALS patients and families, defend their rights at governments, and provide free-of-charge, highly sophisticated aid goods for mobility and communication.

Jerry Williams
Founder and President, Myositis Support & Understanding (MSU)

"At MSU, one of our goals is to provide the myositis community with education, information, and access to research. We are proud to partner with Orphazyme towards the shared goal of a successful clinical trial and ultimately, a treatment for those living with sIBM. We thank Orphazyme for their willingness to work with and learn from myositis patients, non-profits, and community organizations.

Tanya Collin-Histed
CEO, International Gaucher Alliance (IGA)

"The IGA's vison is a world where all Gaucher patients have access to the treatment and care they need and where there is a possibility of a cure. Despite being 28 years on from the introduction of the first treatment for Gaucher disease patients, there are still unmet needs, both clinical and accessibility. The IGA has been a partner with Orphazyme from the very early stages of their clinical development program and we look forward to the potential that this therapy could bring to target the unmet need of the neurological patient community.

Toni Mathieson
Chief Executive, Niemann-Pick UK (NPUK)

"Collaboration between industry, patient groups, and other stakeholders is essential in bringing new therapies to patients. Orphazyme's commitment to developing effective solutions for our rare patient community cannot be overstated and has brought great hope and encouragement to those living with Niemann-Pick type C.

PARTNERSHIPS

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.

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 bearing, 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.

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 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 com-pounds purchased. Please refer to Orphazyme's Prospectus of November 2017 for further details.

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

FINANCIAL REVIEW

INCOME STATEMENT

The net result for the financial year 2018 was a loss of DKK 229.6 million compared to DKK 126.2 million in 2017. The increase is primarily due to increased research and development activities, the hiring of new employees, as well as production of arimoclomol.

RESEARCH AND DEVELOPMENT COSTS

Research and development expenses totaled DKK 196.5 million in 2018 compared to DKK 99.0 million in 2017. The increase was mainly due to the on-going NPC trial, the ramp-up of the sIBM Phase II/III trial, the initiation of the Phase II trial for Gaucher disease in June, and the initiation of the Phase III trial for ALS in August. These expanded clinical trial activities also increased related clinical development costs, including an increase in the manufacturing of the drug substance arimoclomol for the clinical trials, as well the hiring of new employees to support our expanding clinical development activities.

GENERAL AND ADMINISTRATIVE EXPENSES

General and administrative expenses totaled DKK 35.1 million in 2018 compared to DKK 32.0 million in 2017. The increase is mainly due to increased employee costs, the initiation of pre-commercial activities as well as increased investor relations activities. Furthermore, the costs are also impacted by the hiring of new members of management and additional staff to some functions in-house. In 2017, IPO-related costs accounted for a significant part of the general and administrative expenses.

NET FINANCIAL ITEMS

Net financials totaled an expense of DKK 3.5 million in 2018 compared to an income of DKK 0.7 million in 2017. 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 2018 compared to DKK 5.5 million in 2017. The Income tax benefit for the two years represents 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

As of December 31, 2018, Orphazyme had cash DKK 394.7 million compared to DKK 631.7 million as of December 31, 2017. The decrease in cash results from the increase in research and development spend, as described above.

Equity

As of December 31, 2018, equity amounted to DKK 388.3 million compared with DKK 615.7 million as of December 31, 2017. The decrease mainly reflects the financial result for the year.

CASH FLOWS

Cash flow from operating activities

Net cash flow from operating activities amounted to an outflow of DKK 234.8 million in the year ended December 31, 2018 compared to DKK 95.4 million in the year ended December 31, 2017. Net cash flow from operating activities is attributable primarily to the initiation and progression of clinical development activities, as well as general and administrative expenses.

Cash flow from investing activities

Net cash outflow from investing activities amounted to an outflow of DKK 2.3 million in the year ended December 31, 2018 compared to DKK 1.5 million in the year ended December 31, 2017. Investing activities comprise the capitalization of a milestone payment as a license, investment in equipment for research and development purposes, as well as refurbishment of our leased premises in Copenhagen.

Cash flow from financing activities

There was no net cash flow from financing activities in the year ended December 31, 2018 compared to DKK 714.3 million in the year ended December 31, 2017. The financing activity in 2017 was related to the IPO on Nasdaq Copenhagen.

RISK MANAGEMENT

RISKS THAT THREATEN THE ACHIEVEMENT OF OUR KEY OBJECTIVES

Key objective	Risks that threaten the achievement of our key objectives	Our actions to mitigate the risks
To successfully conduct and complete the on-going clinical 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 obtain regulatory approvals 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 and we conduct inspections and internal quality audits to maximize quality, safety, and efficacy. We maintain frequent interactions with regulatory authorities to ensure that we are moving in the right direction to advance our programs towards approval in the most expedient manner.
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.	In anticipation of positive feedback from discussions with regulatory authorities and eventual authorizations, we are refining and finalizing our commercialization strategy. Currently our commercial structure and operations are focused on NPC and on building out an expandable organization as our arimoclomol development program progresses. During 2018 we established a subsidiary in the Boston area in the U.S. with the intent to drive commercialization activities out of there. We have been performing market assessments and working with patient organizations to identify Outside of our key markets being Europe and the U.S., we currently intend to partner with local or regional distributors or license partners to make our products widely available.
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 THAT THREATEN OUR BUSINESS

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, filing activities, commercialization activities, or other critical activities.	With the current capital resources, we expect to be able to fund our operating plans through our planned filing activities. 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 to be able to complete all development and preparation of commercialization activities, while at the same time pursuing filing and registration 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 have adopted new procedures in order to comply with the EU General Data Protection Regulation. Our IT-security level is being scrutinized and new procedures are being implemented in order to reduce the risk of cybercrime. We have set up a special IT Working Group to monitor security as part of the validation of systems and programs used in performing our work.
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 has increased regulatory resources to allow adequate time to interact with regulators and actively monitor the current regulatory environment and ensure our compliance. Our focus in the coming year is on preparations for filing for regulatory approval and therefore compliance is a focus area. We have internal training requirements for all employees and when entering into contracts with external suppliers, we ensure they have an adequate level of measures in place to comply with relevant regulatory requirements.

CORPORATE GOVERNANCE

In order to maintain the trust of the Company's stakeholders, Orphazyme is committed to ensuring transparent and good corporate governance. As a company listed on Nasdaq Copenhagen, Orphazyme is subject to the Recommendations on Corporate Governance from November 2017.

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.

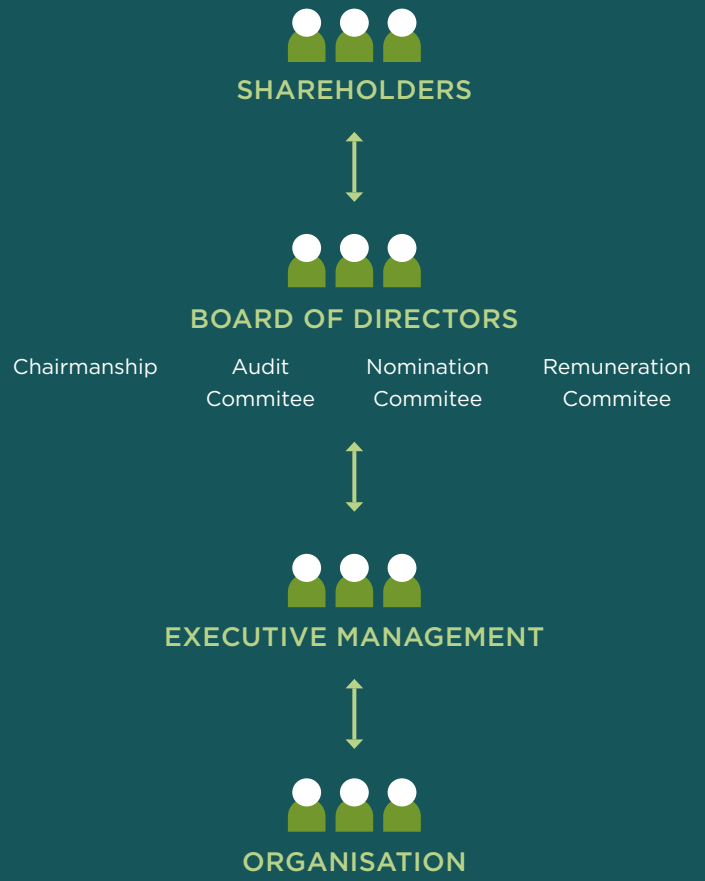
Orphazyme's corporate governance statement includes a summary of the company's governance structure, Orphazyme's position on the Recommendations on Corporate Governance as well as a complete list of the company's comments to each recommendation.

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

Corporate Governance Statement

BOARD OF DIRECTORS

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.



BOARD OF DIRECTORS

Name	Position	Independent[1]	Year of first appointment	Expiration of term
Georges Gemayel	Chairman	Independent	2012	2019
Bo Jesper Hansen	Deputy Chairman	Independent	2010	2019
Martin Bonde	Member	Independent	2010	2019
Sten Verland	Member	Independent	2010	2019
Rémi Droller	Member	Independent	2015	2019
Martijn Kleijwegt	Member	Independent	2017	2019
Catherine Moukheibir	Member	Independent	2017	2019
Anders Hedegaard	Member	Independent	2017	2019

[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).

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 Charters Summary

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 November 2017 (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

As a biopharmaceutical company, we are aware of our 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 demonstrate respect for key moral principles and comply with international regulations and good practice guidelines to make sure that our product is safe and meets strict quality guidelines. As most of our business activities are outsourced, we work closely with our partners to ensure compliance with these regulations and guidelines.

We believe that our ethical behavior strengthens Orphazyme's status as an attractive workplace for both current and future employees.

Diversity Policy

BUSINESS MODEL

Orphazyme is a Danish biopharmaceutical company with approximately 65 employees. The office in Copenhagen serves as headquarters and focuses on coordination and execution of the drug development process and on administration of pre-clinical and clinical trials.

The key activity of Orphazyme is to develop reatments for rare, or orphan, diseases with protein misfolding where it can apply its specialized know-how in heat-shock proteins (HSPs). Our objective is to successfully conduct and complete the planned and on-going trials of arimoclomol for the treatment of 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. We use external suppliers in order to both manufacture and tabletize the medicine, as well as for distribution.

RISK ANALYSIS

Risk is defined as the potential negative effect that can be experienced by the business or any of Orphazyme's stakeholders. Risk is seen as a combination between impact and likelihood of any given subject. Orphazyme's potential risk of impacting social and employee conditions, human rights and anti-corruption is estimated to be limited, mainly due to the strictly regulated business environment the company operates in. Orphazyme's potential risk of impacting the environment and climate is estimated to be very limited mainly as a result of the modest number of employees and the use of external suppliers for all production. Orphazyme has prepared a policy for social and employee conditions, human rights and anti-corruption. Insofar as a specific risk has been identified in any of these areas, it will be addressed by the relevant policy.

CSR REPORTING AREAS

ENVIRONMENT AND CLIMATE

Orphazyme does not have Environment or Climate policies. The Company acknowledges the challenges linked to climate change, and as a result Orphazyme operates an office focused on efficient management of office materials. However, based on the fact that the Company is operating from rented offices and conducts its business in a highly regulated industry and climate, Orphazyme's potential impact on the environment and climate is viewed as minimal. As a result, specific Environment and Climate policies have not been developed at this time.

SOCIAL AND STAFF MATTERS

The office in Copenhagen serves as our headquarters and focuses on coordination and execution of the drug development process and on the conduct of preclinical and clinical trials administration. As of December 31, 2018, the organization comprised:

- Senior Management (5 persons comprising Executive Management and Key Employees)
- Administration (2 persons)
- Clinical development (12 persons)
- Chemistry, manufacturing, and controls/quality assurance (3 persons)
- Regulatory (3 persons)

- Research (11 persons)
- Finance and legal (4 persons)

Policies

Orphazyme A/S knows that all employees are critical to the success of the Company and its programs. The Company is a diverse workplace committed to maintaining a working environment that is free of discrimination, harassment, and bullying 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. 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. The Board of Directors' target is to include at least two female board members by the end of 2021, which we intend to achieve. However, during 2018 we believe the board composition was ideally suited to our current activities and have not made changes.

Orphazyme values the personal development of its employees and offers employees the opportunity to participate in conferences and courses in order to strengthen their professional competences. Policies regarding staff matters are further elaborated in the Employee Handbook.

Activities and results

In 2018 Orphazyme continued to actively encourage all potential employees to apply for employment opportunities, irrespective of gender, age, race, religion, or ethnicity. Moreover, Orphazyme continues to introduce all new employees to the Employee Handbook in order to ensure a workplace that respects diversity and other values inherent in the Company.

We encourage diversity, including age, ethnicity, nationality, religion, education, and skills, and seek to be an attractive workplace for both women and men, endeavoring to ensure equal opportunities for furthering their careers and attaining and occupying management positions.

In 2018, Orphazyme increased its efforts to offer its employees opportunities for personal learning and development by providing internal training as well as external continuing education courses. The Company maintains its focus on a healthy work-life balance by encouraging employees to take all holidays to which they are entitled, and by providing opportunities such as working from home if needed. Furthermore, in 2018 Orphazyme continued to promote a healthy lifestyle by offering weekly employer-paid exercise options.

In 2018, Orphazyme has been able to retain a highly satisfactory employee turnover rate.

HUMAN RIGHTS

Policies

Orphazyme acknowledges and supports the maintenance of internationally declared human rights and bases its work on the UN Universal Declaration of Human Rights and the interpretation that it is the responsibility of the State to protect, and the companies' responsibility to respect these rights.

Orphazyme conducts its business in accordance with the ethical and scientific principles governing clinical research on human subjects, as set out in the Declaration of Helsinki and the International Conference on Harmonization (ICH) guidelines on Good Clinical Practice (GCP)

Orphazyme interprets human rights to comprise respect for diversity. Therefore, it is the aim of the Company to ensure that employees have the possibility to function in a workplace without harassment regardless of gender, age, race, religion or ethnicity.

Activities and results

All protocols in 2018 have been carried out in accordance with the current ICH-GCP guidelines. Furthermore, in 2018 Orphazyme included in its supplier contracts a new condition that requires all suppliers to comply with the Declaration of Helsinki.

In 2018, Orphazyme introduced a whistleblower policy in order to allow reporting of potential violations of the respect for diversity in the workplace. The policy was implemented via email distribution and mandatory meetings for all employees. In 2018, there have been no reports via the whistbleblower scheme concerning violations of the respect for diversity in the workplace or other human rights violations.

ANTI-CORRUPTION AND BRIBERY

Policies

Orphazyme is committed to maintaining the highest standards of ethical conduct and will not tolerate the use of bribery or corruption to achieve its business objectives. Employees must decline any expensive gifts, money, trips, or other such offerings from business contacts. This also includes receiving services from suppliers without paying for them.

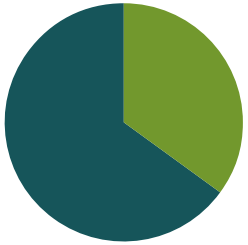
Activities and results

In 2018, Orphazyme introduced a whistleblower policy in order to allow reporting of potential violations of laws and serious violations of internal policies and procedures, including fraud and anti-corruption. The policy was implemented via email distribution and mandatory joint meetings for all employees. Moreover, Orphazyme continued, via its Employee Handbook, to familiarize all new employees with the company policies regarding anti-corruption.

KEY EMPLOYEE RATIOS

	2018		2017	
	Male	**Female**	**Male**	**Female**
Orphazyme A/S	35%	65%	34%	66%
Executive Management and Key Employees	100%	0%	100%	0%
Director level and above	30%	70%	30%	70%
Below director level	29%	71%	22%	78%

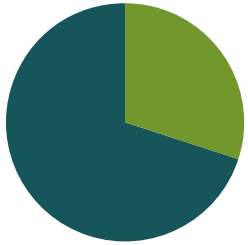
Year-end FTEs - 2018



- Male 20
- Female 37

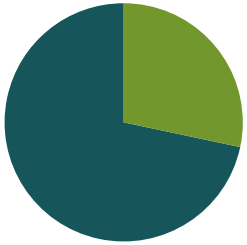
Director level and above - 2018

(not including Executive Management and Key Employees)



- Male 3
- Female 7

Below director level - 2018



- Male 12
- Female 30

This year, Orphazyme has continued to only enter into agreements with consultants that have signed the acknowledgement of Orphazyme's commitment to maintaining the highest standards of ethical conduct.

Orphazyme has not identified any cases of anti-corruption or bribery incidents throughout the year nor have any incidents been reported via the whistleblower scheme.

SHAREHOLDERS AND SHARE INFORMATION

OWNERSHIP

Since November 16, 2017, Orphazyme has been 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, 2018, the number of registered shareholders totaled 3,690 shareholders holding a total of 15,775,628 shares, which represented 74.43% of the total share capital of 19,928,184. In January 2019, there was a capital increase of 26,060 shares related to the issue of bonus shares to Kansas Life Sciences Development Company, Inc. and UCL Business PLC, resulting in a total share capital of 19,965,624 as of March 1, 2019, the date of this annual report.

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 of December 31, 2018.

MAJOR SHAREHOLDERS, DECEMBER 31, 2018

Major shareholder	Company address	Share capital % at December 31, 2018
Novo Holdings A/S	Tuborg Havnevej 19, 2900 Hellerup, Denmark	14.97%[1]
LSP V Coöperatieve U.A.	Wilhelmina Tower 7th Floor, Delflandlaan 1, 1062 EA Amsterdam, Netherlands	13.6%[2]
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] As disclosed by Novo Holdings on January 30, 2019, [2] 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.)

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 poten-tially 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, Oddo, and Redeye.

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

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

Carnegie, Danske Equities, Oddo, and Redeye

SHARE PERFORMANCE

FINANCIAL CALENDER

- **Annual General Meeting:** Wednesday, March 27, 2019
- **Interim Report First Half 2019**: Wednesday, August 28, 2019

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

NEWS

2018 COMPANY ANNOUNCEMENTS

10 **DECEMBER 10**
Orphazyme expects to announce results of full data set for Niemann-Pick disease Type C (NPC) Phase II/III trial in Q1 2019

8 **NOVEMBER 8**
Financial calendar 2019

28 **SEPTEMBER 28**
Orphazyme reports encouraging arimoclomol clinical trial top-line data in Niemann-Pick disease Type C (NPC)

28 **AUGUST 28**
Orphazyme announces Interim Report First Half 2018

10 **AUGUST 10**
Orphazyme announces enrollment of first patient in Phase III clinical trial of arimoclomol for ALS

6 **JULY 6**
Major shareholder announcement

21 **JUNE 21**
Enrollment of first patient in Phase II clinical trial for Gaucher disease

4 **JUNE 4**
New phantom share-based incentive program

29 **MAY 29**
Orphazyme Capital Markets Day 2018 – from biology to bedside

12 **APRIL 12**
Resolution passed at the Annual General Meeting

19 **MARCH 19**
Notice to convene Annual General Meeting

15 **MARCH 15**
Orphazyme announces Annual Report 2017

29 **JANUARY 29**
Capital increase of 11,380 shares (equivalent to approximately 0.06% of the existing shares) in Orphazyme A/S as a result of an issue of bonus shares to KLSDC and UCLB

19 **JANUARY 19**
Arimoclomol for NPC receives Rare Pediatric Disease Designation

NEWS

2018 INVESTOR NEWS

27 **NOVEMBER 27**
Publication of manuscript on preclinical Proof-of-Concept of arimoclomol in Gaucher disease

20 **NOVEMBER 20**
Orphazyme to present at Redeye Life Science Day

15 **OCTOBER 15**
Orphazyme endorses patient advocacy consensus guidelines, highlights importance of collaboration with patient communities

20 **SEPTEMBER 20**
Orphazyme to present at 25th NPUK Annual Family Conference 2018

18 **SEPTEMBER 18**
Orphazyme to present at InvestorDagen 2018

6 **SEPTEMBER 6**
Orphazyme to present at BioCentury 25th Annual NewsMakers in the Biotech Industry

22 **AUGUST 22**
Orphazyme Interim Condensed Consolidated Financial Statements First Half 2018 Presentation

4 **JUNE 4**
Orphazyme presents at 2018 Parseghian Scientific Conference for Niemann-Pick disease Type C

17 **APRIL 17**
Orphazyme announces establishment of US subsidiary in Massachusetts

12 **MARCH 12**
Orphazyme Annual Report 2017 Presentation

9 **MARCH 9**
Orphazyme to present at Cowen & Co. 38th Annual Healthcare Conference

28 **FEBRUARY 28**
Orphazyme recognizes Rare Disease Day 2018

6 **FEBRUARY 6**
Poster presentation at WORLDSymposium™ 2018 – arimoclomol as clinical candidate for treatment of Gaucher disease

24 **JANUARY 24**
Publication of manuscript related to Phase II trial results for arimoclomol in patients with SOD1 ALS

CORPORATE INFORMATION

COMMERCIAL BANKERS

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

Nordea
Vesterbrogade 8
DK-1620 Copenhagen

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 March 27, 2019 at 5.00 PM CET at:
COBIS, Ole Maaløes Vej 3,
DK-2200 Copenhagen N

BOARD OF DIRECTORS

GEORGES GEMAYEL
Chairman of the Board

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

Special competencies
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. Dr Gemayel has significant management and executive experience from the global pharmaceutical industry.

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

BO JESPER HANSEN
Deputy Chairman of the Board

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

Special competencies
Bo Jesper Hansen holds an MD and a PhD in Medicine from the University of Copenhagen. Dr Hansen has extensive experience in orphan drugs, both from the operations and supervisory point of view and has broad and current know-how of the biotechnology environment.

Management duties
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.

BOARD OF DIRECTORS

MARTIN BONDE

Member since: 2010
Born in: 1963
Nationality: Danish
Committees: Nomination Committee

Special competencies

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. Dr. Bonde has executive experience and in-depth knowledge of the biotechnology environment in the Nordic countries.

Management duties

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 member of the Board of Directors of VisioPharm A/S.

RÉMI DROLLER

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

Special competencies

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. Mr. Droller has extensive experience as a biotechnology investor and a proven track-record in negotiating several successful transactions.

Management duties

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

MARTIJN KLEIJWEGT

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

Special competencies

Martijn Kleijwegt holds a Master's degree from the University of Amsterdam. Mr. Kleijwegt has extensive experience as a major European venture-capital investor as well as in-depth experience from the pharmaceutical industry.

Management duties

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.

BOARD OF DIRECTORS

STEN VERLAND

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

Special competencies

Sten Verland holds a Master in Biology and Mathematics and a PhD in Immunology from the University of Copenhagen. Dr. Verland is a serial entrepreneur in biotechnology companies and has extensive investment and managerial experience.

Management duties

Sten Verland is Co-Founder and General Partner of 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, MinervaX ApS, OxThera AB, Danish Venture Capital and Private Equity Asscociation (DVCA), and in companies in or associated with the Sunstone Group.

CATHERINE MOUKHEIBIR

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

Special competencies

Catherine Moukheibir holds a Master in Economics and an MBA, both from Yale University. Ms Moukheibir's has in-depth experience from the pharmaceutical and banking industries and a successful track record in leading Audit Commitees of publicly-traded companies.

Management duties

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), and Genkyotex SA (publ).

ANDERS HEDEGAARD

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

Special competencies

Anders Hedegaard holds a Master of Science in Chemical Engineerng and Biochemistry from the Technical University of Denmark. Mr. Hedegaard has extensive knowledge of the healthcare industry, both in product development and commercialization.

Management duties

Anders Hedegaard is currently Chief Executive Officer of Rodenstock Group.

EXECUTIVE MANAGEMENT

ANDERS HINSBY, PHD

Chief Executive Officer, Co-Founder

Born in: 1973
Nationality: Danish

Special competencies

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

ANDERS VADSHOLT, MBA

Chief Financial Officer

Joined in: 2016
Born in: 1969
Nationality: Danish

Special competencies

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

KEY EMPLOYEES

THOMAS KIRKEGAARD JENSEN, PHD

Chief Scientific Officer, Co-Founder

Born in: 1977
Nationality: Danish

Special competencies

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.

THOMAS BLAETTLER, MD

Chief Medical Officer

Joined in: 2016
Born in: 1967
Nationality: Swiss

Special competencies

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

PAUL MERRIGAN, MBA

Chief Commercial Officer

Joined in: 2018
Born in: 1960
Nationality: American

Special competencies

Executive MBA, Boston University. 30+ years' experience from the biotech and biopharmaceutical industry. Previously at Catalyst Pharmaceuticals, Aegerion Pharmaceuticals, Genzyme, Genentech, Marion Laboratories, and Pfizer.

GLOSSARY

European Medicines Agency (EMA)
Regulatory agency in Europe that facilitates development and access to medicines, evaluates applications for marketing authorization and monitors the safety of medicines.

Fast-Track Designation
“Fast Track” is a process designed by the US Food and Drug Administration (FDA) to enable and accelerate the development and review of drugs for diseases with an unmet medical need, getting new drugs to patients earlier.

Heat-Shock Proteins
Heat-Shock Proteins (HSPs) are molecular chaperones constituting a natural system that makes other proteins work correctly and guard against toxicity arising from misfolded proteins and dysfunctional cellular recycling systems.

Marketing Authorization Application (MAA)
A submission to apply for marketing approval for a drug from EMA.

Orphan Drug Designation
This program provides orphan status to drugs and biologics, which are defined as those intended for the safe and effective treatment, diagnosis or prevention of rare diseases.

Priority Review
FDA designation used for drugs that, if approved, would be significant improvements in the safety or effectiveness of the treatment, diagnosis, or prevention of serious conditions when compared to standard applications.

Rare Pediatric Disease Designation
The Rare Pediatric Disease Designation is granted by the FDA to drugs that have a potential to treat rare pediatric diseases. Orphazyme has received such a designation for arimoclomol. The designation entails the potential receipt of a so-called Priority Voucher upon marketing authorization, if certain criteria are met. The voucher can be redeemed to provide Priority Review of a subsequent marketing application for a different product.

U.S. Food and Drug Administration (FDA)
U.S. regulatory agency responsible for ensuring the safety, efficacy and security of human and veterinary drugs, biological products and medical devices.

ORPHAZYME

2018 CONSOLIDATED FINANCIAL STATEMENTS

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CONSOLIDATED FINANCIAL STATEMENTS

CONSOLIDATED STATEMENTS OF PROFIT OR LOSS AND OTHER COMPREHENSIVE INCOME

For the years ended December 31

Note		2018 TDKK	2017 TDKK
2.1	Research and development expenses	(196,525)	(99,048)
2.3	General and administrative expenses	(35,127)	(31,994)
	Operating loss	**(231,652)**	**(131,042)**
2.6	Financial income	5	26
2.6	Financial expenses	(3,453)	(688)
	Loss before tax	**(235,100)**	**(131,704)**
2.7	Income tax benefit	5,500	5,500
	Net loss for the year	**(229,600)**	**(126,204)**
	Items that will be reclassified subsequently to the Statement of Profit or Loss:		
	Exchange difference from translation of foreign operation, net of tax DKK 0	42	-
	Total comprehensive loss	**(229,558)**	**(126,204)**
	Loss per share, basic and diluted	(11.50)	(10.43)

CONSOLIDATED STATEMENTS OF FINANCIAL POSITION

As of December 31

Note		2018 TDKK	2017 TDKK
	ASSETS		
	Non-current assets		
3.1	Licenses	10,744	9,853
3.2	Property, plant, and equipment	1,940	1,851
2.7	Corporation tax receivable	2,750	2,750
3.3	Prepayments and deposits	2,531	410
	Total non-currents assets	**17,965**	**14,864**
	Current assets		
2.7	Corporation tax receivable	5,500	5,500
3.3	Prepayments and other receivables	23,178	10,718
3.5	Cash	394,706	631,735
	Total current assets	**423,384**	**647,953**
	TOTAL ASSETS	**441,349**	**662,817**
	EQUITY AND LIABILITIES		
	Equity		
4.2	Share capital	19,939	19,928
4.2	Share premium	924,021	924,021
	Other reserves	9,112	9,972
	Accumulated deficit	(564,823)	(338,219)
	Total equity	**388,249**	**615,702**
	Non-current liabilities		
3.4	Other non-current liabilities	105	-
	Total non-current liabilities	**105**	**-**
	Current liabilities		
3.4	Trade payables and accruals	42,183	40,232
3.4	Other liabilities	10,812	6,883
	Total current liabilities	**52,995**	**47,115**
	TOTAL EQUITY AND LIABILITIES	**441,349**	**662,817**

CONSOLIDATED STATEMENTS OF CHANGES IN SHAREHOLDERS' EQUITY

Note		Share capital TDKK	Share premium TDKK	Other Reserves: Foreign currency translation reserve TDKK	Other Reserves: Share-based compensation – aquisition of intangible assets TDKK	Accumulated deficit TDKK	Total TDKK
	Balance as of December 31, 2016	**3,361**	**226,285**	**-**	**-**	**(212,137)**	**17,509**
	Net loss for the year					(126,204)	(126,204)
	Other comprehensive loss						
	Total other comprehensive income/(loss)			-	-	(126,204)	(126,204)
	Transactions with owners:						
3.1	Contribution of a license agreement				9,972		9,972
2.5	Share issue in connection with conversion of former preference shares into ordinary shares	6,488	(6,488)				-
4.2	Proceeds from IPO	7,500	592,500				600,000
2.3	Costs related to IPO		(42,605)				(42,605)
4.2	Capital increase	1,741	155,010				156,751
2.5	Exercise of warrants for cash	838	323				1,161
4.2	Costs related to non-IPO related capital increases		(1,004)				(1,004)
2.5	Share-based compensation expense					122	122
	Total transaction with owners	16,567	697,736	-	9,972	122	724,397
	Balance as of December 31, 2017	**19,928**	**924,021**	**-**	**9,972**	**(338,219)**	**615,702**
	Net loss for the year					(229,600)	(229,600)
	Other comprehensive loss			42		-	42
	Total other comprehensive loss			42	-	(229,600)	(229,558)
	Transactions with owners:						
3.1	Capital increase in connection with issuance of bonus shares	11	-	-	(902)	891	-
2.4	Share-based compensation expense					2,105	2,105
	Total transactions with owners	11	-	-	(902)	2,996	2,105
	Balance as of December 31, 2018	**19,939**	**924,021**	**42**	**9,070**	**(564,823)**	**388,249**

See accompanying notes to these financial statements.

CONSOLIDATED STATEMENTS OF CASH FLOWS

For the years ended December 31

Note		2018 TDKK	2017 TDKK
	Operating loss	(231,652)	(131,042)
	Reversal of non-cash items:		
2.5	Equity-settled share-based compensation expense	2,105	122
3.1, 3.2	Depreciation and amortization	1,366	627
	Exchange rate adjustments	(491)	-
	Gain (loss) on sale or disposal of assets	-	119
	Change in working capital:		
3.3	Change in prepayments, deposits, and other receivables	(14,578)	(2,778)
3.4	Change in trade payables and accruals	1,909	32,688
3.4	Change in other liabilities	4,034	-
2.7	Corporation taxes received	5,500	5,500
2.6	Interest received (paid)	(2,957)	(662)
	Net cash used in operating activities	**(234,764)**	**(95,426)**
	Investing activities		
3.1	Purchase of intangible assets	(1,603)	-
3.2	Purchase of property, plant, and equipment	(743)	(1,491)
	Net cash used in investing activities	**(2,346)**	**(1,491)**
	Financing activities		
4.2	Proceeds from IPO	-	600,000
4.2	Costs related to IPO	-	(42,605)
4.2	Capital contributions from shareholders	-	157,912
4.2	Costs related to capital contributions	-	(1,004)
	Net cash provided by financing activities	**-**	**714,303**
	Net change in cash	(237,110)	617,386
	Effects of changes in exchange rates	81	-
	Cash at the beginning of the year	631,735	14,349
	Cash at the end of the year	**394,706**	**631,735**

See accompanying notes to these financial statements.

SECTION 1 – BASIS OF PREPARATION

Section 1 provides a summary of the significant accounting policies applied by the Group, Management's key accounting estimates and judgements, and new IFRS standards applicable to the Group. A detailed description of accounting policies and key accounting estimates and judgements related to specific financial statement line items is presented in each note to the relevant line item.

The consolidated financial statements of the Group have been prepared in accordance with International Financial Reporting Standards, or IFRS, as issued by the International Accounting Standards Board (IASB) and in accordance with IFRS as endorsed by the EU as well as additional disclosure requirements under the Danish Financial Statements Act. The consolidated financial statements of Orphazyme A/S for the year ended December 31, 2018 were approved by the Board of Directors on March 1, 2019 and will be submitted to the shareholders of Orphazyme A/S for approval at the Annual General Meeting to be held on March 27, 2019.

1.1 CORPORATE INFORMATION

Orphazyme A/S (the "Company") is a limited liability company incorporated and domiciled in Denmark. The registered office is located in Copenhagen, Denmark. On November 16, 2017, the Company successfully completed its Initial Public Offering (IPO) on Nasdaq Copenhagen by issuing 7,500,000 new ordinary shares for gross proceeds of TDKK 600,000.

In April 2018, a fully-owned subsidiary, Orphazyme US, Inc., was incorporated in Massachusetts, USA (together with Orphazyme A/S, "Orphazyme" or the "Group"). Orphazyme US, Inc. will directly support the US market to establish closer relationships with the medical, patient, and financial communities as Orphazyme expands its development programs and global reach.

1.2 SIGNIFICANT ACCOUNTING POLICIES

The consolidated financial statements have been prepared on a historical cost basis except for share-based compensation, which is measured at fair value.

The consolidated financial statements are presented in Danish Kroner, or DKK, which is both the functional and presentation currency of the Parent Company. Where indicated, amounts are rounded to the nearest thousand, or TDKK. The functional currency of Orphazyme US, Inc. is the US dollar (USD).

Principles of consolidation

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

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

Orphazyme US, Inc. has adopted the accounting policies of the Parent Company and therefore the Group's consolidated financial statements have been prepared by combining similar accounting items on a line-by-line basis. On consolidation, intercompany income and expenses, intercompany receivables, and payables, and unrealized gains and losses on transactions between the consolidated companies are eliminated.

Translation of foreign currencies

Items included in the financial statements of each of the Orphazyme entities are measured using the currency of the primary economic environment in which the entity operates, or functional currency. On initial recognition, transactions denominated in foreign currencies are translated at the foreign exchange spot rate at the transaction date. For monetary assets and liabilities, differences arising between the foreign exchange spot rates at the transaction date and the date of settlement or period-end exchange rates are recognized in the Statement of Profit or Loss as financial income or financial expenses.

On consolidation, the assets and liabilities of Orphazyme US, Inc are translated from USD to DKK at the exchange rate in effect at the balance sheet date and the Statement of Profit or Loss and Other Comprehensive Income is translated from USD to DKK at the date of the underlying transaction or average exchange rate of the period if there are no significant fluctuations in exchange rate throughout the period. The exchange rate differences arising on translation for consolidation are recognized in other comprehensive income (loss).

Segment information

Although Orphazyme established a US subsidiary in 2018, the Group is managed and operated as one business unit that is reflected in the 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 Group's internal reporting. For the years ended December 31, 2018 and 2017, the Group generated no revenue and all non-current assets were located in Denmark.

1.3 KEY ACCOUNTING ESTIMATES AND JUDGEMENTS

The use of reasonable estimates and judgements is an essential part of the preparation of the consolidated financial statements. Given the uncertainties inherent in the Group's business activities, Management must make certain key accounting estimates and judgements, which affect the application of accounting policies and therefore the reported amounts of assets, liabilities, revenue, expenses, and disclosures in the consolidated financial statements. The key accounting estimates and judgements identified are those that have a significant risk of resulting in a material adjustment to the consolidated financial statements.

Management bases its estimates on historical experience, assumptions, and information currently available and deemed to be reasonable at the time the consolidated financial statements are prepared. However, actual amounts may differ from the estimated amounts as more detailed information becomes available. Estimates and assumptions are reviewed on an ongoing basis and, if necessary, changes are recognized in the period in which the estimate is revised. Management has made key accounting estimates and judgements in the following areas:

- Key estimate of research and development expenses associated with clinical trials (Note 2.1) and related prepayments (Note 3.3) and accruals (Note 3.4)
- Key estimate of inputs and assumptions used in share-based compensation valuation models (Note 2.5)
- Key estimate of the fair value of licenses (Note 3.1)
- Key judgement regarding IPO and IPO-related costs in 2017 (Note 2.3)
- Key judgement regarding the recognition of deferred tax assets related to taxable losses to be carried forward (Note 2.7)

Please refer to the specific referenced notes for further information on the key accounting estimates and judgements as well as assumptions applied.

1.4 NEW IFRS STANDARDS APPLICABLE TO THE GROUP

On January 1, 2018, the Group adopted IFRS 9, *Financial Instruments*, which did not have a significant impact on the consolidated financial statements. In addition, there have been amendments to IFRS 2, *Share-Based Payment*, which the Group adopted, but did not have any impact on the Group's consolidated financial statements. The Group has not early adopted any other standard, interpretation, or amendment that has been issued but is not yet effective. IFRS 15, *Revenue from Contracts with Customers*, was effective on January 1, 2018, however, as the Group does not generate revenue, the standard is not applicable at this time.

Standards issued but not yet effective

The standards and interpretations that are issued, but not yet effective, up to the date of issuance of the Group's consolidated financial statements are disclosed below. Only the standards and interpretations that are expected to have an impact on the Group's financial position, performance, and/or disclosures are listed. The Group intends to adopt these standards, as applicable, when they become effective.

IFRS 16 Leases

IFRS 16 was issued in January 2016 and 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 is

1.4 NEW IFRS STANDARDS APPLICABLE TO THE GROUP (CONTINUED)

effective on January 1, 2019 and 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 Group will adopt the new standard on the required effective date by using the modified retrospective approach, meaning that comparative information is not restated. The cumulative effect of initially applying IFRS 16 will be presented as an adjustment to opening accumulated deficit in equity.

The change in lease accounting requires capitalization of the Group's right to use assets (i.e. the property lease of the headquarters in Copenhagen). Assuming that no new leases are entered into and no amendments to existing leases are made, upon implementation of IFRS 16 on January 1, 2019 the Group is expected to recognize a lease liability of approximately TDKK 13,006 and a corresponding asset representing the right to use the underlying asset during the lease term. The expected accumulated effect on total assets at January 1, 2019 approximates TDKK 13,006, and there will be no effect on equity. Following the implementation, the Group will separately recognize interest expense on the lease liability and depreciation on the right to use asset. The Group's Statement of Profit or Loss for 2019 is expected to be impacted as follows: Operating loss will decrease by approximately TDKK 98, financial expenses will increase by approximately TDKK 398 and net loss before tax and net loss for the year will increase by approximately TDKK 300.

IFRIC Interpretation 23 *Uncertainty over Income Tax Treatment*

The Interpretation addresses the accounting for income taxes when tax treatments involve uncertainty that affects the application of IAS 12 *Income Taxes* and does not apply to taxes or levies outside the scope of IAS 12, nor does it specifically include requirements relating to interest and penalties associated with uncertain tax treatments.

The Interpretation specifically addresses the following:

- Whether an entity considers uncertain tax treatments separately
- The assumptions an entity makes about the examination of tax treatments by taxation authorities
- How an entity determines taxable profit (tax loss), tax bases, unused tax losses, unused tax credits and tax rates
- How an entity considers changes in facts and circumstances

An entity has to determine whether to consider each uncertain tax treatment separately or together with one or more other uncertain tax treatments. The approach that better predicts the resolution of the uncertainty should be followed. The interpretation is effective for annual reporting periods beginning on or after January 1, 2019, but certain transition reliefs are available. The adoption of this Interpretation will not have a significant effect on the Group.

SECTION 2 – RESULT OF THE YEAR

Section 2 presents details related to Orphazyme's Statement of Profit or Loss and Other Comprehensive Income, including Research and Development expenses and General and Administrative expenses, Government Grants, Employee costs and Share-based Compensation costs. The Group does not yet generate revenue. In addition, this section comprises Financial Income, Financial Expenses, and Income Taxes.

2.1 RESEARCH AND DEVELOPMENT EXPENSES

§ ACCOUNTING POLICIES

Research expenses comprise costs incurred during the very early stages of the drug development cycle from initial drug discovery until the drug is ready for administration to humans. The activities initially focus on identifying a single drug candidate with a profile that will support a decision to initiate development activities. Before selection of the final drug candidate, it is tested in animals to gather efficacy, toxicity and pharmacokinetic information.

Development expenses comprise costs incurred during the different phases of clinical drug development starting in phase 1, when the drug is administered to humans for the first time, through phases 2 and 3, and subsequent activities to obtain marketing authorizations, which will permit Orphazyme to eventually market and sell the drug products.

In line with industry practice, Orphazyme expenses all research costs. Development costs that do not meet the definition of an asset are also expensed as incurred. Due to regulatory and other uncertainties inherent in the development of new products, development costs do not qualify for capitalization as intangible assets until marketing approval by a regulatory authority is obtained or highly probable.

Clinical trial costs are a significant component of research and development expenses. The Company's clinical trials are performed by third-party Clinical Research Organizations (CROs) and in order to estimate the amount of costs to charge to expense Management has developed expense models for each clinical trial based on estimates and assumptions.

The clinical trials generally have three distinctive stages plus pass-through costs:

- Start-up stage: initial setting up of the trial
- Treatment stage: site and trial management during the dosing period
- Wrap-up stage: close down and reporting of the trial

For each clinical trial for which actual services delivered by the CRO are not provided on a regular current basis, the Company reviews the approved budgets for the clinical trial from the original executed agreements and categorizes the individual costs according to the three stages described above. The start-up activities, which include site recruitment, regulatory applications and investigator meetings, usually are performed reasonably uniformly throughout the start-up stage and the related costs are expensed ratably over this stage, which reflects the manner in which related services are performed by the CRO. The start-up stage is followed by the treatment stage, during which patients are dosed with the drug under study and results are monitored and measured. The costs incurred in this stage of the

2.1 RESEARCH AND DEVELOPMENT COSTS (CONTIUNED)

trial, which comprises the major portion of the total cost of the clinical trial, is mainly driven by the number of enrolled patients undergoing treatment. The Company estimates the costs attributable to activities performed in this stage of the trial on a per-patient basis. These costs are expensed over the treatment stage as patients are enrolled and undergo treatment, as reported by the CRO. After the last patient has been treated, the trial begins to be closed down and activities are performed related to data quality assurance and analysis. These activities are performed reasonably uniformly throughout the wrap-up stage and are expensed ratably over this last stage. Other costs, such as central laboratory costs and drug supply costs, are expensed as incurred, which is typically when the service has been performed or the goods delivered.

CROs invoice the Company upon the occurrence of predetermined milestones (such as the enrollment of patients); however, the timing of these invoices and the Company's related payments often do not correspond directly to the level of performance of contracted activities. To the extent payments are made by the Company in advance of the related activities performed by the CROs, they are included in prepayments to vendors (see Note 3.3) and expensed in accordance with the expense model discussed above. To the extent that the payments are made by the Company following the performance of the related activities, the expense is reflected as an accrual (see Note 3.4) in accordance with the expense model.

Key estimate of research and development expenses associated with clinical trials

Accounting for clinical trial costs related to activities performed by Clinical Research Organizations (CROs) and other external vendors requires Management to exercise judgment in making significant estimates in regard to the timing of the expense recognition of these costs. The diverse nature of services being provided by CROs, the different compensation arrangements that exist for each type of service, and the limitation in the availability of information related to when certain clinical activities are performed add complexity to the estimation of the timing of expense recognition for services rendered by CROs and other vendors in connection with clinical trials.

Research and development expenses include costs arising from research and clinical development activities including employee costs for research and development personnel (i.e. salaries, bonuses, employer contributions to pension schemes, share-based compensation), legal expenses related to the protection, defense and enforcement of the Company's intellectual property, as well as rent expense associated with facilities used for research and development purposes.

The following table presents Research and Development expenses recognized for the years ended December 31, 2018 and 2017:

Research and development costs

	2018 TDKK	2017 TDKK
External costs	152,820	71,169
Employee costs (Note 2.4)	40,281	25,648
Facility costs	2,132	1,492
Depreciation and amortization (Notes 3.3 and 3.4)	1,292	739
Total Research and Development Expenses	**196,525**	**99,048**

2.2 GOVERNMENT GRANTS

§ ACCOUNTING POLICIES

Government grants are recognized when there is reasonable assurance that the funding will be received and all underlying conditions will be fulfilled. Income from grants is recognized in the Statement of Profit or Loss as a reduction of the related expenses being reimbursed in the period when the related expenses are incurred.

Government grants comprise research funding from the Danish government and the European Union. The grants received by Orphazyme provide reimbursement for certain project-specific research and development expenses, including wages and salaries.

2.1 GOVERNMENT GRANTS (CONTIUNED)

During the year ended December 31, 2018, Orphazyme has received TDKK 2.126 (2017: TDKK 5.307) in government grant funding, which has been recognized as a reduction of research and development expenses. As of the year ended December 31, 2018, the total amount still receivable under these grants is TDKK 1.237 (2017: TDKK 512) and is classified as Current Other Receivables in the Statement of Financial Position, as all remaining funding from grants is receivable within the next year (Note 3.3).

One grant has been paid to Orphazyme in advance and income in the amount of TDKK 299 (2017: TDKK 520) related to this grant has been deferred and presented in the Statement of Financial Position as current other liabilities (Note 3.4).

All the grants received are subject to repayment clauses upon breach of conditions to maintain the terms under which the grant was awarded. Orphazyme has complied with and anticipates continuing to fully comply with all such terms.

2.3 GENERAL AND ADMINISTRATIVE EXPENSES

§ ACCOUNTING POLICIES

General and administrative expenses include salaries for administrative employees and Executive Management, remuneration to the Board of Directors, share-based compensation costs, rent associated with facilities not used for research and development purposes, investor relations, and costs incurred in connection with the Company's Initial Public Offering on Nasdaq Copenhagen in November 2017, which have not been set off against equity. In addition, we have commenced pre-commercial activities during 2018, including the establishment of our U.S. subsidiary. These pre-commercial activities include the creation of an Early Access Program for NPC, tradename costs, market and pricing studies and related costs.

The following table presents general and administrative costs for the years ended December 31, 2018 and 2017:

Key judgement regarding IPO and IPO-related costs in 2017
IPO and IPO-related costs are costs that have been incurred in connection with the Company's Initial Public Offering on Nasdaq Copenhagen in November 2017 and mainly cover fees to bookrunners, banks, and advisors. Certain costs related to preparing the prospectus and issuing new shares have been charged to equity based on the percentage of the newly issued shares at the IPO. The remaining costs have been charged to expense as incurred. 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 similar costs have been expensed as incurred.

	2018 TDKK	2017 TDKK
External costs	12,471	8,445
IPO costs	-	13,456
Employee costs (Note 2.4)	15,803	7,023
Travel and related expenses	4,115	3,063
Pre-commercial activities	2,664	-
Depreciation (Note 3.2)	74	7
Total general and administrative expenses	**35,127**	**31,994**

2.4 EMPLOYEE COSTS

§ ACCOUNTING POLICIES

Employee costs primarily comprise salaries, bonuses, social security contributions, share-based compensation, vacation and sick leave as well as pension contributions. The cost of these benefits is recognized as an expense as services are received. All employee pension plans are defined contribution plans and not defined benefit plans.

Executive Management consists of the Company's Chief Executive Officer and the Chief Financial Officer. Executive Management is also identical to the registered management of the Company.

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 this short-term incentive programs may not exceed 100% of the annual fixed salary of the participants. For the financial year 2018, the Company expensed TDKK 1,173 (2017: TDKK 1,507) for cash bonuses and 0 (2017: TDKK 351) for IPO bonuses for the Executive Management.

Employees are subject to a discretionary bonus subject to certain predefined and individual goals as determined by the Board of Directors. For the financial year, the Company expensed TDKK 2.243 for cash bonuses to employees. In connection with the completion of the IPO on Nasdaq Copenhagen in November 2017, all employees of Orphazyme (including the Executive Management) received a cash bonus corresponding to 10% of their respective annual base salaries (excluding pension contributions).

The following table presents Employee Costs for the years ended December 31, 2018 and 2017:

	2018 TDKK	2017 TDKK
Employee costs, excluding Executive Management and Board		
Salaries	38,915	21,631
Cash bonus	3,410	3,618
Share-based compensation	1,006	44
Pensions	2,686	968
Other social security contributions	322	177
Other staff costs	966	338
Total employee costs, excluding Executive Management and Board	**47,305**	**26,776**
Executive Management remuneration		
Salaries	3,328	2,759
Cash bonus	1,173	1,473
Share-based compensation	1,139	78
Pensions	372	-
Other social security contributions	4	2
Other staff costs	-	-
Total Executive Management remuneration	**6,016**	**4,312**
Board of Directors remuneration	**2,763**	**1,583**
Total employee costs	**56,084**	**32,671**
Recognized as follows in the Statement of Profit or Loss:		
Research and development expenses	40,281	25,648
General and administrative expenses	15,803	7,023
Total employee costs	**56,084**	**32,671**
Average number of full-time employees	**46**	**26**
Year-end number of full-time employees	**57**	**34**

2.4 EMPLOYEE COSTS (CONTINUED)

Remuneration paid to members of the Board of Directors is made up of board and committee fees, a travel allowance, and reimbursement of out-of-pocket expenses and is recognized as general and administrative expenses in the Statement of Profit or Loss. There are no incentive plans granted to the Board of Directors. The following table lists Board of Directors remuneration for the years ended December 31, 2018 and 2017:

	2018 TDKK	2017 TDKK
Board and Committee fees	2,584	1,211
Travel allowance	179	-
Other fees	-	372
Total Board of Directors remuneration	**2,763**	**1,583**

2.5 SHARE-BASED COMPENSATION COSTS

§ ACCOUNTING POLICIES

Equity-settled awards

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

The fair value of equity-settled awards with service conditions and non-market performance conditions is recognized as compensation expense pro rata over the service period to the extent such awards are estimated to vest. The compensation expense is recognized together with a corresponding increase in equity over the period in which the performance and/or service conditions are fulfilled. The cumulative expense for the Group's share-based compensation awards recognized at each reporting date until the vesting date reflects the extent to which the vesting period has expired and Management's best estimate of the number of instruments that will ultimately vest. The expense or credit in the Statement of Profit or Loss for a period represents the movement in cumulative expense recognized as at the beginning and end of that period.

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.

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 fair value of the award is expensed immediately in the Statement of Profit or Loss.

Cash-settled awards

The phantom share-based incentive program established by the Group in June 2018 is cash-settled. A liability is recognized for the fair value of these awards, which is measured initially and at each reporting date up to and including the settlement date, with changes recognized at each reporting date. The fair value is expensed over the period until vesting date with recognition of a corresponding liability. The fair value is determined using the Monte-Carlo model, further details of which are presented below. The fair value of the cash-settled awards, which vest subject to obtaining a specified share price (i.e. market condition), is reported as compensation expense regardless of whether the share price condition is met if all other vesting conditions are met. For these awards, fair value is determined taking into account the probability of meeting the share price target. No expense is recognized for awards that do not ultimately vest.

2.5 SHARE-BASED COMPENSATION COSTS (CONTINUED)

Key estimate of inputs and assumptions used in share-based compensation valuation models

Estimating the fair value of the Group's share-based compensation programs requires determination of the most appropriate valuation model, which depends on the terms and conditions of the respective award. This estimate also requires making assumptions to determine the most appropriate inputs to the valuation model, including the expected life of the award, expected volatility, dividend pay-out ratio, and risk-free interest rate.

Employees and Executive Management of the Group receive remuneration in the form of both equity-settled and cash-settled awards.

Pre-IPO Warrants (equity-settled)

In 2017 and prior to the IPO, without cancelling or modifying former warrant programs, the Company issued 551,573 warrants under a new warrant program for which a mechanism was put in place to ensure that the respective warrant holders can only exercise warrants from either the former programs or the new program. Orphazyme therefore had multiple warrant programs that ran in parallel. The exercise price of the 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 was determined based on (i) the grant date fair value of the former programs under the original vesting terms, plus (ii) the incremental fair value of the new warrant program, as at the grant date (being the fair value of the new programs less the fair value of the former programs at that date), over the vesting terms of the new program. In addition, prior to the IPO, Orphazyme issued 279,019 warrants of which 130,541 warrants were exercisable subject to completion of an IPO. These warrants vested gradually over 4 years subject to continued employment or upon an IPO or a change in control event. The fair value of these warrants granted amounted to TDKK 52 and Management determined that the incremental value was insignificant, and no expense was recognized. Consequently, all these warrants vested upon the completion of the IPO in November 2017.

The table below summarizes the activity related to the pre-IPO warrants for the years ended December 31, 2017. All warrants outstanding were exercised in November 2017 upon completion of the IPO and therefore there was no activity in 2018 related to these pre-IPO warrants. The average share price upon exercise was DKK 80.

	Executive Management	Key Employees	Board of Directors	Consultants	Total Warrants	Warrants exercisable
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	**-**	**-**	**-**	**-**	**-**	**-**

Post-IPO long-term incentive program (equity settled)

In connection with the completion of the IPO on Nasdaq Copenhagen in November 2017, the Executive Management and Key Employees were offered to subscribe for Offer Shares ("Investment Shares") at the Offer Price for a maximum amount corresponding to approximately 15% (CMO) and 20% (CEO, CFO, and CSO) of their respective current annual base salaries.

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

2.5 SHARE-BASED COMPENSATION COSTS (CONTINUED)

The participants in the LTIP 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 the potential increase in the price of Orphazyme's shares at the time of exercise 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 shares for the CEO and four shares for the other participants multiplied by 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 end of the vesting period. Among other things, vesting is also subject to the participants having maintained ownership of their Investment Shares and continued employment. Based on the number of Investment Shares subscribed for, a total maximum of 86,700 Performance Shares may be issued at the end of the vesting period.

In addition, subject to Board approval, 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 the subscription date of the Investment Shares. The number of Matching Shares shall be equal to the number of Investment Shares subscribed for and vesting will be subject to the participants having maintained ownership of their Investment Shares and continued employment during the one-year vesting period. Based on the number of Investment Shares subscribed for, a total of 14,875 Matching Shares may be issued at the end of the vesting period in November 2018. As of December 31, 2018, those Matching Shares had vested in full but had not yet been issued to the participants. The remaining Matching Shares are expected to vest in April 2019.

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

Grant Date	**Apr 10, 2018**	**Nov 21, 2017**
Fair value at the measurement date (TDKK)	714	3,895
Dividen yield (%)	0	0
Expected volatility (%)	41.8 %	44.6 %
Risk-free interest rate (%)	(0.28 %)	(0.43 %)
Expected life of awards (years)	3.58	3.88
Weighted average share price (DKK)	67.5	80

The risk-free interest rate has been estimated on the basis of Danish government bonds with similar maturities. Expected volatility has been determined on the basis of the historic volatility of comparable listed companies.

2.5 SHARE-BASED COMPENSATION COSTS (CONTINUED)

The table below summarizes the activity related to the LTIP for the years ended December 31, 2018 and 2017:

	Executive Management	Key Employees	Total Awards	Awards exercisable
Outstanding at December 31, 2016	**-**	**-**	**-**	**-**
Granted	9,000	5,875	14,875	
Exercised	-	-	-	
Expired	-	-	-	
Forfeited	-	-	-	
Outstanding at December , 2017	**9,000**	**5,875**	**14,875**	**-**
Granted	-	4,300	4,300	
Exercised	-	-	-	
Expired	-	-	-	
Forfeited	-	-	-	
Outstanding at December 31, 2018	**9,000**	**10,175**	**19,175**	**14,875**

The weighted average remaining contractual life for LTIP awards outstanding at December 31, 2018 was 2.88 years (2017: 3.88 years). The exercise price for each LTIP award outstanding as of December 31, 2018 was DKK 1 (2017: DKK 1).

For the year ended December 31, 2018, TDKK 2.105 (2017: TDKK 122) was recognized as compensation expense related to the LTIP, with a corresponding amount recognized in equity. Of the total expense, TDKK 1.139 (2017: TDKK 78) is attributed to the Executive Management. No LTIP awards were granted to the Board of Directors.

Phantom share-based incentive program (cash-settled)

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

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

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

2.5 SHARE-BASED COMPENSATION COSTS (CONTINUED)

The employee's cash awards are capped and cannot exceed a gross amount of DKK 37,500 or DKK 75,000 per employee, depending on the number of phantom shares allocated to the respective employee under the program. Based on the number of participants in the Phantom Shares Program as of December 31, 2018, the program is expected to consist of up to 6,300 phantom shares in total.

As the Phantom Shares Program is cash-settled, the fair value of the phantom shares granted as part of the program is estimated at each reporting date. As of December 31, 2018, the fair value of the phantom shares was estimated at approximately TDKK 73. For the year ended December 31, 2018, TDKK 39 was recognized as compensation expense related to the Phantom Shares Program, with a corresponding amount recognized as a long-term liability (Note 3.4).

The following table presents the inputs to the Monte-Carlo model used to estimate the fair value of the phantom shares revalued as of December 31, 2018:

Valuation Date	Dec 31, 2018
Fair value at the measurement date (TDKK)	73
Dividen yield (%)	-
Expected volatility (%)	52.3%
Risk-free interest rate (%)	(0.34%)
Expected life of awards (years)	4.08
Weighted average share price (DKK)	43.35

The risk-free interest rate has been estimated on the basis of Danish government bonds with similar maturities. Expected volatility has been determined on the basis of the historic volatility of comparable listed companies.

As of December 31, 2018, all phantom shares granted under the Phantom Shares Program were only granted to employees of Orphazyme. No phantom shares were forfeited or expired, and none of the phantom shares were eligible for exercise.

Bonus shares issued to KLSDC and UCL in connection with a license agreement
Please see Note 3.1.

2.6 FINANCIAL INCOME AND FINANCIAL EXPENSES

§ ACCOUNTING POLICIES

Financial income and expenses Include interest income and expense, gains and losses due to changes in foreign exchange rates and other immaterial miscellaneous items.

For the year ended December 31, 2018, the Company recognized TDKK 5 (2017: TDKK26) as Financial Income due to interest; and the Company recognized TDKK 3,453 (2017: TDKK 688) as Financial Expenses due to foreign currency loss and bank charges.

2.7 INCOME TAXES

§ ACCOUNTING POLICIES

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.

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.

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.

2.6 INCOME TAXES (CONTINUED)

Key judgement regarding the recognition of the deferred tax assets related to taxable losses to be carried forward

Orphazyme is subject to income taxes in Denmark and in the U.S.A. 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. Significant judgment is required to determine the amount of deferred tax assets that may 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 to successfully commercialize and defend its intellectual property. After consideration of these factors, Management has concluded that as regulatory approval has not yet been obtained as of December 31, 2018, the deferred income tax assets related to taxable losses carried forward do not meet the criteria for being recognized as assets in the Statement of Financial Position.

The Company's tax losses can be carried forward infinitely subject to the general rules on limited deductibility due to ownership changes. In Denmark, the Company's ability to use tax loss carry forwards in any one year is limited to 100% of the first MDKK 7.5 of taxable income plus 60% of taxable income above MDKK 7.5. For the years ended December 31, 2018 and 2017, the Company has unrecognized net tax loss carry-forwards in the Danish entity in the amount of MDKK 280 and MDKK 235, respectively.

As of December 31, 2018, there are no tax audits in process nor has management been notified of any pending tax audit. As of December 31, 2018, the tax years that remain open for audit by the Danish tax authorities include 2013 through 2016.

Management periodically evaluates positions taken in tax returns with respect to situations in which applicable tax regulations are subject to interpretation or uncertainty and establishes provisions, where appropriate. To date, there have not been any provisions established for uncertain tax positions.

The following table presents the total income tax benefit for the years ended December 31, 2018 and 2017:

	2018 TDKK	2017 TDKK
Current tax benefit on net loss	51,722	28,975
Tax credit research and development expenses	5,500	5,500
Change in unrecognized deferred tax before tax credit	(51,850)	(27,592)
Permanent deviations	128	(1,383)
Total income tax benefit for the period	**5,500**	**5,500**

2.6 INCOME TAXES (CONTINUED)

The following table presents the reconciliation of the effective tax rate to the statutory corporate income tax rate in Denmark.

	2018 TDKK	2017 TDKK
Net loss before tax	(235,100)	(131,704)
Corporate income tax rate in Denmark	22%	22%
Computed income tax benefit	**51,722**	**28,975**
Tax effect of:		
Other non-deductible expenses, including IPO-related costs and share-based compensation	128	(1,383)
Deferred tax asset not recognized after tax credit	(46,350)	(22,092)
Total income tax benefit for the period	**5,500**	**5,500**

The following table presents the carrying amount of deferred tax in the Statement of Financial Position:

	2018 TDKK	2017 TDKK
Tax deductible losses	61,647	51,644
Deferred tax on intangible assets	35,887	-
Other temporary differences	738	277
	98,272	51,921
Deferred tax asset not recognized	98,272	51,921
Carrying amount included on statement of financial position	**-**	**-**

SECTION 3 – OPERATING ASSETS AND LIABILITIES

Section 3 presents details of the operating assets and liabilities that form the basis of Orphazyme's activities, including Licenses, Property, Plant and Equipment, Prepayments, Deposits, and Other Receivables, Financial Assets and Liabilities, Cash, and Contractual Obligations and Contingencies.

3.1 LICENSES

§ ACCOUNTING POLICIES

License rights to develop and commercialize products that are accounted for as intangible assets and acquired separately are measured on initial recognition at cost. For acquisition of intangible rights involving equity-settled share-based payment transactions, Management measures the fair value of the rights received and the corresponding increase in equity by reference to the fair value of the rights received, unless that fair value cannot be estimated reliably. If Management cannot estimate reliably the fair value of the rights received, it measures the fair value and the corresponding increase in equity 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. 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.

Key estimate of the fair value of licenses

Licenses contains an agreement entered into with the University of Kansas and University College London, in which the Company will obtain access to data and know-how generated in the course of research in connection with the sIBM trial. Consideration for the license is to be paid out by issuing new shares to the contract partners for a value corresponding to the costs incurred during the preceding calendar year. The valuation of the license upon the execution of the agreement involves uncertainty and was estimated by Management based on the expected costs over the contract period. In addition, the estimation of the duration of a license agreement at times involves uncertainty if termination is dependent on a time limit after successful commercialization. Management has considered potential commercialization dates and will re-assess this estimate on an ongoing basis.

3.1 LICENSES (CONTINUED)

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, which includes arimoclomol, to Orphazyme. Under the terms of the Asset Purchase Agreement, Orphazyme agreed to make future payments to CytRx that were contingent upon the achievement of specified clinical, regulatory, and sales milestones. The Asset Purchase Agreement further includes royalty payments to be made by Orphazyme based on a specified percentage of any eventual net sales of products containing one of the compounds purchased. In August 2018, the Company made a milestone payment of USD 250,000 (TDKK 1,603) upon the enrollment of the first patient in the ALS clinical trial. The Company has capitalized this amount as an acquired license right, as Management assesses that the consideration paid reflects market expectations about the probability that future economic benefits will flow to the Company. The acquired license is not being amortized until approval of the underlying asset has been received from regulatory authorities.

In 2017, the Company entered into a license agreement with KU Center for Technology Commercialization, Inc., University of Kansas, Kansas Life Sciences Development Company, Inc., ("KLSDC") and UCL Business PLC ("UCL") granting Orphazyme the right 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 worldwide. The total consideration for the license is to be paid out in bonus shares to KLSDC and UCL up to an aggregate value of USD 2.5 million (DKK 15.8 million), depending on the amount of grants awarded to KLSDC and UCL for use in the trial. At the time the license agreement was executed, Management estimated the aggregate amount of the funding to be received by KLSDC and UCL to be USD 1.583 thousand (TDKK 9.972), which has been recognized as an intangible asset (License) with a corresponding increase in equity reserves (Share-based compensation-acquisition of intangible assets).

Consideration to KLSDC and UCL is payable in shares of the Company ("Bonus Shares") each January and is based on incurred costs reported by KLSDC and UCL for the previous year. As at December 31, 2017 the aggregate costs incurred by KLSDC and UCL amounted to USD 143 thousand (TDKK 902), and a total of 11,380 Bonus Shares were issued to KLSDC and UCL in January 2018, based on the average 30-day closing price of Orphazyme's shares. In addition, at the time of the share issuance the equity reserve was decreased by TDKK 902, which represents the market value of the shares issued.

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.

The license is being amortized over the duration of the license agreement, which has been estimated to be approximately 14 years. Amortization expense for the years ended December 31, 2018 and 2017 amounts to TDKK 712 and TDKK 119, respectively, and is recognized within research and development expenses.

The following table presents the cost and respective amortization of the licenses held by Orphazyme:

	TDKK
Cost at December 31, 2016	-
Additions	9,972
Cost at December 31, 2017	9,972
Additions	1,603
Cost at December 31, 2018	**11,575**
Accumulated amortization at December 31, 2016	-
Amortization expense	119
Accumulated amortization at December 31, 2017	119
Amortization expense	712
Accumulated amortization at December 31, 2018	**831**
Net carrying value at	
December 31, 2017	9,853
December 31, 2018	**10,744**

3.2 PROPERTY, PLANT, AND EQUIPMENT

§ ACCOUNTING POLICIES

Property, plant, and equipment includes IT, lab and other equipment, furniture and leasehold improvements that 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 the asset is ready for use. The residual value of equipment is not material. Depreciation is calculated on a straight-line basis over the expected useful life of the asset, being 3-5 for equipment and furniture. Leasehold improvements are depreciated over the shorter of the useful life of the improvement or the remaining lease term. The useful life of assets and method of depreciation 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. In addition, the Company has fully depreciated equipment still in use.

Property, plant, and equipment is required to be tested for impairment when there are impairment indicators present. 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.

The following table presents the Company's Property, plant and equipment as of the years presented:

	Furniture and equipment TDKK	Leasehold improvements TDKK	Total TDKK
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
Additions	687	56	743
Disposals	-	-	-
Cost at December 31, 2018	**4,419**	**402**	**4,821**
Accumulated depreciation at December 31, 2016	1,609	493	2,102
Depreciation expense	574	53	627
Impairment	-	-	-
Disposals	-	(502)	(502)
Accumulated depreciation at December 31, 2017	2,183	44	2,227
Depreciation expense	578	76	654
Impairment	-	-	-
Disposals	-	-	-
Accumulated depreciation at December 31, 2018	**2,761**	**120**	**2,881**
Net carrying value at			
December 31, 2017	1,549	302	1,851
December 31, 2018	**1,658**	**282**	**1,940**

There has been no impairment of property, plant and equipment for the years ended December 31, 2018 and 2017. Depreciation expense is included within operating loss as follows:

	2018 TDKK	2017 TDKK
Research and development expenses	580	620
General and administrative expenses	74	7
Total depreciation expense	**654**	**627**

3.3 PREPAYMENTS, DEPOSITS AND OTHER RECEIVABLES

§ ACCOUNTING POLICIES

Prepayments
Prepayments include advance payments made to vendors that will be incurred and expensed in subsequent financial reporting periods. When the period for full expense recognition is longer than one year from the balance sheet date, the portion to be expensed subsequent to one year is classified as non-current.

Deposits
Deposits include advance payments made to vendors to be settled upon completion of the underlying contract. When the contract term is longer than one year from the balance sheet date, the deposit is classified as non-current.

Key estimate of prepayments related to clinical trial development costs
As explained in Note 2.1, Orphazyme incurs substantial costs associated with clinical trials related to its development programs and there is a high degree of estimation involved in accounting for clinical trial development costs. In particular, certain CROs and vendors are paid upfront in connection with clinical activities and Management is required to estimate the timing of the prepayment release to expense. This expense for the year is estimated by using an expense model, as described in Note 2.1.

Other receivables
Other receivables include current and non-current amounts due to the Company.

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.

The following items comprised Non-current Prepayments and Deposits as of December 31, 2018 and 2017:

	2018 TDKK	2017 TDKK
Deposits with vendors	1,266	-
Prepayments to vendors	633	-
Leasehold deposit	632	410
Total non-current Prepayments and Deposits	**2,531**	**410**

Non-current Prepayments and Deposits mainly includes a deposit with a CRO for advance payment of pass-through costs in connection with a clinical trial; prepaid insurance; and the lease deposit on our headquarters in Copenhagen.

Current Prepayments and Other Receivables are specified below:

	2018 TDKK	2017 TDKK
Prepayments to vendors	14,233	4,847
Grant income receivable	1,237	512
VAT receivable, net	1,468	5,216
Foreign VAT receivable	6,116	-
Other current receivables	124	143
Total current Prepayments and Other Receivables	**23,178**	**10,718**

Current Prepayments to vendors includes prepayments made to CROs for clinical trial costs of TDKK 8,572 (2017: TDKK 3,488).

3.4 FINANCIAL ASSETS AND LIABILITIES

§ ACCOUNTING POLICIES

Financial assets

Initial recognition and measurement

Financial assets that meet certain criteria are classified at initial recognition as subsequently measured at amortized cost, fair value through other comprehensive income (OCI), or fair value through profit or loss. The Group does not hold any financial assets meeting these classification criteria except cash and certain types of other receivables. Generally, the Company's financial assets are available to support current operations and amounts expected to be realized within the next twelve months are classified in the Statement of Financial Position as current assets.

The Group'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. 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.

Subsequent measurement

Historically, the Group's receivables are due within a twelve-month period and therefore the impact of using the effective interest rate method on the Group's financial statements has been immaterial.

Financial asset impairment

The Group assesses at the end of each reporting period whether there has been objective evidence that a financial asset may be impaired. Impairment losses are recognized if there is objective evidence of impairment and the evidence indicates that estimated future cash flows will be negatively impacted. The Group did not assess an impairment of a financial asset for either of the years ended December 31, 2018 or 2017.

Financial liabilities

Trade payables and accruals

Trade payables and accruals relate to the Group's purchase of products and services from various vendors in the normal course of business.

Other liabilities

Other payables are measured at net realizable value. The amount payable to employees for the Phantom Shares Program (Note 2.5) is classified as non-current and is measured at fair value.

Key estimate of accruals related to clinical trial development costs

As explained in Note 2.1, Orphazyme incurs substantial costs associated with clinical trials related to its development programs and there is a high degree of estimation involved in accounting for clinical trial development costs. As described in Note 2.1, Management uses an expense model to estimate the timing of expenses recognition in each period and related accruals at the end of the year.

The Group's financial assets include cash (Note 3.5) and other receivables related to Government Grants (Note 2.2) and Corporate Tax Receivable (Note 2.7). The Group 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, nor has the Group placed any assets as security for loans at either December 31, 2018 or 2017.

As of the years ended December 31, 2018 and 2017, the Group's financial liabilities comprise trade payables and accruals as follows:

	2018 TDKK	2017 TDKK
Trade payables	18,090	13,436
Accruals	24,093	26,796
Total Trade Payables and Accruals	**42,183**	**40,232**

3.4 FINANCIAL ASSETS AND LIABILITIES (CONTINUED)

As of the year ended December 31, 2018, Accruals includes an amount of TDKK 13,376 (2017: TDKK 8,520) for clinical trial costs.

As of the years ended December 31, 2018 and 2017, total other liabilities is comprised as follows

	2018 TDKK	2017 TDKK
Deferred grant income	299	520
Remuneration to the Board of Directors	1,836	113
Payroll and employee-related costs	8,677	6,251
Total other liabilities	**10,812**	**6,883**

Certain amounts included in prior year trade payables and accruals and other liabilities have been reclassified for consistency with current year presentation.

In addition, the Group has the following non-current liabilities as of the years ended December 31, 2018 and 2017:

	2018 TDKK	2017 TDKK
Accrual for milestone payment to vendor	66	-
Phantom shares liability to employees	39	-
Non-current Liabilities	**105**	**-**

3.5 CASH

§ ACCOUNTING POLICIES

Cash includes cash on hand and in banks that is subject to the risk of changes in value (Note 4.4).

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 Group's cash at the beginning and end of the year, including any effects of exchange rate changes.

Cash flows used in operating activities converts items in the Statement of Profit or Loss from the accrual basis of accounting to the cash basis of accounting. Non-cash items such as foreign exchange gains and losses, depreciation, amortization, and changes in working capital are reversed from the net loss for the year and actual cash receipts and payments are included.

Cash flows from investing activities shows payments related primarily to the purchase of licenses and property, plant, and equipment.

Cash flows from financing activities shows proceeds from share issuances, net of transaction costs, including the proceeds from Orphazyme's IPO on Nasdaq Copenhagen in 2017.

As of the years ended December 31, 2018 and 2017, the Group's cash balance denominated in foreign currencies was as follows:

	2018 TDKK	2017 TDKK
DKK	392,196	631,324
EUR	596	93
USD	1,886	288
GBP	28	29
Total cash	**394,706**	**631,735**

3.6 CONTRACTUAL OBLIGATIONS AND CONTINGENCIES

§ ACCOUNTING POLICIES

Leases

The Group is party to lease agreements only in which it is a lessee and not a lessor. A lease is classified at the inception date as a finance lease or an operating lease. Lease contracts where the lessor retains the significant risks and rewards associated with ownership of the asset are classified as operating leases. A lease that transfers substantially all the risks and rewards associated with ownership to the Group is classified as a finance lease. The Group has not entered into any finance leases. When entering into new contracts, the Group considers whether the arrangement contains a lease and should be accounted for as a lease.

An operating lease is a lease other than a finance lease. Operating lease payments are recognized as an operating expense in the Statement of Profit or Loss on a straight-line basis over the lease term. Lease payments related to facilities used for research purposes are recognized in Research and Development expenses. Lease payments related to facilities not used for research purposes are recognized in General and Administrative expenses.

On January 1, 2019, the Group adopted IFRS 16, *Leases* (Note 1.4).

For the years ended December 31, 2018 and 2017, the total lease expense recognized was TDKK 1,910 and TDKK 1,671, respectively.

Contractual obligations

The Group has entered into an operating lease for its headquarters location in Denmark with a non-cancelable lease term of six months, which as of December 31, 2018 established a contractual commitment of TDKK 1,333 (2017: TDKK 902). In addition, the Group has contractual obligations related to contracts with CROs and other vendors for research and development activities that have been initiated and are non-cancelable as of December 31, 2018, which establishes contractual commitments of approximately DKK 178 million (2017: DKK 48 million).

Contingent Assets and Contingent Liabilities

The Group will recognize assets or liabilities in the future that are dependent on achieving certain milestones that may or may not be in the Group's control.

Under the terms of the asset purchase agreement with CytRx described in Note 3.1, Orphazyme agreed to make future royalty payments to CytRx based on a specified percentage of any eventual net sales of products containing one of the compounds purchased. In addition, under the terms of the license agreement with KLSDC and UCL described in Note 3.1, 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 any products sold, which will exceed any royalty payments due to third parties. Orphazyme has no obligation to pay royalties prior to the occurrence of the sale of products and accordingly, neither a liability for the royalties to be paid nor contingent consideration to be received as part of the rights acquired has been recognized.

SECTION 4 – OTHER DISCLOSURES

Section 4 presents details of other disclosures relevant to the consolidated financial statements of the Group, including Capital Management, Equity, Earnings per Share, Financial Risk, Related Parties, Fees to Statutory Auditors, and Significant Events after the Reporting Period.

4.1 CAPITAL MANAGEMENT

For the purpose of the Group's capital management, capital includes issued capital, share premium and all other equity reserves attributable to the equity holders of the Group. The primary objective of the Group'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 in order to maintain investor, creditor and market confidence, and a continuous advancement of the Group's intellectual property, product pipeline, and business. Cash 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, 2018, the Group held cash totaling TDKK 394,706 that will be sufficient to provide adequate funding to allow the Group 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.

4.2 EQUITY

The following table summarizes the Company's share activity:

	Class A ordinary shares	Class B preferred shares	Class C preferred shares	Ordinary shares
December 31, 2016	125,000	2,050,208	1,185,333	-
Capital increases			1,741,669	
Conversion of shares prior to IPO	(125,000)	(2,050,208)	(2,927,002)	5,102,210
Non-cash bonus shares				6,487,882
Exercise of warrants				838,092
Initial Public Offering				7,500,000
December 31, 2017	**-**	**-**	**-**	**19,928,184**
Issuance of bonus shares as part of license agreement (Note 3.1)				11,380
December 31, 2018	**-**	**-**	**-**	**19,939,564**

4.2 EQUITY (CONTINUED)

In first quarter 2017, the Company completed a capital increase by issuing 534,007 Class C 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.

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

In preparation for the IPO, the capital structure of Orphazyme was changed in November 2017 by merging the three then-existing share classes (A, B and C) into one combined ordinary share class by issuance of bonus shares in order to replace the cancelled 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 ordinary 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.

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

In November 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.

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.

In January 2018, the Company issued 11,380 bonus shares ("2018 Bonus Shares") using free reserves of the Company to KU Center for Technology Commercialization, Inc., University of Kansas, Kansas Life Sciences Development Company, Inc. ("KLSDC"), and UCL Business PLC ("UCL") under the terms of the license agreement entered into in October 2017 (Note 3.1).

Following this share capital increase, the total nominal share capital of the Company as of December 31, 2018 was DKK 19,939,564, divided into 19,939,564 ordinary shares each with a nominal value of DKK 1.

4.3 LOSS PER SHARE

Basic loss per share for the year is calculated by dividing the net loss for the year by the weighted average number of ordinary shares outstanding during the year. The diluted loss per share is calculated by dividing the net loss for the year by the weighted average number of ordinary shares outstanding during the period increased by the dilutive effect of the assumed issuance of outstanding share-based awards. As a result of the Group incurring losses for each of the years ended December 31, 2018 and 2017, the potential shares issuable related to outstanding share-based awards have been excluded from the calculation of diluted per share amounts, as the effect of such shares is anti-dilutive.

On November 2, 2017, in preparation for the IPO, the capital structure of Orphazyme was adjusted by merging the three then-existing share classes (A, B and C) into one combined ordinary share class by issuance of 6,487,882 bonus shares ("2017 Bonus Shares") in order to replace the cancelled preference shares.

In January 2018, the Company issued 11,380 bonus shares ("2018 Bonus Shares") to KLSDC and UCL under the terms of the license agreement entered into in October 2017 with the parties (Note 3.1).

In January 2019, the Company issued 26.050 bonus shares ("2019 Bonus Shares") to KLSDC and UCL under the terms of the same license agreement mentioned above.

Basic and diluted loss per share for the years presented have been adjusted retrospectively to include the 2017 Bonus Shares, the 2018 Bonus Shares, and the 2019 Bonus Shares in the number of weighted average shares outstanding for the years ended December 31, 2018 and 2017.

4.3 LOSS PER SHARE (CONTINUED)

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 years ended December 31, 2018 and 2017:

	2018	2017
Net loss for the year (TDKK)	(229,600)	(126,204)
Weighted-average shares outstanding	19,965,614	12,101,543
Loss per share	**(11.50)**	**(10.43)**

4.4 FINANCIAL RISKS

The Group's activities expose it to a number of financial risks whereby future events, which can be outside the control of the Group, could have a material effect on its 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 Group has not historically hedged its financial risks.

Liquidity Risk

The Group's liquidity risk is assessed to be low.
Management continuously assesses the Group's capital structure and whether its liquidity reserves will allow it to achieve the business objectives. Following the IPO in November 2017, the Group's cash reserves have allowed it to continue to advance its development activities.

Foreign Currency Risk

The Group's foreign currency risk is assessed to be high.
The Group's functional currency is the DKK and it conducts cross border transactions where the functional currency is not always used. Accordingly, future changes in the exchange rates of the DKK against the EUR, the USD and/or the GBP will

4.4 FINANCIAL RISKS (CONTINUED)

expose the Group to currency gains or losses that will impact the reported amounts of assets, liabilities, income and expenses and the impact could be material.

Interest Rate Risk

The Group's interest rate risk is assessed to be low.
The Group has no interest-bearing debt. Due to the current interest level in Denmark, the Group incurs negative interest on bank deposits.

Credit Risk

The Group's credit risk is assessed to be low.
The Group'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 is held primarily at two banks in Denmark with Moody's long-term credit ratings exceeding of A1.

The Group has prepared a sensitivity analysis in order to assess the potential impact on the Group's net loss for possible fluctuations in the EUR, USD, and GBP exchange rates against the DKK and the impact for the possible fluctuations in the interest rate on bank deposits in Denmark and in the U.S. The methods and assumptions used are consistent with prior year and consider increases and decreases in the Group's three main currencies, as well as reasonable fluctuations in the interest rate on its bank deposits. Based on the sensitivity analyses, the impact on the Group's net loss for the year ended December 31, 2018 would be as follows:

Currency	Currency fluctuation	Effect 2018 TDKK	Effect 2017 TDKK
EUR	+/- 2%	713	215
USD	+/-10%	1,278	936
GBP	+/-10%	218	358

Cash deposits	Interest rate fluctuation	Effect 2018 TDKK	Effect 2017 TDKK
DKK	+/- 1%	20	6313
EUR	+/- 1%	-	1
USD	+/- 1%	-	-
GBP	+/- 1%	-	3

4.5 RELATED PARTIES

Orphazyme A/S, incorporated in Denmark, is the ultimate parent company of the Group, which wholly owns Orphazyme US, Inc. These two entities are considered related parties. Orphazyme A/S is not ultimately controlled by any of its investors. Major investors owning more than 10% of the Company are considered related parties.

For the years ended December 31, 2018 and 2017, the following related party transactions were identified:

- Remuneration to Executive Management (Note 2.4)
- Remuneration to the Board of Directors (Note 2.4)
- Capital increases in 2017 (Note 4.2)
- Consulting services from a shareholder in the amount of TDKK 355 in 2018.
- Consulting services from board members in the amount of TDKK 372 in 2017.

As of December 31, 2018 and 2017, the Company did not have any amounts receivable from related parties and therefore has not recorded any impairment. 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.

None of the board members have been granted share-based compensation awards for 2018 or 2017.

For the years ended December 31, 2018 and 2017, Executive Management and members of the Board of Directors had the following shareholding in Orphazyme A/S:

	Number of shares owned 2018	Number of shares owned 2017	Investment Shares (LTIP) 2018	Investment Shares (LTIP) 2017
Anders Hinsby	204,596	204,596	5,000	5,000
Anders Vadsholt	127,806	127,806	4,000	4,000

Members of the Board of Directors:	Number of shares 2018	Number of shares 2017
Georges Gemayel	87,758	87,758
Bo Jesper Hansen	79,945	79,945
Martijn Kleijwegt	-	-
Martin Bonde	46,009	46,009
Martin Rahbek Kornum[1]	-	41,476
Rémi Droller	-	-
Sten Verland	-	-
Anders Hedegaard	6,250	6,250
Cathrine Moukheibir	7,980	7,980

[1] Not a member of the Board of Directors as of December 31, 2018.

4.6 FEES TO STATUTORY AUDITORS

The following table presents the fees to our statutory auditors, Ernst & Young Godkendt Revisionspartnerselskab, recognized in general and administrative expenses in the Statement of Profit or Loss for the years ended December 31, 2018 and 2017:

	2018 TDKK	2017 TDKK
Audit fee for statutory audit	320	250
Fee for statutory audit, previous year	-	142
Other assurance engagements	156	30
Other assistance	50	2,735
Total fees to statutory auditors	526	3,157

4.7 SIGNIFICANT EVENTS AFTER THE REPORTING PERIOD

As described in Note 3.1, as part of the license agreement with KLSDC and UCL, consideration to KLSDC and UCL is payable in shares of the Company ("Bonus Shares") each January and is based on incurred costs reported by KLSDC and UCL for the previous year. As at December 31, 2018 the aggregate costs incurred by KLSDC and UCL amounted to USD 190 thousand (TDKK 1,197), and a total of 26,060 Bonus Shares ("2019 Bonus Shares") were issued to KLSDC and UCL in January 2019, based on the average 30-day closing price of Orphazyme's shares. In addition, at the time of the share issuance the equity reserve was decreased by TDKK 1,197, which represents the market value of the shares issued.

Other than the issuance of the 2019 Bonus Shares, there have been no other significant events after December 31, 2018.

ORPHAZYME

2018 PARENT COMPANY FINANCIAL STATEMENTS

STATEMENTS OF PROFIT OR LOSS AND OTHER COMPREHENSIVE INCOME

PARENT COMPANY

For the years ended December 31

Note		2018 TDKK	2017 TDKK
2.1*	Research and development expenses	(196,525)	(99,048)
2.1	General and administrative expenses	(35,127)	(31,994)
	Operating loss	**(231,652)**	**(131,042)**
2.6*	Financial income	5	26
2.6*	Financial expenses	(3,453)	(688)
	Loss before tax	**(235,100)**	**(131,704)**
2.7*	Income tax benefit	5,500	5,500
	Net loss for the period	**(229,600)**	**(126,204)**
	Items that will be reclassified subsequently to the Statement of Profit or Loss:		
	Exchange difference from translation of foreign operation, net of tax DKK 0	(51)	-
	Total comprehensive loss	**(229,651)**	**(126,204)**
	Loss per share, basic and diluted	(11.50)	(10.46)

*Please refer to respective notes in the Consolidated Financial Statements of the Group

STATEMENTS OF FINANCIAL POSITION

PARENT COMPANY

As of December 31

Note		2018 TDKK	2017 TDKK
	ASSETS		
	Non-current assets		
3.1*	Licenses	10,744	9,853
3.2*	Property, plant, and equipment	1,940	1,851
2.7*	Corporation tax receivable	2,750	2,750
3.3*	Prepayments and deposits	2,531	410
2.3	Investment in subsidiary	1,207	
	Total non-currents assets	**19,172**	**14,864**
	Current assets		
2.7*	Corporation tax receivable	5,500	5,500
2.4	Prepayments and other receivables	23,081	10,718
2.6	Cash	393,123	631,735
	Total current assets	**421,704**	**647,953**
	TOTAL ASSETS	**440,876**	**662,817**
	EQUITY AND LIABILITIES		
	Equity		
4.2*	Share capital	19,939	19,928
4.2*	Share premium	924,021	924,021
	Other reserves	9,019	9,972
	Accumulated deficit	(564,823)	(338,219)
	Total equity	**388,156**	**615,702**
	Non-current liabilities		
2.5	Other non-current liabilities	105	-
2.5	Intercompany payables	826	
	Total non-current liabilities	**931**	**-**
	Current liabilities		
2.5	Trade payables and accruals	40,977	40,232
2.5	Other liabilities	10,812	6,883
	Total current liabilities	**51,789**	**47,115**
	TOTAL EQUITY AND LIABILITIES	**440,876**	**662,817**

*Please refer to respective notes in the Consolidated Financial Statements of the Group

STATEMENTS OF CHANGES IN SHAREHOLDERS' EQUITY

PARENT COMPANY

Note		Share capital TDKK	Share premium TDKK	Foreign Currency Translation Reserve TDKK	Share-based compensation - acquistion of intangible assets TDKK	Accumulated deficit TDKK	Total TDKK
	Balance as of December 31, 2016	**3,361**	**226,285**	**-**	**-**	**(212,137)**	**17,509**
	Net loss for the year					(126,204)	(126,204)
	Other comprehensive loss						
	Total other comprehensive income/(loss)			-	-	(126,204)	(126,204)
	Transactions with owners:						
3.1*	Contribution of a license agreement						
2.5*	Share issue in connection with conversion of former preference shares into ordinary shares	6,488	(6,488)				-
4.2*	Proceeds from IPO	7,500	592,500				600,000
2.3*	Costs related to IPO	-	(42,605)				(42,605)
4.2*	Capital increase	1,741	155,010				156,751
2.5*	Exercise of warrants for cash	838	323				1,161
4.2*	Costs related to non-IPO related capital increases	-	(1,004)				(1,004)
2.5*	Share-based compensation expense					122	122
	Total transaction with owners	16,567	697,736	-	9,972	122	724,397
	Balance as of December 31, 2017	**19,928**	**924,021**	**-**	**9,972**	**(338,219)**	**615,702**
	Net loss for the year					(229,600)	(229,600)
	Other comprehensive loss			(51)		-	(51)
	Total other comprehensive loss			(51)	-	(229,600)	(229,651)
	Transactions with owners:						
3.1*	Capital increase in connection with issuance of Bonus Shares	11			(902)	891	-
2.5*	Share-based compensation expense					2,105	2,105
	Total transaction with owners	11	-	-	(902)	2,996	2,105
	Balance as of December 31, 2018	**19,939**	**924,021**	**(51)**	**9,070**	**(564,823)**	**388,156**

*Please refer to respective notes in the Consolidated Financial Statements of the Group

STATEMENTS OF CASH FLOWS

PARENT COMPANY

For the years ended December 31

Note		2018 TDKK	2017 TDKK
	Operating loss	(231,652)	(131,042)
	Reversal of non-cash items:		
2.5*	Equity-settled share-based compensation expense	2,105	122
3.1*, 3.2*	Depreciation and amortization	1,366	627
	Exchange rate adjustments	(491)	-
	Gain (Loss) on sale or disposal of assets	-	119
	Change in working capital:		
2.4	Change in Prepayments, deposits, and other receivables	(14,484)	(2,778)
2.5	Change in Trade payables and accruals	745	32,688
2.5	Change in Other liabilities	4,034	-
2.5	Change in Intercompany payables	825	-
2.7*	Corporation taxes received	5,500	5,500
2.6*	Interest received (paid)	(2,957)	(662)
	Net cash used in operating activities	**(235,009)**	**(95,426)**
	Investing activities		
3.1*	Purchase of intangible assets	(1,603)	-
3.2*	Purchase of property, plant and equipment	(743)	(1,491)
	Capital increase in subsidiaries	(1,207)	-
	Net cash used in investing activities	**(3,553)**	**(1,491)**
	Financing activities		
4.2*	Proceeds from IPO	-	600,000
4.2*	Costs related to IPO	-	(42,605)
4.2*	Capital contributions from shareholders	-	157,912
4.2*	Costs related to capital contributions	-	(1,004)
	Net cash provided by financing activities	**-**	**714,303**
	Net change in cash	**(238,562)**	617,386
	Effects of changes in exchange rates	(50)	-
	Cash at the beginning of the year	631,735	14,349
	Cash at the end of the year	**393,123**	**631,735**

*Please refer to respective notes in the Consolidated Financial Statements of the Group

SECTION 1 – BASIS OF PREPARATION

The financial statements of Orphazyme A/S (the "Parent Company") have been prepared in accordance with International Financial Reporting Standards, or IFRS, as adopted by the EU and additional disclosure requirements under the Danish Financial Statements Act. The Parent Company financial statements of Orphazyme A/S for the year ended December 31, 2018 were approved by the Board of Directors on March 1, 2019 and will be submitted to the shareholders of Orphazyme A/S for approval at the Annual General Meeting to be held on March 27, 2019.

1.1 CORPORATE INFORMATION

Orphazyme A/S is a limited liability company incorporated and domiciled in Denmark. The registered office is located in Copenhagen, Denmark. On November 16, 2017, the Company successfully completed its Initial Public Offering (IPO) on Nasdaq Copenhagen by issuing 7,500,000 new ordinary shares for gross proceeds of TDKK 600,000.

In April 2018, a fully-owned subsidiary, Orphazyme US, Inc., was incorporated in Massachusetts, USA. Orphazyme US, Inc. will directly support the US market to establish closer relationships with the medical, patient, and financial communities as Orphazyme expands its development programs and global reach.

1.2 SIGNIFICANT ACCOUNTING POLICIES APPLICABLE TO THE PARENT COMPANY

As applicable to the Orphazyme A/S, the Parent Company applies the same accounting policies as disclosed in the Group's consolidated financial statements. Therefore, only accounting policies specific to the Parent Company or that differ from the accounting policies applied by the Group are disclosed in these notes to the financial statement. If an accounting policy is not specifically mentioned, the Group accounting policy is applied.

A description of Management's key accounting estimates and judgements as well as new IFRS standards are disclosed in the Group financial statements and also apply to the Parent Company.

The Parent Company financial statements are presented in Danish Kroner, or DKK, which is both the functional and presentation currency of the Parent Company. Where indicated, amounts are rounded to the nearest thousand, or TDKK.

SECTION 2 – NOTES

The notes applicable to the financial statements of the Parent Company are the same as those presented in the Group Consolidated Financial Statements, except for those notes presented in this Section 2.

2.1 GENERAL AND ADMINISTRATIVE EXPENSES

	2018	2017
External costs	12,065	8,445
Intercompany expenses	5,510	-
IPO costs	-	13,456
Employee Costs (Note 2.2)	11,960	7,023
Travel and related expenses	3,893	3,063
Pre-Commercial activities	1,625	-
Depreciation	74	7
Total General and Administrative Expenses	**35,127**	**31,994**

Intercompany expenses includes general and administrative expenses incurred on behalf of the US subsidiary.

2.2 EMPLOYEE COSTS

	2018 TDKK	2017 TDKK
Employee Costs, excluding Executive Management and Board		
Salaries	36,324	21,631
Cash bonus	2,243	3,618
Share-based compensation	1,006	44
Pensions	2,686	968
Other social security contributions	322	177
Other staff costs	881	338
Total employee costs, excluding Executive Management and Board	**43,462**	**26,776**
Executive Management remuneration		
Salaries	3,328	2,759
Cash bonus	1,173	1,473
Share-based compensation	1,139	78
Pensions	372	-
Other social security contributions	4	2
Other staff costs	-	-
Total Executive Management remuneration	**6,016**	**4,312**
Board of Directors remuneration	**2,763**	**1,583**
Total Employee Costs	**52,241**	**32,671**
Recognized as follows in the Statement of Profit or Loss:		
Research and development expenses	40,281	25,648
General and administrative expenses	11,960	7,023
Total Employee Costs	**52,241**	**32,671**
Average number of full-time employees	**44**	**26**
Year-end number of full-time employees	**55**	**34**

2.2 EMPLOYEE COSTS (CONTINUED)

	2018 TDKK	2017 TDKK
Board and Committee fees	2,584	1,211
Travel allowance	179	-
Other fees	-	372
Total Board of Directors remuneration	**2,763**	**1,583**

2.3 INVESTMENT IN GROUP COMPANIES

§ ACCOUNTING POLICIES

Investments in subsidiaries are measured in the Parent Company financial statements at the lower of cost or recoverable amount. Any distributed dividends are recognized in the income statmement of the Parent Company.

	2018 TDKK	2017 TDKK
January	-	-
Cost at December 31	**1,207**	-

	Registered office	Ownership interest (%)	Share capital (TUSD)	Equity (TUSD)	Net profit (TUSD)
Orphazyme US, Inc.	Delaware, USA	100	200	312	-

2.4 PREPAYMENTS, DEPOSITS AND OTHER RECEIVABLES

	2018 TDKK	2017 TDKK
Deposits with vendors	1,266	-
Prepayments to vendors	633	-
Leasehold deposit	632	410
Total non-current prepayments and deposits	**2,531**	**410**

	2018 TDKK	2017 TDKK
Prepayments to vendors	14,135	4,847
Grant income receivable	1,237	512
VAT receivable, net	1,468	5,216
Foreign VAT receivable	6,116	-
Other current receivables	125	143
Total current prepayments and other receivables	**23,081**	**10,718**

2.5 FINANCIAL ASSETS AND LIABILITIES

	2018 TDKK	2017 TDKK
Trade payables	17,531	13,436
Accruals	23,446	26,796
Total trade payables and accruals	**40,977**	**40,232**

	2018 TDKK	2017 TDKK
Deferred grant income	299	519
Remuneration to the Board of Directors	1,836	113
Payroll and employee-related	8,677	6,251
Total other liabilities	**10,812**	**6,883**

	2018 TDKK	2017 TDKK
Accrual for milestone payment to vendor	66	-
Phantom shares liability to employees	39	
Intercompany payables	826	-
Total non-current liabilities	**931**	**-**

2.6 CASH

	2018 TDKK	2017 TDKK
DKK	392,196	631,324
EUR	596	93
USD	303	288
GBP	28	30
Total cash	**393,123**	**631,735**

ORPHAZYME

STATEMENTS AND SIGNATURES

STATEMENT BY THE BOARD OF DIRECTORS AND EXECUTIVE MANAGEMENT

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, 2018.

The consolidated financial statements of the Group and the Parent Company's financial statements have been prepared in accordance with International Financial Reporting Standards (IFRS) as issued by the International Accounting Standards Board (IASB) and in accordance with IFRS as endorsed by the EU as well as additional disclosure requirements under the Danish Financial Statements Act.

In our opinion, the Group's consolidated financial statements and the Parent Company financial statements provide a fair presentation of the assets, liabilities, and financial position at December 31, 2018 and of the results of the Group's and Parent Company's operations and cash flows for the financial year January 1–December 31, 2018.

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

We recommend that the Annual Report be adopted at the Annual General Meeting on March 27, 2019.

BOARD OF DIRECTORS

Georges Gemayel
Chairman of the Board

Bo Jesper Hansen
Deputy Chairman of the Board

Martin Bonde

Anders Hedegaard

Rémi Droller

Martin Kleijwegt

Sten Verland

Catherine Moukheibir

EXECUTIVE MANAGEMENT

Anders Hinsby
Chief Executive Officer, Co-Founder

Anders Vadsholt
Chief Financial Officer

INDEPENDENT AUDITORS' REPORT

TO THE SHAREHOLDERS OF ORPHAZYME A/S

OPINION

We have audited the consolidated financial statements and the parent company financial statements of Orphazyme A/S for the financial year January 1 – December 31, 2018, which comprise statement of profit or loss and other comprehensive income, statement of financial position, statement of changes in shareholders' equity, statement of cash flow and notes, including accounting policies, for the Group and the Parent Company. The consolidated financial statements and the parent company financial statements are prepared in accordance with International Financial Reporting Standards (IFRS) as issued by the International Accounting Standards Board (IASB) and in accordance with IFRS as endorsed by the EU, as well as additional disclosure requirements under the Danish Financial Statements Act.

In our opinion, the consolidated financial statements and the parent company financial statements give a true and fair view of the financial position of the Group and the Parent Company at December 31, 2018 and of the results of the Group's and the Parent Company's operations and cash flows for the financial year January 1 – December 31, 2018 in accordance with International Financial Reporting Standards (IFRS) as issued by the International Accounting Standards Board (IASB) and in accordance with IFRS as endorsed by the EU as well as additional disclosure requirements under 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 of the consolidated financial statements and the parent company financial statements" (hereinafter collectively referred to as "the 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 Group 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

We were initially appointed as auditor of Orphazyme A/S on December 4, 2015 for the financial year July 1 to December 31, 2015. We have been reappointed annually by resolution of the general meeting for a total consecutive period of 4 years up until the financial year 2018.

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 2018. 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	How our audit addressed the key audit matter
Accrual for costs incurred through Clinical Research Organisations (CROs) *Orphazyme has entered into several clinical research contracts with Clinical Research Organisations (CROs) that perform research and development for the Company.* *Due to the materiality of these arrangements and since they cover multiple periods, recognition of the transactions in the correct period requires Management to make significant estimates.* *We focused on this area due to materiality and because the arrangements and the related accounting treatment of accruals accounting for CRO costs require significant estimation by Management.* *Refer to Note 2.1 and 3.4 to the Consolidated Financial Statements.*	*We have evaluated relevant processes including Management's controls to ensure that CRO contracts are recognised and measured appropriately on an ongoing basis.* *We obtained Management's calculations for CRO accruals at year-end and reconciled inputs and key assumptions to underlying terms and conditions in the agreements and relevant internal and external sources – and assessed the accuracy of the accruals for CRO services received – including assessed the accuracy of the accruals in previous periods. Furthermore, the CRO accrual models have been tested for clerical accuracy.* *We assessed whether the disclosures in relation to CRO accruals were appropriate and met the requirements of the 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 consolidated financial statements and parent company financial statements that give a true and fair view in accordance with International Financial Reporting Standards (IFRS) as issued by the International Accounting Standards Board (IASB) and in accordance with IFRS as endorsed by the EU as well as additional disclosure requirements under 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 Group's and the Parent 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 Group or the Parent 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 Group's and the Parent 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 Group's and the Parent 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 Group and the Parent 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.
- Obtain sufficient appropriate audit evidence regarding the financial information of the entities or business activities within the Group to express an opinion on the consolidated financial statements. We are responsible for the direction, supervision and performance of the group audit. We remain solely responsible for our audit opinion.

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 consolidated financial statements and the parent company 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 1, 2019
Ernst & Young
Godkendt Revisionspartnerselskab
CVR no. 30 70 02 28

Christian Schwenn Johansen
State Authorised Public Accountant
mne33234

Lars Hansen
State Authorised Public Accountant
mne24828

Orphazyme A/S
Ole Maaløes Vej 3
DK-2200 Copenhagen N

Approval at Annual General Meeting (AGM): March 27, 2019
Chairman of AGM: Rikke Schiøtt Petersen, Gorrissen Federspiel Advokatpartnerselskab