

## **Zymenex A/S**

Roskildevej 12 C, 3400 Hillerød  
CVR no. 27 39 47 52

### **Annual report for 2015**

This annual report has been adopted at the  
company's annual general meeting on

*5 May 2016*

Chairman of the meeting

**Lars Terp**  
Advokat

A handwritten signature in blue ink, appearing to be "Lars Terp", written over the printed name and title.



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**The company**

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Zymenex A/S  
Roskildevej 12 C  
3400 Hillerød  
Tel.: 48 25 00 54  
Registered office: Hillerød  
CVR no.: 27 39 47 52

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**Board of Directors**

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Jens Fogh, chairman  
Claes Magnus Andersson  
Natallia Misuna  
Paolo Patri

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**Executive Board**

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CEO Paolo Patri

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**Auditors**

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KPMG  
Statsautoriseret Revisionspartnerselskab

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**Parent company**

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Zymenex Holding A/S, Hillerød

**Statement of the Board of Directors and Executive Board on the annual report**

Board of Directors and Executive Board have on this day considered and adopted the annual report for the financial year 01.01.15 - 31.12.15 for Zymenex A/S.

The annual report is presented in accordance with the Danish Financial Statements Act (Årsregnskabsloven).

In our opinion, the financial statements give a true and fair view of the company's assets, liabilities, financial position and results.

We believe that the management's review gives a true and fair review of the matters dealt with in the review.

The annual report is submitted for adoption by the general meeting.

Hillerød, March 22, 2016


**Executive Board**

  
Paolo Patri  
CEO

**Board of Directors**

  
Jens Fogh  
Chairman

  
Paolo Patri

  
Claes Magnus Andersson

  
Natallia Misuna



**To the shareholder of Zymenex A/S**

## **REPORT ON THE FINANCIAL STATEMENTS**

We have audited the financial statements of Zymenex A/S for the financial year 01.01.15 - 31.12.15, which comprise the income statement, balance sheet, accounting policies and notes. The financial statements are prepared in accordance with 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.

### **Auditor's 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 regulation. 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 auditor's 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 auditor considers 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 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 assets, liabilities and financial position at 31.12.15 and of the results of the company's operations for the financial year 01.01.15 - 31.12.15 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 further 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, March 22, 2016

**KPMG**

Statsautoriseret Revisionspartnerselskab  
CVR no. 25 57 81 98

Lau Bent Baun

State Authorised Public Accountant





### **Main activities**

As in previous years, Zymenex A/S main activity has been the development of enzyme replacement therapy for the treatment of rare genetic diseases.

The Zymenex pipeline has been focused on Lamazym (rhLAMAN) for the treatment of the lysosomal disease.

### **Project status**

Zymenex has focused the company's research and development on the recombinant enzyme, Lamazym (rhLAMAN), for enzyme replacement therapy (ERT) for the lysosomal alpha-Mannosidosis storage disease.

This disease is caused by a deficiency of the Laman-enzyme and affects approximately 500 patients world-wide. The project is in late clinical development. The disease is life-threatening, and there is no treatment today. Previously, Zymenex has been a partner in a number of European research groups under the EU grant "Euraman" for the development of the enzyme and has together with this scientific group, published proof-of-principle data (Hum. Mol. Gen., vol. 13, no. 18, July 21, 2004). Zymenex and the research group concluded the EU grant "Hueman" under the 6th framework in October 2009 and has since October 2010, been working together with research groups and clinicians in an EU Framework 7 supported project group "ALPHA-MAN" with the continued development of Lamazym (rhLaman). In 2011, both Phase 1 and Phase 2a clinical studies were conducted and in 2012 Phase 2b was completed with positive results in both biochemical and clinical parameters. The project is now in Phase 3.

Lamazym has Orphan Drug designation in both Europe and USA.

Zymenex has licensed the rights to develop, register, manufacture, promote, distribute and sell certain of its Lamazym Compound to Chiesi Farmaceutici S.p.A. through a license agreement dated December 23, 2014.

Chiesi Farmaceutici S.p.A. has obtained a license from Zymenex to use the Patent and the Know-How for the purpose of developing, manufacturing and selling the product in relation to Lamazyme.

At the end of 2015, employees numbered 12, 5 of which work in the Zymenex A/S Swedish affiliate.



### **Development in the company's financial activities and affairs**

The income statement for the period 01.01.15 - 31.12.15 showed a profit of DKK 656,421 against DKK 1,066 thousand for the period 01.01.14 - 31.12.14. The balance sheet showed negative equity of DKK 64,099,563.

Pharmaceutical development is in general an area with extensive risk. This includes delays in the development process, lack of effect of the product candidate in clinical trials or lack of approval from authorities in connection with production or marketing of the product candidate.

### **Financial position**

Chiesi Farmaceutici S.p.A. that in August 2013 acquired Zymenex Holding A/S and its subsidiaries, including Zymenex A/S, has issued a letter of support stating that they will continue to provide financial support to enable Zymenex Holding A/S, Zymenex A/S and ACE Biosciences A/S to continue their development, trading and investments and to meet all their financial obligations as they fall due for payment for the foreseeable future and at least for the period until 31 December 2016.

### **Important events occurring after the end of the financial year**

After the end of the reporting year, no events have occurred that can change the company's financial position significantly.



Note	2015 DKK	2014 DKK '000
Research and development costs	-52.501.446	-67.882
<b>Gross result</b>	<b>-52.501.446</b>	<b>-67.882</b>
Administrative expenses	-7.046.267	-6.474
Other operating income	63.120.576	79.487
<b>Operating profit/loss</b>	<b>3.572.863</b>	<b>5.131</b>
2 Other financial income	13.722	219
3 Other financial expenses	-3.588.161	-4.284
<b>Total net financials</b>	<b>-3.574.439</b>	<b>-4.065</b>
<b>Profit/loss before tax</b>	<b>-1.576</b>	<b>1.066</b>
4 Tax on profit/loss for the year	657.997	0
<b>Profit/loss for the year</b>	<b>656.421</b>	<b>1.066</b>
<b>Proposed appropriation account</b>		
Retained earnings	656.421	1.066
<b>Total</b>	<b>656.421</b>	<b>1.066</b>

<b>ASSETS</b>		31.12.15	31.12.14
Note		DKK	DKK '000
	Plant and machinery	2.010.293	807
	Other plant, fixtures and fittings, tools and equipment	222.560	112
<b>5</b>	<b>Total property, plant and equipment</b>	<b>2.232.853</b>	<b>919</b>
<b>6</b>	<b>Other receivables</b>	<b>114.697</b>	<b>114</b>
	<b>Total investments</b>	<b>114.697</b>	<b>114</b>
	<b>Total non-current assets</b>	<b>2.347.550</b>	<b>1.033</b>
	Receivables from group enterprises	4.433.887	4.177
	Receivable joint taxation contributions	657.997	1.275
	Other receivables	577.409	1.630
	Prepayments	1.111.355	1.210
	<b>Total receivables</b>	<b>6.780.648</b>	<b>8.292</b>
	<b>Cash</b>	<b>8.826.262</b>	<b>9.161</b>
	<b>Total current assets</b>	<b>15.606.910</b>	<b>17.453</b>
	<b>Total assets</b>	<b>17.954.460</b>	<b>18.486</b>

**EQUITY AND LIABILITIES**

Note		31.12.15 DKK	31.12.14 DKK '000
	Share capital	2.500.000	2.500
	Retained earnings	-66.599.563	-67.256
7	<b>Total equity</b>	<b>-64.099.563</b>	<b>-64.756</b>
	Payables group enterprises	74.363.606	70.470
8	<b>Total long-term payables</b>	<b>74.363.606</b>	<b>70.470</b>
	Trade payables	1.768.826	8.480
	Other payables	5.921.591	4.292
	<b>Total short-term payables</b>	<b>7.690.417</b>	<b>12.772</b>
	<b>Total payables</b>	<b>82.054.023</b>	<b>83.242</b>
	<b>Total equity and liabilities</b>	<b>17.954.460</b>	<b>18.486</b>

- 9 Contingent liabilities  
 10 Contractual obligations  
 11 Ownership



## **GENERAL**

The annual report has been presented in accordance with the provisions of the Danish Financial Statements Act for reporting class B enterprises.

The accounting policies have been applied consistently with previous years.

### **Basis of recognition and measurement**

Income is recognised in the income statement as earned, including value adjustments of financial assets and liabilities. All expenses, including depreciation, amortisation, impairment losses and write-downs, are also recognised in the income statement.

Assets are recognised in the balance sheet when it is probable that future economic benefits will flow to the company and the value of such assets can be measured reliably. Liabilities are recognised in the balance sheet when it is probable that future economic benefits will flow from the company and the value of such liabilities can be measured reliably. On initial recognition, assets and liabilities are measured at cost. Subsequently, assets and liabilities are measured as described for each item below.

Certain financial assets and liabilities are measured at amortised cost applying a constant effective rate of interest over the term of the assets and liabilities. Amortised cost is determined as original cost less any principal repayments and less/plus accumulated amortisation of the difference between cost and nominal value.

On recognition and measurement, account is taken of foreseeable losses and risks arising before the time at which the annual report is presented and proving or disproving matters arising on the balance sheet date.

### **FOREIGN CURRENCY**

The annual report is presented in Danish kroner.

On initial recognition, transactions denominated in foreign currency are translated at the exchange rate applicable at the transaction date. Exchange rate differences between the exchange rate applicable at the transaction date and the exchange rate at the date of payment are recognised in the income statement as a financial item. Receivables, payables and other monetary items denominated in foreign currencies are translated using the exchange rate applicable at the balance sheet date. The difference between the exchange rate applicable at the balance sheet date and at the date at which the receivable or liability arose or was recognised in the latest annual report is recognised in the income statement under financial income or expenses. Non-current assets, inventories and other non-monetary assets acquired in foreign currency are translated using historical exchange rates.

On recognition of integrated foreign entities, monetary items are translated using the exchange rate applicable at the balance sheet date. Non-monetary items are translated using the exchange rate applicable at the time of acquisition or the time of subsequent revaluation, impairment or write-down of the asset. The items in the income statement are translated using the exchange rate applicable at the transaction date, while items derived from non-monetary items are translated using the historical exchange rates for such non-monetary items.

## **INCOME STATEMENT**

### **Research and development costs**

Research and development costs include cost related to research and development activities, including salaries and wages and other indirect costs.

Research and development costs, include ongoing cost of patent rights, recognized in the income statement as and when incurred.

### **Administrative expenses**

Administrative expenses comprise expenses in connection with administrative employees, management, office premises, office expenses etc., including depreciation, amortisation and wages and salaries.

### **Other operating income**

Other operating income comprises items of a secondary nature in relation to the enterprise's activities, including proceeds from the disposal of intangible assets and property, plant and equipment.



**Depreciation**

The depreciation of property, plant and equipment aim at systematic depreciation over the expected useful lives of the assets. The following useful lives and residual values are applied by the company:

	Useful lives, years	Residual value, per cent
Plant and machinery	5	0
Other plant, fixtures and fittings, tools and equipment	3-5	0

New acquisitions of production plant and machinery and other fixtures and fittings, tools and equipment with a cost of less than DKK 12,800 each are expensed in the income statement in the year of acquisition.

**Net financials**

Interest income and interest expenses, foreign currency translation adjustments as well as realised and unrealised capital gains and losses on securities are recognised under net financials.

**Tax**

The current and deferred taxes for the year are recognised in the income statement as taxes for the year with the portion attributable to the net profit or loss for the year, and directly in equity with the portion attributable to amounts recognised directly in equity.

The company is taxed jointly with the Danish consolidated enterprises.

In connection with the settlement of joint taxation contributions, the current Danish income tax is allocated between the jointly taxed enterprises in proportion to their taxable incomes. This means that enterprises with a tax loss receive joint taxation contributions from enterprises which have been able to use this loss to reduce their own taxable profit.

## **BALANCE SHEET**

### **Property, plant and equipment**

Property, plant and equipment are measured in the balance sheet at the lower of cost less accumulated depreciation and recoverable amount.

Cost comprises the purchase price and any costs directly related to the purchase until the date when the asset is available for use.

Grants received for the production of property, plant and equipment are recognised as deferred income. For depreciable assets, the grant is recognised in step with the depreciation of the asset.

Assets held under finance leases are recognised at the time of acquisition in the balance sheet at the lower of fair value and present value of future lease payments. Assets held under finance leases are subsequently recognised and measured as other property, plant and equipment.

Gains and losses from the disposal of property, plant and equipment are determined as the difference between the selling price less selling costs and the carrying amount on the date of disposal.

### **Investments**

Other receivables comprise deposits, which are valued at amortised cost.

### **Impairment of assets**

The carrying amount of non-current assets which are not measured at fair value is assessed annually for indications of impairment over and above what is reflected in depreciation/amortisation.

If there are indications of impairment, an impairment test is conducted of individual assets or groups of assets. The assets or groups of assets are impaired to the lower of recoverable amount and carrying amount.

The higher of net selling price and value in use is used as the recoverable amount. The value in use is determined as the present value of expected net cash flows from the use of the asset or group of assets as well as expected net cash flows from the sale of the asset or group of assets after the expiry of their useful lives.



### **Receivables**

Receivables are measured at amortised cost, which usually corresponds to the nominal value, less write-downs for bad debts.

Write-downs for bad debts are determined on the basis of an assessment of the individual receivables.

### **Prepayments**

Prepayments comprise costs incurred in respect of the next financial year.

### **Cash**

Cash consist of bank deposits and cash at bank and in hand.

### **Current and deferred taxes**

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

Joint taxation contributions payable and receivable are recognised as income tax in the balance sheet under receivables or payables.

Deferred tax liabilities and deferred tax assets are computed on the basis of all temporary differences between the carrying amount and tax base of assets and liabilities. However, deferred tax is not recognised on temporary differences relating to goodwill which is non-amortisable for tax purposes and other items where temporary differences, except for acquisitions, have arisen at the date of acquisition without affecting either the net profit or loss for the year or the taxable income. In cases where the tax value can be determined according to different taxation rules, deferred tax is measured on the basis of the management's intended use of the asset or settlement of the liability.

Deferred tax assets are recognised, following an assessment, at the expected realisable value through offsetting against deferred tax liabilities or tax on future earnings.

Deferred tax is measured on the basis of the tax rules and at the tax rates which, according to the legislation in force at the balance sheet date, will be applicable when the deferred tax is expected to crystallise as current tax.



### **Liabilities**

Non-current liabilities are measured at cost at the time of contracting such payables (raising of the loan). The liabilities are subsequently measured at amortised cost where capital losses and loan expenses are recognised in the income statement as a financial expense over the term of the liability on the basis of the calculated effective interest rate in force at the time of contracting the liability.

Current liabilities are also measured at amortised cost, which usually corresponds to the nominal value of the liability.



### 1. Capital position and financial development

In the financial statements for the financial year 01.01.15 - 31.12.15, it is important to note the following estimates and assessments as they have had significant influence on the assets and liabilities recognised in the financial statements:

Chiesi Farmaceutici S.p.A. that in August 2013 acquired Zymenex Holding A/S and its subsidiaries, including Zymenex A/S, has issued a letter of support stating that they will continue to provide financial support to enable Zymenex Holding A/S, Zymenex A/S and ACE Biosciences A/S to continue their development, trading and investments and to meet all their financial obligations as they fall due for payment for the foreseeable future and at least for the period until 31 December 2016. The refunding is included in other operating income.

	2015 DKK	2014 DKK '000
<b>2. Other financial income</b>		
Financial income from group enterprises	0	216
Foreign exchange gains	13.722	3
<b>Total</b>	<b>13.722</b>	<b>219</b>

### 3. Other financial expenses

Financial expenses for group enterprises	3.404.354	4.060
Interest expenses	25.184	45
Foreign exchange losses	158.623	179
<b>Total</b>	<b>3.588.161</b>	<b>4.284</b>



	2015 DKK	2014 DKK '000
<b>4. Taxes</b>		
Tax on profit for the year	-657.997	0
Total	-657.997	0

**5. Property, plant and equipment**

Figures in DKK	Plant and machinery	Fixtures and fittings, tools and equipment
Cost as at 31.12.14	4.382.034	416.484
Additions during the year	1.992.587	123.791
Cost as at 31.12.15	6.374.621	540.275
Depreciation and impairment losses as at 31.12.14	3.574.571	304.968
Depreciation during the year	789.757	12.747
Depreciation and impairment losses as at 31.12.15	4.364.328	317.715
Carrying amount as at 31.12.15	2.010.293	222.560

**6. Other receivables**

Figures in DKK	Other receivables
Cost as at 31.12.14	113.918
Additions during the year	779
Cost as at 31.12.15	114.697
Carrying amount as at 31.12.15	114.697

**7. Equity**

Figures in DKK	Share capital	Retained earnings
<i>Statement of changes in equity for the period 01.01.14 - 31.12.14</i>		
Balance as at 31.12.13	2.500.000	-68.321.765
Proposed distribution of net profit	0	1.065.781
Balance as at 31.12.14	2.500.000	-67.255.984

*Statement of changes in equity for the period 01.01.15 - 31.12.15*

Statement of changes in equity for the period as at 01.01.15	2.500.000	-67.255.984
Proposed distribution of net profit	0	656.421
Statement of changes in equity for the period as at 31.12.15	2.500.000	-66.599.563

There have been no changes in share capital during the four preceding financial years.

The share capital consists of:

	Quantity	Nominal value
Shares	2.500.000	1

**8. Payables**

	Repayment first year DKK	Outstanding debt after 5 years DKK	Total payables at 31.12.15 DKK	Total payables at 31.12.14 DKK '000
Payables group enterprises	0	74.363.606	74.363.606	70.470
Total	0	74.363.606	74.363.606	70.470

**9. Contingent liabilities**

The company is taxed jointly with the other Danish companies in the group, and, as from the 2013 financial year, the company is liable for tax claims on a pro rata basis and must comply with any obligations to withhold tax at source on interest, royalties and dividends for the jointly taxed companies. The maximum liability totals an amount corresponding to the share of the capital in the company which is owned directly or indirectly by the ultimate parent. As at the balance sheet date, the total liability amounts to DKK 0.

**10. Contractual obligations**

The company has entered into operating lease agreements with the non-cancellable period of up to 27 months. At the balance sheet date is estimated lease obligations total t.DKK 1.739.



**11. Ownership**

Basis of influence

Controlling influence:

Chiesi Farmaceutici S.p.A.

Parent Company

Zymenex A/S is included in the consolidated financial statements of the parent, Chiesi Farmaceutici S.p.A.