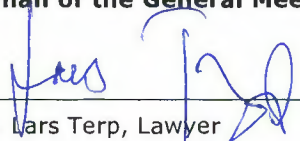


**Ace BioSciences A/S**  
c/o Bech-Bruun (AJU/LAT)  
Langelinie Allé 35  
2100 København Ø  
Central Business Registration No  
25616715

## **Annual report 2018**

The Annual General Meeting adopted the annual report on 15.04.2019

### **Chairman of the General Meeting**

  
Name: Lars Terp, Lawyer

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## Entity details

### Entity

Ace BioSciences A/S  
c/o Bech-Bruun (AJU/LAT) Langelinie Allé 35  
2100 København Ø

Central Business Registration No: 25616715

Registered in: København Ø

Financial year: 01.01.2018 - 31.12.2018

Phone: 48250054

### Board of Directors

Mark Parry-Billings, Chairman

Claes Magnus Andersson

Nina Teerés Herne

### Executive Board

Nina Teerés Herne, 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 Ace BioSciences A/S for the financial year 01.01.2018 - 31.12.2018.

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 31.12.2018 and of the results of its operations for the financial year 01.01.2018 - 31.12.2018.

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

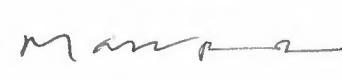
We recommend the annual report for adoption at the Annual General Meeting.

København, 22.03.2019

### Executive Board

x   
Nina Teerés Herne  
CEO

### Board of Directors

x   
Mark Parry-Billings  
Chairman

x   
Claes Magnus Andersson

x   
Nina Teerés Herne

## Independent auditor's report

### To the shareholders of Ace BioSciences A/S

#### Opinion

We have audited the financial statements of Ace BioSciences A/S for the financial year 01.01.2018 - 31.12.2018, 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 31.12.2018 and of the results of its operations for the financial year 01.01.2018 - 31.12.2018 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, 22.03.2019

### **Deloitte**

Statsautoriseret Revisionspartnerselskab

Central Business Registration No: 33963556



Flemming Larsen

State Authorised Public Accountant

Identification number (MNE) mne27790

## Management commentary

### Primary activities

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

The ACE pipeline has been focused mainly on projects for the therapeutic indications of lysosomal disease Globoid Cell Leukodystrophy (Krabbe Disease) and Aspartylglucosaminuria (AGU disease).

### Development in activities and finances

The income statement for the period 01.01.18 – 31.12.18 showed a profit of t.DKK 658 against t.DKK 545 for the period 01.01.17 – 31.12.17. The balance sheet showed equity of t.DKK 15.179 as at 31.12.18.

Mid-February 2018 the board of directors determined to cease the activity of Ace BioSciences A/S, and transfer relevant activity to one or more entities within the Chiesi Farmaceutici S.p.A. Group. The transfer was effective June 2018. The company has no commercial activity. Management will commence a solvent liquidation of the company. Management believes that the company can be liquidated by means of a solvent liquidation and has therefore prepared the 2018 financial statements using the accounting policies applied in prior periods.

### 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 2018

|   | <u>Notes</u> | <u>2018</u><br><u>DKK'000</u> | <u>2017</u><br><u>DKK'000</u> |
|---|--------------|-------------------------------|-------------------------------|
| Revenue                                     |              | 14.243                        | 17.972                        |
| <b>Gross profit/loss</b>                    |              | <b>14.243</b>                 | <b>17.972</b>                 |
| Research and development costs              |              | (10.248)                      | (14.670)                      |
| Administrative costs                        |              | (2.882)                       | (2.052)                       |
| Other operating income                      |              | 719                           | 0                             |
| <b>Operating profit/loss</b>                |              | <b>1.832</b>                  | <b>1.250</b>                  |
| Other financial income                      |              | 0                             | 55                            |
| Other financial expenses                    | 2            | (1.066)                       | (434)                         |
| <b>Profit/loss before tax</b>               |              | <b>766</b>                    | <b>871</b>                    |
| Tax on profit/loss for the year             |              | (108)                         | (326)                         |
| <b>Profit/loss for the year</b>             |              | <b>658</b>                    | <b>545</b>                    |
| <b>Proposed distribution of profit/loss</b> |              |                               |                               |
| Retained earnings                           |              | 658                           | 545                           |
|   |              | <b>658</b>                    | <b>545</b>                    |

## Balance sheet at 31.12.2018

|  | <u>Notes</u> | <u>2018</u><br><u>DKK'000</u> | <u>2017</u><br><u>DKK'000</u> |
|--|--------------|-------------------------------|-------------------------------|
| Other fixtures and fittings, tools and equipment |              | 0                             | 1.495                         |
| <b>Property, plant and equipment</b>             | 3            | <u>0</u>                      | <u>1.495</u>                  |
| <b>Fixed assets</b>                              |              | <u>0</u>                      | <u>1.495</u>                  |
| Receivables from group enterprises               |              | 787                           | 4.426                         |
| Other receivables                                |              | 910                           | 900                           |
| Income tax receivable                            |              | 20                            | 0                             |
| Joint taxation contribution receivable           | 4            | 7.336                         | 7.536                         |
| <b>Receivables</b>                               |              | <u>9.053</u>                  | <u>12.862</u>                 |
| <b>Cash</b>                                      |              | <u>9.231</u>                  | <u>1.697</u>                  |
| <b>Current assets</b>                            |              | <u>18.284</u>                 | <u>14.559</u>                 |
| <b>Assets</b>                                    |              | <u>18.284</u>                 | <u>16.054</u>                 |

## Balance sheet at 31.12.2018

|   | <u>Notes</u> | <u>2018</u><br><u>DKK'000</u> | <u>2017</u><br><u>DKK'000</u> |
|---|--------------|-------------------------------|-------------------------------|
| Contributed capital                                 | 5            | 500                           | 500                           |
| Retained earnings                                   |              | 14.679                        | 12.660                        |
| <b>Equity</b>                                       |              | <b>15.179</b>                 | <b>13.160</b>                 |
| Trade payables                                      |              | 26                            | 333                           |
| Payables to group enterprises                       |              | 2.937                         | 1.102                         |
| Income tax payable                                  |              | 0                             | 272                           |
| Other payables                                      |              | 142                           | 1.187                         |
| <b>Current liabilities other than provisions</b>    |              | <b>3.105</b>                  | <b>2.894</b>                  |
| <b>Liabilities other than provisions</b>            |              | <b>3.105</b>                  | <b>2.894</b>                  |
| <b>Equity and liabilities</b>                       |              | <b>18.284</b>                 | <b>16.054</b>                 |
| Uncertainty relating to recognition and measurement | 1            |                               |                               |
| Unrecognised rental and lease commitments           | 6            |                               |                               |
| Contingent liabilities                              | 7            |                               |                               |

## Statement of changes in equity for 2018

|                           | <b>Contributed<br/>capital<br/>DKK'000</b> | <b>Retained<br/>earnings<br/>DKK'000</b> | <b>Total<br/>DKK'000</b> |
|---------------------------|--|--|--------------------------|
| Equity beginning of year  | 500  | 12.660                                   | 13.160                   |
| Exchange rate adjustments | 0  | 1.361                                    | 1.361                    |
| Profit/loss for the year  | 0  | 658                                      | 658                      |
| <b>Equity end of year</b> | <b>500</b>                                 | <b>14.679</b>                            | <b>15.179</b>            |

## Notes

### 1. Uncertainty relating to recognition and measurement

Management believes that the company can be liquidated by means of a solvent liquidation and has therefore prepared the 2017 financial statements using the accounting policies applied in prior periods.

|   | <b>2018</b>    | <b>2017</b>   |
|---|----------------|---|
|   | <b>DKK'000</b> | <b>DKK'000</b>  |
| <b>2. Other financial expenses</b>                        |                |   |
| Interest expenses   | 1.066          | 434   |
|   | <b>1.066</b>   | <b>434</b>  |
|   |                | <b>Other<br/>fixtures and<br/>fittings,<br/>tools and<br/>equipment<br/>DKK'000</b> |
| <b>3. Property, plant and equipment</b>                   |                |   |
| Cost beginning of year                                    |                | 2.214   |
| Disposals   |                | (2.214)   |
| <b>Cost end of year</b>                                   |                | <b>0</b>  |
| Depreciation and impairment losses beginning of the year  |                | (719)   |
| Depreciation for the year                                 |                | (175)   |
| Reversal regarding disposals                              |                | 894   |
| <b>Depreciation and impairment losses end of the year</b> |                | <b>0</b>  |
| <b>Carrying amount end of year</b>                        |                | <b>0</b>  |

### 4. Joint taxation contribution receivable

The company is taxed jointly with 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.

## Notes

|                               | <u>Number</u> | <u>Par value<br/>DKK'000</u> | <u>Nominal<br/>value<br/>DKK'000</u> |
|-------------------------------|---------------|------------------------------|--------------------------------------|
| <b>5. Contributed capital</b> |               |                              |                                      |
| Share Capital                 | 500           | 1                            | 500                                  |
|                               | <b>500</b>    |                              | <b>500</b>                           |

### 6. Unrecognised rental and lease commitments

The Entity has not entered into any rental or lease agreements as of 31.12.2018.

### 7. Contingent liabilities

The Entity participates in a Danish joint taxation arrangement in which Zymenx Holding A/S serves as the administration company. According to the joint taxation provisions of the Danish Corporation Tax Act, the Entity is therefore alternatively liable from the financial year 2013 for income taxes etc for the jointly taxed entities, but only for the share by which the Entity is included in the Group, and from 1 July 2012 also for obligations, if any, relating to the withholding of tax on interest, royalties and dividends for these entities. The total known net liability of the jointly taxed entities under the joint taxation arrangement is evident from the administration company's financial statements.

## 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.

Management believes that the contemplated liquidation does not impact the measurement of assets and liabilities as at 31.12.2018

### 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 parent company Chiesi Farmaceutici for the research and development cost as well as administration cost that the company have based on a cost plus agreement.

#### Research and development costs

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

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

#### Administrative costs

Administrative costs comprise expenses incurred for the Entity's administrative functions, including wages and salaries for administrative staff and Management, stationery and office supplies as well as amortisation, depreciation and impairment losses relating to property, plant and equipment.

## Accounting policies

### Other operating income

Other operating income comprises income of a secondary nature as viewed in relation to the Entity's primary activities.

### 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 securities, payables and transactions in foreign currencies, amortisation of financial assets 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 securities, payables and transactions in foreign currencies, amortisation of financial liabilities as well as tax surcharge under the Danish Tax Prepayment Scheme etc.

### 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

#### Property, plant and equipment

Plant and machinery as well as other fixtures and fittings, tools and equipment are measured at cost less accumulated depreciation and impairment losses.

Cost comprises the acquisition price, costs directly attributable to the acquisition and preparation costs of the asset until the time when it is ready to be put into operation.

The basis of depreciation is cost less estimated residual value after the end of useful life. Straight-line depreciation is made on the basis of the following estimated useful lives of the assets:

|  |           |
|--|-----------|
| Plant and machinery                              | 5 years   |
| Other fixtures and fittings, tools and equipment | 3-5 years |

Estimated useful lives and residual values are reassessed annually.

Items of property, plant and equipment are written down to the lower of recoverable amount and carrying amount.

### Receivables



## Accounting policies

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

### **Income tax payable or receivable**

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.

### **Cash**

Cash comprises cash in hand and bank deposits.

### **Other financial liabilities**

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