

Zymenex Holding A/S
Roskildevej 12 C
3400 Hillerød
Central Business Registration No
21002348

Annual report 2016

The Annual General Meeting adopted the annual report on 27.04.2017

Chairman of the General Meeting

Name: Lars Terp, Lawyer

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Entity details

Entity

Zymenex Holding A/S
Roskildevej 12 C
3400 Hillerød

Central Business Registration No: 21002348

Registered in: Hillerød

Financial year: 01.01.2016 - 31.12.2016

Phone: 48250054

Board of Directors

Jens Morten Fogh, Chairman
Claes Magnus Andersson
Paolo Patri

Executive Board

Paolo Patri, CEO

Auditors

Deloitte Statsautoriseret Revisionspartnerselskab
Weidekampsgade 6
Postboks 1600
0900 København C

Statement by Management on the annual report

The Board of Directors and the Executive Board have today considered and approved the annual report of Zymenex Holding A/S for the financial year 01.01.2016 - 31.12.2016.

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

In our opinion, the financial statements give a true and fair view of the Entity's financial position at 31.12.2016 and of the results of its operations for the financial year 01.01.2016 - 31.12.2016.

We believe that the management commentary contains a fair review of the affairs and conditions referred to therein.

We recommend the annual report for adoption at the Annual General Meeting.

Hillerød, 20.04.2017

Executive Board

Paolo Patri
CEO

Board of Directors

Jens Morten Fogh
Chairman

Claes Magnus Andersson

Paolo Patri

Independent auditor's report

To the shareholders of Zymenex Holding A/S

Opinion

We have audited the financial statements of Zymenex Holding A/S for the financial year 01.01.2016 - 31.12.2016, which comprise the 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.

In our opinion, the financial statements give a true and fair view of the Entity's financial position at 31.12.2016 and of the results of its operations for the financial year 01.01.2016 - 31.12.2016 in accordance with 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 this auditor's report. We are independent of the Entity in accordance with the International Ethics Standards Board of Accountants' Code of Ethics for Professional Accountants (IESBA Code) and the additional requirements applicable in Denmark, and we have fulfilled our other ethical responsibilities in accordance with these requirements. We believe that the audit evidence we have obtained is sufficient and appropriate to provide a basis for our opinion.

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 the Danish Financial Statements Act, and for such internal control as Management determines is necessary to enable the preparation of financial statements that are free from material misstatement, whether due to fraud or error.

In preparing the financial statements, Management is responsible for assessing the Entity's ability to continue as a going concern, for disclosing, as applicable, matters related to going concern, and for using the going concern basis of accounting in preparing the financial statements unless Management either intends to liquidate the Entity 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 the 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 these financial statements.

As part of an audit conducted in accordance with ISAs and the additional requirements applicable in Denmark, we exercise professional judgement and maintain professional scepticism throughout the audit. We also:

Independent auditor's report

- 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 Entity'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 Entity's ability to continue as a going concern. If we conclude that a material uncertainty exists, we are required to draw attention in our auditor's report to the related disclosures in the financial statements or, if such disclosures are inadequate, to modify our opinion. Our conclusions are based on the audit evidence obtained up to the date of our auditor's report. However, future events or conditions may cause the Entity to cease to continue as a going concern.
- Evaluate the overall presentation, structure and content of the financial statements, including the disclosures in the notes, and whether the financial statements represent the underlying transactions and events in a manner that gives a true and fair view.

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.

Statement on the management commentary

Management is responsible for the management commentary.

Our opinion on the financial statements does not cover the management commentary, and we do not express any form of assurance conclusion thereon.

In connection with our audit of the financial statements, our responsibility is to read the management commentary and, in doing so, consider whether the management commentary is materially inconsistent with the financial statements or our knowledge obtained in the audit or otherwise appears to be materially misstated.

Moreover, it is our responsibility to consider whether the management commentary provides the information required under the Danish Financial Statements Act.

Independent auditor's report

Based on the work we have performed, we conclude that the management commentary is in accordance with the financial statements and has been prepared in accordance with the requirements of the Danish Financial Statements Act. We did not identify any material misstatement of the management commentary.

København, 20.04.2017

Deloitte

Statsautoriseret Revisionspartnerselskab
Central Business Registration No: 33963556

Flemming Larsen
State Authorised Public Accountant

Management commentary

Primary 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 Branches, Zymenex Holding A/S Danmark Filial Sverige, Zymenex A/S Danmark Filial Sverige and EAC-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 Group's pipeline has been focused on Velmanase Alfa, Lamzede (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 disease is life-threatening and there is no treatment today. Previously, Zymenex has been a partner of European research groups under the EU grant "Euraman" for the development of the enzyme and together with this scientific group has 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 a EU Framework 7 supported project group "ALPHA-MAN" with the continued development of Lamzede (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. Final phase 3 Clinical Trial results were presented in Q2 2016 and filing to EMA for obtaining marketing authorisation was performed in Q3 2016. Lamzede 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 phase.

At the end of 2016, headcount numbered 13 and 1 ad-interim worker, hereof 8 worked in the Swedish branches.

Development in activities and finances

The income statement for the period 01.01.16 – 31.12.16 showed a profit of DKK 2,300,156 against DKK 2,714,790 for the period 01.01.15 – 31.12.15. The balance sheet showed equity of DKK 305.061.331 as at 31.12.16.

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.

At the annual general meeting held in subsidiary A/S, the board of directors proposed to distribute a receivable with group companies DKK 153 million to Zymenex Holding A/S.

At the annual general meeting held at the same date, the board of directors propose to distribute DKK 148 million to parent company of Zymenex Holding A/S.

Management commentary

Events after the balance sheet date

No events have occurred after the balance sheet date to this date, which would influence the evaluation of this annual report.

Income statement for 2016

	<u>Notes</u>	<u>2016 DKK</u>	<u>2015 DKK</u>
Revenue		209.419	959.930
Gross profit/loss		209.419	959.930
Administrative costs		(209.419)	(959.931)
Operating profit/loss		0	(1)
Other financial income	1	2.960.090	3.596.891
Other financial expenses	2	(9.305)	(48.145)
Profit/loss before tax		2.950.785	3.548.745
Tax on profit/loss for the year		(650.629)	(833.955)
Profit/loss for the year		2.300.156	2.714.790
Proposed distribution of profit/loss			
Ordinary dividend for the financial year		148.000.000	8.900.000
Retained earnings		(145.699.844)	(6.185.210)
		2.300.156	2.714.790

Balance sheet at 31.12.2016

	<u>Notes</u>	<u>2016 DKK</u>	<u>2015 DKK</u>
Investments in group enterprises		235.358.077	235.358.077
Receivables from group enterprises		<u>69.126.266</u>	<u>74.525.306</u>
Fixed asset investments	3	<u>304.484.343</u>	<u>309.883.383</u>
Fixed assets		<u>304.484.343</u>	<u>309.883.383</u>
Cash		<u>7.236.409</u>	<u>3.473.847</u>
Current assets		<u>7.236.409</u>	<u>3.473.847</u>
Assets		<u>311.720.752</u>	<u>313.357.230</u>

Balance sheet at 31.12.2016

	<u>Notes</u>	<u>2016 DKK</u>	<u>2015 DKK</u>
Contributed capital	4	157.000.000	157.000.000
Retained earnings		61.331	145.761.175
Proposed dividend		<u>148.000.000</u>	<u>8.900.000</u>
Equity		<u>305.061.331</u>	<u>311.661.175</u>
Payables to group enterprises		4.924.139	0
Joint taxation contribution payable	5	1.484.584	833.955
Other payables		<u>250.698</u>	<u>862.100</u>
Current liabilities other than provisions		<u>6.659.421</u>	<u>1.696.055</u>
Liabilities other than provisions		<u>6.659.421</u>	<u>1.696.055</u>
Equity and liabilities		<u>311.720.752</u>	<u>313.357.230</u>
Unrecognised rental and lease commitments	6		
Contingent liabilities	7		

Statement of changes in equity for 2016

	Contributed capital DKK	Retained earnings DKK	Proposed dividend DKK	Total DKK
Equity beginning of year	157.000.000	145.761.175	8.900.000	311.661.175
Ordinary dividend paid	0	0	(8.900.000)	(8.900.000)
Proposed dividend	0	0	148.000.000	148.000.000
Profit/loss for the year	0	(145.699.844)	0	(145.699.844)
Equity end of year	157.000.000	61.331	148.000.000	305.061.331

Notes

	2016	2015
	DKK	DKK
1. Other financial income		
Financial income arising from group enterprises	2.958.094	3.593.907
Exchange rate adjustments	1.996	2.984
	2.960.090	3.596.891

	2016	2015
	DKK	DKK
2. Other financial expenses		
Interest expenses	5.461	45.564
Exchange rate adjustments	3.844	2.581
	9.305	48.145

3. Fixed asset investments

	Registered in	Corpo- rate form	Equity inte- rest %	Equity DKK	Profit/loss DKK
Investments in group enterprises comprise:					
Zymenex A/S	Hillerød	A/S	100,0	230.260.000	294.360.000
Ace BioSciences A/S	Hillerød	A/S	100,0	12.250.000	8.375.000

Receivables from group enterprises, there have not been agreed any final settlement date for the intercompany receivable. The subsidiary company are making payments when it have adequate Cash.

Notes

	<u>Number</u>	<u>Par value DKK</u>	<u>Nominal value DKK</u>
4. Contributed capital			
Share Capital	<u>157.000.000</u>	1	<u>157.000.000</u>
	<u>157.000.000</u>		<u>157.000.000</u>

5. Joint taxation contribution payable

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.

6. Unrecognised rental and lease commitments

The Entity has not entered into any operating lease agreements.

7. Contingent liabilities

The Entity serves as an administration company in a Danish joint taxation arrangement. According to the joint taxation provisions of the Danish Corporation Tax Act, the Entity is therefore liable from the financial year 2013 for income taxes etc for the jointly taxed entities, and from 1 July 2012 also for obligations, if any, relating to the withholding of tax on interest, royalties and dividends for these entities.

Accounting policies

Reporting class

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

The accounting policies applied to these financial statements are consistent with those applied last year.

Recognition and measurement

Assets are recognised in the balance sheet when it is probable as a result of a prior event that future economic benefits will flow to the Entity, and the value of the asset can be measured reliably.

Liabilities are recognised in the balance sheet when the Entity has a legal or constructive obligation as a result of a prior event, and it is probable that future economic benefits will flow out of the Entity, and the value of the liability can be measured reliably.

On initial recognition, assets and liabilities are measured at cost. Measurement subsequent to initial recognition is effected as described below for each financial statement item.

Anticipated risks and losses that arise before the time of presentation of the annual report and that confirm or invalidate affairs and conditions existing at the balance sheet date are considered at recognition and measurement.

Income is recognised in the income statement when earned, whereas costs are recognised by the amounts attributable to this financial year.

Income statement

Revenue

Revenue consist of founding from the subsidiaries for the administrations cost.

Administrative costs

Administrative costs comprise expenses incurred for the Entity's administrative function.

Other financial income

Other financial income comprises dividends etc received on other investments, interest income, including interest income on receivables from group enterprises, net capital gains on, payables and transactions in foreign currencies, as well as tax relief under the Danish Tax Prepayment Scheme etc.

Other financial expenses

Other financial expenses comprise interest expenses, including interest expenses on payables to group enterprises, net capital losses on payables and transactions in foreign currencies, as well as tax surcharge under the Danish Tax Prepayment Scheme etc.

Accounting policies

Tax on profit/loss for the year

Tax for the year, which consists of current tax for the year and changes in deferred tax, is recognised in the income statement by the portion attributable to the profit for the year and recognised directly in equity by the portion attributable to entries directly in equity.

Balance sheet

Investments in group enterprises

Investments in group enterprises are measured at cost and are written down to the lower of recoverable amount and carrying amount.

Receivables

Receivables are measured at amortised cost, usually equalling nominal value less writedowns for bad and doubtful debts.

Cash

Cash comprises cash in hand and bank deposits.

Dividend

Dividend is recognised as a liability at the time of adoption at the general meeting. Proposed dividend for the financial year is disclosed as a separate item in equity. Extraordinary dividend adopted in the financial year is recognised directly in equity when distributed and disclosed as a separate item in Management's proposal for distribution of profit/loss.

Other financial liabilities

Other financial liabilities are measured at amortised cost, which usually corresponds to nominal value.

Income tax receivable or payable

Current tax payable or receivable is recognised in the balance sheet, stated as tax computed on this year's taxable income, adjusted for prepaid tax