

# Orphazyme ApS

Ole Maaløes Vej 3, 2200 Copenhagen N


CVR no. 32 26 63 55



## Annual report for the period 1 July - 31 December 2015

Approved at the Company's annual general meeting on 1 June 2016

Chairman:



.....  
Georges Gemayel



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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 1 July - 31 December 2015.

The annual report is prepared in accordance with the Danish Financial Statements Act.

In our opinion, the financial statements give a true and fair view of the Company's financial position at 31 December 2015 and of the results of the Company's operations for the financial year 1 July - 31 December 2015.

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.

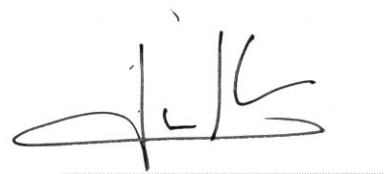
Copenhagen, 1 June 2016  
Executive Board:

  
Anders Mørkeberg Hinsby

Board of Directors:

  
Georges Gemayel  
Chairman

  
Patrick Johan Hendrik Krol

  
Rémi Pascal Louis Droller

  
Nanna Liebach Lüneborg

  
Martin Bonde

  
Martin Rahbek Korum

  
Sten Verland

  
Bo Jesper Hansen

## Independent auditors' report

To the shareholders of Orphazyme ApS

### Independent auditors' report on the financial statements

We have audited the financial statements of Orphazyme ApS for the financial year 1 July - 31 December 2015, which comprise an income statement, balance sheet, statement of changes in equity and notes, including a summary of significant accounting policies. The financial statements are prepared in accordance with the Danish Financial Statements Act.

### *Management's responsibility for the financial statements*

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

### *Auditors' responsibility*

Our responsibility is to express an opinion on the financial statements based on our audit. We conducted our audit in accordance with International Standards on Auditing and additional requirements under Danish audit regulations. This requires that we comply with ethical requirements and plan and perform the audit to obtain reasonable assurance as to whether the financial statements are free from material misstatement.

An audit involves performing procedures to obtain audit evidence about the amounts and disclosures in the financial statements. The procedures selected depend on the auditors' judgement, including the assessment of the risks of material misstatement of the financial statements, whether due to fraud or error. In making those risk assessments, the auditors consider internal control relevant to the Company's preparation of financial statements that give a true and fair view 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. An audit also includes evaluating the appropriateness of accounting policies used and the reasonableness of accounting estimates made by Management, as well as evaluating the overall presentation of the financial statements.

We believe that the audit evidence we have obtained is sufficient and appropriate to provide a basis for our opinion.

Our audit has not resulted in any qualification.

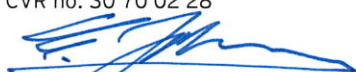
### *Opinion*

In our opinion, the financial statements give a true and fair view of the Company's financial position at 31 December 2015 and of the results of its operations for the financial year 1 July - 31 December 2015 in accordance with the Danish Financial Statements Act.

### Statement on the Management's review

Pursuant to the Danish Financial Statements Act, we have read the Management's review. We have not performed any other procedures in addition to the audit of the financial statements. On this basis, it is our opinion that the information provided in the Management's review is consistent with the financial statements.

Copenhagen, 1 June 2016  
ERNST & YOUNG  
Godkendt Revisionspartnerselskab  
CVR no. 30 70 02 28



Christian Schwenn Johansen  
State Authorised Public Accountant

## Management's review

### Company details

Name	Orphazyme ApS
Address, Postal code, City	Ole Maaløes Vej 3, 2200 Copenhagen N
CVR No.	32 26 63 55
Established	19 June 2009
Registered office	Copenhagen
Financial year	1 July - 31 December
Website	<a href="http://www.orphazyme.com">www.orphazyme.com</a>
E-mail	<a href="mailto:contact@orphazyme.com">contact@orphazyme.com</a>
Telephone	+45 70 70 29 80
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, P O Box 250, 2000 Frederiksberg, Denmark
Bankers	Dansk Bank Nordea



## **Management's review**

### **Operating review**

#### **The Company's business review**

Orphazyme is dedicated to the development of a novel class of therapeutics for the treatment of genetic and degenerative diseases. Orphazyme's programs are in preclinic / clinical development as potential treatments for lysosomal storage diseases and other neurodegenerative diseases such as ALS.

Orphazyme's lead project is to develop new medicine for the treatment of lysosomal storage diseases, a family of more than 50 genetic disorders that occur in 1:10.000 births. The Company applies proprietary know-how and technology based on the cytoprotective properties of heat-shock proteins.

Arimoclomol effectively treats cells from patients suffering from different lysosomal storage diseases, for which there is no current treatment available. Arimoclomol has also exhibited therapeutic effect in animal disease models further substantiating its therapeutic potential. A clinical program in Niemann Pick disease type C, a debilitating and ultimately fatal lysosomal storage disease, is currently enrolling and patients will enter the interventional part with arimoclomol in 2016. Moreover, Arimoclomol is currently in clinical development in a genetically defined subpopulation of patients suffering from the motorneuron disease ALS and will start development in sIBM towards the end of 2016.

#### **Financial review**

The income statement for 2015 shows a loss of DKK 26,210,613 against a loss of DKK 31,958,124 last year, and the balance sheet at 31 December 2015 shows equity of DKK 75,025,888. Management considers the Company's financial performance in the year satisfactory.

#### **Post balance sheet events**

No significant events have occurred subsequent to the financial year.

## Financial statements for the period 1 July - 31 December

### Income statement

Note	DKK	2015 6 months	2014/15 12 months
2	Administrative expenses	-2,052,450	-4,055,946
	<b>Ordinary operating profit/loss</b>	-2,052,450	-4,055,946
	Research costs	-26,948,247	-33,046,514
	<b>Operating profit/loss</b>	-29,000,697	-37,102,460
3	Financial income	80,093	34,264
4	Financial expenses	-40,009	-764,928
	<b>Profit/loss before tax</b>	-28,960,613	-37,833,124
5	Tax for the year	2,750,000	5,875,000
	<b>Profit/loss for the year</b>	-26,210,613	-31,958,124
<b>Proposed profit appropriation/distribution of loss</b>			
	Retained earnings/accumulated loss	-26,210,613	-31,958,124
		-26,210,613	-31,958,124

## Financial statements for the period 1 July - 31 December

### Balance sheet

Note	DKK	31/12 2015	30/6 2015
	<b>ASSETS</b>		
	<b>Non-current assets</b>		
	<b>Property, plant and equipment</b>		
	Fixtures and fittings, other plant and equipment	1,211,755	1,419,407
	Leasehold improvements	274,874	328,903
		<u>1,486,629</u>	<u>1,748,310</u>
	<b>Investments</b>		
	Corporation tax receivable	2,750,000	5,875,000
	Deposits	211,161	183,978
		<u>2,961,161</u>	<u>6,058,978</u>
	<b>Total non-current assets</b>	<u>4,447,790</u>	<u>7,807,288</u>
	<b>Current assets</b>		
	<b>Receivables</b>		
	Corporation tax receivable	5,875,000	6,250,000
	Other receivables	6,600,171	3,036,163
	Prepayments	205,903	290,275
		<u>12,681,074</u>	<u>9,576,438</u>
	<b>Cash</b>	<u>68,014,297</u>	<u>78,161,075</u>
	<b>Total current assets</b>	<u>80,695,371</u>	<u>87,737,513</u>
	<b>TOTAL ASSETS</b>	<u><u>85,143,161</u></u>	<u><u>95,544,801</u></u>



## Financial statements for the period 1 July - 31 December

### Balance sheet

Note	DKK	31/12 2015	30/6 2015
	<b>EQUITY AND LIABILITIES</b>		
	<b>Equity</b>		
6	Share capital	3,345,755	3,218,031
	Retained earnings	71,680,133	86,523,310
	<b>Total equity</b>	<b>75,025,888</b>	<b>89,741,341</b>
	<b>Liabilities other than provisions</b>		
	<b>Current liabilities other than provisions</b>		
	Bank debt	0	245,151
	Trade payables	7,557,981	3,394,496
	Other payables	2,559,292	2,163,813
		<b>10,117,273</b>	<b>5,803,460</b>
	<b>Total liabilities other than provisions</b>	<b>10,117,273</b>	<b>5,803,460</b>
	<b>TOTAL EQUITY AND LIABILITIES</b>	<b>85,143,161</b>	<b>95,544,801</b>
1	Accounting policies		
7	Collateral		
8	Contractual obligations and contingencies, etc.		

## Financial statements for the period 1 July - 31 December

### Statement of changes in equity

DKK	Share capital	Retained earnings	Total
Equity at 1 July 2015	3,218,031	86,523,310	89,741,341
Capital increase	127,724	11,397,436	11,525,160
Expenses, capital increase	0	-30,000	-30,000
Profit/loss for the year	0	-26,210,613	-26,210,613
Equity at 31 December 2015	<u>3,345,755</u>	<u>71,680,133</u>	<u>75,025,888</u>

## Financial statements for the period 1 July - 31 December

### Notes to the financial statements

#### 1 Accounting policies

The annual report of Orphazyme ApS for 2015 has been prepared in accordance with the provisions applying to reporting class B enterprises under the Danish Financial Statements Act.

##### Changes in accounting policies

The income statement is now presented by function. Previously, the income statement was presented by nature.

In Management's opinion, these changes result in a more true and fair view of the Company's activities.

The changes have not affected the results of operations, equity or balance sheet total.

The Company changed its financial year to follow the calendar year. Current financial year covers only six months, while the comparative figures cover 12 months.

The financial statements have otherwise been presented in accordance with the same accounting policies as were applied last year. Comparative figures have been restated to reflect the policy change.

##### Reporting currency

The financial statements are presented in Danish kroner.

##### Foreign currency translation

On initial recognition, transactions denominated in foreign currencies are translated at the exchange rates at the transaction date. Foreign exchange differences arising between the exchange rates at the transaction date and at the date of payment are recognised 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 exchange rates at the balance sheet date. The difference between the exchange rates at the balance sheet date and at the date at which the receivable or payable arose or was recognised in the latest financial statements is recognised in the income statement as financial income or financial expenses.

##### 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 recognised 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 recognised in the income statement under "Research costs" once it is probable that all criteria for being given the grant are satisfied. Grants which must be repaid under certain circumstances are recognised only in so far as they are not expected to be repaid.

### Income statement

#### Gross profit

In the income statement, revenue, production expenses and other operating income have been aggregated into one item called 'gross margin', cf. section 32 of the Danish Financial Statements Act.

## Financial statements for the period 1 July - 31 December

### Notes to the financial statements

#### 1 Accounting policies (continued)

##### Administrative expenses

Administrative expenses include expenses incurred in the year for purposes of managing and administering the company, including expenses relating to administrative staff, management, office premises/expenses as well as amortisation/depreciation of assets used for administrative purposes.

##### Research costs

The item includes research costs not satisfying the criteria for capitalisation, including expenses relating to clinical trials, patent, staff costs, etc.

##### Financial income and expenses

Financial income and expenses are recognised in the income statements at the amounts that concern the financial year. Net financials include interest income and expenses as well as allowances and surcharges under the advance-payment-of-tax scheme, etc.

##### Tax

Tax for the year includes current tax on the year's expected taxable income and the year's deferred tax adjustments. The portion of the tax for the year that relates to the profit/loss for the year is recognised in the income statement, whereas the portion that relates to transactions taken to equity is recognised in equity.

Tax of the year includes tax credit for costs incurred in connection with research and development activities under the Danish Tax Credit Regime.

### Balance sheet

##### Property, plant and equipment

Items of property, plant and equipment 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.

##### Other securities and investments

Rental deposits are measured at cost.

##### Impairment of non-current assets

Intangible assets, property, plant and equipment and investments are subject to an annual test for indications of impairment other than the decrease in value reflected by depreciation or amortisation. Impairment tests are conducted in respect of individual assets or groups of assets generating separate cash flows when there are indications of impairment. The assets are written down to the higher of the value in use and net realisable value (recoverable amount) of the asset or group of assets if this is lower than the carrying amount. As for group of assets, impairment losses are first recognised in respect of goodwill and thereafter proportionately in respect of the other assets.

##### Receivables

Receivables are measured at amortised cost.

An impairment loss is recognised if there is objective indication that a receivable or a group of receivables is impaired. If there is objective indication that an individual receivable has been impaired, write-down is made on an individual basis.

## Financial statements for the period 1 July - 31 December

### Notes to the financial statements

#### 1 Accounting policies (continued)

##### Prepayments

Prepayments recognised under 'Assets' comprise prepaid expenses regarding subsequent financial reporting years.

##### Cash at hand and in bank

Cash at hand and in bank comprise cash and short-term marketable securities which are subject to an insignificant risk of changes in value.

##### Corporation tax

Current tax payable and receivable is recognised in the balance sheet as tax computed on the taxable income for the year, adjusted for tax on the taxable income of prior years and for tax paid on account.

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.

Deferred tax is measured according to the tax rules and at the tax rates applicable at the balance sheet date when the deferred tax is expected to crystallise as current tax. Deferred tax assets are recognised at the expected value of their utilisation; either as a set-off against tax on future income or as a set-off against deferred tax liabilities in the same legal tax entity. Changes in deferred tax due to changes in the tax rate are recognised in the income statement.

##### Other payables

Other payables are measured at net realisable value.

DKK		2015 6 months	2014/15 12 months
2	<b>Staff costs</b>		
	Wages/salaries	6,909,979	10,566,745
	Pensions	466,551	796,885
	Other social security costs	22,140	33,030
	Other staff costs	10,795	30,441
		<u>7,409,465</u>	<u>11,427,101</u>

Staff costs are recognised as follows in the financial statements:

Administration	1,353,470	2,135,789
Research	6,055,995	9,291,312
	<u>7,409,465</u>	<u>11,427,101</u>



## Financial statements for the period 1 July - 31 December

### Notes to the financial statements

DKK		2015 6 months	2014/15 12 months
<b>3</b>	<b>Financial income</b>		
	Other interest income	6,196	34,264
	Exchange gain	73,897	0
		<u>80,093</u>	<u>34,264</u>
<b>4</b>	<b>Financial expenses</b>		
	Interest expenses, associates	0	607,491
	Other interest expenses	25,527	0
	Exchange losses	0	129,255
	Other financial expenses	14,482	28,182
		<u>40,009</u>	<u>764,928</u>
<b>5</b>	<b>Tax for the year</b>		
	Estimated tax charge for the year	-2,750,000	-5,875,000
		<u>-2,750,000</u>	<u>-5,875,000</u>
<b>6</b>	<b>Share capital</b>		
	DKK	31/12 2015	30/6 2015
	The share capital consists of the following:		
	125,000 A shares of DKK 1.00 each	125,000	125,000
	2,050,208 B shares of DKK 1.00 each	2,050,208	2,050,208
	1,170,547 C shares of DKK 1.00 each	1,170,547	1,042,823
		<u>3,345,755</u>	<u>3,218,031</u>

The share capital of the Company is divided into 4 classes, an A class, a B class, a C class and a W class with a preferential position for the B and C class.

Until 6 January 2020, the Board of Directors shall be authorised to decide on one or more issues of warrants with the rights of subscription of 1 A share per warrant in the Company in the total amount of up to DKK 383,000 with addition of DKK 245,000 without pre-emptive subscription right for the Company's shareholders. The Board of Directors has issued a total of 462,701 warrants under the authorisation with a right to subscribe for up to nominally DKK 462,701 A shares. Consequently, warrants with a right to subscribe further A shares in the nominal amount of DKK 165,299 may be issued under the authorisation.

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



## Financial statements for the period 1 July - 31 December

### Notes to the financial statements

#### 6 Share capital (continued)

Analysis of changes in the share capital over the past 5 years:

DKK	2015	2014/15	2013/14	2012/13	2011/12
Opening balance	3,218,031	2,175,208	2,175,208	1,629,422	565,136
Capital increase	127,724	1,042,823	0	545,786	1,064,286
	<u>3,345,755</u>	<u>3,218,031</u>	<u>2,175,208</u>	<u>2,175,208</u>	<u>1,629,422</u>

In connection with the capital increase, the Company incurred expenses totalling DKK 30,000.

#### 7 Collateral

The Company has not placed any assets or other as security for loans at 31/12 2015.

#### 8 Contractual obligations and contingencies, etc.

##### Other financial obligations

Other rent and lease liabilities:

DKK	31/12 2015	30/6 2015
Rent and lease liabilities	<u>569,000</u>	<u>557,000</u>