

Zymenex Holding A/S

Roskildevej 12 C, 3400 Hillerød
CVR no. 21 00 23 48

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

A blue ink signature of Lars Terp, consisting of a stylized first name and a more complex surname.

Lars Terp
Advokat



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The company

Zymenex Holding A/S
Roskildevej 12 C
3400 Hillerød
Tel.: 48 25 00 54
Registered office: Hillerød
CVR no.: 21 00 23 48

Board of Directors

Jens Fogh, chairman
Paolo Patri
Claes Magnus Andersson
Natallia Misuna

Executive Board

CEO Paolo Patri

Auditors

KPMG
Statsautoniseret Revisionspartnerselskab

Parent company

Chiesi Farmaceutici S.p.A.

Zymenex Holding A/S

Company information etc.



Subsidiaries

Zymenex A/S, Hillerød
ACE Biosciences 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 Holding 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


Natalia Misuna


Paolo Patri


Claes Magnus Andersson

To the shareholder of Zymenex Holding A/S

REPORT ON THE FINANCIAL STATEMENTS

We have audited the financial statements of Zymenex Holding 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 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

Zymenex Holding A/S is a holding company for the Zymenex Group. The Zymenex Group consists of Zymenex Holding A/S, Zymenex A/S and ACE BioSciences A/S and three Swedish affiliates, Zymenex Holding A/S Danmark Filial Sverige, Zymenex A/S Danmark Filial Sverige and ECA-DK Bio, Filial av ACE BioSciences A/S, Danmark.

As in previous years the main activity of the group has been the development of enzyme replacement therapy for the treatment of rare genetic diseases.

Project status

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

The Zymenex Group also develops enzyme replacement therapy (ERT) for the lysosomal disease Globoid Cell Leukodystrophy (Krabbe Disease) and for Aspartylglucosaminuria (AGU disease). Both projects are in pre-clinical development.

At the end of 2015, employees numbered 15, 8 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 2,714,790 against DKK 3,778 thousand for the period 01.01.14 - 31.12.14. The balance sheet showed equity of DKK 311,661,175.

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.

Important events occurring after the end of the financial year

No events materially affecting the financial position of the company have occurred after the end of the financial year.

The company's expected development

Chiesi Farmaceutici S.p.A. that in August 2013 acquired Zymenex Holding A/S and its subsidiaries, 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.

Note	2015 DKK	2014 DKK
Administrative expenses	-959.931	-152
Other operating income	959.930	151
Operating profit/loss	-1	-1
1 Other financial income	3.596.891	5.383
2 Other financial expenses	-48.145	-725
Total net financials	3.548.746	4.658
Profit/loss before tax	3.548.745	4.657
3 Tax on profit/loss for the year	-833.955	-879
Profit for the year	2.714.790	3.778
Proposed appropriation account		
Proposed dividend for the financial year	8.900.000	29.774
Retained earnings	-6.185.210	-25.996
Total	2.714.790	3.778

ASSETS

Note		31.12.15 DKK	31.12.14 DKK
4	Investments in group enterprises	235.358.077	235.358
	Receivables from group enterprises	74.525.306	70.470
	Total investments	309.883.383	305.828
	Total non-current assets	309.883.383	305.828
	Receivables from group enterprises	0	33.827
	Other receivables	0	371
	Total receivables	0	34.198
	Cash	3.473.847	380
	Total current assets	3.473.847	34.578
	Total assets	313.357.230	340.406

EQUITY AND LIABILITIES

Note		31.12.15 DKK	31.12.14 DKK
	Share capital	157.000.000	157.000
	Retained earnings	145.761.175	151.946
	Proposed dividend for the financial year	8.900.000	29.774
5	Total equity	311.661.175	338.720
	Trade payables	0	120
	Payables group enterprises	833.955	1.482
	Other payables	862.100	84
	Total short-term payables	1.696.055	1.686
	Total payables	1.696.055	1.686
	Total equity and liabilities	313.357.230	340.406

⁶ Contingent liabilities

⁷ Related parties



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.

In accordance with Section 110 of the Danish Financial Statements Act, the company has decided not to prepare consolidated financial statements.

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.

INCOME STATEMENT

Gross profit

Gross profit comprises administrative expenses and other operating income.

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 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.

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.

Amortisation of capital losses and loan costs relating to financial assets and liabilities is recognised on an ongoing basis as financial expenses and financial income, respectively.

Dividend from equity investments recognised at cost is recognised as income in the parent in the financial year in which the dividend is declared.

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. The parent is the administration company for the joint taxation and thus settles all income tax payments with the tax authorities.

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

Investments

Investments in subsidiaries are measured in the balance sheet at cost less any impairment.

Gains or losses on the divestment of subsidiaries are determined as the difference between the consideration and the carrying amount.



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.

Cash

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

Equity

The proposed dividend for the financial year is recognised as a special item under equity.

Current and deferred taxes

As the administration company, the company assumes liability for the jointly taxed enterprises' income tax payments to the tax authorities in step with the joint taxation contributions being paid by the jointly taxed enterprises.

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

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

	2015 DKK	2014 DKK
1. Other financial income		
Financial income from group enterprises	3.593.907	5.329
Interest income	1	8
Foreign exchange gains	2.983	46
Total	3.596.891	5.383
2. Other financial expenses		
Financial expenses for group enterprises	0	715
Interest expenses	45.564	5
Foreign exchange losses	2.581	5
Total	48.145	725
3. Taxes		
Tax on profit for the year	833.955	879
Total	833.955	879

31.12.15	31.12.14
DKK	DKK

4. Investments in group enterprises

Cost as at 31.12.14	235.358.077	235.358
Cost as at 31.12.15	235.358.077	235.358
Carrying amount as at 31.12.15	235.358.077	235.358

Group enterprises

Name	Ownership interest	Equity	Net profit for the year
Zymenex A/S, Hillerød	100%	-64.099.563	656.421
ACE Biosciences A/S, Hillerød	100%	3.830.037	414.117

5. Equity

Figures in DKK	Share capital	Share premium	Retained earnings	Proposed dividend for the financial year
<i>Statement of changes in equity for the period 01.01.14 - 31.12.14</i>				
Balance as at 31.12.13	156.922.087	0	146.771.529	0
Capital contributed on establishment	77.913	0	0	0
Capital increase	0	31.170.982	0	0
Proposed distribution of net profit	0	0	-25.996.126	29.774.400
Transfer from share premium	0	-31.170.982	31.170.982	0
Balance as at 31.12.14	157.000.000	0	151.946.385	29.774.400

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	157.000.000	0	151.946.385	29.774.400
Dividend paid	0	0	0	-29.774.400
Proposed distribution of net profit	0	0	-6.185.210	8.900.000
Statement of changes in equity for the period as at 31.12.15	157.000.000	0	145.761.175	8.900.000

There have been changes in share capital in 2013 TDKK 14.896 and in 2014 TDKK 78.

The share capital consists of:

	Quantity	Nominal value
Shares	157.000.000	1



6. 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 together with the other jointly taxed companies for the total income tax and must comply with any obligations to withhold tax at source on interest, royalties and dividends for the jointly taxed companies. As at the balance sheet date, the total liability amounts to DKK 0. The liability furthermore includes any subsequent corrections to the calculated tax liability as a consequence of changes made to the jointly taxable income etc.

7. Ownership

Basis of influence

Controlling influence:

Chiesi Farmaceutici S.p.A.

Parent company

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