# Orphazyme ApS

Ole Maaløes Vej 3, 2200 Copenhagen N CVR no. 32 26 63 55

Annual report for the period January 1 – December 31, 2016

Approved at the annual general meeting of shareholders on May 20, 2017 Chairman:

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# Statement by the Board of Directors and the Executive Board

The Board of Directors and the Executive Board have today discussed and approved the annual report of Orphazyme ApS for the financial year January 1 - December 31, 2016.

The annual report is prepared in accordance with IFRS as adopted in EU and additional Danish Disclosure requirements.

In our opinion, the financial statements give a true and fair view of the Company's financial position at December 31, 2016 and of the results of the Company's operations and cash flows for the financial year January 1 - December 31, 2016.

Further, in our opinion, the Management's review gives a fair review of the matters discussed in the Management's review.

We recommend that the annual report be approved at the annual general meeting.

Copenhagen, Executive Board:

Anders Mørkeberg Hinsb

Board of Directors:

Georges Gemayel Chairman

Nanna Liebach Lüneborg

Patrick Johan Hendrik Krol

Martin Bond

Bo Jesper Hansen

Rémi Pascal Louis Droller

Martin Rahbek Kornum



#### Independent auditor's report

To the shareholders of Orphazyme ApS

#### Opinion

We have audited the financial statements of Orphazyme ApS (the Company) for the financial year January 1 – December 31, 2016 with comparative figures for the financial years July 1 – December 31, 2015 and July 1, 2014 - June 30, 2015 (the "financial statements"), which comprise a statement of profit or loss and other comprehensive income, statement of financial position, statement of changes in shareholders' equity, statement of cash flows and notes, including accounting policies. The financial statements are prepared in accordance with International Financial Reporting Standards as adopted by the EU and additional requirements of the Danish Financial Statements Act.

In our opinion, the financial statements give a true and fair view of the financial position of the Company at December 31, 2016 with comparative figures at December 31, 2015 and June 30, 2015 and of the results of the Company's operations and cash flows for the financial year January 1 – December 31, 2016 with comparative figures for the financial years July 1 – December 31, 2015 and July 1, 2014 – June 30, 2015 in accordance with International Financial Reporting Standards as adopted by the EU and additional requirements of the Danish Financial Statements Act.

#### 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 financial statements" section of our report. We are independent of the Company in accordance with the International Ethics Standards Board for Accountants' Code of Ethics for Professional Accountants (IESBA Code) and additional requirements applicable in Denmark, and we have fulfilled our other ethical responsibilities in accordance with these rules and requirements. We believe that the audit evidence we have obtained is sufficient and appropriate to provide a basis for our opinion.

## 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 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 our procedures, 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 misstatements of the Management's review.

#### Management's responsibilities for the financial statements

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



#### Independent auditor's report

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

#### Auditor's responsibilities for the audit of the financial statements

Our objectives are to obtain reasonable assurance about 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 skepticism throughout the audit. We also:

- Identify and assess the risks of material misstatement of the financial statements, whether due to fraud or error, design and perform audit procedures responsive to those risks and obtain audit evidence that is sufficient and appropriate to provide a basis for our opinion. The risk of not detecting a material misstatement resulting from fraud is higher than for one resulting from error, as fraud may involve collusion, forgery, intentional omissions, misrepresentations or the override of internal control.
- Obtain an understanding of internal control relevant to the audit in order to design audit procedures that are appropriate in the circumstances, but not for the purpose of expressing an opinion on the effectiveness of the Company's internal control.
- Evaluate the appropriateness of accounting policies used and the reasonableness of accounting estimates and related disclosures made by Management.
- Conclude on the appropriateness of Management's use of the going concern basis of accounting in preparing the financial statements and, based on the audit evidence obtained, whether a material uncertainty exists related to events or conditions that may cast significant doubt on the Company's ability to continue as a going concern. If we conclude that a material uncertainty exists, we are required to draw attention in our auditor's report to the related disclosures in the financial statements or, if such disclosures are inadequate, to modify our opinion. Our conclusion is based on the audit evidence obtained up to the date of our auditor's report. However, future events or conditions may cause the Company to cease to continue as a going concern.
- Evaluate the overall presentation, structure and contents of the financial statements, including the note disclosures, and whether the financial statements represent the underlying transactions and events in a manner that gives a true and fair view.



## Independent auditor's report

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.

Copenhagen, May 30, 2017 Ernst & Young Godkendt Revisionspartnerselskab

Christian Schwenn Johansen

State Authorised Public Accountant Lars Hansen State Authorised Public Accountant

## Management's review

## Company details

Name

Orphazyme ApS

Address, Postal code, City

Ole Maaløes Vej 3, 2200 Copenhagen N, Denmark

CVR No. Established Registered office Financial year

32 26 63 55 19 June 2009

Copenhagen January 1 - December 31

Website E-mail

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**Board of Directors** 

Georges Gemayel, Chairman Patrick Johan Hendrik Krol Rémi Pascal Louis Droller Nanna Liebach Lüneborg

Martin Bonde

Martin Rahbek Kornum

Sten Verland Bo Jesper Hansen

**Executive Board** 

Anders Mørkeberg Hinsby

Auditors

Ernst & Young Godkendt Revisionspartnerselskab

Osvald Helmuths Vej 4, PO Box 250, 2000 Frederiksberg,

Denmark

Bankers

Dansk Bank Nordea

#### **About Orphazyme ApS**

Orphazyme ApS is a Danish biotech company developing paradigm-changing medicines for the treatment of protein misfolding diseases with well-characterised mechanisms of disease.

The lead program is in development as a treatment for neuromuscular and lysosomal storage diseases. The lead programme is in Phase 3 clinical development in a lysosomal storage disease called Niemann-Pick disease type C and late stage clinical trials in two protein aggregation diseases: sporadic inclusion body myositis and SOD1-ALS in 2017 and early 2018, respectively.

During 2016 Orphazyme expanded its management team by the hiring of Dr Thomas Blaettler and Mr Anders Vadsholt as company Chief Medical Officer and Chief Financial Officer, respectively.

Orphazyme is owned by an investor syndicate comprising Novo A/S, Sunstone Capital, Aescap Venture, Kurma Partners, LSP, ALS Investment Fund and Idinvest Partners. For more information, please visit www.orphazyme.com.

#### **About Arimoclomol**

Arimoclomol is a new chemical entity with a very favorable safety and tolerability record in humans: Seven Phase I clinical studies have been conducted in healthy volunteers. Arimoclomol is administered orally, three times daily and can be easily dissolved in liquids or food for best possible patient comfort and compliance.

## **Operating review**

2016 was a busy and exciting year for Orphazyme.

In the beginning of June 2016 the U.S. Food and Drug Administration (FDA) gave clearance to use our drug arimoclomol in a study as a new treatment for Niemann-Pick disease type C. Arimoclomol is a new chemical entity with a very favorable safety and tolerability record in humans.

End of June 2016 FDA granted a Fast Track designation for the study. A fast track program is a process designed to facilitate the development, and expedite the review of drugs to treat serious conditions and fill an unmet medical need. The purpose is to get important new drugs to the patient earlier.

Late October 2016 the multi-centre, double-blind, placebo-controlled interventional study was expanded to cover three sites in the USA. Today there are 15 sites across Europe and the USA. The full enrolment was completed in 2017.

The primary endpoint of the study is the change in disease severity in patients treated with arimoclomol compared to placebo, which, supported by biochemical readouts, will provide the basis for an application for market authorisation at the end of the study. The trial is estimated to complete in the second quarter

In December 2016, the results of a phase II trial with patients having rapidly progressive ALS due to mutations in the SOD1 gene, was published. The results showed promise for the treatment of ALS. Patients were treated with Arimoclomol for up to 12 months. The primary outcome was safety and tolerability; secondary outcome was efficacy, measured in terms of survival and functional decline.

On the organizational plan, Dr Blaettler MD was appointed as a new Chief Medical Officer and Mr. Vadsholt was appointed Chief Financial Officer. Dr Blaettler is a certified neurologist with extensive clinical development and leadership experience, most recently from Roche. Mr. Vadsholt is a senior biotech executive, who previously was CEO and CFO of Topotarget A/S. With Thomas Blaettler and Anders Vadsholt now on board, we will be well equipped to design and implement long-term clinical and financial strategies.

When we began our company, our focus was on making a positive difference for patients with rare diseases with a high unmet need. In line with this, we are today working to create new therapeutic approaches for a number of different diseases, including our lead candidate within Niemann-Pick type C disease, but also diseases like Gaucher's disease, sporadic Inclusion Body Myositis (sIBM) and Amyotrophic Lateral Sclerosis (ALS). We look forward to report on our progress in order to dramatically improve the life of patients.

## Financial review

The income statement for the twelve months ended December 31, 2016 shows a loss of MDKK 57.9 against a loss of MDKK 26.7 for the six months ended December 31, 2015 and a loss of MDKK 32.6 for the twelve months ended June 30, 2015. The balance sheet at December 31, 2016 shows equity of MDKK 17.5 compared to MDKK 74.1 for the six months ended December 31, 2015 and MDKK 89.4 for the twelve months ended June 30, 2015.

#### Post balance sheet events

Subsequent to December 31, 2016, the company extended its 2015 financing round by issuing new shares in exchange for cash for a total of MDKK 48. Please see Note 16 for further details of this transaction.

On March 8, 2017 the Company further completed a MDKK 104 financing round by issuing new shares to LSP V Coöperatieve U.A. and ALS Investment Fund to further expand the clinical development program. The current financing will support clinical development in two additional protein misfolding diseases: Gaucher disease and SOD1-ALS.

In March 2017 enrolment of patients completed for phase 3 clinical trials in Niemann-Pick Type C Disease.

# Statement of Profit or Loss and Other Comprehensive Income

Note		Twelve months ended December 31, 2016 TDKK	Six months ended December 31, 2015 TDKK	Twelve months ended June 30, 2015 TDKK
4,5,9	Research and development	(55,817)	(25,478)	(31,604)
5,9	expenses Administrative expenses	(7,703)	(4,044)	(5,494)
0.7	Operating loss	(63,520)	(29,522)	(37,098)
7	Financial income	182	80	34
8	Financial expenses	(97)	(40)	(1,403)
	Loss before tax	(63,435)	(29,482)	(38,467)
11	Income tax benefit	5,500	2,750	5,875
	Net loss for the period	(57,935)	(26,732)	(32,592)
	Other comprehensive income/(loss)	-		
	Total comprehensive loss	(57,935)	(26,732)	(32,592)
12	Loss per share, basic and diluted			
	Class C preferred shares	(16.71)	(7.98)	(11.15)
	Class B preferred shares	(17.12)	(8.20)	(12.53)
	Class A ordinary shares	(26.08)	(12.47)	(20.29)

See accompanying notes to these financial statements.

# **Statement of Financial Position**

Note		December 31, 2016 TDKK	December 31, 2015 TDKK	June 30, 2015 TDKK	July 1, 2014 TDKK
	ASSETS				
,	Non-current assets				
9	Property, plant and equipment	987	1,487	1,748	1,694
11	Corporation tax receivable	2,750	2,750	5,875	-
	Deposits	310	211	184	179
	Total non-current assets	4,047	4,448	7,807	1,873
	Current assets				
11	Corporation tax receivable	5,500	5,875	6,250	7,500
	Other receivables	3,421	644	408	269
	Prepayments	4,624	5,971	2,721	190
	Cash and cash equivalents	14,349	68,014	78,161	25,732
	<b>Total current assets</b>	27,894	80,504	87,540	33,691
	TOTAL ASSETS	31,941	84,952	95,347	35,564
	EQUITY AND LIABILITIES				
	Equity				
10	Share capital	3,361	3,346	3,218	2,175
10	Share premium	226,285	224,999	213,632	124,250
	Accumulated deficit	(212,137)	(154,202)	(127,470)	(94,878)
	Total equity	17,509	74,143	89,380	31,547
	Current liabilities				
	Bank debt	-	-	245	1
	Trade payables	4,718	2,447	955	997
	Other payables	9,714	8,362	4,767	3,019
	Total current liabilities	14,432	10,809	5,967	4,017
	TOTAL EQUITY AND LIABILITIES	31,941	84,952	95,347	35,564

<sup>1</sup> Accounting policies

See accompanying notes to these financial statements.

<sup>14</sup> Contractual obligations and contingencies

Statement of Changes in Shareholders' Equity

	Note	Share capital TDKK	Share premium TDKK	Accumulated deficit TDKK	Total TDKK
Balance as of July 1, 2014		2,175	124,250	(94,878)	31,547
Opening effects from conversion to IFRS	17		E.	0	1
Net loss for the period		1		(32,592)	(32,592)
Other comprehensive loss for the period		1		1	ı
Transactions with owners					
Capital increase		827	73,618	1	74,445
Conversion of debt		216	15,949	1	16,165
Expenses, capital increase		ī	(182)	1	(185)
Share-based payment expense	9	•	•		ì
Balance as of June 30, 2015		3,218	213,632	(127,470)	89,380
Net loss for the period			t	(26,732)	(26,732)
Other comprehensive loss for the period		ı	ĭ	1	
Transactions with owners					
Capital increase		128	11,397	T	11,525
Expenses, capital increase		ī	(30)	T	(30)
Share-based payment expense	9			T	ı
Balance as of December 31, 2015		3,346	224,999	(154,202)	74,143
Net loss for the period		1		(57,935)	(57,935)
Other comprehensive loss for the period		1		1	1
Transactions with owners					
Capital increase		15	1,316	ī	1,331
Expenses, capital increase		1	(30)	Ε.	(30)
Share-based payment expense	9	1	,		•
Balance as of December 31, 2016		3,361	226,285	(212,137)	17,509

See accompanying notes to these financial statements

# **Statement of Cash Flows**

	Note	Twelve months ended December 31, 2016 TDKK	Six months ended December 31, 2015 TDKK	Twelve months ended June 30, 2015 TDKK
Operating activities		· · · · · · · · · · · · · · · · · · ·		
Net loss before tax		(63,435)	(29,482)	(38,467)
Adjustments to reconcile loss before tax to cash flows from operating activities  Share-based payment expense	6			0
Depreciation and write-down	9	706	286	504
Gain/loss on sale and disposal of assets	9	,	200	504
Change in other receivables		33 (2,876)	(263)	(145)
Change in other receivables  Change in prepayments			(3,250)	(2,531)
Change in trade payables		1,347 2,271	1,492	(42)
Change in other payables		1,352	3,595	1,748
Cash flows from taxes		5,875	5,595 6,250	1,250
Interest paid		5,6/5	0,250	1,245
Net cash used in operating activities		(54,727)	(21,372)	(36,438)
		(01)// //		
Investing activities				
Investment in property, plant and equipment	9	(238)	(25)	(558)
Net cash used in investing activities		(238)	(25)	(558)
Financing activities				
Capital contributions from shareholders	10	1,330	11,525	74,445
Cash from convertible loan	10	-,550		14,920
Bank loans		_	(245)	245
Expenses related to capital contributions		(30)	(30)	(185)
Net cash provided by financing activities		1,300	11,250	89,425
Net change in cash and cash equivalents		(53,665)	(10,147)	52,429
Net foreign exchange differences		-	-	_
Cash and cash equivalents at the beginning of the period		68,014	78,161	25,732
Cash and cash equivalents at the end of the period		14,349	68,014	78,161

See accompanying notes to these financial statements

#### Notes to the financial statements

#### Corporate information

Orphazyme ApS (the "Company") is a limited liability company incorporated and domiciled in Denmark. The registered office is located in Copenhagen, Denmark. The company has no subsidiaries, and consequently, the financial statements are standalone financial statements. The financial statements for the year ended December 31, 2016 were authorized for approval at the Annual General Meeting to be held on May 29, 2017, with a resolution of the Board of Directors on May 29, 2017.

## Note 1 - Accounting policies

#### Basis of preparation

The financial statements of the Company have been prepared in accordance with International Financial Reporting Standards, or IFRS, as adopted by the EU. In prior years, the annual reports were prepared in accordance with the provisions applying to reporting class B enterprises under the Danish Financial Statements Act. Upon adoption of IFRS, standards and interpretations which are mandatory for reporting periods beginning on or after January 1, 2016 have been applied. The impact on reported financial position and financial performance is discussed in note 17.

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

In 2015, the Company changed its financial year to follow the calendar year, having previously used a financial year spanning July 1 to June 30, as the Company's peers also follows the calendar year. The current period in the statement of loss comprises the twelve-month period ended December 31, 2016 while the comparative figures comprise the six-month period ended December 31, 2015 and the twelve-month period ended June 30, 2015 and the figures are therefore not directly comparable. The company has voluntarily decided to disclose the comparative figures for the financial year 2014/15.

The opening balance as of July 1, 2014 has been prepared in applying the rules under IFRS. This has not resulted in any adjustments to the items in the opening balance.

## Segment information

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

## Foreign currency transactions and balances

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

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

## Share-based payment

Employees and Management of the company receive remuneration in the form of equity settled awards whereby services are rendered as consideration for warrants. The fair value of these equity-settled awards is determined at the date of grant resulting in a fixed fair value at grant date that is not adjusted for future changes in the fair value of the equity awards that may occur over the service period. Fair value of warrants and options is determined using the Black Scholes model.

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

The fair value of equity-settled awards is reported as compensation expense pro rata over the service period to the extent such awards are estimated to vest. No cost is recognized for awards that do not ultimately vest.

**Employee** benefits

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

#### Leases

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

#### **Public grants**

Public grants given to cover expenses are recognized in the income statement as a reduction to research and development expenses over the periods to which the costs relate. The terms of the grants do not obligate the Company to repay any of portion of the grant.

## Financial instrument valuation hierarchy

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

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

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

## Property, plant and equipment

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

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

## Corporation tax receivable

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

Deferred tax is measured using the balance sheet liability method on all temporary differences between the carrying amount and the tax value of assets and liabilities, with the exception of temporary differences occurring at the time of acquisition and liabilities neither effecting the result of operation nor the taxable income. The Company has no deferred tax balances as of December 31, 2016 or 2015 or as of June 30, 2015.

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

#### **Deposits**

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

Prepayments

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

#### Other receivables

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

#### Financial assets

Initial recognition and measurement

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

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

The Company has no derivative financial assets nor has there been a change in classification of a financial asset after initial recognition and measurements as discussed herein. Financial assets are not acquired for trading or speculative purposes. The Company has not placed any assets as security for loans at either December 31, 2016 or 2015 or at June 30, 2015.

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

Subsequent measurement

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

Financial asset impairment

The Company assesses at the end of each reporting period whether there has been objective evidence that a financial asset or Company of financial assets may be impaired. Impairment losses are incurred if there is objective evidence of impairment and the evidence indicates that estimated future cash flows will be negatively impacted. For financial assets held at amortized costs, the amount of impairment loss to be recognized in the financial statements is measured as the difference between the carrying value of the financial asset and the present value of the expected cash flows of the financial asset using the original effective interest rate. The Company did not experience an impairment of a financial asset for either the twelve months ended at June 30, 2015, the six months ended December 31, 2015 or the twelve months ended December 31, 2016.

#### Other receivables

Receivables are measured at amortized cost. An impairment loss is recognized if there is an objective indication that a receivable or a group of receivables is impaired. Receivables that are concluded to be impaired are written down on an individual basis.

Cash and cash equivalents

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

#### Financial liabilities

The Company's financial liabilities historically have included bank debt, trade payables and other payables.

#### Bank debt

Bank debt of TDKK 245 as of June 30, 2015 had been repaid as of December 31, 2015. The Company has no bank debt as of December 31, 2016.

Trade payables

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

Convertible debt

The Company in the past has issued convertible loans that meet certain technical requirements, including (but not limited to) settlement of the conversion option for a fixed number of the Company's ordinary shares, that are initially recognized at fair value, net of transaction costs incurred. Subsequently these convertible loans are measured at amortized cost and accounted for using the effective interest rate method. Gains and losses are recognized in the statement of profit or loss within other finance costs when the convertible loans are derecognized as well as through the effective interest rate amortization process. Amortized cost is calculated by taking into account any discount or premium from the face value of the convertible loan plus direct and incremental transaction costs incurred in connection with issuance of the convertible loan.

#### Other payables

Other payables are measured at net realizable value.

## Statement of Profit or Loss and Other Comprehensive Income

#### Revenue

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

#### Research and development costs

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

#### Administrative expenses

Administrative expenses include salaries for administrative staff and management, costs of share-based payment and rent associated with facilities not used for research purposes.

#### Financial income and expense

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

#### Income tax benefit

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

## Statement of cash flows

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

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

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

Cash flows from financing activities are comprised of repayment of bank debt and proceeds from capital increases net of transaction costs.

# Note 2 - Significant accounting judgments, estimates and assumptions

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

# Judgments made in applying accounting policies

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

- Estimation of accruals and prepaid costs for clinical research trials

- Judgment in respect of recognition of deferred taxes related to taxable losses to be carried forward (note 11)

#### Estimates and assumptions

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

#### Estimation of accruals and prepaid costs for clinical trial development costs

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

Accounting for clinical trials relating to activities performed by clinical research organizations (CROs) and other external vendors requires management to exercise significant estimates in regards to the timing and accounting for these costs. The diverse nature of services being provided by CROs and other arrangements, the different compensation arrangements that exists for each type of service and the limitations in respect of information related to certain clinical activities adds complexity to the estimation of accruals for services rendered by CROs and other vendors in connection with clinical trials. Furthermore, certain CROs and vendors are paid upfront in connection with clinical activities. In estimating the relevant periods etc., the company evaluates the start-up, treatment and wrap-up periods, compensation arrangements and services rendered attributable to each clinical trial. Estimated costs are regularly tested against payment plans and trial completion assumptions.

The company has recognized accruals related to clinical trial development costs of TDKK 0, TDKK 330 and TDKK 1,448 for the twelve months ended December 31, 2016, the six months ended December 31, 2015 and the twelve months ended June 30, 2015, respectively.

The company has recognized prepaid costs related to clinical trial development costs of TDKK 3,794, TDKK 5,765 and TDKK 2,430 for the twelve months December 31, 2016, the six months ended December 31, 2015 and the twelve months ended June 30, 2015, respectively.

## Judgement related to deferred taxes related to taxable losses to be carried forward

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

The company has net tax loss carry-forwards that are not recognized of MDKK 135, MDKK 97 and MDKK 81 for the twelve months December 31, 2016, the six months ended December 31, 2015 and the twelve months ended June 30, 2015, respectively.

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

Reference is made to note 11.

#### Note 3 - Standards issued but not yet effective

The IASB has issued a number of new standards that become effective on or after 1 January 2017. Management's current expectation is that the new standards will be adopted by the company at the effective date. Depending on the stage of development of the company as of this point in time, the following new standards could have an impact of the financial statements:

#### IFRS 9 - Financial instruments

This standard addresses the accounting for financial assets and liabilities including their recognition, classification and measurement and hedge accounting. The Company does not anticipate adopting IFRS 9 before the mandatory effective date of January 1, 2018. The impact on the Company's financial statements of the future adoption of IFRS 9 will be determined based on facts and circumstances that exist at the time of adoption that cannot be predicted currently.

IFRS 16 - Leases

This standard introduces a single lessee accounting model and requires a lessee to recognize assets and liabilities for all leases with a term of more than twelve months, unless the underlying asset is of low value. A lessee is required to recognize a right of use asset representing its right to use the underlying leased asset and a lease liability representing its obligation to make lease payments. IFRS 16 has an effective date of January 1, 2019. The impact on the Group's financial statements of the future adoption of IFRS 16 will be determined based on facts and circumstances that exist at the time of adoption that cannot be predicted currently.

Note 4- Government grant

Government grants comprise research funding from the Danish government and EU. Government grants are recognized in the period where the expenses funded by the grants have been incurred. Government grants are recognized as a reduction in research and development expenses as the grants are considered to be cost refunds. Income from government grants amount to TDKK 2,786, TDKK 1,003 and TDKK 0 respectively in 2016, 2015 and 2014/2015.

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

Note 5 - Staff costs

	Twelve months ended December 31, 2016 TDKK	Six months ended December 31, 2015 TDKK	Twelve months ended June 30, 2015 TDKK
Staff costs			
Wages/salaries	11,777	5,128	7,749
Share-based payment	-	-	-
Pensions	-	=	-
Other social security costs	117	30	57
Other staff costs	372	161	228
Total staff costs	12,266	5,319	8,034
Key management remuneration			
Wages/salaries	4,486	1,603	2,989
Share based payment	-	-	-
Pensions	60	-	-
Other social security costs	36	3	6
Other staff costs		-	-
Total key management remuneration	4,582	1,606	2,995
Total staff and key management costs	16,848	6,925	11,029
Staff and key management costs are recogniz	ed as follows in the financi	al statements:	
Administrative expenses	2,431	685	1,232
Research and development expenses	14,417	6,240	9,797
Total staff and key management costs	16,848	6,925	11,029

The amounts disclosed in the table above are the amounts recognized as an expense during the reporting periods. Key management consists of the Company's Chief Executive Officer, Chief Scientific Officer, Chief Financial Officer and Chief Medical Officer for the twelve months ended December 31, 2016. For the six months ended December 31, 2015 and the twelve months ended June 30, 2014 key management consisted of the Company's Chief Executive Officer and Chief Scientific Officer. See Note 14 for compensation paid to the members of the board of directors.

#### Note 6 - Warrants

The company has issued warrants to employees, consultants providing similar services and key management. The warrants can be settled by subscribing for A-shares of the company at an exercise price of DKK 44 per share. Due to the liquidation preference to B and C shares as discussed in note 10, the exercise price for the warrants were significantly above the fair value of one A-share at the respective issuance dates.

Management has applied a Black Scholes option valuation model to determine fair value of the warrants. Fair

value of the warrants granted in 2014/2015 and 2015 respectively amounts to TDKK 3 and TDKK 12 respectively.

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

The table below summarizes the activity related to the warrants for the year ended December 31, 2016, the sixmonth period ended December 31, 2015 and the twelve months ended June 30, 2015:

	key mana- gement	Emp- loyees	Board of Di- rectors	Con- sultants	Total War- rants	War- rants exer- cisable
Outstanding at June 30,	153,278	54,692	76,638	-	284,608	154,564
2014 Granted	7.	27,746	20,437	9,700	57,883	
Exercised	-	-	=	-	-	
Expired	=	4	_	_	-	
Forfeited	-	(12,773)	-	-	(12,773)	
Outstanding at June 30, 2015	153,278	69,665	97,075	9,700	329,718	221,719
Granted	58,601	21,789	27,047	_	107,437	
Exercised	-	5		-	-	
Expired	-	-	-	-	-	
Forfeited		(15,278)	-	-	(15,278)	
<b>Outstanding at December</b>	211,879	76,176	124,122	9,700	421,877	266,621
<b>31, 2015</b> Granted				-		
Exercised	-	-	_	-		
Expired	-	-	-	-		
Outstanding at December 31, 2016	211,879	76,176	124,122	9,700	421,877	324,078

The weighted average remaining contractual life of the warrants outstanding as of December 31, 2016, December 31, 2015 and June 30, 2015 was 1.6 years, 1.7 years and 1.7 years, respectively.

#### Note 7 - Financial Income

	Twelve months ended December 31, 2016 TDKK	Six months ended December 31, 2015 TDKK	Twelve months ended June 30, 2015 TDKK
Exchange gain	180	74	-
Other interest income	2	6	34
Total financial income	182	80	34

## Note 8 - Financial expenses

	Twelve months ended December 31, 2016 TDKK	Six months ended December 31, 2015 TDKK	Twelve months ended June 30, 2015 TDKK
Interest expense	71	26	14
Other financial expenses	26	14	15
Interest expense, convertible debt (note 15)	0	0	1,245
Exchange losses	0	0	129
Total financial expenses	97	40	1,403

Note 9 - Property, plant and equipment

	Fixtures and fittings, other plant and equipment TDKK	Leasehold improvements TDKK	Total TDKK
Cost at June 30, 2014	1,825	481	2,306
Additions	499	59	558
Cost at June 30, 2015	2,324	540	2,864
Additions	25		25
Cost at December 31, 2015	2,349	540	2,889
Additions	218	20	238
Disposals	0	(38)	(38)
Cost at December 31, 2016	2,567	522	3,089
Accum. depreciation at June 30, 2014	501	111	612
Depreciation expense	404	100	504
Accum. depreciation at June 30, 2015	905	211	1,116
Depreciation expense	232	54	286
Accum. depreciation at December 31, 2015	1,137	265	1,402
Depreciation expense	472	100	572
Write-down	0	134	134
Disposals	0	(6)	(6)
Accum. depreciation at December 31, 2016	1,609	493	2,102
Net book value at			
June 30, 2014	1,324	370	1,694
June 30, 2015	1,419	329	1,748
December 31, 2015	1,212	275	1,487
December 31, 2016	958	29	987

At the end of 2016 the Company started the process of moving to new premises. In connection with this the book value of the leasehold improvement has been impaired and written down to its expected net realizable value as of December 31, 2016. The write down has been recognized under research and development expenses in the statement of profit or loss and other comprehensive income. There has been no write down of property, plant and equipment in the six months ended December 31, 2015 and twelve months ended June 30, 2015.

Depreciation expense is included within operating loss as follows:

	Twelve months ended December 31, 2016 TDKK	Six months ended December 31, 2015 TDKK	Twelve months ended June 30, 2015 TDKK
Administrative expenses	-	-	-
Research and development expenses	472	286	504
Total depreciation expense	472	286	504

## Note 10 - Equity

The following table summarizes the Company's share activity:

	Class A	Class B	Class C
	ordinary	preferred	preferred
	shares	shares	shares
June 30, 2014	125,000	2,050,208	_

Capital increase
Converted debt
June 30, 2015
Capital increase
December 31, 2015
Capital increase
December 31, 2016

-	827,165 215,658
2,050,208	1,042,823
	127,724
2,050,208	1,170,547
	14,786
2,050,208	1,185,333
	2,050,208 2,050,208

The share capital of the Company is divided into 3 classes, an A class, a B class and a C class. A class shares are ordinary shares. The B and C class shares both receive preference in all distributions for all amounts up to the amount paid in upon subscription plus an additional 10% compounded interest per year, with C class having the most senior preference. No distributions shall be made to the B class shares until the C class distributions have been satisfied, and likewise no distributions shall be made to the ordinary shares until the B class distributions have been satisfied. Once the B and C class distributions have been satisfied, any remaining distributions will be distributed on a pro rata basis amongst all issued shares of the Company.

Any B and C class share may at the request of its holder at any time be converted into an A class share (Conversion rate 1:1). The Board is authorized to adopt any changes to the Articles of Association required to implement any conversion request.

On January 6, 2015, the company issued 1,704,554 indemnification warrants to the investors subscribing for C class shares at this date. The warrants entitle the holders to subscribe for additional C class shares at DKK 1 per share if certain liabilities of the company prove higher than warranted by the company in the Investment agreement. The warrants expiry on January 6, 2018 and it is not expected to be realised. The warrants are considered as an adjustment mechanism to the subscription price and not as separate derivative liabilities.

On February 5, 2015, the share capital was increased by 827,165 C class shares through issuance of shares for TDKK 74,445 in cash and issuance of 215,658 C class shares through conversion of convertible debt instruments with a principal amount of TDKK 15,527. The convertible bonds were issued on June 27, 2014 for proceeds of TDKK 14,920 that were received during July 2014. The conversion ratio was the lower of DKK 76.44 per share and the subscription price less 20% in a subsequent capital increase in excess of MEUR 10. The coupon rate was 8% p.a. Due to the variability in the conversion price, the conversion option was classified as a derivative financial instrument. Upon exercise, fair value of the conversion option TDKK 638 (for further details please refer to note 17) was transferred to equity and presented as part of the proceeds from the capital increase as they fulfill the requirements according to IFRS. In connection with the capital increase, the Company incurred expenses totaling TDKK 185.

As part of the subscription of C class shares in the six months ended December 31, 2015, the subscribers received 1,704,554 indemnification warrants to subscribe for additional C class shares at DKK 1 per share. The warrants may only be exercised in the event of the occurrence of one or more Warranty Claims as defined in the Investment Agreement dated December 30, 2014. Exercise of the warrants is further subject to the submission to the Company by the Owner of written notice of one or more Warranty Claims on or before January 6, 2016, or, in the event of a Warranty Claim which relates to tax issues, before January 6, 2018. The number of warrants exercisable is determined by the amount of the Claim. The warrants are accounted for as equity instruments.

On December 15, 2015, the share capital was increased by 127,724 C class shares though issuance of shares for TDKK 11,525 in cash. In connection with the capital increase, the Company incurred expenses totaling TDKK 30.

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

Until January 6, 2020, the Board of Directors are authorized to decide on one or more issues of warrants with the rights of subscription of 1 A class share per warrant in the Company in the total amount of up to TDKK 383 with addition of TDKK 245 without pre-emptive subscription right for the Company's shareholders. The Board of Directors has issued a total of 462,701 warrants under the authorization with a right to subscribe for up to nominally DKK 462,701 A class shares. Consequently, warrants with a right to subscribe further A class shares in the nominal amount of DKK 165,299 may be issued under the authorization.

The Company has never paid a dividend on ordinary shares and does not expect to pay dividends for the foreseeable future.

Subsequent to December 31, 2016, the company has finished a capital increase by issuing 534,007 C class

shares to existing shareholders for a net proceeds received of MDKK 48 in January 2017. Furthermore, the Company completed a MDKK 104 financing round by issuing new C class shares to new shareholders in March 2017.

Note 11 - Income tax and deferred tax

	Twelve months ended December 31, 2016 TDKK	Six months ended December 31, 2015 TDKK	Twelve months ended June 30, 2015 TDKK
Current tax on benefit on net loss	13,962	6,928	9,040
Adjustment to prior years	0	0	0
Tax credit research and development expenses	5,500	2,750	5,875
Change in unrecognized deferred tax before tax credit	(13,951)	(6,923)	(9,031)
Change in unrecognized deferred tax before tax credit	(11)	(5)	(9)
Total income tax benefit for the period	5,500	2,750	5,875
Reconciliation of effective tax rate to Dar	nish statutory tax rate December 31, 2016 TDKK	December 31, 2015 TDKK	June 30, 2015 TDKK
Net loss before tax	(63,465)	(29,482)	(38,467)
Corporate income tax rate in Denmark	22%	23.5%	23.5%
Computed income tax benefit	13,962	6,928	9,040
Tax effect of: Adjustment to prior years	0	O	0
Other non-deductible expenses, including share-based compensation	(11)	(5)	(9)
Deferred tax asset not recognized	(8,451)	(4,173)	(3,156)
Total income tax benefit for the period	5,500	2,750	5,875
Deferred tax in the statement of financia	l position December 31, 2016 TDKK	December 31, 2015 TDKK	June 30, 2015 TDKK
Tax deductible losses	29,716	21,425	17,848
Other temporary differences	(113)	(41)	(80)
	29,603	21,384	17,768
Deferred tax asset not recognized	(29,603)	(21,384)	(17,768)
Carrying amount included on statement of financial position	0	0	0

The Company had net tax loss carry-forwards in Denmark for income tax purposes of MDKK 135, MDKK 97 and MDKK 81 million as of December 31, 2016 and 2015 and June 30, 2015.

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

The tax loss carry forwards have no expiry date. The Company's ability to use tax loss carry forwards in any one year is limited to 100% of the first MDKK7.5 of taxable income plus 60% of taxable income above MDKK 7.5.

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

Significant judgment

The development of therapeutic products within the biopharmaceutical industry is subject to significant risks and uncertainties and there is no assurance a therapeutic product will be successfully developed. As the result of this uncertainty and since the Company has reported significant losses since inception, has no commercial products or revenues and does not expect to generate revenues or profits for the foreseeable future, management has concluded that deferred tax assets should not be recognized as of December 31, 2016 or at any other prior date. The tax assets are currently not deemed to meet the criteria for recognition as management is not able to provide convincing positive evidence that taxable profits will be available in the future to utilize the benefit from the tax assets.

As of December 31, 2016 there are no tax audits in process nor has management been notified of any pending tax audit. As of December 31, 2016, the tax years that remain open for audit by the Danish tax authorities include 2012 through 2015.

#### Note 12 - Loss per share

The following reflects the net loss attributable to both preferred and ordinary shareholders and share data used in the basic and diluted earnings/(loss) per share computations for the twelve months ended December 31, 2016 and 2015 and the six months ended December 31, 2015:

	Twelve months ended December 31, 2016 TDKK	Six months ended December 31, 2015 TDKK	Twelve months ended June 30, 2015 TDKK
Loss for the period	(57,935)	(26,732)	(32,592)
Preference dividend attributable to B shares	(18,367)	(8,746)	(15,902)
Preference dividend attributable to C shares	(10,982)	(4,702)	(3,574)
Loss attributable to all share classes	(87,284)	(40,180)	(52,068)
Weighted average A shares outstanding	125,000	125,000	125,000
Weighted average B shares outstanding	2,050,208	2,050,208	2,050,208
Weighted average C shares outstanding	1,171,410	1,048,145	391,059
Total weighted average shares outstanding	3,346,618	3,223,353	2,566,267
Loss attributable to all share classes	(87,284)	(40,180)	(52,068)
Total weighted average shares outstanding	3,346,618	3,223,353	2,566,267
Loss per share, A shares	(26.08)	(12.47)	(20.29)
Preference dividend attributable to B shares	18,367	8,746	15,902
Pro rata share of loss	(53,471)	(25,557)	(41,597)
Earnings/(loss) attributable to B shares	(35,104)	(16,811)	(25,695)
Weighted average B shares outstanding	2,050,208	2,050,208	2,050,208
Earnings/(loss) per share, B shares	(17.12)	(8.20)	(12.53)
Preference dividend attributable to C shares	10,982	4,702	3,574
Pro rata share of loss	(30,551)	(13,066)	(7,934)
Earnings/(loss) attributable to C shares	(19,569)	(8,364)	(4,360)
Weighted average C shares outstanding	1,171,410	1,048,145	391,059
Earnings/(loss) per share, C shares	(16.71)	(7.98)	(11.15)

Basic loss per share amounts are calculated by dividing the net earnings/(loss) for the period attributable to each share class by the weighted average number of shares outstanding during each the period. Due to the fact that the Company has incurred losses for each period presented, the potential A class shares issuable related to outstanding warrants have been excluded from the calculation of diluted loss per share as the effect of such shares is anti-dilutive. There are not outstanding warrants related to the B or C class shares, therefore, basic and diluted loss per share are the same for each period presented.

Subsequent to December 31, 2016, the company has finished two capital increases increasing the number of shares with a total of 534,007, which will have an impact on the calculation of earnings per share from 2017 and forward.

#### Note 13 - Capital management

For the purpose of the Company's capital management, capital includes issued capital, share premium and all other equity reserves attributable to the equity holders of the Company. The primary objective of the Company's capital management is to maximize shareholder value. The board of directors' policy is to maintain a strong capital base so as to maintain investor, creditor and market confidence, and a continuous advancement of the Company's intellectual property, product pipeline and business. Cash, cash equivalents and financial assets are monitored on a regular basis by management and the board of directors in assessing current and long-term capital needs. As of December 31, 2016 the Company held cash and cash equivalents totaling MDKK 14.3 that together with the subsequent capital increases will be sufficient to provide adequate funding to allow the Company to meet its planned operating activities, including increased levels of research and development activities, in the normal course of business for the next twelve months. The Company currently has no significant planned capital expenditures.

Subsequent to December 31, 2016, the company has finished two capital increases for a total of MDKK 152, where MDKK 65 has been paid in cash and thereby increase the cash and cash equivalents and MDKK 87 is recognized as a receivable. MDKK 52 is due when the Board of Directors requests the funds. The remaining MDKK 35 is due upon on reaching a specific milestone where MDKK 9 is immediately due and the remaining MDKK 26 is due when the Board of Directors requests the funds.

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

#### Foreign Currency

The Company maintains operations in Denmark and uses the DKK as its functional currency. The Company conducts cross border transactions where the functional currency is not always used. Accordingly, future changes in the exchange rates of the DKK, the EUR, the USD and/or the GBP will expose the Company to currency gains or losses that will impact the reported amounts of assets, liabilities, income and expenses and the impact could be material. For the year ended December 31, 2016 and 2015 and the six months ended December 31, 2015 the impact on the Company's statement of loss for possible changes in the EUR, USD and GBP exchange rates against the Company's functional currency of DKK would be as follows:

Currency	Possible change	Twelve months ended December 31, 2016 TDKK	Six months ended December 31, 2015 TDKK	Twelve months ended June 30, 2015 TDKK
EUR	+/2%	+177 / (177)	+611 / (611)	+704 / (704)
USD	+/10%	(21) / +21	+26 / (26)	+591 / (591)
GBP	+/10%	(327) / +327	(153 / +153	+486 / (486)

## **Interest Rate Risk**

The Company has no interest bearing debt.

#### Credit Risk

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

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

#### Note 14 - Contractual obligations and contingencies

## Contractual obligations

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

Contractual obligations	Twelve months ended December 31, 2016 TDKK	Six months ended December 31, 2015 TDKK	Twelve months ended June 30, 2015 TDKK
o − 1 years	418	284	278
1 – 5 years	-	-	-
More than 5 years	-	-	2
Total	418	284	278

Total expense under non-cancelable contractual obligations was TDKK 848, TDKK 397 and TDKK 647 for the twelve months December 31, 2016, the six months ended December 31, 2015 and the twelve months ended June 30, 2015, respectively.

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

#### **Contingencies**

Contingencies are assets and liabilities that arise from past events but whose existence will only be confirmed by the occurrence or non-occurrence of future events that in some situations are beyond the Company's control. As of December 31, 2016 and 2015 and June 30, 2015 there are no contingent assets or liabilities.

## Note 15 - Related party disclosures

The Company is not ultimately controlled by any of the investors. See Note 5 for additional related party transactions, related to the remuneration paid to key management. In addition to Novo A/S, Kurma Biofund II, Idinvest, Sunstone Life Science Ventures Fund II K/S and Cooperative Aescap Venture I U.A. all owns more than 5%.

There have been no transactions between related parties in the 12 months ended December 31, 2016 and the six months ended December 31, 2015 besides capital increases as described in note 10. In the twelve months ended June 30, 2015 in addition to capital increases there was also a convertible debt, that was converted to shares in the Company. The interest expense on the convertible debt was TDKK 1,245 up to the date of conversion. Reference is made to note 17.

## Terms and conditions of transactions with related parties

Amounts due to related parties are uncollateralized and interest free. There have been no guarantees provided or received for any related party receivables or payables. For the twelve months ended December 31, 2016, the six months ending December 31, 2015 and the twelve months ended June 30, 2015 the Company has not recorded any impairment of receivables relating to amounts owed by related parties. There are no related party receivables at any of the balance sheet dates.

## Transactions with key management

The Company has not granted any loans, guarantees, or other commitments to or on behalf of any of the members of the board of directors or key management personnel.

Other than the remuneration described in Note 5, no other significant transactions have taken place with key management personnel during the period presented herein.

## Compensation paid to members of the board of directors

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

	Twelve months ended December 31, 2016 TDKK	Six months ended December 31, 2015 TDKK	Twelve months ended June 30, 2015 TDKK
Director fees	524	795	522
Warrants	0	0	0
Other fees	0	0	0
Total compensation paid to the board of directors	524	795	522

Certain members of the board of directors have received warrants in the Company (reference is made to note

6). Besides warrants and the director fees presented below, they have not received any other remuneration for their services

Transactions with shareholders and affiliates

There have been no transactions with shareholders or affiliates of shareholders during the twelve months ending December 31, 2016, the six months ended December 31, 2015 or the twelve months ending June 30, 2015, except for the capital increase disclosed in Note 10. In the twelve months ended June 30, 2015 the Company finished a capital increase with Cooperative Aescap Venture I U.A., Novo A/S and Sunstone Life Science Ventures Fund II K/S for a total of MDKK 74.

#### Note 16 - Subsequent events

Management has evaluated its financial statements for potential subsequent events occurring after the balance sheet date of December 31, 2016 but prior to the date that these financial statements were issued. During Q1 2017, the Company's 2015 financing round was extended, with 534,007 C class preferred shares issued to existing shareholders for total net proceeds received of MDKK 48 including issuance costs of TDKK 79.

On March 8, 2017 the Company completed a MDKK 104 financing round by issuing new shares to LSP V Coöperatieve U.A. and ALS Investment Fund. MDKK 69 has been invested and MDKK 35 is dependent on certain future milestones.

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

## Note 17 - First time adoption of IFRS

These financial statements are the first financial statements presented in accordance with IFRS as adopted by the EU. The date of transition is July 1, 2014. As the company is presenting financial statements in accordance with IFRS as adopted by the EU for the first time, estimates for each balance sheet date has been revisited and updated with the most recent information.

Under IFRS, fair value of warrants issued to employees as remuneration for their services is recognized as an expense over the vesting period. Under previous GAAP, no expense has been recognized for these warrants.

Convertible bonds issued and converted into equity in 2014/2015 comprises a conversion option with a conversion price which was not fixed. Consequently, the conversion option is a derivative financial liability which shall be accounted for separately from the host debt contract and measured at fair value through profit or loss. Under previous GAAP, the convertible bond was classified in full as a financial liability.

The change in accounting policy has resulted in an increase in financial expenses of TDKK 638 in 2014/2015. It has not affected equity on any of the balance sheet dates presented because the bond was issued and converted within the same financial year.