santhera

THEIR FUTURE - OUR FOCUS

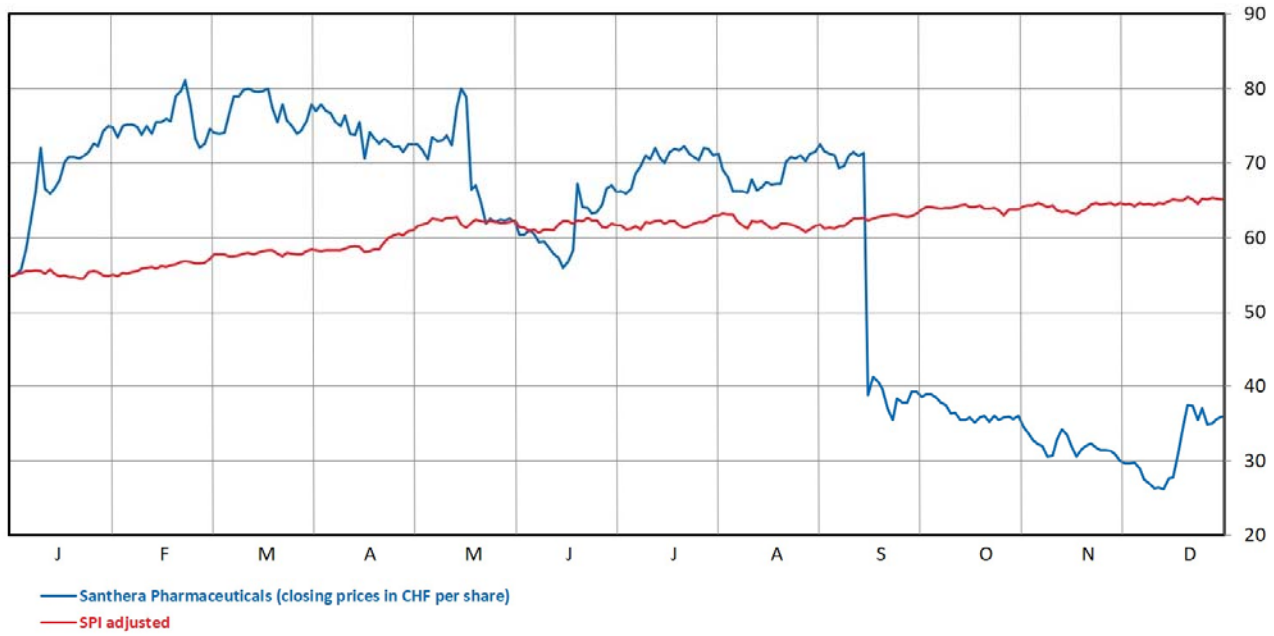
Annual Report 2017

Financial Key Figures

IFRS consolidated, in CHF thousands	**2017**	2016
Net sales	**22,943**	19,033
Operating expenses	**–69,563**	–48,638
Operating result	**–50,454**	–33,127
Net result	**–51,532**	–35,415
Basic and diluted net result per share (in CHF)	**–8.22**	–5.65
Freely available liquid funds at December 31 *	**58,206**	49,815
Net change in cash and cash equivalents	**–4,620**	–27,044

* Cash and cash equivalents and short-term financial assets

Share Price Development in 2017



High	CHF 81.10 (February 22, 2017)
Low	CHF 26.15 (December 11, 2017)
Share price performance in 2017	–32.2%
Share price at year-end	CHF 35.95
Market capitalization at year-end	CHF 226 million
Average trading volume	53,143 shares/day

(based on closing share prices)

Contents

Letter to Our Shareholders

Dear Shareholders,

We are pleased with the progress Santhera made on many fronts in 2017, although not obtaining approval for Duchenne muscular dystrophy in Europe was very disappointing. One of last year's highlights was certainly the strong sales performance for Raxone® (idebenone) in Leber's hereditary optic neuropathy (**LHON**) with a net revenue growth of 21% to CHF 22.9 million, reaching the upper end of our guidance and a clear reflection of our commercial success.

With ambitious growth plans, securing adequate financial resources remains a key task which we achieved with the successful placement of CHF 60 million senior unsecured convertible bonds, executed in February 2017. Strong top-line growth coupled with the available funds provides us the financial flexibility to execute our development, regulatory, commercial and corporate development projects as currently planned.

Across Europe, our country organizations made good progress with the rollout of Raxone, complemented by the new distribution agreements with Pharmathen for Greece and Cyprus and Megapharm for Israel.

Last year Raxone was approved for the treatment of visual impairment in adolescent and adult patients with LHON by the Ministry of Health Israel which underlines our intention of making this treatment also available to patients outside the EU.

Santhera Pharmaceuticals (USA), Inc., our subsidiary in the Boston area, solidified Santhera's presence in the US market and elevated the overseas awareness for our Company.

In addition to a positive impact on top-line sales for LHON, strong and established local operations will put us in an optimal position for a future launch of idebenone in Duchenne muscular dystrophy (**DMD**). With this in mind, we intensified collaborations with clinical opinion leaders and patient advocacy groups, implemented several disease awareness programs and launched a U.S. Expanded Access Program (**EAP**) – referred to as BreatheDMD – with idebenone for patients with DMD.

Following the negative decision from the Committee for Medicinal Products for Human Use (**CHMP**) of the European Medicines Agency on our Type-II-extension application for idebenone for DMD, we will collect additional clinical data in preparation for a new Marketing Authorization Application (**MAA**) for patients unable to use glucocorticoids.

Convinced of the benefits of idebenone in DMD and with the intention to making this medicine available to a larger patient group, we are also conducting the Phase III SIDEROS trial to assess the efficacy in patients with DMD who receive concomitant glucocorticoid treatment. Subject to a positive outcome, this could allow for a label extension to include all patients suffering from DMD, irrespective of their glucocorticoid use status.

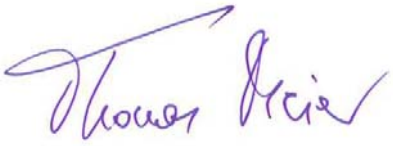
Broadening our clinical stage pipeline and extending the range of indications while maintaining our focus on providing therapies for rare diseases is a prime goal. We took an important step towards achieving this objective with the in-licensing of POL6014, a drug candidate being evaluated for cystic fibrosis (**CF**) and other pulmonary diseases, from Polyphor in February 2018. Developing this promising compound gives us the option to address multiple pulmonary indications with a high unmet medical need. As our development program in DMD already specifically highlights respiratory needs and complications, we gained tremendous know-how in investigating pulmonary function. POL6014 adds a third therapeutic pillar and going forward, Santhera will be focusing on rare neuro-ophthalmological, neuromuscular and pulmonary diseases.

Our Company has grown to over 100 employees located in Europe and the US. We are grateful for their commitment and efforts which form the basis of Santhera's success. Our vision is a unified view of what we stand for and living our values will shape our Company and will help us attract and retain the best talents.

Outlook and Guidance

Santhera will continue to grow its international business and advance its pipeline programs. Based on the sales performance in the first months of the current year the Company reiterates its guidance for net sales of Raxone in the currently approved indication LHON to reach CHF 28-30 million in 2018. The development priorities for 2018 are the clinical studies and data collection to support regulatory filings for idebenone in DMD and advancing the development of the other clinical stage candidates in our pipeline.

We thank you, our esteemed Shareholders, for the confidence you put in our Company and for your continued support.



Elmar Schnee
Chairman

Thomas Meier
Chief Executive Officer

FINANCIAL AND OPERATIONS HIGHLIGHTS

Santhera with Solid Top-line Growth in 2017

Sales growth and successful financing ensure the implementation of Santhera's pipeline and commercial strategies and activities as planned.

Strong sales growth from Raxone for LHON

In 2017, Santhera reported net revenues from product sales of Raxone for LHON of CHF 22.9 million which corresponds to a growth of 21% year-on-year (2016: CHF 19.0 million). The roll-out of Raxone in the approved indication is progressing as planned and the product is currently sold in 20 European countries. By end of 2017, full reimbursement for Raxone in LHON was achieved for 8 European countries. In an additional 12 European countries, Raxone is made available to patients by special reimbursement schemes.

Sales up 21% year-on-year

Operating and net results reflect investment predominantly in clinical development and preparation for regulatory submissions as well as commercial operations

Late stage clinical studies such as the Phase III SIDEROS trial, post-authorization studies for LHON and the preparation of regulatory filings in Europe and the US led to higher development expenses of CHF 26.6 million (2016: CHF 17.7 million). Commercial operations in Europe were expanded to support marketing of Raxone for LHON. In February, US operations were established in the Boston metropolitan area to foster relationships with patient advocacy groups and clinicians, supporting ongoing studies in the US, assembling a NDA filing for DMD and preparing for market entry. This resulted in higher marketing and sales expenses of CHF 28.5 million (2016: CHF 21.1 million) as well as general and administrative expenses of CHF 14.4 million (2016: CHF 9.8 million). In summary, total operating expenses were CHF 69.6 million (2016: CHF 48.6 million) and the operating result amounted to CHF −50.5 million (2016: CHF −33.1 million). For the full-year 2017, Santhera reported a net result of CHF −51.5 million (2016: CHF −35.4 million), in line with the Company's guidance.

Well financed to implement business strategy as planned

In February 2017, Santhera successfully placed CHF 60 million senior unsecured convertible bonds due 2022. These funds are being primarily used for investment into clinical trials with idebenone in DMD to facilitate regulatory filings, for the commercialization of Raxone in the currently approved indication LHON, to advance the pipeline and for other corporate and business development purposes.

Financial strength secures development and commercial plans

As of December 31, 2017, freely available liquid funds (cash and cash equivalents and short-term financial assets) amounted to CHF 58.2 million (December 31, 2016: CHF 49.8 million). In addition, the Company reported CHF 7.5 million of restricted cash designated for the interest payments related to the convertible bonds during the first three years.

OUR VISION

Santhera – Our Promise, Our Values, Our Brand

Santhera is a young and dynamic international pharma company, developing and commercializing innovative treatments for rare diseases.

Our **vision** is to improve the lives of people with rare diseases, by delivering therapeutic options where none previously existed.

santhera
THEIR FUTURE - OUR FOCUS

respect
is the foundation of any partnership

Everything we do at Santhera, we do with **respect**. For the patients that inspire us with their courage, for the scientists at the cutting edge of therapeutic breakthroughs, for all our stakeholders in this important and rewarding enterprise, and for the partnerships with our colleagues.

passion
drives our entrepreneurial aspirations

Passion is the cornerstone of Santhera's aspirations to improve patients' lives. Our focus is on individuals with rare diseases – small groups of patients often overlooked by the wider pharmaceutical industry. We feel strongly that all patients deserve the best care, regardless of the prevalence of their condition.

commitment
to improve patients' lives

The area of rare diseases presents many challenges, and our mission to improve the lives of patients with rare diseases requires great resolve and dedication. Only by ensuring our ongoing **commitment** will we be able to overcome the challenge of bringing new therapies to market.

collaboration
ensures our success

A core pillar that gives the other values cohesion and depth. By fostering a strong team spirit at Santhera, and by combining our efforts with trusted external partners – from clinicians to scientists to patient organizations – we can achieve success through **collaboration**.

accountability
is the basis of our integrity

Where passion gives us drive, **accountability** gives us direction. Our results-driven approach to research, development and commerce with integrity at its heart, ensures we will deliver benefits to all our stakeholders, including effective solutions for the patients affected by rare and devastating diseases.

OUR FOCUS

We Care for Patients with Rare Diseases

Rare diseases, also called orphan diseases, are disorders that affect a small proportion of the population. They often have an underlying genetic defect, are life-altering conditions and difficult to diagnose because of their rarity and a wide variety of symptoms. They do not only touch on patients' individual lives but also impact families, caregivers and society as a whole.

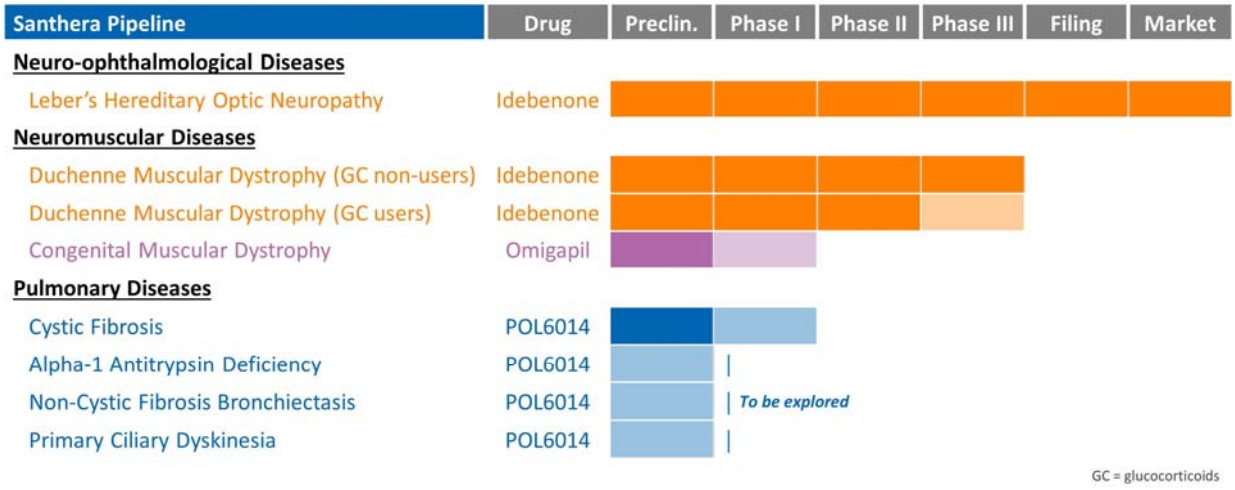
Passionate about providing treatment options for orphan diseases, Santhera focuses its efforts on the development of therapies for rare **neuro-ophthalmological**, **neuromuscular** and **pulmonary** diseases with high unmet medical needs:

NEURO-OPHTHALMOLOGICAL

NEUROMUSCULAR

PULMONARY

The portfolio comprises clinical stage and marketed treatments. Raxone® (idebenone) is authorized in the European Union, Norway, Iceland, Liechtenstein and Israel for the treatment of Leber's hereditary optic neuropathy (**LHON**) and currently commercialized in 20 countries. The most advanced pipeline product, idebenone, is in clinical Phase III for the treatment of Duchenne muscular dystrophy (**DMD**). Omigapil is in clinical trials for the treatment of congenital muscular dystrophy (**CMD**) and POL6014 is being developed in cystic fibrosis (**CF**) and may have benefits in additional neutrophilic pulmonary diseases.



OUR FOCUS

Adopting a Patient-Centric Vision

Rare diseases present unique challenges and adopting a patient-centric perspective is important. The path chosen by Santhera to tune in to patients' needs is by engaging in **disease awareness campaigns** and **working with patient advocacy groups and representatives**.

Give patients a voice

For LHON and DMD, Santhera has launched various initiatives to support the specific patient and caregiver needs, foster disease awareness and education, support diagnosis, improve treatment and care, and work towards better patient outcomes.

At Santhera's initiative, the *1st European Patient Groups Advisory Board meeting for Leber's Hereditary Optic Neuropathy (LHON)* brought together patient groups from 7 different European countries. Besides community-building, the panel discussed best practices and issued recommendations to better fulfill the unmet needs of people affected by LHON.

In cooperation with ophthalmology centers and patient advocacy groups, Santhera is engaging in awareness and educational campaigns, supported by **Fabrizio Sottile**, an Italian Paralympic champion swimmer and ambassador for LHON.

Raising awareness for respiratory function in DMD

Santhera launched educational disease awareness campaigns which underscore the importance of respiratory care for people living with DMD residing in the US. The mission is to help people living with DMD and their families receive practical, in-depth information in one centralized location to help manage respiratory complications. Because knowledge provides assurance and comfort.

"I applaud Santhera for developing such an educational resource focused on the respiratory aspects of Duchenne muscular dystrophy. DMD, one of the most common and devastating types of muscular degeneration, is accompanied by respiratory function decline that typically leads to substantial life limitation," said **Oscar H Mayer**, MD, pediatric pulmonologist focused on treating neuromuscular diseases. For more information visit: www.takeabreath.com.

PRODUCTS AND PIPELINE

Raxone® (idebenone) in LHON

Leber's hereditary optic neuropathy (**LHON**) is a heritable genetic disease which presents itself as rapid, painless loss of central vision, usually leading to permanent bilateral blindness within a few months of the onset of symptoms.

Normal vision

Impaired vision with LHON

Raxone is the first and only treatment approved for LHON

Only one treatment has been approved in Europe for LHON: Raxone (idebenone), an oral medication, for the treatment of visual impairment in adolescent and adult patients with LHON. Raxone can slow down vision loss and promote a recovery of vision.

Phase IV LEROS trial recruiting patients in Europe and USA

The Phase IV open-label, post-approval LEROS trial further assesses the efficacy and safety of Raxone as long-term treatment of patients with LHON. The trial is currently recruiting patients in centers in Europe and the USA. Additional clinical data from this trial, especially from participating US centers, could potentially be used to support a regulatory submission of idebenone in LHON to the FDA.

First Consensus Statement on the Clinical and Therapeutic Management of LHON

In October 2017, the first International Consensus Statement on the Clinical and Therapeutic Management of LHON has been published in the Journal of Neuro-Ophthalmology[1]. The Consensus Statement reflects the independent views from a panel of world-leading experts in the management of patients with LHON, based on the evidence currently available. Amongst other clinical aspects, the Consensus Statement focuses on the use of Raxone (idebenone), the first and only treatment approved by the European Medicines Agency (**EMA**) for the treatment of this heritable form of vision loss.

1 Journal of Neuro-Ophthalmology, Volume 37, Issue 4, p 371–381, December 2017

PRODUCTS AND PIPELINE

Idebenone in DMD

Duchenne muscular dystrophy (**DMD**) is one of the most common and devastating types of muscle degeneration and results in rapidly progressive muscle weakness and wasting, leading to early morbidity and mortality. Weakness of respiratory muscles impairs patients' ability to breathe and leads to respiratory infections, need for antibiotics and hospitalizations. Glucocorticoids, routinely used in DMD, delay the onset of respiratory dysfunction, but do not prevent it, and there is currently no approved treatment to slow the loss of respiratory function. Studies estimate that a majority of patients with DMD die from respiratory complications.

Clinical data show that idebenone preserves the respiratory function and reduces respiratory complications in patients with DMD.

Santhera is focusing on the preservation of respiratory function and the prevention of disease-related respiratory complications in its DMD development program:

Idebenone in DMD patients not able to take glucocorticoids

Cornerstone of the clinical development program in DMD is the positive Phase III DELOS trial which demonstrated statistically significant and clinically relevant evidence that idebenone slows the decline of respiratory function and reduces the risk of bronchopulmonary complications and hospitalization in patients with DMD not using glucocorticoids. Santhera currently collects additional data to support a Marketing Authorization Application (**MAA**) in Europe and prepares a filing also for the US.

Idebenone in DMD patients using glucocorticoids – Phase III SIDEROS trial ongoing

The SIDEROS study is a Phase III clinical trial, evaluating the efficacy of idebenone compared to placebo, in delaying the loss of respiratory function in patients with DMD receiving concomitant glucocorticoid treatment. The SIDEROS trial is enrolling patients in approximately 60 clinical centers in Europe, the US and Israel. Positive results would support the use of idebenone in DMD patients irrespective of their glucocorticoid use und could support a corresponding label extension. Details of the study can be viewed at www.siderosdmd.com.

U.S. Expanded Access Program with idebenone for patients with DMD

The Expanded Access Program (**EAP**) named BreatheDMD allows eligible DMD patients in respiratory function decline to obtain access to idebenone, at no cost, through a growing network of research centers across the U.S. EAPs are permitted by the U.S. Food and Drug Administration (**FDA**). Such programs allow eligible patients with serious or life-threatening diseases or conditions, where there is a lack of satisfactory therapeutic alternatives, to gain access to a medicine under investigation before it is approved by regulatory authorities. More information is available at www.breatheDMD.com.

PRODUCTS AND PIPELINE

Omigapil in CMD

Congenital muscular dystrophy (**CMD**) refers to a variety of inherited neuromuscular conditions characterized by different forms of progressive loss of muscle tissue. Severe forms can affect newborns or young children with life-threatening progressive muscle weakness ("floppy infant syndrome"). Complications associated with the disorder such as loss of body weight, skeletal deformations and respiratory distress result in immobility at a young age and early mortality.

No pharmacological therapy is currently available, or in advanced clinical development, to slow down or stop progression of CMD. Treatment options are confined to respiratory support and orthopedic surgery as well as supplementary nutrition to avoid malnutrition.

First investigational drug for CMD, currently in phase I

Omigapil – an investigational product for CMD

Santhera has inlicensed omigapil from Novartis for development and commercialization in CMD and initiated a clinical development program. Nonclinical studies in a disease-relevant model showed that omigapil inhibits cell death and reduces body weight loss and skeletal deformation, while increasing locomotive activity and protecting from early mortality.

CALLISTO Phase I trial completed

Santhera evaluated omigapil in the Phase I study (**CALLISTO**) which investigates the safety, tolerability and pharmacokinetic profile of a new liquid formulation of omigapil in pediatric and adolescent patients with CMD. The CALLISTO trial which was conducted at the US NIH's National Institute of Neurological Disorders and Stroke (**NINDS**) in Bethesda, Maryland, has been completed and results on the pharmacokinetic profile, safety and tolerability are currently being analyzed and are planned to be published in Q2 2018. Santhera plans to discuss the results from this first intervention trial in CMD with clinical experts and regulators before defining the best development path forward.

Omigapil has Fast Track Designation from the US Food and Drug Administration (**FDA**) for the treatment of CMD and Orphan Drug Designation for CMD in both the EU and the US.

PRODUCTS AND PIPELINE

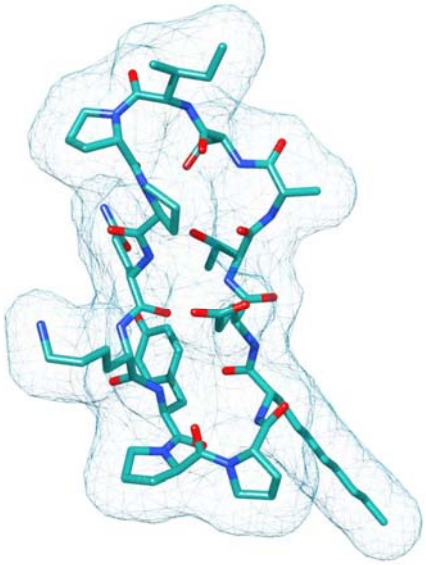
POL6014 in CF and Other Pulmonary Diseases

Cystic fibrosis (**CF**) is a rare, hereditary, life-threatening, progressive disease affecting approximately 70,000 patients in the U.S. and Europe and is characterized by persistent lung infection and chronic inflammation thereby limiting the ability to breathe over time. There is a high unmet medical need for treatments which reduce the inherent chronic lung inflammation in patients with cystic fibrosis and other neutrophilic pulmonary diseases.

Worldwide exclusive license to POL6014 broadens Santhera's clinical stage pipeline

In February 2018, Santhera has entered into a license agreement with Polyphor Ltd. for POL6014, a clinical stage candidate with the potential to treat CF and other neutrophilic pulmonary diseases.

Santhera will assume the global development, regulatory filings and commercialization of POL6014. With POL6014, Santhera is expanding its product pipeline into a third therapeutic area, pulmonary diseases.



Molecular structure of the cyclic peptide POL6014
(courtesy of Polyphor Ltd)

POL6014 is a first-in-class hNE inhibitor that targets chronic inflammation and addresses a high unmet medical need

POL6014 is a highly potent and selective inhibitor of human neutrophil elastase (**hNE**). Inhibition of hNE is expected to stop or slow the damage to lung tissue and may help to improve the overall quality of life for individuals with CF. POL6014 could be potentially used in combination with existing treatments.

POL6014 will be administered by inhalation via an optimized eFlow® nebulizer, a well-accepted rapid nebulizer system routinely used by patients with CF, leading to high concentrations in the lung and favoring local activity in the lung with low systemic exposure.

Santhera to exploit therapeutic potential in CF and other rare pulmonary diseases

POL6014 has been shown to be effective in various nonclinical studies and has successfully been tested in two completed safety and tolerability Phase I trials: one in a first-in-man trial in healthy volunteers and one in a single ascending dose (**SAD**) safety and tolerability trial in cystic fibrosis patients.

Santhera intends to start a multiple ascending dose (**MAD**) trial in cystic fibrosis patients by Q3 2018, a trial that was already designed by Polyphor and is financially supported by the Cystic Fibrosis Foundation Therapeutics Inc. (**CFFT**), USA. Upon dose selection, a Phase II trial could be started in 2019.

In parallel, Santhera will also collaborate with experts to explore not only CF, but potentially other rare pulmonary disorders for which POL6014 offers a treatment option (e.g. non-cystic fibrosis bronchiectasis (**NCFB**), alpha-1 antitrypsin deficiency (**AATD**) and primary ciliary dyskinesia (**PCD**)).

Consolidated Financial Statements

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Consolidated Balance Sheet

As of December 31, in CHF thousands	Notes	2017	2016
Assets			
Tangible assets	5	2,157	517
Intangible assets	6	23,560	26,549
Financial assets long-term		713	270
Restricted cash long-term	10	4,500	0
Deferred tax assets	13	1,242	1,106
Noncurrent assets		**32,172**	**28,442**
Prepaid expenses and other assets	7	853	583
Inventories	8	10,147	7,676
Trade and other receivables	9	5,402	4,276
Financial assets short-term	12	13,011	0
Restricted cash short-term	10	3,000	0
Cash and cash equivalents	10	45,195	49,815
Current assets		**77,608**	**62,350**
Total assets		**109,780**	**90,792**
Equity and liabilities			
Share capital	11	6,289	6,280
Capital reserves and share premium		392,002	382,322
Retained earnings		−360,081	−308,549
Employee benefit reserve		−4,905	−4,734
Treasury shares	11	−335	−172
Translation differences		−714	−796
Total equity		**32,256**	**74,351**
Convertible bonds	12	53,111	0
Derivative financial instruments	12	2,792	0
Pension liabilities	21	8,375	6,183
Total noncurrent liabilities		**64,278**	**6,183**
Trade and other payables	14	4,734	4,458
Accrued expenses	15	8,512	5,800
Total current liabilities		**13,246**	**10,258**
Total liabilities		**77,524**	**16,441**
Total equity and liabilities		**109,780**	**90,792**

Consolidated Income Statement

For the year ended December 31, in CHF thousands	Notes	2017	2016
Net sales	18	22,943	19,033
Cost of goods sold		−4,104	−3,883
Of which amortization intangible assets		*−3,039*	*−3,039*
Other operating income	19	270	361
Development	20	−26,561	−17,675
Marketing and sales	20	−28,522	−21,051
General and administrative	20	−14,416	−9,805
Other operating expenses	20	−64	−107
Operating expenses	**20**	**−69,563**	**−48,638**
Operating result		**−50,454**	**−33,127**
Financial income	22	4,134	928
Financial expenses	22	−4,955	−995
Result before taxes		**−51,275**	**−33,194**
Income taxes	23	−257	−2,221
Net result		**−51,532**	**−35,415**
Basic earnings/loss per share (in CHF)	24	−8.22	−5.65
Diluted earnings/loss per share (in CHF)	24	−8.22	−5.65

Consolidated Statement of Comprehensive Income

For the year ended December 31, in CHF thousands	Notes	2017	2016
Net result		**–51,532**	**–35,415**
Items never to be reclassified to net income in subsequent periods:			
Actuarial gains/losses on defined benefit plans	21	–171	–1,776
Items to be reclassified to net income in subsequent periods:			
Currency translation differences		82	–18
Other comprehensive result		**–89**	**–1,794**
Total comprehensive result		**–51,621**	**–37,209**

Consolidated Cash Flow Statement

For the year ended December 31, in CHF thousands	Notes	2017	2016
Result before taxes		**–51,275**	**–33,194**
Depreciation of tangible assets	5	257	168
Amortization of intangible assets	6	3,125	3,096
Expenses for equity rights plans	17, 20	9,687	4,683
Change in fair value of derivatives		–2,540	0
Change in fair value of financial assets short-term		–96	0
Change in pension liabilities	21	2,021	450
Taxes paid		–392	–266
Change in net working capital		315	–2,131
Total financial result	22	821	67
Interest received	22	5	5
Interest paid	22	–1,561	–15
Cash flow from operating activities		**–39,633**	**–27,137**
Investments in tangible assets	5	–1,261	–289
Investments in intangible assets	6	–136	–86
Investments in other short-term financial assets	12	–12,915	0
Investments in other long-term financial assets		–427	–84
Change in restricted cash	10	–7,500	0
Cash flow from investing activities		**–22,239**	**–459**
Capital increases from options exercised	11	34	385
Proceeds from sale of treasury shares	11	9,372	418
Purchase of treasury shares	11	–9,567	–172
Proceeds from convertible bonds	12	57,269	0
Cash flow from financing activities		**57,108**	**631**
Effects of exchange rate changes on cash and cash equivalents		144	–79
Net increase/decrease in cash and cash equivalents		**–4,620**	**–27,044**
Cash and cash equivalents at January 1		49,815	76,859
Cash and cash equivalents at December 31		**45,195**	**49,815**

Consolidated Statement of Changes in Equity

In CHF thousands	Notes	Share capital	Capital reserves and share premium	Retained earnings	Employee benefit reserve	Treasury shares	Translation differences	**Total**
Balance at January 1, 2016		**6,263**	**377,031**	**−273,134**	**−2,958**	**−177**	**−778**	**106,247**
Net result		0	0	−35,415	0	0	0	**−35,415**
Other comprehensive result	21	0	0	0	−1,776	0	−18	**−1,794**
Total comprehensive result for the period		**0**	**0**	**−35,415**	**−1,776**	**0**	**−18**	**−37,209**
Transactions for equity rights plans	17, 20	0	4,682	0	0	0	0	**4,682**
Capital increase from options exercise	11	17	368	0	0	0	0	**385**
Change in treasury shares		0	241	0	0	5	0	**246**
Balance at December 31, 2016		**6,280**	**382,322**	**−308,549**	**−4,734**	**−172**	**−796**	**74,351**
Balance at January 1, 2017		**6,280**	**382,322**	**−308,549**	**−4,734**	**−172**	**−796**	**74,351**
Net result		0	0	−51,532	0	0	0	**−51,532**
Other comprehensive result	21	0	0	0	−171	0	82	**−89**
Total comprehensive result for the period		**0**	**0**	**−51,532**	**−171**	**0**	**82**	**−51,621**
Transactions for equity rights plans	17, 20	0	9,687	0	0	0	0	**9,687**
Capital increase from options exercise	11	9	25	0	0	0	0	**34**
Change in treasury shares	11	0	−32	0	0	−163	0	**−195**
Balance at December 31, 2017		**6,289**	**392,002**	**−360,081**	**−4,905**	**−335**	**−714**	**32,256**

Notes to the Consolidated Financial Statements

1 General Information

Santhera Pharmaceuticals Holding AG (the **Company**, together with its subsidiaries **Santhera** or **Group**) is a Swiss specialty pharmaceutical company focused on the development and commercialization of products for the treatment of neuro-ophthalmological, neuromuscular and pulmonary diseases, areas which include many orphan and niche indications with high unmet medical need.

The Company, having its primary listing of its registered shares (**Shares**) on the SIX Swiss Exchange (**SIX**), is a Swiss stock corporation and the parent company of the Group. Its purpose is to acquire, dispose and manage investments. The Company has its business offices at Hohenrainstrasse 24 in 4133 Pratteln, Switzerland. The legal domicile will remain at Hammerstrasse 49 in 4410 Liestal, Switzerland, until the forthcoming Annual General Meeting when the shareholders will vote on its relocation to Pratteln.

The consolidated financial statements were approved for publication by the Board of Directors (**Board**) on March 19, 2018. They are subject to approval by the Annual General Meeting of Shareholders (**AGM**) on April 12, 2018.

2 Summary of Significant Accounting Policies

The principal accounting policies applied in the preparation of these financial statements are set out below. These policies have been consistently applied to all the years presented, unless otherwise stated.

Basis of preparation

The consolidated financial statements of Santhera have been prepared in accordance with International Financial Reporting Standards (**IFRS**).

The consolidated financial statements are based on the financial statements of the individual Santhera companies prepared for the same reporting period using consistent accounting policies. The consolidated financial statements are prepared using the historical cost convention except for the re-valuation to fair value of certain financial assets and financial liabilities.

The presentation currency is Swiss francs (**CHF**). All figures included in these financial statements and notes to the financial statements are rounded to the nearest CHF 1,000 except where otherwise indicated.

Consolidation

Subsidiaries in which the Company has a direct or indirect controlling interest are consolidated. Control exists when the investor is exposed, or has rights, to variable returns from its investment with the investee and has the ability to affect those returns through its power over the investee. Control is normally evidenced when the Company owns, either directly or indirectly, more than 50% of the voting rights or potential voting rights of a company's share capital that are currently exercisable.

The consolidated financial statements of Santhera include the accounts of Santhera Pharmaceuticals Holding AG, Liestal, Switzerland, and its wholly owned subsidiaries Santhera Pharmaceuticals (Schweiz) AG, Pratteln, Switzerland; Santhera Pharmaceuticals (USA), Inc., Burlington, US; Santhera Pharmaceuticals (Canada), Inc., Montréal, Canada; Santhera Pharmaceuticals (Deutschland) GmbH, Lörrach, Germany; and Oy Santhera Pharmaceuticals (Finland) Ltd, Helsinki, Finland. The accounts further include the wholly owned subsidiaries of Santhera Pharmaceuticals (Schweiz) AG: Santhera Pharmaceuticals (Liechtenstein) AG, Ruggell, Fürstentum Liechtenstein; Santhera (Italy) S.r.l., Milano, Italy; Santhera (Germany) GmbH, München, Germany; Santhera (Netherlands) B.V., Nieuwegein, The Netherlands; and Santhera (UK) Limited, London, United Kingdom.

Consolidation commences from the date on which control is transferred to the Company, and subsidiaries are no longer consolidated from the date that control ceases. Intercompany balances and transactions between Group companies are eliminated. Intercompany transactions solely result from providing services, financing and selling goods to other Group companies.

Changes in accounting policies

New, revised or amended IFRS standards and interpretations 2017

The following new, revised or amended standards that became effective on January 1, 2017 did not have any significant impact on the consolidated financial statements.

- IAS 7 Statement of Cash Flows: Disclosure Initiative
  The amendments require entities to provide disclosure of changes in their liabilities arising from financing activities, including both changes arising from cash flows and non-cash changes.
- IAS 12 Income Taxes (Amendments) Recognition of Deferred Tax Assets for Unrealized Losses
- Annual Improvements to IFRSs 2014–2016 Cycle

New, revised or amended IFRS standards and interpretations issued but not yet effective

The following new, revised or amended standards have been published but are not yet effective and have not been early adopted by the Group.

- IFRS 9, Financial Instruments (effective January 1, 2018)
  IFRS 9 introduces a single approach for the classification and measurement of financial assets according to their cash flow characteristics and the business model they are managed in, and provides a new impairment model based on expected credit losses. IFRS 9 also includes new regulations regarding the application of hedge accounting to better reflect an entity's risk management activities especially with regard to managing nonfinancial risks. The Group adopted the new standard on the required effective date as of January 1, 2018, and did not restate comparative information. During 2017, the Group has performed an impact assessment of all three aspects of IFRS 9. Upon implementation of IFRS 9, the assessment did not show a significant impact.

- IFRS 9 requires the Group to record expected credit losses on all of its trade receivables, either on a 12-month or lifetime basis. The Group applies the simplified approach and records lifetime expected losses.
- IFRS 15 Revenue from Contracts with Customers (effective January 1, 2018)
  According to the new standard, revenue is recognized to depict the transfer of promised goods or services to a customer in an amount that reflects the consideration to which the Company expects to be entitled to in exchange for those goods or services. Revenue is recognized when, or as the customer obtains control of the goods or services.
  The Group is focused on the development and commercialization of products for the treatment of mitochondrial and neuromuscular diseases. It has identified the sale of product as the only performance obligation and revenue stream from its contracts with customers. For these contracts the adoption of IFRS 15 did not have any impact on the Group's revenue and profit or loss and no transition adjustment was recorded upon adoption as of January 1, 2018.

- IFRS 16 Leases (effective January 1, 2019)
  The new standard eliminates the current classification model for lessee's lease contracts as either operating or finance leases and, instead, introduces a single lessee accounting model requiring lessees to recognize right-of-use assets and lease liabilities for leases with a term of more than twelve months. This brings the previous off-balance sheet leases on the balance sheet in a manner largely comparable to current finance lease accounting. A lessee can choose to apply the standard using either a full retrospective or a modified retrospective approach. Adoption of IFRS 16 will result in the Group recognizing right of use assets and lease liabilities for all contracts that are, or contain, a lease. For leases currently classified as operating leases, under current accounting requirements the Group does not recognize related assets or liabilities, and instead spreads the lease payments on a straight-line basis over the lease term, disclosing in the notes to its annual consolidated financial statements the total commitment. The Group is expecting that current leasing arrangements relating to real estate and cars will be capitalized under IFRS 16. In 2018, the Group will continue to assess the potential effect of IFRS 16 on its consolidated financial statements.
  The Group does not expect any other standards issued by the IASB, but not yet effective, to have a material impact on the Group's financial statements.

Segment reporting

Santhera has one operating segment, namely the development and commercialization of products for the treatment of neuro-ophthalmological, neuromuscular and pulmonary diseases. The Board, the Executive Management and senior managers, being the Chief Operating Decision Makers (**CODM**), assess the reporting data and allocate resources as one segment on a consolidated level according to operating expenses by function. Santhera generates revenue from sales of Raxone for the treatment of LHON. Geographic revenue information is based on location of the customer or licensee.

Foreign currency translations

The consolidated financial statements are presented in CHF. The functional currency of each of Santhera's companies is the currency of the primary economic environment in which the local entity

operates. Transactions in foreign currencies are accounted for at the rates prevailing at the dates of the transaction. Translation differences from financial transactions are included in the financial result.

Gains and losses resulting from the translation of foreign currency transactions and from the adjustment of foreign currency monetary assets and liabilities at the reporting date are recognized in the income statement.

Assets and liabilities of foreign entities are translated into CHF using the balance sheet exchange rates at year-end. Income and expenses are translated into CHF at average exchange rates. The exchange differences arising on the retranslation are accounted for in the statements of comprehensive income/equity.

Intangible assets

Patents, licenses, trademarks and other intangible assets are capitalized as intangible assets when it is probable that future economic benefits will be generated. Such assets are in general amortized on a straight-line basis over their useful lives. Estimated useful life is the lower of legal duration or economic useful life. The estimated useful life of the intangible assets is regularly reviewed and if necessary, the future amortization charge is accelerated. For pharmaceutical products, the estimated useful life normally corresponds to the remaining lifetime of their patent or orphan drug protection (up to 20 years).

IT software

Acquired IT software licenses are capitalized on the basis of the costs incurred to acquire and implement the specific software. These costs are amortized on a straight-line basis over their estimated useful lives (2 to 5 years).

Tangible assets

Tangible assets are stated at cost less accumulated depreciation and any impairment losses. Depreciation is calculated on a straight-line basis over the estimated useful life of the asset or the shorter lease term, as follows:

	Useful life
Equipment	4 to 10 years
IT hardware	2 to 5 years
Leasehold improvements	2 to 10 years

Impairment of assets

Assets include intangible assets not yet available for use, intangible assets with finite useful lives and tangible assets. In general, and in accordance with the terms of IFRS, assets not in use are capitalized at cost in the balance sheet and reviewed for impairment at least annually. Impairment testing is performed at the same time every year or whenever there is an indication that the asset may be impaired. Once an intangible asset starts to be used, amortization starts. Testing for indicators of impairment is done at the end of each reporting period.

Trade and other receivables

Receivables which generally have 30 to 60 days payment terms are stated at their nominal value less an allowance for any uncollectible amount if required. An allowance for doubtful debts is made when collection is deemed no longer probable.

Inventories

Inventories are stated at the lower of cost or net realizable value using the weighted average cost formula.

Financial assets

Generally, Santhera classifies its financial assets in the following categories:

Financial assets at fair value through profit or loss

This category has two subcategories: financial assets held for trading and those designated at fair value through profit or loss upon initial recognition. A financial asset is classified in this category if acquired principally for the purpose of selling in the short-term. Assets in this category are classified as current assets if they are either held for trading or are expected to be realized within 12 months of the reporting date. Valuation is at fair value through profit or loss. Financial assets at fair value through profit or loss are subsequently carried at fair value. Realized and unrealized gains and losses arising from changes in the fair value are included in the income statement in the period in which they arise.

Loans and receivables

Loans and receivables are nonderivative financial assets with fixed or determinable payments that are not quoted in an active market. They arise when Santhera provides money, goods or services directly to a debtor with no intention of trading the receivable. They are included in current assets, except for maturities longer than 12 months after the balance sheet date. These are classified as noncurrent assets. Loans and receivables are measured at amortized cost using the effective interest method.

Leases

Leases of assets under which Santhera essentially assumes all the rewards and risks of ownership are classified as finance leases. Finance leases are capitalized as assets and liabilities at the commencement of the lease at the fair value of the leased item or, if lower, at the present value of the minimum lease payments. The assets acquired under these contracts are depreciated over the shorter of the estimated useful life of the asset or the lease term.

Leases of assets under which the risks and rewards of ownership are effectively retained by the lessor are classified as operating leases, and payments made are charged to the income statement on a straight-line basis.

Cash and cash equivalents

This item includes cash on hand and at banks, deposits held at call with banks and other short-term highly liquid investments with original maturities of three months or less.

Restricted cash

Cash set aside in escrow and not available to finance Santhera's day-to-day operations is shown under this category. Maturities of less than 12 months are considered short-term; those of more than 12 months are long-term.

Share capital

Common shares are classified as equity. Incremental costs directly attributable to the issue of new common shares or options are shown in equity in the capital reserves and share premium as a deduction, net of tax, from the proceeds.

Treasury shares

Treasury shares are purchased at cost and recognized as deduction from equity. Income or loss from subsequent sale is presented in equity.

Financial liabilities

Santhera classifies its financial liabilities into two categories:

Financial liabilities at fair value through profit or loss

This category includes derivatives with negative replacement values. They are initially recognized at their fair value. Any subsequent change in fair value is recognized in the income statement in the period the changes occur.

Other liabilities measured at amortized costs

This category principally covers debt instruments and trade and other payables. They are initially recognized at fair value and subsequently measured at amortized costs using the effective interest method. Any difference between the net proceeds received and the principal value due on redemption is amortized over the duration of the debt instrument and is recognized as part of interest expense in the income statement.

Income taxes

The income tax charge is based on profit for the year and includes deferred taxes. Deferred taxes are calculated using the liability method. Deferred income taxes reflect the net tax effects of temporary differences between the carrying amounts of assets and liabilities for financial reporting purposes and the amounts used for income tax purposes. Deferred tax assets and liabilities are measured using the tax rates expected to apply to taxable income in the years in which those temporary differences are expected to be recovered or settled based on enacted or substantially enacted tax rates as of the balance sheet date.

The amount of deferred tax liabilities and deferred tax assets reflects the tax consequences on the balance sheet date of the Company's expectation of recovery or settlement of such carrying amount of its assets and liabilities.

Deferred tax assets and liabilities are not discounted and are classified as noncurrent assets (liabilities) in the balance sheet. They are offset against each other if they relate to the same taxable entity and tax authority.

Deferred tax assets are recognized when it is probable that sufficient taxable profits will be available against which the deferred tax assets can be utilized. At each balance sheet date, the Company reassesses unrecognized deferred tax assets and the carrying amount of deferred tax assets. The Company recognizes a previously unrecognized deferred tax asset to the extent that it has become probable that future taxable profit will allow the deferred tax asset to be recovered. The Company conversely reduces the carrying amount of a deferred tax asset to the extent that it is no longer probable that sufficient taxable profit will be available to allow the benefit of part or the entire deferred tax asset to be utilized. Deferred tax is provided on temporary differences arising on investments in subsidiaries, associates and joint ventures, except where the timing of the reversal of the temporary difference can be controlled and it is probable that the difference will not reverse in the foreseeable future.

Earnings/loss per share

Basic earnings/loss per share are calculated by dividing the net profit/loss attributable to owners of ordinary Shares of the Company by the weighted average number of Shares outstanding during the reporting period. Diluted earnings per share are calculated by dividing the net profit attributable to owners of ordinary Shares of the Company by the weighted average number of shares issued and outstanding during the reporting period adjusted for Shares held as treasury shares (purchased at market), the number of potential shares from stock option plans and the convertible bonds.

Employee benefits

Post-retirement benefits

Santhera operates both defined benefit and defined contribution pension schemes.

- Defined benefit scheme:

  Santhera's pension plan in Switzerland is classified as a defined benefit plan. Payments under this scheme are made directly to the pension fund for the account of each insured person. Typically, on retirement, an employee will receive an amount of the accumulated defined benefit obligation depending on several factors such as the total individual amount paid in, age and implied life expectancy. The compensation will be in the form of a lifelong pension or a lump sum payment. The scheme also covers disability as a consequence of illness and death-in-service.

  The liability recognized in the balance sheet in respect of defined benefit pension plans is the present value of the defined benefit obligation at the balance sheet date less the fair value of plan assets, adjusted for the effects of the asset ceiling, when relevant.

  The defined benefit obligation is calculated annually by independent actuaries using the projected unit credit method. The present value of the defined benefit obligation is determined by discounting the estimated future cash outflows using interest rates of high-quality corporate bonds that are denominated in the currency in which the benefits will be paid and that have terms to maturity approximating the terms of the related pension liability.

- Defined contribution schemes:

  Defined contribution schemes are also funded through direct payments for the account of each insured person. Upon retirement, an employee will receive an amount of the accumulated contributions in the form of a lifelong pension or a lump sum payment. No further obligations arise from these schemes other than the fixed periodic contributions to the plan.

Share-based compensation

Santhera has established stock option and share appreciation rights (**SAR**) plans to align the long-term interests of the members of the Board, the Executive Management, employees and selected consultants who are eligible to participate. Under all plans, options and share appreciation rights are equity-settled. The fair value of options and SAR is determined at the grant date and recognized as personnel expense over the period Santhera receives services for each award. Where stock option awards are modified as a minimum, the expenses are recognized as if no terms had been modified; modifications which increase the fair value of options are expensed additionally. Unless determined otherwise by the Board, terminations of employment by the employer are treated as forfeiture and any previously accumulated share-based payment expenses for unvested awards are reversed.

Provisions

Provisions are recognized when Santhera has a present obligation (legal or constructive) as a result of a past event, where it is more probable than not that a cash outflow will be required to fulfill the obligation and where a reliable estimate can be made of the amount of the obligation.

If the effect of the time value of money is material, provisions are determined by discounting the expected future outflows.

Revenue recognition

Revenue comprises the fair value of the sale of goods and services, net of value-added tax, rebates, discounts, returns and after eliminating intercompany sales. Revenue is recognized when title, risks and rewards of the products are transferred to customers.

Revenue from outlicensing

Outlicensing agreements are concluded with third parties, where the counterparty has to pay license fees. In situations where no further performance commitment exists, revenue is recognized on the earlier of when payments are received or collection is assured. Where continuous involvement for a certain period is required in the form of technology transfer or technical support, revenues are recognized over the period in question.

Revenue associated with up-front payments or performance milestones

Such revenue is recognized in accordance with respective agreements.

Revenue from royalties

Royalty payments are recognized on an accrual basis in accordance with the respective agreements.

Interest income

Interest income is recognized on a pro rata temporis basis using the effective interest method.

Development / intangible assets

Development expenses are charged to the income statement as incurred. They are capitalized as intangible assets when it is probable that future economic benefits will flow to Santhera. Such intangible assets are amortized on a straight-line basis over the period of the expected benefit when the asset becomes available for use, and are reviewed for impairment indicators at each balance sheet date. Assets not available for use are tested annually for impairment.

3 Critical Accounting Estimates, Assumptions and Judgments

The preparation of financial statements in conformity with IFRS requires the use of certain critical accounting estimates. It also requires management to exercise its judgment in the process of applying Santhera's accounting policies. Santhera makes estimates and assumptions concerning the future. The resulting accounting will not necessarily equal the related actual outcome. The following areas involve assumptions and estimates that can have a significant impact on the consolidated financial statements:

- Assessment of the Group's ability to continue as a going concern.
- Measurement and impairment testing of intangible assets.
- Measurement and testing for net realizable value of inventory, see note 8 *"Inventories"*.
- Valuation of derivative financial instruments in connection with the convertible bonds, see note 12 "*Financial Assets and Liabilities"*
- Personnel expenses from share-based payments in accordance with IFRS 2, i.e. estimates regarding the valuation of equity rights plans when granted, see note 17 "*Equity Rights Plans"*.
- Actuarial valuations in the context of defined benefit pension plans where various assumptions on e.g. discount rates, salary increase rates and mortality rates, etc. bear significant uncertainties due to the long-term nature of the plans, see note 21 *"Employee Expenses and Benefits"*.

4 Exchange Rates of Principal Currencies

	Income statement in CHF average rates		**Balance sheet in CHF** year-end rates	
	2017	**2016**	**2017**	**2016**
1 Euro (**EUR**)	1.1114	1.0902	1.1691	1.0737
1 US dollar (**USD**)	0.9847	0.9851	0.9753	1.0160
1 British pound (**GBP**)	1.2683	1.3352	1.3173	1.2498
1 Canadian dollar (**CAD**)	0.7590	0.7435	0.7777	0.7532

5 Tangible Assets

In CHF thousands	Equipment	IT hardware	Leasehold improvements	2017
Cost				
At January 1	**237**	**756**	**91**	**1,084**
Additions	114	396	1,376	**1,886**
Disposals	–2	–47	0	**–49**
Exchange differences	8	6	2	**16**
Reclassification	5	–5	0	**0**
At December 31	**362**	**1,106**	**1,469**	**2,937**
Accumulated depreciation				
At January 1	**179**	**344**	**44**	**567**
Additions	32	215	10	**257**
Disposals	–2	–47	0	**–49**
Exchange differences	2	3	0	**5**
Reclassification	1	–1	0	**0**
At December 31	**212**	**514**	**54**	**780**
Net book value	**150**	**592**	**1,415**	**2,157**

In CHF thousands	Equipment	IT hardware	Leasehold improvements	2016
Cost				
At January 1	**225**	**567**	**45**	**837**
Additions	17	226	46	**289**
Disposals	–3	–36	0	**–39**
Exchange differences	–1	–2	0	**–3**
Reclassification	–1	1	0	**0**
At December 31	**237**	**756**	**91**	**1,084**
Accumulated depreciation				
At January 1	**166**	**231**	**42**	**439**
Additions	16	150	2	**168**
Disposals	–3	–36	0	**–39**
Exchange differences	0	–1	0	**–1**
At December 31	**179**	**344**	**44**	**567**
Net book value	**58**	**412**	**47**	**517**

6 Intangible Assets

In CHF thousands	Idebenone	Fipamezole	IT software/ patents	2017
Cost				
At January 1	**30,387**	**3,918**	**535**	**34,840**
Additions	0	0	136	**136**
Disposals	0	0	–17	**–17**
At December 31	**30,387**	**3,918**	**654**	**34,959**
Accumulated amortization				
At January 1	**4,052**	**3,918**	**321**	**8,291**
Additions	3,039	0	86	**3,125**
Disposals	0	0	–17	**–17**
At December 31	**7,091**	**3,918**	**390**	**11,399**
Net book value	**23,296**	**0**	**264**	**23,560**

In CHF thousands	Idebenone	Fipamezole	IT software/ patents	2016
Cost				
At January 1	**30,387**	**3,918**	**477**	**34,782**
Additions	0	0	86	**86**
Disposals	0	0	–28	**–28**
At December 31	**30,387**	**3,918**	**535**	**34,840**
Accumulated amortization				
At January 1	**1,013**	**3,918**	**292**	**5,223**
Additions	3,039	0	57	**3,096**
Disposals	0	0	–28	**–28**
At December 31	**4,052**	**3,918**	**321**	**8,291**
Net book value	**26,335**	**0**	**214**	**26,549**

During 2017 there was no trigger for impairment of intangible assets. "Idebenone" represents the main intangible asset of Santhera. It has become available for use in September 2015 and has an estimated useful life of 10 years. Prior to that it was not available for use and did not generate cash inflows.

7 Prepaid Expenses and Other Assets

In CHF thousands	2017	2016
Prepayments	853	487
Other assets	0	96
Total at December 31	**853**	**583**

8 Inventories

In CHF thousands	2017	2016
Raw material (active pharmaceutical ingredients)	7,488	5,052
Semi-finished goods	2,335	2,369
Finished goods	324	255
Total at December 31	**10,147**	**7,676**

9 Trade and Other Receivables

In CHF thousands	2017	2016
Trade receivables	4,194	3,412
Other receivables	1,208	864
Total at December 31	**5,402**	**4,276**

Trade receivables in 2017 result from product sales, see note 18 *"Segment and Geographic Information"*. Other receivables consist mainly of amounts due from the government for tax reimbursements (e.g. VAT). They are due within 30 to 120 days and bear no interest. As of December 31, 2017, an allowance for doubtful debts of TCHF 55 was recognized on the trade receivables (no allowance was booked as of December 31, 2016).

10 Cash and Cash Equivalents and Restricted Cash

10.1 Cash and cash equivalents

In CHF thousands	2017	2016
Cash at banks and on hand		
In CHF	34,730	44,358
In EUR	8,152	4,661
In GBP	697	546
In USD	1,496	149
In CAD	120	67
Other currencies	0	34
Total at December 31	**45,195**	49,815
Of which: Short-term deposits		
In CHF	21,007	31,000

10.2 Restricted cash

in CHF thousands	Dec. 31, 2017	Dec. 31, 2016
Long-term	4,500	0
Short-term	3,000	0
Total at period end	**7,500**	0

Restricted cash is designated for interest payments due related to the convertible bonds during the first 3 years (starting 2017). These funds are kept in an escrow account with the bond agent.

11 Share Capital

Ordinary share capital

As of January 1, 2016, the share capital amounted to CHF 6,262,798, divided into 6,262,798 shares ("Shares") at a nominal value of CHF 1 each. During 2016, 17,059 Shares were issued from conditional capital upon the exercise of stock options. As a result, as of December 31, 2016, the share capital amounted to CHF 6,279,857, divided into 6,279,857 Shares at a nominal value of CHF 1 each.

During 2017, 8,698 Shares were issued from conditional capital upon the exercise of stock options. As a result, as of December 31, 2017, the share capital amounted to CHF 6,288,555, divided into 6,288,555 Shares at a nominal value of CHF 1 each.

Treasury shares

In the second half of 2016, Santhera entered into an agreement for market making with a well-known bank. Independently, the bank buys and sells Shares on the market on behalf of the Company. On December 31, 2017, Santhera held 9,921 treasury Shares (2016: 3,616 treasury Shares).

Authorized share capital

On the occasion of the AGM on May 11, 2016, the shareholders approved the increase of the authorized share capital as well as an extension. The Board is authorized to increase the share capital at any time until May 10, 2018, through the issuance of up to 1,500,000 Shares with a nominal value of CHF 1 each. An increase in instalments is permitted. For each such increase, the Board has to determine the issue price, the type of payment, the date of issuance of new Shares, the conditions for the exercise of pre-emptive rights and the beginning date for dividend entitlement.

Conditional share capital

At the AGM on April 4, 2017, the shareholders approved a maximum increase of the share capital by an aggregate amount of CHF 700,000 (2016: CHF 550,000) through the issuance of a maximum of 700,000 (2016: 550,000) Shares with a nominal value of CHF 1 each. The Shares can be issued through the exercise of equity rights which are granted according to respective regulations of the Board.

In addition, the shareholders approved a maximum increase of the share capital by an aggregate amount of CHF 930,000 (2016: CHF 650,000) through the issuance of a maximum of 930,000 (2016: 650,000) Shares with a nominal value of CHF 1 per Share by the exercise of option and/or conversion rights which can be granted in connection with the issuance of bonds, similar obligations or other financial instruments by the Company or another Group company, and/or by the exercise of options which are granted by the Company or another Group company. In the case of the issue of bonds, similar obligations or other financial instruments linked with option and/or conversion rights, and in the case of the issue of option rights, the pre-emptive right of shareholders is excluded.

As of December 31, 2017, the Company had a conditional share capital, pursuant to the above provisions, whereby the share capital may be increased by

- a maximum amount of CHF 691,302 (2016: CHF 532,941) through the issuance of up to 691,302 (2016: 532,941) Shares, under the exclusion of shareholders' pre-emptive rights, for option rights being exercised under the Company's stock option plans, see note 17 *"Equity Rights Plans"*, and
- a maximum amount of CHF 930,000 (2016: CHF 650,000) by issuing up to 930,000 (2016: 650,000) Shares, through the exercise of warrants/options and/or notes granted in connection with bonds or similar debt instruments linked with option and/or conversion rights granted by the Company.

12 Financial Assets and Liabilities

Santhera measures certain financial instruments at fair value. Fair values are categorized into the following hierarchy based on the inputs used to measure them:

Level 1: Quoted prices (unadjusted) in active markets for identical assets or liabilities.

Level 2: Inputs other than quoted prices included within Level 1 that are observable for the asset or liability, either directly (as prices) or indirectly (derived from prices).

Level 3: Unobservable inputs for the asset or liability. These inputs reflect the best estimates of Santhera based on criteria that market participants would use to determine prices for assets or liabilities at the reporting date.

12.1 Financial assets short-term

Financial assets (units in a fund) are classified as held for trading. They are measured at fair value through profit or loss and based on quoted prices (Level 1). A net gain of TCHF 96 resulted during the reporting period (no such financial assets were held during the same period in 2016).

12.2 Financial liabilities

On February 17, 2017, Santhera issued senior unsecured convertible bonds in the nominal amount of CHF 60 million. Transaction costs of CHF 2.7 million lead to a net amount of CHF 57.3 million (consisting of senior unsecured convertible bonds (CHF 52.0 million) and embedded derivative financial instruments (CHF 5.3 million)). The bonds, listed on the SIX, are interest bearing (5%) with a maximum term of 5 years and are convertible into registered Shares of Santhera with a nominal value of CHF 1 each. The initial conversion price is fixed at CHF 86.4006 and will be reset after the first year if the volume weighted average price (**VWAP**) of the Shares during a specified period of time will be below the reference share price (CHF 71.9969). In February 2018, the conversion price was reset to CHF 64.80. In addition, Santhera may call the convertible bonds at any time on or after the second anniversary of the issue date at par, plus accrued interest, if any, if the VWAP of the Shares is at least 160% of the conversion price. The convertible bonds are measured at amortized costs applying the effective interest method. The fair value of the bond at December 31, 2017, amounts to CHF 51.6 million (no convertible bonds in 2016).

The embedded financial derivatives (conversion right, reset mechanism and early redemption option) are valued by an independent consultant initially and at period end at fair value, applying a simulation-based valuation approach. The valuation of the embedded derivatives is based on input parameters, classified as Level 3. The simulation is mainly based on the historical volatility of Santhera shares. The period of volatility data used is measured according to the remaining life of the convertible bonds. The volatility used as per December 31, 2017, was 87.2%.

The value of the derivatives on February 17, 2017, amounted to CHF 5.3 million and at period end to CHF 2.8 million; the change in the fair value was recognized in financial income (TCHF 2,540).

Sensitivity analysis:

	Increase/decrease in volatility assumption	Effect on result before taxes in CHF thousands
Change in volatility	+5%	175
	−5%	−181

Changes in liabilities arising from financing activities

In CHF thousands	2017	
	Convertible bonds	Derivative financial instruments
January 1, 2017	**0**	**0**
Proceeds from convertible bonds	60,000	0
Transaction costs relating to convertible bonds	−2,731	0
Cash flows	**57,269**	**0**
Non-cash changes		
Initial recognition derivative financial instruments	−5,332	5,332
Change in fair value of derivative financial instruments	0	−2,540
Effective interest/amortized cost calculation	1,174	0
December 31, 2017	**53,111**	**2,792**

13 Deferred Taxes

Net deferred taxes recorded

In CHF thousands	2017	2016
Temporary differences on inventory	1,242	1,067
Temporary difference on accruals	0	39
Deferred tax assets recognized	**1,242**	**1,106**
Temporary differences on intangible assets	1,905	2,154
Temporary differences on intercompany loans	13,449	13,449
Temporary differences on convertible bonds	321	0
Tax loss carryforwards	–15,675	–15,603
Deferred tax liabilities recognized	**0**	**0**
Tax loss carryforwards	339,492	301,667
Of which recorded	–197,008	–195,583
Of which unrecorded	**142,484**	**106,084**
Expiring in		
1 year	22,671	5,832
2 years	30,569	22,671
3 years	4,223	27,366
4 years	0	4,223
5 years	0	0
More than 5 years	54,552	17,942
Without expiration	30,469	28,050
Total unrecorded tax loss carryforwards	**142,484**	**106,084**

Due to the uncertainty surrounding the future results of operations and the uncertainty as to whether Santhera can use the loss carryforwards for tax purposes, deferred tax assets on tax loss carryforwards were only considered to the extent that they offset taxable temporary differences within the same taxable entity. As there are no temporary differences associated with investments in subsidiaries, no deferred tax liability has to be recognized. No deferred tax assets are calculated on temporary differences related to pension obligations from IAS 19 (TCHF 8,375 at December 31, 2017, and TCHF 6,183 at December 31, 2016, respectively).

14 Trade and Other Payables

In CHF thousands	2017	2016
Trade payables	3,585	3,574
Other payables (nonfinancial)	1,149	884
Total at December 31	**4,734**	**4,458**

All positions are noninterest-bearing and usually settled within 30 to 60 days.

15 Accrued Expenses

In CHF thousands	2017	2016
Development programs	1,547	749
Liabilities to employees	3,429	2,013
Accruals for pricing and reimbursement	839	1,107
Accrued marketing and sales expenses	469	953
Accruals for audit, consulting and other	688	696
Accruals for interest expenses	1,108	0
Accruals for income taxes	432	282
Total at December 31	**8,512**	**5,800**

16 Commitments and Contingent Liabilities

Commitments

Commitments for operating lease (noncancelable)

In CHF thousands	2017	2016
Within 1 year	1,176	734
After 1 year through to 5 years	1,177	804
After 5 years	34	0
Total at December 31	**2,387**	**1,538**

Contingent liabilities

Collaboration and license agreement with Takeda

In September 2013, Santhera announced an agreement with Takeda Pharmaceutical Company Ltd, Osaka, Japan (**Takeda**) to license back all previously granted rights in DMD and Friedreich's ataxia (**FA**) in order to increase its strategic flexibility. In return, Takeda is eligible to obtain a percentage from future licensing and/or sales income generated by Santhera in DMD of up to EUR 7.0 million. In addition, Santhera has obtained the right to cross-reference Takeda's idebenone data for regulatory use in any indication and in any territory. If Santhera makes use of such cross-reference right, Takeda is eligible

to obtain a percentage from future licensing and/or sales income generated by Santhera in such indications of up to EUR 3.0 million. Lastly, both companies agreed to terminate a similar agreement for FA signed in 2005 and Santhera's contingent liability of EUR 1.0 million payable to Takeda has been waived. Takeda is eligible to receive up to EUR 1.0 million as a percentage from future income generated by Santhera to offset this waiver.

Agreement with the University of Leuven

In March 2005, Santhera entered into an agreement with Katholieke Universiteit Leuven, Leuven, Belgium (**KU Leuven**), under which KU Leuven assigned to Santhera its patents and patent applications relating to the use of idebenone to treat various forms of muscular-dystrophy-related disorders, particularly DMD. Based on this agreement, Santhera has filed patent applications in major territories covering the use of idebenone for the treatment of DMD.

KU Leuven is entitled to a success fee of up to EUR 0.4 million if and when Santhera commercializes any product in a major market, which includes the EU, the US or Japan and certain countries within the EU. In addition, in the event Santhera commercializes the product itself, KU Leuven is entitled to receive 5% royalties on net sales. In the event Santhera grants commercialization rights to a third party, KU Leuven will receive 15% of all the consideration received by Santhera from such third party.

License agreement with Novartis

On June 30, 2007, Santhera entered into an agreement with Novartis Pharma AG, Basel, Switzerland (**Novartis**), under which it inlicensed omigapil. Santhera develops omigapil for the treatment of congenital muscular dystrophy (**CMD**). Additional payments will be due to Novartis a) upon start of a pivotal clinical trial, b) upon regulatory approval in a major market country, and c) after reaching certain commercialization milestones. Santhera will also have to pay royalties to Novartis calculated on net sales.

Agreement with the National Institutes of Health

In June 2013, Santhera has obtained an exclusive license from the National Institutes of Health, Bethesda/Maryland, US (**NIH**), to its rights on a patent granted in the US for the use of idebenone for the treatment of primary progressive multiple sclerosis (**PPMS**). Under the terms of the agreement, Santhera would have to make certain milestone payments to the NIH not exceeding USD 1.4 million in total. Furthermore, the NIH is eligible to a royalty fee of 3% on net sales and 15% of considerations received in case Santhera sublicenses the program.

Contracts for clinical development and other

As part of its ordinary course of business, Santhera has entered into several contracts for e.g. clinical or technical development services. Commitments are within current market prices and can be terminated at the Company's discretion.

Contingent liabilities summary

Santhera believes that the disclosures above and accruals (see note 15 *"Accrued Expenses"*) are adequate based upon currently available information. However, given the inherent difficulties in estimating liabilities relating to clinical development, regulatory, tax, possible litigation and certain other matters due to uncertainty concerning both the amount and timing of future expenditures, additional costs may be incurred materially beyond the amounts accrued.

17 Equity Rights Plans

Santhera has established equity rights plans to align the long-term interests of the members of the Board, the Executive Management and employees. Rights granted under these plans are equity-settled.

17.1 Stock Option Plans

Executive Incentive Plan (EIP)

In November 2006, under the EIP, the members of the Executive Management were granted stock options to acquire 101,065 Shares, as a management incentive. Each of these stock options entitled its holder to purchase one Share at an exercise price of CHF 1. The vesting period of the options was one year. At the end of the option term, i.e. after a period of ten years as from the grant date, all unexercised stock options will expire without value. The EIP is administered under the responsibility of the Board. No further grants can be made under the EIP.

Options outstanding, exercised or forfeited under the EIP

Number of options	2017				2016			
Plan	Exercised	Forfeited	Expired	Outstanding	Exercised	Forfeited	Expired	Outstanding
EIP	0	0	0	0	1,210	0	0	0

Employee Stock Option Plans

The Company adopted the ESOP 2004, ESOP 2008, ESOP 2010 and ESOP 2015 (collectively the **ESOP**) to provide incentives to the Executive Management, employees and consultants helping to ensure their commitment to Santhera over the long-term. Option grants were made periodically at the discretion of the Board or as contractually agreed with employees. The ESOP contain customary provisions in respect of the adjustment or cancellation of stock options upon termination of employment, retirement, death, disability and certain corporate transactions. All stock option plans are administered under the responsibility of the Board. Each stock option entitles its holder to purchase one Share of the Company at an exercise price defined to be either a) equal to the volume-weighted average share price in the three preceding months for Swiss employees, or b) the closing share price on the SIX Swiss Exchange (**SIX**) at each grant date. In general, 50% of the stock options vest on the second anniversary, 25% on the third anniversary and the remaining 25% on the fourth anniversary of the grant date. At the end of the option term, i.e. after a period of 10 years as from the grant date, unexercised stock options expire without value. Subject to the provisions of the ESOP 2004, vested stock options of employees leaving the Company in good faith do not lapse. Under the ESOP 2008 and ESOP 2010 vested stock options of employees leaving the Company in good faith expire six months after the termination date of the employment. Under the ESOP 2015 vested stock options of employees leaving the Company in good faith do not expire. Unvested stock options of employees leaving the Company are forfeited under all stock option plans. No further grants can be made under the ESOP.

Options outstanding, exercised, forfeited or expired under ESOPs

Number of options						2017
	At January 1	Exercised	Granted	Forfeited	Expired	At December 31
ESOP 2004	753	0	0	0	–753	0
ESOP 2010	38,249	–8,698	0	0	0	29,551
ESOP 2015	260,801	0	0	–15,472	0	245,329
Total	**299,803**	**–8,698**	**0**	**–15,472**	**–753**	**274,880**

Number of options						2016
	At January 1	Exercised	Granted	Forfeited	Expired	At December 31
ESOP 2004	26,091	–4,825	0	0	–20,513	753
ESOP 2008	1,500	–1,500	0	0	0	0
ESOP 2010	47,773	–9,524	0	0	0	38,249
ESOP 2015	140,260	0	135,830	–15,289	0	260,801
Total	**215,624**	**–15,849**	**135,830**	**–15,289**	**–20,513**	**299,803**

Board Stock Option Plans

The Company adopted the BSOP 2011 and BSOP 2015 (collectively the **BSOP**) to provide incentives to members of the Board. The BSOPs contain the same customary provisions as under the ESOP described above. Each stock option entitles its holder to purchase one Share of the Company at an exercise price defined to be either a) equal to the volume-weighted average share price in the three preceding months, or b) the closing share price on the SIX at each grant date. In general, 50% of the stock options vest on the second anniversary, 25% on the third anniversary and the remaining 25% on the fourth anniversary of the grant date. At the end of the option term, i.e. after a period of 10 years as from the grant date, unexercised stock options expire without value. Under the BSOP 2011 vested stock options of Board members leaving the Board in good faith expire six months after the termination date of them being a member of the Board while unvested stock options of Board members leaving the Board in good faith are forfeited. Under the BSOP 2015 vested and unvested stock options of Board members leaving the Board in good faith do not expire. No further grants can be made under the BSOP.

Options outstanding, exercised, forfeited or expired under BSOPs

Number of options						2017
	At January 1	Exercised	Granted	Forfeited	Expired	At December 31
BSOP 2015	13,562	0	0	0	0	13,562
Total	**13,562**	**0**	**0**	**0**	**0**	**13,562**

Number of options						2016
	At January 1	Exercised	Granted	Forfeited	Expired	At December 31
BSOP 2015	7,000	0	6,562	0	0	13,562
Total	**7,000**	**0**	**6,562**	**0**	**0**	**13,562**

Since July 1, 2016, no more stock options are available for future grants under the ESOP 2015 and/or the BSOP 2015. Stock option plans were replaced by Share Appreciation Rights (**SAR**), see note 17.2 *"Share Appreciation Rights Plans".*

Fair value calculations for stock options granted

The fair value of stock options is determined at each grant date by using the Hull-White pricing model. The calculation of the option value was performed by applying the following parameters:

	2016
Market price of stock	CHF 37.05 to 91.25
Exercise prices	CHF 69.30 to 89.45
Weighted average fair value of options granted	CHF 24.18
Expected volatility[1]	38% to 39%
CHF risk-free interest rate	0.0% p.a.
Option term[2]	10 years
Expected dividend yield	0%

[1] The expected volatility was determined on the basis of selected biotech companies.

[2] After expiration of the vesting period, the stock options become American-style options and may be exercised any time until the end of the option term. The option-pricing model takes into consideration certain assumptions about potential early exercises.

Number of stock options outstanding and exercisable

Number of options	2017	2016
Outstanding at January 1	**313,365**	**223,834**
Granted	0	142,392
Exercised[1]	–8,698	–17,059
Forfeited	–15,472	–15,289
Expired	–753	–20,513
Outstanding at December 31	**288,442**	**313,365**
Exercisable at December 31	**102,642**	**36,327**

[1] The average closing share price of options exercised during the reporting period 2017 was CHF 39.05 (2016: CHF 68.12).

The value of stock options granted is recognized as personnel expense over the period Santhera receives services. In 2017, previously granted stock options resulted in personnel expenses of TCHF 2,778 (TCHF 374 related to Development, TCHF 1,525 related to Marketing and sales (**M&S**) and TCHF 879 to General and administrative (**G&A**)) and in 2016, such grants resulted in personnel expenses of TCHF 3,311 (TCHF 418 related to Development, TCHF 1,766 related to M&S and TCHF 1,127 to G&A).

Terms of options outstanding at December 31

Exercise price range for options (in CHF)	Number outstanding	Weighted average remaining contractual life (years)	**2017 Number exercisable**	Number outstanding	Weighted average remaining contractual life (years)	**2016 Number exercisable**
from 3.85 to 4.53	25,001	5.13	25,001	33,699	6.66	33,574
at 22.25	4,550	6.50	3,275	4,550	7.48	2,000
at 69.30	14,800	8.25	0	18,800	9.23	0
from 82.10 to 114.50	244,091	7.73	74,366	256,316	8.61	753
Total	**288,442**	**7.61**	**102,642**	**313,365**	**8.42**	**36,327**

17.2 Share Appreciation Rights Plans

Starting with July 1, 2016, Santhera switched from stock option plans to Share Appreciation Rights Plans (**SARP**). It introduced a Board Share Appreciation Plan 2016 (**BSARP 2016**) and an Employee Share Appreciation Rights Plan 2016 (**ESARP 2016**). In 2017 Santhera has introduced a Board Share Appreciation Rights Plan (**BSARP 2017**) for the members of its Board and an Employee Share Appreciation Rights Plan (**ESARP 2017**) for the Executive Management, employees and consultants. SAR grants are made periodically at the discretion of the Board or as contractually agreed with employees. The SARP contain customary provisions in respect of the adjustment or cancellation of SARs upon termination of employment, retirement, death, disability and certain corporate transactions. All SARPs are administered under the responsibility of the Board. In general, 50% of the SARs vest on the second anniversary, 25% on the third anniversary and the remaining 25% on the fourth anniversary of the grant date. SARP introduced in 2017 (BSARP 2017 and ESARP 2017) foresee vesting of 1/3 of the SAR on the first anniversary; the remaining 2/3 vest by each following quarter end through the second and third year after the grant date

(8 times 1/12 of the SAR granted). At the end of the SAR term, i.e. after a period of 10 years as from the grant date, unexercised SARs expire without value. Upon exercise of one SAR, participants receive the difference between the price of one Share at the time of exercise and the base value ("exercise price" as defined upon grant), in Shares. Subsequently, participants may sell their Shares.

SAR outstanding, exercised, forfeited or expired under SARP

Number of SAR						2017
	At January 1	Exercised	Granted	Forfeited	Expired	At December 31
ESARP 2016	56,581	0	63,889	–34,881	0	85,589
BSARP 2017	0	0	15,120	0	0	15,120
ESARP 2017	0	0	271,234	–11,833	0	259,401
Total	**56,581**	**0**	**350,243**	**–46,714**	**0**	**360,110**

Number of SARs						2016
	At January 1	Exercised	Granted	Forfeited	Expired	At December 31
ESARP 2016	0	0	56,581	0	0	56,581
Total	**0**	**0**	**56,581**	**0**	**0**	**56,581**

Fair value calculations for SAR granted

The fair value of SAR is determined at each grant date by using the Hull-White pricing model. The calculation of the SAR value was performed by applying the following parameters:

	2017	2016
Market price of stock	CHF 29.00 to 82.00	CHF 37.05 to 91.25
Exercise prices	CHF 38.60 to 77.80	CHF 51.75 to 76.50
Weighted average fair value of SAR granted	CHF 25.94	CHF 22.12
Expected volatility[1]	38%	38% to 39%
CHF risk-free interest rate	0.0% p.a.	0.0% p.a.
SAR term[2]	10 years	10 years
Expected dividend yield	0%	0%

[1] The expected volatility was determined on the basis of selected biotech companies.

[2] After expiration of the vesting period, the SARs become rights similar to American-style options and may be exercised any time until the end of the SAR term. The SAR pricing model takes into consideration certain assumptions about potential early exercises.

Number of SAR outstanding and exercisable

Number of SAR	2017	2016
Outstanding at January 1	**56,581**	**0**
Granted	350,243	56,581
Exercised	0	0
Forfeited	–46,714	0
Expired	0	0
Outstanding at December 31	**360,110**	**56,581**
Exercisable at December 31	**0**	**0**

The value of SAR granted is recognized as personnel expense over the period Santhera receives services. In 2017, SAR grants resulted in personnel expenses of TCHF 4,517 (TCHF 1,513 related to Development, TCHF 1,513 related to M&S and TCHF 1,491 to G&A) and in 2016, such grants resulted in personnel expenses of TCHF 150 (TCHF 15 related to Development, TCHF 135 related to M&S and TCHF 0 to G&A).

Santhera plans to allocate up to 444,164 SAR in the first quarter of 2018 (in the first quarter 2017, it was planned to allocate up to 198,162 SAR). These SAR form part of the long-term incentive (**LTI**) award to employees for the year ended December 31, 2017. Although these SAR were not legally granted in 2017, Executive Management considers it appropriate to recognize expenses in 2017 as employees have been rendering services in 2017 in expectation of the annual LTI allocation. Personnel expenses in 2017 for this amounted to TCHF 2,392 (TCHF 825 related to Development, TCHF 655 related to M&S and TCHF 912 related to G&A) based on an estimate of fair value (in 2016 personnel expenses for this amounted to TCHF 1,222 (TCHF 332 related to Development, TCHF 569 related to M&S and TCHF 321 related to G&A)). The allocation of these SAR is conditional for the Executive Management and becomes unconditional once the compensation is approved on the occasion of the AGM, to be held on April 12, 2018. After the AGM the grant date fair value of the SAR will be determined and the cumulative expense adjusted.

Terms of SAR outstanding at December 31

Exercise price range for SAR (in CHF)	Number outstanding	Weighted average remaining contractual life (years)	2017 **Number exercisable**	Number outstanding	Weighted average remaining contractual life (years)	2016 **Number exercisable**
at 38.70	33,257	9.76	0	n/a	n/a	n/a
from 51.75 to 76.50	283,435	9.01	0	56,581	9.71	0
from 76.50 to 77.80	43,418	9.08	0	n/a	n/a	n/a
Total	**360,110**	**9.04**	**0**	**56,581**	**9.71**	**0**

18 Segment and Geographic Information

Segment information

Santhera operates in one operating segment, the development and commercialization of specialty niche products for the treatment of neuro-ophthalmological, neuromuscular and pulmonary diseases. The Board, the Executive Management and senior managers, being the CODM, assess the reporting data and allocate resources as one segment on a consolidated level according to the operating expenses by function. Santhera generates revenue from sales of Raxone for the treatment of LHON. Geographic revenue information is based on location of the customer.

Geographic information

Net sales

In CHF thousands	2017	2016
EU	22,859	19,002
Rest of the world	84	31
Total	**22,943**	**19,033**

In 2017, net sales amounted to CHF 22.9 million (2016: CHF 19.0 million). Raxone was sold in 20 European countries, with the majority of sales reached in France and Germany (2016: 15 European countries).

Noncurrent assets (excluding financial instruments, restricted cash and deferred taxes)

In CHF thousands	2017	2016
Switzerland	25,451	26,966
EU	171	100
North America	95	0
Total	**25,717**	**27,066**

19 Other Operating Income

This position consists primarily of reimbursements from scientific programs.

20 Operating Expenses by Nature

In CHF thousands	2017	2016
External Development expenses	−14,762	−12,119
Patent and license expenses	−381	−280
Marketing expenses	−13,018	−10,121
Employee expenses	−35,488	−21,403
Of which non-cash-relevant expenses for equity rights plans	*−9,687*	*−4,683*
Other administrative expenses	−4,727	−3,796
Depreciation and amortization	−343	−225
Lease expenses	−780	−587
Other operating expenses	−64	−107
Total operating expenses	**−69,563**	**−48,638**

21 Employee Expenses and Benefits

Employee expenses

In CHF thousands	2017	2016
Wages and salaries	−18,372	−12,397
Social security and other personnel-related expenses[1]	−7,429	−4,323
Of which non-cash-relevant adjustments of pension fund	*−2,021*	*−450*
Expenses for equity rights plans	−9,687	−4,683
Total employee costs	**−35,488**	**−21,403**

Average number of full-time equivalents[2]	**92.9**	**65.1**
Full-time equivalents at year-end	**106.2**	**74.4**
Total headcount at year-end	**112**	**80**

[1] Thereof TCHF 306 were expensed for defined contribution plans in North America and some EU countries (2016: TCHF 124).

[2] For the calculation of full-time equivalents, only employees with part-time and full-time permanent working contracts are taken into consideration.

Pension plan

In accordance with the Swiss pension fund law "Federal Act on Occupational Old Age, Survivors' and Invalidity Pension Provision" (**OPA**), all employees of Santhera Pharmaceuticals Holding AG, Liestal, and Santhera Pharmaceuticals (Schweiz) AG, Pratteln, both in Switzerland, have to be affiliated with a collective independent pension fund. These funds provide for retirement benefits, as well as risk benefits (death and disability). The plans qualify as defined benefit plans under IAS 19 and the assets cannot revert to the employer. Contributions to the plans are such that the employee contributes 40% and the employer the rest. Contributions are computed as percentage of the salary, depending on age. In order to manage these risks, Santhera had an agreement with AXA Foundation for occupational benefits (**AXA**

foundation) during 2017. As of January 1, 2018, Santhera has entered into an agreement with PKG Pensionskasse (**PKG**) and changed its pension fund provider, effective as of this date. The AXA foundation and PKG are responsible for the governance of the plan; their boards are composed of an equal number of representatives from the employers and employees chosen from all affiliated companies. AXA foundation and PKG have set up investment guidelines, defining in particular the strategic allocation with margins. AXA foundation has reinsured its risks (investment risk, mortality risk, etc.) with AXA Life Ltd, Winterthur, Switzerland (**AXA**), whereas PKG has only insured the risks disability and death before retirement with PKRück AG, Vaduz, Fürstentum Liechtenstein. The accumulated savings capital is allocated to each insured individual and consists of annual contributions, savings credits and interest credits. In certain situations, additional payments or increased periodic contributions by the employer may become due based on the pension plans funded status as measured under Swiss pension rules (**OPA**).

An independent actuary has performed the respective calculations as required by IAS 19:

Changes in defined benefit obligations

In CHF thousands	2017	2016
Present value of obligation, January 1	**21,279**	**15,797**
Current employer service cost	1,614	1,038
Past service expense[1]	1,240	0
Interest cost	138	142
Employee contributions	514	382
Benefits paid / transfer payments	–620	2,259
Insurance premiums	–266	–192
Remeasurements[2]	253	1,853
Present value of obligation, December 31	**24,152**	**21,279**

[1] Increase of obligation due to increase of the conversion rates for the over-mandatory part of the retirement capital under the new PKG plan rules (based on the agreements signed in November 2017).

[2] Details of remeasurements:

In CHF thousands	2017	2016
Effect of changes in demographic assumptions	0	–435
Actuarial gain/loss due to changes in financial assumptions	–400	599
Actuarial gain/loss due to experience adjustments	653	1,689
Subtotal gain/loss	**253**	**1,853**
Return/loss on plan assets (excluding interest income)	–82	–77
Total remeasurements in other comprehensive income gain/loss	**171**	**1,776**

Changes in plan assets

In CHF thousands	2017	2016
Fair value of assets, January 1	**15,096**	**11,840**
Interest income on assets	101	110
Employer contributions	870	620
Employee contributions	514	382
Benefits paid / transfer payments	–620	2,259
Insurance premiums	–266	–192
Remeasurements (return/loss on plan assets (excluding interest income))	82	77
Fair value of assets, December 31	**15,777**	**15,096**

Net defined benefit asset/obligation

In CHF thousands	2017	2016
Present value of obligation, December 31	24,152	21,279
Fair value of assets, December 31	15,777	15,096
Net defined asset/obligation	**–8,375**	**–6,183**

Asset breakdown

In CHF thousands	2017		2016	
	Quoted market price	Not quoted market price	Quoted market price	Not quoted market price
Insurance contract	0	15,777	0	15,096
Total value of assets	**0**	**15,777**	**0**	**15,096**

An asset breakdown is not available. The assets of Santhera's defined benefit plan have no quoted market price since AXA fully insures them as an insurance contract.

The weighted average assumptions to determine benefit obligations and defined benefit cost were as follows:

In %	2017	2016
Discount rate	0.70	0.65
Expected future salary increases	1.50	1.50

Sensitivity analysis for 2017:

In CHF thousands	Defined benefit obligation		Gross (net) service cost	
	Increase assumption	Decrease assumption	Increase assumption	Decrease assumption
Discount rate +/−0.25%	−1,027	1,105	−129	137
Salary increase +0.25%	156	–	0	–
Life expectancy +1 year	448	–	35	–

Sensitivity analysis for 2016:

In CHF thousands	Defined benefit obligation		Gross (net) service cost	
	Increase assumption	Decrease assumption	Increase assumption	Decrease assumption
Discount rate +/−0.25%	−896	961	−117	125
Salary increase +0.25%	130	–	−1	–
Life expectancy +1 year	431	–	32	–

Mortality rate:

Life expectancy at age 65	2017	2016
Male	22.5	22.4
Female	24.5	24.4

The expected employer contributions for fiscal year 2018 amount to approximately TCHF 936 (2016: TCHF 876). Benefit obligations of pensioners amounted to TCHF 3,103 at December 31, 2017 (2016: none). The duration of the plan liabilities calculated is 20.3 years as of December 31, 2017 (2016: 21.6 years).

22 Financial Income/Expenses

Financial income

In CHF thousands	2017	2016
Interests on cash and cash equivalents	5	5
Change in fair value of financial derivative instruments	2,540	0
Income from financial assets	267	0
Realized and unrealized foreign exchange gains	1,322	923
Total	**4,134**	**928**

Financial expenses

In CHF thousands	2017	2016
Interest expenses	−3,843	−15
Expenses from financial assets	−197	0
Realized and unrealized foreign exchange losses	−915	−980
Total	**−4,955**	**−995**

23 Income Taxes

In CHF thousands	2017	2016
Current income tax income/expense	−390	−266
Deferred tax income/expense	133	−1,955
Total	**−257**	**−2,221**

The following is a theoretical reconciliation of tax expense and the accounting profit multiplied by expected income tax rate of principal:

In CHF thousands	2017	2016
Result before taxes	−51,275	−33,194
Tax expense/income at expected group tax rate of 9.3%	4,769	3,087
Effect of tax rate difference group versus local	−422	−724
Effect of nondeductible expenses	−784	−372
Prior year DTA (deferred tax assets) decrease	−289	−249
Utilization of previously unrecognized tax losses	24	20
Recognition of previously unrecognized DTL (deferred tax liabilities)	0	−13,449
Recognition of DTA on previously unrecognized tax losses	0	13,449
Unrecognized deferred taxes	−3,555	−3,983
Effective tax income/expense	**−257**	**−2,221**

According to currently applicable Swiss tax law, the period to offset tax loss carryforwards against taxable profit is limited to seven years. According to currently applicable German tax law, tax loss carryforwards can, besides other conditions, be offset against taxable profit for an unlimited period but only to an amount of EUR 1.0 million and in addition for 60% of further amounts beyond this threshold per annum.

24 Earnings/Loss per Share

Basic earnings/loss per share is calculated by dividing the net profit/net loss attributable to equity holders by the weighted average number of Shares issued and outstanding during the reporting period, excluding Shares held as treasury shares (purchased at market).

	2017	2016
Net result attributable to shareholders (in CHF)	−51,531,770	−35,414,845
Weighted average number of shares issued and outstanding	6,269,813	6,273,460
Basic and diluted net result per share (in CHF)	**−8.22**	**−5.65**

For the years ended December 31, 2017 and 2016, basic and diluted net result per share is based on the weighted average number of Shares issued and outstanding and excludes Shares to be issued upon the future exercise of equity rights and upon conversion of the convertible bonds, as they would be anti-dilutive. In case Santhera shows a profit in the future, equity rights and convertible bonds upon conversion may have a dilutive effect on the net profit per Share and will need to be considered for the purpose of this calculation.

25 Related Party Transactions

Board and Executive Management compensation

Total compensation of Board and Executive Management

In CHF thousands	2017	2016
Compensation, wages and salaries	3,344	2,546
Post-employment benefits (pension fund contributions)	272	221
Share-based payment expenses (fair value according to IFRS 2)	3,826	1,508
Total	**7,442**	**4,275**

Transactions with members of the Board and Executive Management

There are no loans outstanding or guarantee commitments granted to members of the Board and Executive Management.

In 2017, no stock options were exercised by members of the Board (2016: no stock options exercised). During 2017, 5,000 stock options were exercised by the Executive Management (2016: 3,999 stock options exercised).

26 Risk Management Objectives and Policies

Santhera Pharmaceuticals Holding AG maintains a Group-wide corporate risk management system consisting of the areas corporate governance, financial internal controls and quality control / quality assurance.

On a regular basis, operational corporate risks are identified and their likelihood and impact assessed (gross risks). By defining and undertaking appropriate measures, these risks are managed accordingly to either reduce or avoid such risk (net risk). The results of this process are discussed at Board meetings.

Those risks as identified within the area of accounting and financial reporting as well as related control processes are further covered by the Company's Group-wide internal control system.

Santhera conducts development activities primarily in Switzerland, the EU and the US and is exposed to a variety of financial risks, such as, but not limited to, foreign exchange rate risk, credit risk, liquidity risk, cash flow and interest rate risk. Part of Santhera's overall risk management focuses on financial risks and the unpredictability of financial markets seeking to minimize potential adverse effects on the financial performance of the Group. Special guidelines and policies approved by the Board exist for overall risk management, financial internal controls and treasury management and are monitored by the Executive Management and the Board on a regular basis. The risk of foreign exchange rate fluctuations on the expenses can partly be managed by entering into foreign exchange derivative contracts. In accordance with the relevant treasury guidelines, Santhera only concludes contracts with selected high-quality financial institutions of good reputation and is not allowed to engage in speculative transactions. In addition, Santhera's treasury guidelines limit the Company to engage in money market deposits or similar instruments with a maturity beyond 6 months.

Foreign exchange rate risk

Santhera holds cash amounts in five major currencies CHF, EUR, USD, GBP and CAD to cover the majority of future expected expenses. In addition, in order to reduce its foreign exchange rate exposure, Santhera occasionally enters into derivative currency contracts (forwards, options, structured derivatives) to hedge against additional major foreign currency exchange rate fluctuations. Evaluations based on market values are performed regularly. Any fair value changes of such currency positions are recorded accordingly in the income statement. Santhera's primary exposure to financial risk is due to fluctuation of exchange rates between CHF, EUR, USD, GBP and CAD. No derivative currency contracts are outstanding as of December 31, 2017 and 2016.

The following table demonstrates the sensitivity to a reasonable possible change in the EUR exchange rate, with all other variables held constant, of the Group's result before taxes. There is no impact on the Group's equity.

	Increase/decrease foreign currency rate	Effect on result before taxes in CHF thousands
EUR positions		
2017	+5%	+411
	−5%	−411
2016	+5%	+187
	−5%	−187

Interest rate risk

Santhera earns interest income on cash and cash equivalents and its profit and loss may be influenced by changes in market interest rates. Santhera holds its cash on deposit/current accounts or invests cash through deposits in line with its treasury guidelines to follow its financial needs over time.

The following calculation demonstrates the sensitivity to a reasonable change in interest rates, with all other variables held constant, of the Group's result before taxes. There is no impact on the Group's equity.

As of the end of 2017, variances of +/−50 basis points were calculated, resulting in fluctuations of +/−TCHF 263 before tax (end of 2016: +/−50 basis points resulting in fluctuations of +/−TCHF 249 before tax).

Additionally, Santhera's interest rate risk arises from long-term debt. Debt issued at fixed rates exposes the Group to fair value interest rate risk.

Credit risk

Santhera has a certain concentration of credit risk. Short-term investments are invested as cash on deposit or in low-risk money market funds. No investment or contract with any single counterparty, except cash on deposit subject to the criteria above, comprises more than 30% of cash and cash equivalents at the date of investment.

Santhera has policies in place to ensure that sales of products or entered partnerships are made to or entered with customers or partners with an appropriate credit history and a commitment to ethical business practices. The maximum credit risk exposure is limited to the carrying amount of its financial assets including derivatives.

Liquidity risk

Prudent liquidity risk management implies maintaining sufficient cash and cash equivalents. Currently, the Company is financed through equity and convertible bonds (see note 12 *"Financial Assets and Liabilities"*). Santhera's treasury calculates on a rolling basis the needs for aligning the current expenses against the need for optimized financial investments.

Contractual undiscounted cash flows

Year ended December 31, 2017 In CHF thousands	On demand	Less than 3 months	3 to 12 months	1 to 5 years	Total	Book value
Convertible bonds	0	1,500	1,500	70,500	73,500	53,111
Trade payables	0	3,585	0	0	3,585	3,585
Accrued expenses	0	5,083	0	0	5,083	5,083
Total	**0**	**10,168**	**1,500**	**70,500**	**82,168**	**61,779**

Year ended December 31, 2016 In CHF thousands	On demand	Less than 3 months	3 to 12 months	1 to 5 years	Total	Book value
Trade payables	0	3,574	0	0	3,574	3,574
Accrued expenses	0	3,787	0	0	3,787	3,787
Total	**0**	**7,361**	**0**	**0**	**7,361**	**7,361**

Categories of financial instruments

Year ended December 31, 2017 In CHF thousands	Book value	Loans and receivables	Other liabilities at amortized cost	At fair value through profit or loss
Assets				
Financial assets long-term	713	713	0	0
Restricted cash long-term	4,500	4,500	0	0
Trade receivables	4,194	4,194	0	0
Other receivables	149	149	0	0
Financial assets short-term	13,011	13,011	0	0
Restricted cash short-term	3,000	3,000	0	0
Cash and cash equivalents	45,195	45,195	0	0
Total	**70,762**	**70,762**	**0**	**0**
Liabilities				
Convertible bonds	53,111	0	53,111	0
Derivative financial instruments	2,792	0	0	2,792
Trade payables	3,585	0	3,585	0
Accrued expenses	5,083	0	5,083	0
Total	**64,571**	**0**	**61'779**	**2,792**

Year ended December 31, 2016 In CHF thousands	Book value	Loans and receivables	Other liabilities at amortized cost	At fair value through profit or loss
Assets				
Financial assets long-term	270	270	0	0
Trade receivables	3,412	3,412	0	0
Other receivables	55	55	0	0
Cash and cash equivalents	49,815	49,815	0	0
Total	**53,552**	**53,552**	**0**	**0**
Liabilities				
Trade payables	3,574	0	3,574	0
Accrued expenses	3,787	0	3,787	0
Total	**7,361**	**0**	**7,361**	**0**

Capital management

The first priority of Santhera's capital management is to provide adequate cash funds to ensure the financing of successful development and marketing activities so that future profits can be generated by gaining marketing authorization approvals for pharmaceutical products. As a company with currently only one marketed product, the capital management continues to be focused on the cash and cash equivalents position and is governed by specific Group treasury guidelines.

The funds raised in various private financing rounds, private placements in 2008, 2014 and 2015, SEDA (Standby Equity Distribution Agreement), the sale of Shares by an independent broker, convertible bonds as well as funds generated through product sales and revenue from licensing enabled the Group to be adequately financed.

Minor changes in goals and policies of the treasury management have been made in 2017, such as e.g. the extension from 20% to 30% for short-term investments with one counterparty or the possibility of physical cash deposits (there were no changes in 2016).

27 Events after the Reporting Date

On January 26, 2018, Santhera announced that the Committee for Medicinal Products for Human Use (CHMP) of the European Medicines Agency (**EMA**) maintained its negative opinion on the Type II extension application for idebenone in Duchenne muscular dystrophy (**DMD**) following a re-examination procedure. The CHMP concluded that an approval for idebenone in DMD, applied as a Type II variation of the existing marketing authorization, cannot be granted at the present time based on the current existing evidence. Although the positive outcome of the Phase III DELOS trial was acknowledged, the CHMP has invited Santhera to present additional data to further link the observed treatment effects on respiratory function outcomes to patient benefit. Management has considered the impact on the Group's ability to continue as a going concern and potential indicators of impairment on the financial statements as of December 31, 2017.

On February 15, 2018, Santhera announced that it has entered into a license agreement with Polyphor Ltd., Allschwil, Switzerland, for POL6014, a clinical stage selective inhibitor of human neutrophil elastase with the potential to treat cystic fibrosis (**CF**) and other neutrophilic pulmonary diseases such as non-cystic fibrosis bronchiectasis (**NCFB**), alpha-1 antitrypsin deficiency (**AATD**) and primary ciliary dyskinesia (**PCD**).

Santhera will assume the global development, regulatory filings and commercialization of POL6014. The development program has been advanced with financial support by the Cystic Fibrosis Foundation Therapeutics Inc. (**CFFT**), USA, to Polyphor. With POL6014, Santhera is expanding its product pipeline in pulmonary diseases where the Company is already developing its lead product idebenone for respiratory complication in Duchenne muscular dystrophy.

Under the agreement, Santhera obtains the worldwide, exclusive rights to develop and commercialize POL6014, an innovative macrocycle elastase inhibitor, and analogs for an initial payment of CHF 6.5 million, payable in Santhera shares at an agreed valuation of CHF 27.2053 per share and additional cash payments of up to CHF 121 million contingent to future development, regulatory and particularly sales milestones. In addition, Polyphor is entitled to tiered royalty payments from Santhera´s future net sales of POL6014 and to undisclosed milestone payments and royalties provided that Santhera advances the development and market entry of POL6014 in other pulmonary diseases. Santhera issued the 238,924 shares (3.8% of its currently issued shares) required for the initial payment to Polyphor out of its existing authorized share capital.

Building a better working world

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To the General Meeting of
Santhera Pharmaceuticals Holding Ltd, Liestal

Basle, March 19, 2018

Statutory auditor's report on the audit of the consolidated financial statements

Opinion

We have audited the consolidated financial statements of Santhera Pharmaceuticals Holding Ltd and its subsidiaries (the Group), which comprise the consolidated balance sheet as at December 31, 2017, and the consolidated income statement, consolidated statement of comprehensive income, consolidated cash flow statement and consolidated statement of changes in equity for the year then ended, and notes to the consolidated financial statements, including a summary of significant accounting policies.

In our opinion, the consolidated financial statements (pages 16 to 57) give a true and fair view of the consolidated financial position of the Group as at December 31, 2017, and its consolidated financial performance and its consolidated cash flows for the year then ended in accordance with International Financial Reporting Standards (IFRS) and comply with Swiss law.

Basis for opinion

We conducted our audit in accordance with Swiss law, International Standards on Auditing (ISAs) and Swiss Auditing Standards. Our responsibilities under those provisions and standards are further described in the *Auditor's Responsibilities for the Audit of the Consolidated Financial Statements* section of our report.

We are independent of the Group in accordance with the provisions of Swiss law and the requirements of the Swiss audit profession, as well as the IESBA Code of Ethics for Professional Accountants, 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.

Key audit matters

Key audit matters are those matters that, in our professional judgment, were of most significance in our audit of the consolidated financial statements of the current period. These matters were addressed in the context of our audit of the consolidated financial statements as a whole, and in forming our opinion thereon, and we do not provide a separate opinion on these matters. For each matter below, our description of how our audit addressed the matter is provided in that context.

We have fulfilled the responsibilities described in the *Auditor's responsibilities for the audit of the consolidated financial statements* section of our report, including in relation to these matters. Accordingly, our audit included the performance of procedures designed to respond to our assessment of the risks of material misstatement of the consolidated financial statements. The results of our audit procedures, including the procedures performed to address the matters below, provide the basis for our audit opinion on the consolidated financial statements.

Management's assessment of the Group's ability to continue as a going concern

Areas of focus

Based on the stage of the operations combined with recurring development costs and the level of cash inflows from its operating activities, the Group incurred negative cash flows from operations in the past and it expects to continue to incur such costs and negative cash flows in the near future. Management has prepared various budget scenarios and business plans and has assessed the likelihood and impact that uncertain cash flows could have on the Group's ability to continue as going concern. This assessment was based on the evaluation of the available year-end funds of CHF 45.2 million as well as the budget.

Despite the Group's development into a more sales oriented business, funding from non-revenue related sources is still required in the future and we therefore concluded that the assessment of the ability to continue as a going concern represents a key audit matter for our audit.

Our audit response

Our audit procedures related to the key audit matter of the assessment of the going concern included the following procedures:

- We gained an understanding of management's budgeting and forecasting process underlying the going concern assessment.
- We discussed the budget and business plans with management, evaluated the development and the assessments made and also considered how uncertain elements were included in the budget and business plans. We evaluated underlying key assumptions such as expected cash inflow from product sales and expected cash outflow from purchases of inventory, research and development expenses and other operating expenses.
- We tested the sensitivity of the revenue estimates and assessed how they compared to historical revenues.
- We further discussed the base case scenario with management and assessed the likelihood of this scenario.
- We performed inquiries of management about current developments and the ability to execute the elements included in the budget and business plans supporting the evaluation of the going concern assumption. We read the Board of Directors meeting minutes that approved the underlying budget to assess potential contrary information. We performed procedures regarding subsequent events to assess the accuracy of the budget as of the date of the auditor's report.

Accounting for board and employee share appreciation rights

Areas of focus

The Group operates several equity right plans for its employees and board members. Grants are made periodically at the discretion of the board or as contractually agreed with employees. The fair value of the equity rights and ultimately personnel expenses to be recognized are determined based upon assumptions. In the reporting period 2017 expenses recorded in reference to equity right plans amounted to CHF 9.7 million compared to CHF 4.7 million in the same period in 2016.

Refer to note 17 to these financial statements disclosures related to equity rights plans.

Our audit response

We tested the fair value determination for all grants in the year 2017 and assessed the accuracy of the share-based payment expenses recognized. This included, among others, the following procedures:

- We obtained and read documentation related to new share appreciation right plans established during financial year 2017 to understand the terms under which these rights were granted.
- We inspected new grants on a sample basis and referenced the grants to supporting documentation such as the communication to the employees. Further, we reconciled the number of awards granted to the calculation of the expenses and recalculated the amounts to be recognized over the vesting period.
- We assessed management's assumptions used in the calculation of the expenses by comparing these to market and historical data. Assumptions assessed included forfeiture rates.
- We also considered the adequacy of the disclosures made in relation to share-based payments.

Other information in the annual report

The Board of Directors is responsible for the other information in the annual report. The other information comprises all information included in the annual report, but does not include the consolidated financial statements, the stand-alone financial statements, the compensation report and our auditor's reports thereon.

Our opinion on the consolidated financial statements does not cover the other information in the annual report and we do not express any form of assurance conclusion thereon.

In connection with our audit of the consolidated financial statements, our responsibility is to read the other information in the annual report and, in doing so, consider whether the other information is materially inconsistent with the consolidated financial statements or our knowledge obtained in the audit, or otherwise appears to be materially misstated. If, based on the work we have performed, we conclude that there is a material misstatement of this other information, we are required to report that fact. We have nothing to report in this regard.

Responsibility of the Board of Directors for the consolidated financial statements
The Board of Directors is responsible for the preparation of the consolidated financial statements that give a true and fair view in accordance with IFRS and the provisions of Swiss law, and for such internal control as the Board of Directors determines is necessary to enable the preparation of consolidated financial statements that are free from material misstatement, whether due to fraud or error.

In preparing the consolidated financial statements, the Board of Directors is responsible for assessing the Group's ability to continue as a going concern, disclosing, as applicable, matters related to going concern and using the going concern basis of accounting unless the Board of Directors either intends to liquidate the Group or to cease operations, or has no realistic alternative but to do so.

Auditor's responsibilities for the audit of the consolidated financial statements
Our objectives are to obtain reasonable assurance about whether the consolidated 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 Swiss law, ISAs and Swiss Auditing Standards 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 consolidated financial statements.

A further description of our responsibilities for the audit of the consolidated financial statements is located at the website of EXPERTsuisse: http://www.expertsuisse.ch/en/audit-report-for-public-companies. This description forms part of our auditor's report.

Report on other legal and regulatory requirements
In accordance with article 728a para. 1 item 3 CO and the Swiss Auditing Standard 890, we confirm that an internal control system exists, which has been designed for the preparation of consolidated financial statements according to the instructions of the Board of Directors.

We recommend that the consolidated financial statements submitted to you be approved.

Ernst & Young Ltd

/s/ Frederik Schmachtenberg
Licensed audit expert
(Auditor in charge)

/s/ Jan Meyer
Licensed audit expert

Statutory Financial Statements of Santhera Pharmaceuticals Holding AG

Contents

Balance Sheet

As of December 31, in CHF thousands	Notes	2017	2016
Assets			
Cash and cash equivalents		30,738	43,187
Financial assets short-term		13,011	0
Other receivables from third parties		54	24
Prepaid expenses and accrued income		153	46
Restricted cash short-term		3,000	0
Current assets		**46,956**	**43,257**
Loans to shareholdings	3.1	63,293	17,727
Investments in shareholdings	3.2	198	115
Restricted cash long-term		4,500	0
Noncurrent assets		**67,991**	**17,842**
Total assets		**114,947**	**61,099**
Liabilities and equity			
Trade accounts payable to third parties		311	97
Other accounts payable to third parties		35	56
Accrued expenses		1,538	453
Current liabilities		**1,884**	**606**
Senior unsecured convertible bonds[1]	2	60,000	0
Noncurrent liabilities		**60,000**	**0**
Total liabilities		**61,884**	**606**
Share capital	3.3	6,289	6,280
Reserves from capital contributions		*7,450*	*7,425*
Other capital reserves		*2,916*	*2,916*
Statutory capital reserves		10,366	10,341
Accumulated result		*–13,752*	*–6,451*
Results carried forward		*–6,451*	*–5,557*
Net result for the period		*–7,301*	*–895*
Other voluntary reserves (free reserves)		*50,495*	*50,495*
Voluntary accumulated result and other reserves		36,743	44,044
Treasury shares	3.4	–335	–172
Total equity		**53,063**	**60,493**
Total liabilities and equity		**114,947**	**61,099**

[1] interest bearing

Income Statement

For the year ended December 31, in CHF thousands	Notes	2017	2016
Income from shareholdings	3.5	1,325	1,551
Other operating income		11	134
Total operating income		**1,336**	**1,685**
General and administrative expenses	3.6	−5,407	−2,159
Employee costs		−1,122	−792
Other operating expenses		−12	−25
Total operating expenses		**−6,541**	**−2,976**
Operating result		**−5,205**	**−1,291**
Financial income		1,011	358
Financial expenses		−3,189	−18
Financial result		**−2,178**	**340**
Reversal on allowance of investment		82	56
Result before taxes		**−7,301**	**−895**
Direct taxes		0	0
Net result		**−7,301**	**−895**

Notes to the Statutory Financial Statements

1 Introduction

Santhera Pharmaceuticals Holding AG (the Company or Santhera) is the parent company of Santhera Group. The Company has its business offices at Hohenrainstrasse 24 in 4133 Pratteln, Switzerland. The legal domicile will remain at Hammerstrasse 49 in 4410 Liestal, Switzerland, until the forthcoming Annual General Meeting when the shareholders will vote on its relocation to Pratteln.

2 Principles

General

The statutory financial statements of the Company are prepared in accordance with the general accepted accounting principles as set out in Art. 957 to Art. 963b, of the Swiss Code of Obligations (**CO**). Since Santhera prepares consolidated financial statements in accordance with International Financial Reporting Standards (**IFRS**) of the International Accounting Standards Board (**IASB**), a recognized accounting standard, the Company has, in accordance with the CO, elected to forego presenting the statement of cash flows, the additional disclosures and the management report otherwise required by the CO.

Cash

Santhera holds cash balances, denominated mainly in Swiss francs (**CHF**) which include cash deposited in demand bank accounts, money market investment accounts and other liquid investments and interest earned on such cash balances.

Financial assets short-term

Financial assets (units in a fund) are held for trading and measured at fair value. In case of gains and losses from such assets are recognized through the income statement as financial income or financial expense.

Current assets and liabilities

Current assets are recorded at historical cost less adjustments for impairment of value and current liabilities at historical cost.

Loans to shareholdings

These are valued at their acquisition cost adjusted for impairment losses.

Investments in shareholdings

Investments in shareholdings are recorded at acquisition cost less adjustments for impairment of value. Investments in subsidiaries are evaluated for impairment annually and an impairment loss is recorded when the carrying amount of such assets exceeds the fair value. Fair value estimates of investments are predominantly based on the income approach.

Convertible bonds

On February 17, 2017, Santhera issued senior unsecured convertible bonds in the nominal amount of CHF 60 million. The bonds were issued at 100% of the principal amount and will also mature at 100% of that amount on February 17, 2022, unless previously redeemed, converted or repurchased and cancelled. The bonds, listed on the SIX, are interest bearing (5%) with a maximum term of 5 years and are convertible into registered Shares of Santhera with a nominal value of CHF 1 each. The initial conversion price is fixed at CHF 86.4006 and will be reset after the first year if the volume weighted average price (**VWAP**) of the Shares during a specified period of time will be below the reference share price (CHF 71.9969). The new conversion price must not be lower than 75% of the conversion price at issuance. In addition, Santhera may call the convertible bonds at any time on or after the second anniversary of the issue date at par, plus accrued interest, if any, if the VWAP of the Shares is at least 160% of the conversion price.

Treasury shares

Treasury shares are recognized at acquisition cost and deducted from shareholders' equity at the time of acquisition. Santhera holds treasury shares for market making which is maintained by an external bank. In case of a resale, the gain or loss is recognized through the income statement as financial income or financial expenses.

Related parties

In the meaning of the Swiss Accounting Law, related parties are only considered to be shareholders, direct and indirect subsidiaries (shareholdings) and the Board of Directors.

3 Information on Balance Sheet and Income Statement Items

3.1 Loans to shareholdings

Loans are granted to shareholdings primarily to fund the development and marketing activities of the Santhera Group (December 31, 2017: CHF 235.6 million; December 31, 2016: CHF 190.1 million). Until the end of 2015 the balance consisted of fully impaired and subordinated loans to Santhera Pharmaceuticals (Schweiz) AG. To finance the activities in development and the commercialization of LHON, in 2016 the loan granted to Santhera Pharmaceuticals (Schweiz) AG was increased (with the additional loans also being subordinated). As part of the annual reassessment as of December 31, 2017, Executive Management concluded that approximately 30% of the total loan balance is recoverable considering a more positive outlook, both in terms of market success of the developed and launched product (Raxone in LHON) and the development progress in other indications (e.g. idebenone in DMD).

3.2 Investments in shareholdings

In 2017 and 2016, the following companies are direct subsidiaries of Santhera Pharmaceuticals Holding AG (100% ownership and 100% voting rights):

	Share capital at December 31	2017	2016
Santhera Pharmaceuticals (Schweiz) AG Pratteln, Switzerland	CHF	125,000	125,000
Santhera Pharmaceuticals (Deutschland) GmbH Lörrach, Germany	EUR	25,000	25,000
Santhera Pharmaceuticals (USA), Inc. Burlington, US	USD	1,000	1,000
Santhera Pharmaceuticals (Canada), Inc. Montréal, Canada	CAD	1,000	1,000
Oy Santhera Pharmaceuticals (Finland) Ltd Helsinki, Finland	EUR	2,500	2,500

Santhera Pharmaceuticals (Schweiz) AG is the primary operational entity while Santhera Pharmaceuticals (Deutschland) GmbH holds the market authorization for the EU. Oy Santhera Pharmaceuticals (Finland) Ltd is not employing any personnel.

The following companies are 100% direct subsidiaries (100% voting rights) of Santhera Pharmaceuticals (Schweiz) AG:

	Share capital at December 31	2017	2016
Santhera Pharmaceuticals (Liechtenstein) AG Ruggell, Fürstentum Liechtenstein	CHF	50,000	50,000
Santhera (Italy) S.r.l. Milano, Italy	EUR	50,000	50,000
Santhera (Germany) GmbH München, Germany	EUR	50,000	50,000
Santhera (Netherlands) B.V. Nieuwegein, The Netherlands	EUR	50,000	50,000
Santhera (UK) Limited London, United Kingdom	GBP	50,000	50,000

3.3 Share capital

During 2017, the share capital was increased by a total amount of CHF 8,698 to CHF 6,288,555 as of December 31, 2017 (2016: CHF 6,279,857) through the exercise of employee stock options (from conditional share capital).

3.4 Treasury shares

The movement of treasury shares held by Santhera was as follows:

	No of Shares	TCHF
January 1, 2016	8,028	177
Purchase	23,002	1,069
Sale[1]	−27,414	−1,074
December 31, 2016	3,616	172
Purchase	180,083	9,567
Sale	173,778	−9,404
December 31, 2017	9,921	335

[1] In connection with the liquidation of Oy Juvantia Pharma, Turku, Finland (**Juvantia**), acquired in 2009, Santhera received 8,028 shares from former Juvantia shareholders. These treasury shares served as pledge from the former owners of Juvantia for compensation of a potential tax claim related to pre-acquisition activities. The claim was resolved and the shares were sold with a financial profit of TCHF 186.

3.5 Income from shareholdings

Income from shareholdings represents reimbursement for management services provided by the Company to its major shareholding Santhera Pharmaceuticals (Schweiz) AG.

3.6 General and administrative expenses

In CHF thousands	2017	2016
Administrative expenses	1,220	990
Consulting expenses	1,456	1,169
Expenses in connection with convertible bonds	2,731	0
Total	**5,407**	**2,159**

4 Other Information

4.1 Full-time equivalents

The number of full-time equivalents at period end was not above 10 in 2017 and 2016.

4.2 Significant shareholders (>2%)

Pursuant to information from the Company's share register and the disclosure of participations made to the Company in accordance with applicable stock exchange regulation, the following shareholders owned 2% or more of the Company's share capital as registered in the commercial register at December 31, 2017: 6,279,857 shares (December 31, 2016: 6,262,798 shares):

	2017 Shares[1]	2017 %	2016 Shares[2]	2016 %
Iglu Group, Switzerland[3]	557,350	8.9	632,300	10.1
Bertarelli Ernesto, Guichard-Bertarelli Donata and Bertarelli Maria-Iris, Switzerland[3]	545,777	8.7	545,777	8.7
The Goldman Sachs Group, Inc., Corporation Trust Centre[4]	457,309	7.3	n/a	n/a
Roderick Wong (RTW Master Fund, LTD, US)	315,339	5.0	146,365	2.3
Norges Bank (the Central Bank of Norway)[5]	214,258	3.4	n/a	n/a
Consonance Capital Management, US	n/a	<3	597,069	9.5
UBS Fund Management (Switzerland) AG	n/a	<3	195,007	3.1
Lagoda Investments Management, LLC, US	n/a	<3	187,888	3.0
UBS Fund Management (Luxembourg) S.A.	n/a	<3	183,699	2.9
Union Asset Management Holding AG	n/a	<3	175,838	2.8
Visium Balanced Master Fund, Ltd., US[6]	n/a	n/a	n/a	n/a

[1] Including disclosures until December 31, 2017

[2] Including disclosures until December 31, 2016

[3] On January 25, 2017, both Iglu Group and the Bertarelli Group announced that they had formed a new group with combined holdings of 1,179,977 Shares (18.8%).

[4] Purchase positions in connection with securities lending transactions (as of January 3, 2018)

[5] Purchase positions in connection with securities lending transactions, shares held as collateral (as of January 17, 2018)

[6] The fund was liquidated in June 2016 (Bloomberg, June 18, 2016).

4.3 Disclosure of shares and equity rights (share appreciation rights and stock options) held by members of the Board and Executive Management (and their respective related party)

As of December 31, 2017:

	Number of Shares	Number of vested equity rights	Number of unvested equity rights	Total number of equity rights
Board of Directors				
Elmar Schnee, Chairman since April 4, 2017	2,000	0	4,486	4,486
Martin Gertsch, Vice-Chairman	38,109	1,500	8,935	10,435
Jürg Ambühl, Director until April 4, 2017 [1]	30,000	7,281	0	7,281
Philipp Gutzwiller, Director since April 2017	500	0	3,157	3,157
Thomas Meier, Director since April 4, 2017		--- See below ---		
Patrick Vink, Director since April 4, 2017	1,000	0	6,116	6,116
Executive Management				
Thomas Meier, CEO	75,562	8,000	29,663	37,663
Todd Bazemore, Chief Operating Officer US until November 17, 2017 [2]	0	0	0	0
Nicholas Coppard, Head Development until January 31, 2017 [3]	0	3,500	19,216	22,716
Günther Metz, Head Business Development	0	14,000	16,500	30,500
Christoph Rentsch, Chief Financial Officer	0	7,500	28,601	36,101
Kristina Sjöblom Nygren, Chief Medical Officer & Head Development since January 1, 2017	0	0	18,617	18,617
Giovanni Stropoli, Chief Commercial Officer Europe and Rest of World	250	7,500	26,499	33,999
Oliver Strub, General Counsel and Secretary to the Board	0	6,001	16,990	22,991

[1] Number of Shares as of April 4, 2017

[2] Number of Shares as of November 17, 2017

[3] Number of Shares as of January 31, 2017

As of December 31, 2016:

	Number of Shares	Number of vested equity rights	Number of unvested equity rights	Total number of equity rights
Board of Directors				
Martin Gertsch, Chairman	38,109	0	6,281	6,281
Jürg Ambühl, Director	30,000	0	7,281	7,281
Executive Management				
Thomas Meier, CEO	72,902	3,750	14,875	18,625
Todd Bazemore, Chief Operating Officer US[1]	0	0	34,881	34,881
Nicholas Coppard, Head Development	0	0	12,250	12,250
Günther Metz, Head Business Development	0	12,000	7,120	19,120
Christoph Rentsch, Chief Financial Officer	0	0	22,000	22,000
Giovanni Stropoli, Chief Commercial Officer Europe and Rest of World	600	0	20,565	20,565
Oliver Strub, General Counsel and Secretary to the Board	0	9,001	7,240	16,241

[1] Joined the Executive Management on September 6, 2016.

4.4. Disclosure of the allocation of equity rights for Board of Directors, Executive Management and employees of Santhera Group

	2017 Quantity	2017 Value (in TCHF)[1]	2016 Quantity	2016 Value (in TCHF)[1]
Board of Directors	17,913	524	6,562	224
Executive Management	104,033	3,686	65,431	1,349
Employees of Santhera Group	228,297	4,876	126,980	3,121
Total	**350,243**	**9,086**	**198,973**	**4,694**

[1] Value of the equity rights calculated in accordance with the Hull-White model at the date of allocation in accordance with the terms of the award. The tax value of equity rights is 0 until they would be exercised. Such equity rights values are theoretical values and do not reflect income tax values and do also take into consideration certain vesting provisions. For information about the underlying equity rights plans, see note 17 *"Equity Rights Plans"* in the consolidated financial statements. For information about the Company's compensation procedures, consult the Corporate Governance Report and the Compensation Report.

On January 1, 2018, 444,164 share appreciation rights (**SAR**s) were planned to be granted to employees of Santhera (conditionally for the Executive Management). These SARs are part of the bonus award for the year 2017 to employees of the Group. These SARs were granted under ESARP 2016 (see note 17 *"Equity Rights Plans"*).

	Quantity	Value (in TCHF)[1]
Executive Management	103,810	1,452
Employees of Santhera Group	340,354	4,761
Total	**444,164**	**6,213**

[1] Value of the equity rights calculated in accordance with the Hull-White model at the date of allocation in accordance with the terms of the award. The tax value of equity rights is 0 until they would be exercised. Such equity rights values are theoretical values and do not reflect income tax values and do also take into consideration certain vesting provisions. For information about the underlying equity rights plans, see note 17 *"Equity Rights Plans"* in the consolidated financial statements. For information about the Company's compensation procedures, consult the Corporate Governance Report and the Compensation Report.

4.5 Contingencies and guarantees

Guarantee towards Swiss VAT authorities

The Company is part of the value-added tax group of the Swiss affiliated companies of Santhera Pharmaceuticals and is therefore jointly and severally liable to the Swiss federal tax administration for their value-added tax liabilities.

Guarantee towards Santhera Pharmaceuticals (Schweiz) AG

The Company guarantees to pay for the liabilities of its subsidiary Santhera Pharmaceuticals (Schweiz) AG until the Annual General Meeting in 2018.

Declaration of liability towards Arval Deutschland GmbH

The Company guarantees to pay for the liabilities of its subsidiary Santhera (Germany) GmbH for contractual duties and obligations.

4.6 Events after the reporting date

On January 26, 2017, Santhera announced that the Committee for Medicinal Products for Human Use (**CHMP**) of the European Medicines Agency (**EMA**) maintained its negative opinion on the Type II extension application for idebenone in Duchenne muscular dystrophy (**DMD**) following a re-examination procedure. The CHMP concluded that an approval for idebenone in DMD, applied as a Type II variation of the existing marketing authorization, cannot be granted at the present time based on the current existing evidence. Although the positive outcome of the Phase III DELOS trial was acknowledged, the CHMP has invited Santhera to present additional data to further link the observed treatment effects on respiratory function outcomes to patient benefit. Management has considered the impact on the Group's ability to continue as a going concern and potential indicators of impairment on the financial statements as of December 31, 2017.

On February 15, 2018, Santhera announced that it has entered into a license agreement with Polyphor Ltd., Allschwil, Switzerland, for POL6014, a clinical stage selective inhibitor of human neutrophil elastase with the potential to treat cystic fibrosis (**CF**) and other neutrophilic pulmonary diseases such as non-cystic fibrosis bronchiectasis (**NCFB**), alpha-1 antitrypsin deficiency (**AATD**) and primary ciliary dyskinesia (**PCD**).

Santhera will assume the global development, regulatory filings and commercialization of POL6014. The development program has been advanced with financial support by the Cystic Fibrosis Foundation Therapeutics Inc. (**CFFT**), USA, to Polyphor. With POL6014, Santhera is expanding its product pipeline in pulmonary diseases where the Company is already developing its lead product idebenone for respiratory complication in Duchenne muscular dystrophy.

Under the agreement, Santhera obtains the worldwide, exclusive rights to develop and commercialize POL6014, an innovative macrocycle elastase inhibitor, and analogs for an initial payment of CHF 6.5 million, payable in Santhera shares at an agreed valuation of CHF 27.2053 per share and additional cash payments of up to CHF 121 million contingent to future development, regulatory and particularly sales milestones. In addition, Polyphor is entitled to tiered royalty payments from Santhera´s future net sales of POL6014 and to undisclosed milestone payments and royalties provided that Santhera advances the development and market entry of POL6014 in other pulmonary diseases. Santhera issued the 238,924 shares (3.8% of its currently issued shares) required for the initial payment to Polyphor out of its existing authorized share capital.

Proposal of the Board of Directors to the Annual General Meeting

Proposal of the Board for the result to be carried forward, subject to the approval of the Annual General Meeting

In CHF	2017	2016
Result carried forward	−6,451,188	−5,556,524
Net result of the year	−7,300,395	−894,664
Accumulated result	**−13,751,583**	**−6,451,188**
Result to be carried forward	**−13,751,583**	**−6,451,188**

The Board of Directors requests the approval of the Annual General Meeting for the following release and transfer from reserves from capital contribution:

	In CHF
Reserves from capital contribution at December 31, 2016	7,425,157
Share premium from option exercise during 2017	24,790
Reserves from capital contribution	**7,449,947**
Transfer from reserves from capital contribution to other voluntary reserves (free reserves)	−7,000,000
Reserves from capital contribution	**449,947**

Subject to approval by the Annual General Meeting, the other voluntary reserves (free reserves) develop as follows:

	In CHF
Other voluntary reserves (free reserves) at December 31, 2016	50,494,714
Transfer from reserves from capital contribution	7,000,000
Free reserves	**57,494,714**

Building a better working world

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To the General Meeting of
Santhera Pharmaceuticals Holding Ltd, Liestal

Basle, March 19, 2018

Report of the statutory auditor on the financial statements

As statutory auditor, we have audited the financial statements of Santhera Pharmaceuticals Holding Ltd, which comprise the balance sheet, income statement and notes (pages 63 to 73), for the year ended December 31, 2017.

Board of Directors' responsibility
The Board of Directors is responsible for the preparation of the financial statements in accordance with the requirements of Swiss law and the company's articles of incorporation. This responsibility includes designing, implementing and maintaining an internal control system relevant to the preparation of financial statements that are free from material misstatement, whether due to fraud or error. The Board of Directors is further responsible for selecting and applying appropriate accounting policies and making accounting estimates that are reasonable in the circumstances.

Auditor's responsibility
Our responsibility is to express an opinion on these financial statements based on our audit. We conducted our audit in accordance with Swiss law and Swiss Auditing Standards. Those standards require that we plan and perform the audit to obtain reasonable assurance 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 judgment, 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 the internal control system relevant to the entity's preparation of the financial statements 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 system. An audit also includes evaluating the appropriateness of the accounting policies used and the reasonableness of accounting estimates made, 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 audit opinion.

Opinion
In our opinion, the financial statements for the year ended December 31, 2017 comply with Swiss law and the company's articles of incorporation.

Report on key audit matters based on the circular 1/2015 of the Federal Audit Oversight Authority

Key audit matters are those matters that, in our professional judgment, were of most significance in our audit of the financial statements of the current period. These matters were addressed in the context of our audit of the financial statements as a whole, and in forming our opinion thereon, and we do not provide a separate opinion on these matters. For each matter below, our description of how our audit addressed the matter is provided in that context.

We have fulfilled the responsibilities described in the *Auditor's responsibilities* section of our report, including in relation to these matters. Accordingly, our audit included the performance of procedures designed to respond to our assessment of the risks of material misstatement of the financial statements. The results of our audit procedures, including the procedures performed to address the matters below, provide the basis for our audit opinion on the financial statements.

Management's assessment of the Group's ability to continue as a going concern

Area of focus

Based on the stage of the operations combined with recurring development costs and the level of cash inflows from its operating activities, the Group incurred negative cash flows from operations in the past and it expects to continue to incur such costs and negative cash flows in the near future. Management has prepared various budget scenarios and business plans and has assessed the likelihood and impact that uncertain cash flows could have on the Group's ability to continue as going concern. This assessment was based on the evaluation of the available year-end funds of CHF 45.2 million as well as the budget.

Despite the Group's development into a more sales oriented business, funding from non-revenue related sources is still required in the future and we therefore concluded that the assessment of the ability to continue as a going concern represents a key audit matter for our audit.

Our audit response

Our audit procedures related to the key audit matter of the assessment of the going concern included the following procedures:

- We gained an understanding of management's budgeting and forecasting process underlying the going concern assessment.
- We discussed the budget and business plans with management, evaluated the development and the assessments made and also considered how uncertain elements were included in the budget and business plans. We evaluated underlying key assumptions such as expected cash inflow from product sales and expected cash outflow from purchases of inventory, research and development expenses and other operating expenses.
- We tested the sensitivity of the revenue estimates and assessed how they compared to historical revenues.
- We further discussed the base case scenario with management and assessed the likelihood of this scenario.

- We performed inquiries of management about current developments and the ability to execute the elements included in the budget and business plans supporting the evaluation of the going concern assumption. We read the Board of Directors meeting minutes that approved the underlying budget to assess potential contrary information. We performed procedures regarding subsequent events to assess the accuracy of the budget as of the date of the auditor's report.

Valuation of investments in and long-term receivables from shareholdings

Area of focus	Santhera Pharmaceuticals Holding Ltd holds investments in subsidiaries and grants loans to subsidiaries for financing purposes, both of which are assessed for impairment as of the balance sheet date. Management's assessment requires estimation and judgement around assumptions used, including prospective financial information and discount rates. Changes to assumptions could lead to significant changes in the estimated recoverable amount, impacting both potential impairment charges as well as potential reversals of impairment. As such, we considered this matter to be significant to our audit. *Refer to note 3.1 and 3.2 to these financial statements for Santhera Pharmaceuticals Holding Ltd disclosures related to investment in and long-term receivables from shareholdings.*
Our audit response	We evaluated management's impairment assessment, which is based on an income approach, and analysed the underlying key assumptions in relation to prospective financial information as well as discount rates used. We evaluated the historical accuracy of the Company's previous estimates on prospective financial information. We tested the sensitivity of the assessment due to changes to key assumptions and compared these assumptions to externally available information in order to assess management's impairment conclusion.

Report on other legal requirements

We confirm that we meet the legal requirements on licensing according to the Auditor Oversight Act (AOA) and independence (article 728 CO and article 11 AOA) and that there are no circumstances incompatible with our independence.

In accordance with article 728a para. 1 item 3 CO and Swiss Auditing Standard 890, we confirm that an internal control system exists, which has been designed for the preparation of financial statements according to the instructions of the Board of Directors.

We recommend that the financial statements submitted to you be approved.

Ernst & Young Ltd

/s/ Frederik Schmachtenberg
Licensed audit expert
(Auditor in charge)

/s/ Jan Meyer
Licensed audit expert

Compensation Report

Contents

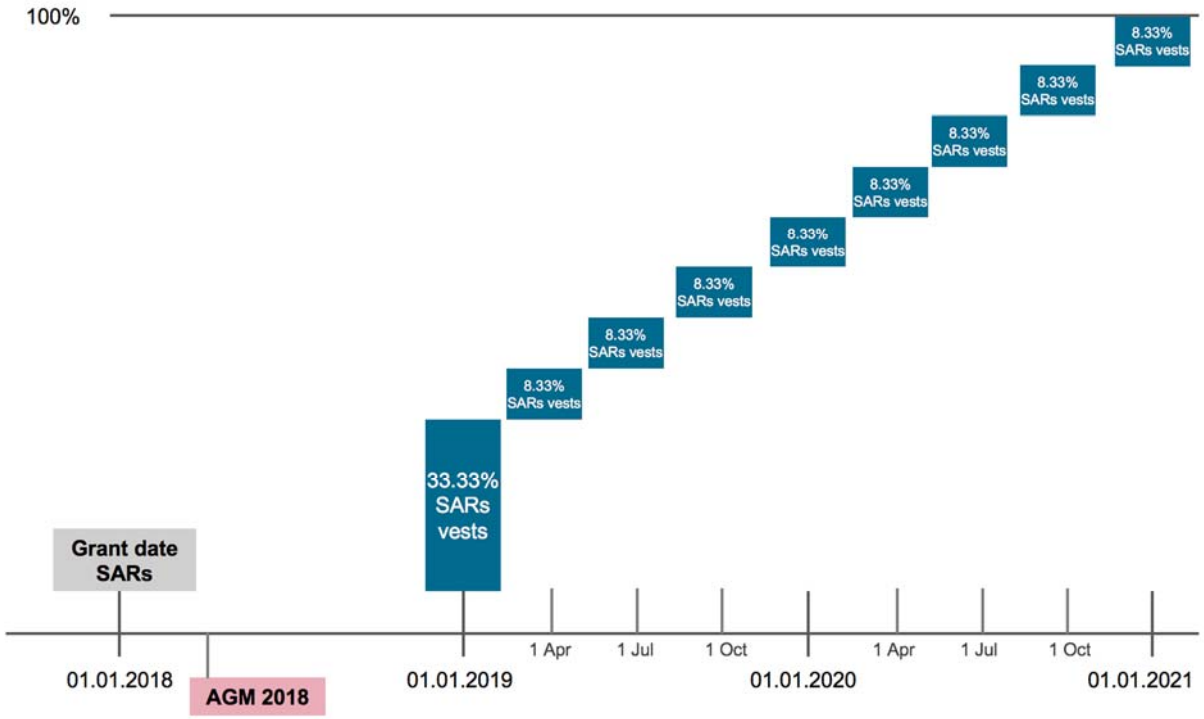
Introduction

This Compensation Report (**Report**) describes the principles of the compensation system of Santhera's Board of Directors (**Board**) and Executive Management (**EM**) members (**Executives**) and how the respective decisions are made. Furthermore, the Report discloses the compensation made to the Board and EM for 2017, as well as shareholdings of the members of the Board and EM members.

Santhera's compensation policy is designed to attract, motivate and retain talent in order to support the achievement of the Company's financial and strategic objectives and also to ensure that the total compensation package is fair and competitive. By combining short- and long-term incentive elements, the Board believes that the compensation system is designed in a way that the interests of the management are aligned with the interests of the Company and its shareholders. The Company's compensation system does not set any unintended enticements or contain any components that could be counterproductive to the objectives of the compensation system. In addition, compensation elements are focused on rewarding the delivery of outstanding and sustainable results without inappropriate risk-taking.

On an ongoing basis, the Compensation Committee (**CC**) reviews and monitors Santhera's compensation policy in light of the Company's business strategy, corporate goals and values, in order to ensure the alignment of employee interests with those of the shareholders. In 2017, changes to the compensation system with respect to the vesting of Share Appreciation Rights (**SAR**) were made:

- The overall vesting period of the plan was reduced from four years to three years.
- In accordance with the new Employee Share Appreciation Rights Plan 2018 (**ESARP 2018**), 33 1/3% of the SAR vest after a period of one year from the grant date, thereafter, each calendar quarter, 8 1/3% of the total SAR vest for seven calendar quarters. The remaining SAR vest on the third anniversary of the grant.



One of the major corporate goals to be achieved in 2017 was to obtain marketing authorization in the EU for idebenone for the treatment of patients with Duchenne muscular dystrophy (**DMD**). As the Company did not reach this goal in 2017 (and other goals only partially), the corporate target achievement was determined by the Board to amount to 60% for the calculation of both the short-term (cash bonus) and the long-term incentive (SAR allocation).

In view of the major efforts being undertaken by the Company, the importance to obtain regulatory approval for DMD in the EU and to retain our employees, the Board asks the shareholders to approve a special allocation of SAR that would be calculated on an additional corporate achievement rate of 30%. Such SAR would only vest if and when a positive CHMP opinion would be obtained and the number of SAR decreases over time.

This special grant will also be made to all other employees of the Company. The terms and conditions of this special grant would be essentially the same as in the ESARP 2017.

The Board believes that the compensation provisions for the Board and EM members serve the best interests of Santhera's shareholders.

The Role and Powers of the Compensation Committee

The Compensation Committee (**CC**) currently consists of the two members of the Board, Patrick Vink as Chairman and Elmar Schnee as Member. The CC annually reviews the compensation system of the members of the Board and EM and ensures that the Company's regulations and Articles of Incorporation remain in compliance with requirements of the Ordinance against Excessive Compensation (**OaEC**), the SIX Swiss Exchange, as well as Swiss and international best corporate governance practices.

According to the Company's Articles, the role of the CC assists the Board with the:

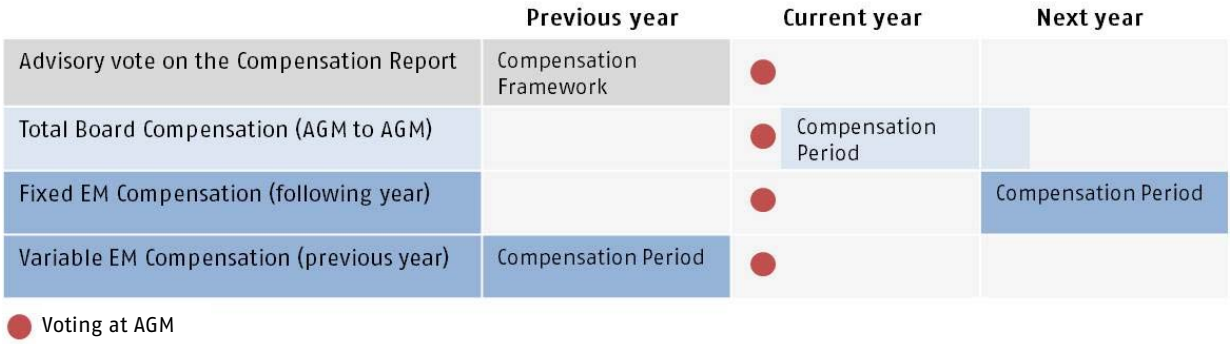
- Determination and review of remuneration policies and guidelines.
- Determination and review of performance objectives.
- Proposals to the AGM concerning the compensation of the Board of Directors and of EM.
- Resolution of other compensation related matters.

The Board may assign other tasks to the CC.

Compensation Periods and Approvals by Shareholders

For the Board, the compensation period starts after the Annual General Meetings (**AGM**) and ends on the day before the AGM of the subsequent year.

For Executive Management, the compensation period starts on January 1 of a given year and ends on December 31 of such year. With respect to the fixed compensation, the approval of the shareholders is prospective and with respect to the variable compensation, such approval is retrospective, allowing the Board to base its respective motions to the shareholders on the achievement of corporate and individual goals by the Executives.



Voting procedures at the 2018 AGM

The Board will propose the following votes on compensation for shareholder approval:

1. Consultative vote on the Compensation Report 2017.
2. Board
   2.1. The maximum total amount of the fixed compensation for the period between the 2018 AGM and the 2019 AGM.
3. Executive Management
   3.1. The maximum total amount of the fixed compensation for the period from January 1, 2019 to December 31, 2019.
   3.2. The maximum total amount of the variable compensation for the period from January 1, 2017 to December 31, 2017.
   3.3. The maximum total amount for a special allocation of SAR to vest only if and when the marketing approval for Raxone in DMD will have been obtained.

The invitation to the AGM contains the text of agenda items, motions and the explanations thereto in detail.

Board of Directors Compensation

The compensation for members of the Board consists of:

- Annual cash fees (50% of the total compensation)
- Annual grant of Share Appreciation Rights (**SAR;** 50% of the total compensation)

Both components, cash fees and SAR allocation, do not depend on the achievement of corporate goals or the individual performance of a Board member. Additionally, the Company assumes the payment of employer's social security contributions due on these amounts. Board members do not receive any variable compensation.

Annual cash fees

At the 2017 AGM, the shareholders approved a total cash compensation for the entire Board of a maximum of CHF 500,500. For the period between the 2017 and the 2018 AGM, including social security contributions, the compensation of the Chairman of the Board is expected to amount to CHF 143,000, the compensation of the Vice Chairman of the Board to CHF 121,000 and the compensation of the other members of the Board to CHF 99,000 each. The CEO of the Company, Thomas Meier, will not receive any additional compensation for his Board membership. The Chairman of the Audit Committee is expected to receive an additional amount of CHF 16,500; and the Chairman of the Compensation Committee an additional CHF 11,000. The members of the Audit Committee and of the Compensation Committee are expected to receive an additional amount of CHF 5,500 each.

Share Appreciation Rights (SAR)

At the 2017 AGM, the shareholders approved a total maximum amount of CHF 500,500 to be granted in SAR for the period until the 2018 AGM. In accordance with the Board Share Appreciation Rights Plan (**BSARP 2017**), 15,120 SARs were granted to the Board members (excluding Thomas Meier, as he does not receive a separate compensation for his Board mandate) as of April 4, 2017. The exercise price was the closing price of Santhera's share on April 4, 2017 and amounted to CHF 77.00 (2016: CHF 82.00). According to BSARP 2017, 33 1/3% of the SAR vest after a period of one year from the grant date, thereafter, each calendar quarter, 8 1/3% of the total SAR vest for seven calendar quarters. The remaining SAR vest on the third anniversary of the grant. During such vesting periods, SAR may lapse subject to certain conditions as defined by the BSARP. The term of the SAR grant is 10 years. Compared to the previous option programs, the first vesting period was shortened from two years to one year (with respect to SAR) to make the plan more attractive for the employees. For more information about the underlying Plan, see note 17 *"Equity Rights Plans"* in the consolidated financial statements.

Maximum Total Compensation

Function	Maximum compensation (CHF)	Number	Total maximum compensation (CHF)
Chairman of the Board (**COB**)	286,000	1	286,000
Vice Chairman of the Board (**VC**)	242,000	1	242,000
Member of the Board	198,000	2*	396,000
Chairman of the AC	33,000	1	33,000
Member of the AC	11,000	1	11,000
Chairman of the CC	22,000	1	22,000
Member of the CC	11,000	1	11,000
Total			**1,001,000**

* Five Board members excluding Chairman, Vice Chairman and Santhera's CEO who will not receive a separate compensation as member of the Board.

Compared to the compensation of the former Board, the current compensation was increased to reflect the increase in complexity and responsibility for the Board members. In addition, an increase from two to five Board members resulted in a considerable increase in compensation by more than 100%.

Disclosure of compensation of members of the Board for the financial years 2017 and 2016 (audited)

In CHF	Annual cash fees	Stock options[1] SAR	Social security[1, 2]	Total compensation	Number of stock options/SAR granted
2017					
Elmar Schnee[3]	111,375	137,501	10,313	259,189	4,486
Martin Gertsch[4]	144,236	127,325	20,994	292,555	4,154
Philipp Gutzwiller[3]	72,569	96,766	13,015	182,350	3,157
Thomas Meier[5]	0	0	0	0	0
Patrick Vink[3] – As Board member	76,389	101,854	13,700	191,943	3,323
Jürg Ambühl	44,600	0	1,900	46,500	0
Total	449,169	463,446	59,922	972,537	15,120
2016					
Martin Gertsch	99,500	112,013	16,023	227,536	3,281
Jürg Ambühl	89,523	112,013	13,111	214,647	3,281
Total	**189,023**	**224,026**	**29,134**	**442,183**	**6,562**

[1] Reflects value of share-based payments in accordance with IFRS 2 at grant, i.e. the value of unvested stock options attributable at grant. The tax value of such stock options is CHF 0 until stock options are exercised. Such stock option values are theoretical values and do not reflect income tax values and do also take into consideration certain vesting provisions.

[2] To be in line with the market practice, the Board has decided to disclose the social security from 2015 onwards not on exercised but on the fair value of allocated options. For all SARs held by Board members as of December 31, 2017, the social security contribution is CHF 0 since the SARS are not in-the-money. The total value of social security payments on options exercised by members of the Board during 2017 is CHF 0 (2016: CHF 0).

[3] Elmar Schnee, Philipp Gutzwiller, Thomas Meier and Patrick Vink were elected as Chairman and members of the Board on April 4, 2017. The Board compensation period for these Board members therefore only covers the last 9 months of 2017. In the first three months of 2017, Patrick Vink had acted as an advisor to the Board. In such capacity, he received an additional amount of CHF 84,510

[4] Martin Gertsch has been a Board member during the entire 2017. His compensation is therefore disclosed for 12 months.

[5] Thomas Meier did not receive any compensation as a Board member.

Comparison of the approved and paid Board compensation during the approval period from one AGM to the next

At the 2017 AGM, the shareholders approved a maximum total amount of fixed compensation for the Board of CHF 500,500 for the period from the AGM 2017 to the AGM 2018. In addition, the shareholders approved the allocation of SAR the fair value of which would be a maximum of CHF 500,500.

The table below represents the approved maximum compensation for the Board, the actual amounts paid in 2017, those still payable until AGM 2018.

	Approved AGM 2017 – AGM 2018	Paid/payable AGM 2017 – AGM 2018
Board fees (CHF)	500,500	474,425
SAR[1] (CHF)	500,500	463,446
Total (CHF)	**1,001,000**	937,871
SAR (number)	n/a	15,120

[1] The shareholders approved a fix amount in CHF which was converted into a number of SAR based on the fair market value of such SAR on the first trading day immediately following the 2017 AGM (CHF 30.6512).

Outlook for Board compensation

At the 2018 AGM, the existing members are proposed to be re-elected as members of the Board. Among those proposed is Thomas Meier, CEO of Santhera, who will not receive a separate compensation as a Board member.

In 2017 the Board has put in place an Audit Committee (**AC**) in addition to a Compensation Committee (**CC**). This will be continued in 2018. For rules and responsibilities of these two committees, see section on DCG 3.5.3 in the Corporate Governance report on page 101.

Both committee chairmanships as well as memberships of the Board and its committees are proposed to be remunerated as per the table on page 82 of this Compensation Report. The proposal does not foresee an increase of the Board compensation.

The total maximum compensation would be made 50% in the form of cash fees (including social security contributions) and 50% in the form of SAR.

To calculate the number of SAR to be allocated, the total SAR amount of approximately CHF 463,000 (CHF 500,500 minus approximately CHF 37,500 for social security deductions) would be divided by the fair value of the SAR on the date of their grant. The fair value is calculated based on the share price on the trading day at the grant day, then applying the Hull-White model (excluding employer's social security contribution).

Executive Management Compensation

The compensation for members of Executive Management currently consists of:

- Fixed compensation.
- Variable compensation:
    - Annual bonus paid in cash.
    - Annual grant of SAR.

Fixed compensation

The fixed compensation for the EM members includes base salary, social security contributions and payments to the pension fund by the Company. The base salary takes into account the position,

responsibilities, experience and skills of an individual EM member. Base salaries are reviewed annually by the CC, taking into account individual performance and the results of the external benchmarking.

The compensation of the EM members is reviewed periodically in order to ensure market competitiveness. For this purpose, the CC mandated Kienbaum to provide an in-depth benchmark analysis of the compensation of the EM members in 2017. The basis for comparison consists of almost 30 Swiss and international Biotech and Pharma companies from which most of our talents are likely to join from.

The benchmark analysis served as a basis for the CC to review the compensation of the EC members and to confirm or revise their target compensation levels for financial years 2017 and 2018. For compensation decisions that had been made previously, the CC relied on the benchmark report provided by Willis Towers Watson.

Annual cash bonus

The annual cash bonus for 2017 is based on the achievement of Company and individual goals and will be paid in April 2018, subject to the shareholders' approval. The Company goals included receipt of a positive opinion by the CHMP with respect to the marketing authorization application of the Company for the treatment of DMD, the achievement of sales targets, the successful completion of a financing, certain business development activities and advancing the Company's SIDEROS study. As the Company did not obtain a positive opinion in 2017 (and achieved other goals only partially), the corporate target achievement was determined by the Board to amount to 60%. The target bonus, i.e. cash bonus to be paid if 100% of corporate and individual objectives are met, is determined individually for each EM member as percentage of the base salary, ranging from 25% to 50%.

The weightings of the corporate and individual goals are individual for each EM member and vary depending on the position. In general, the higher the position of an employee, the more weight is put on the achievement of corporate goals rather than on individual goals. For the Chief Executive Officer (**CEO**), the weighting of the achievement of corporate goals has been 90% and for the other Executives 70%. The final payout is capped at 100% of the target bonus.

Calculation of the individual annual bonus for EM members

The proposal to the shareholders at the 2018 AGM is for a maximum cash bonus payment of CHF 575,000 (of which a maximum of CHF 125,000 for social security contributions).

Share Appreciation rights (SAR)

Under the Employee Share Appreciation Rights Plan (**ESARP**), members of the EM receive an annual grant of a certain number of SAR which is determined by the Board, taking into account the achievement of Company and individual goals.

For the allocation of these SAR for the 2017 goal achievements (achievement of corporate goals of 60% and individual goal achievement of between 90% and 100%), the exercise base value is equal to the closing price of Santhera's share on the first trading day in 2018 (January 3, 2018) and amounts to CHF 36.70 (previous year: CHF 54.85).

The proposal to the shareholders at the 2018 AGM is for a maximum SAR allocation for CHF 1,575,000 which would result in an allocation of a maximum of 104,711 SAR to the EM.

According to the ESARP, 33 1/3% of the SAR vest after a period of one year from the grant date, thereafter, each calendar quarter, 8 1/3% of the total SAR vest for seven calendar quarters. The remaining SAR vest on the third anniversary of the grant. During such vesting periods, SAR may lapse subject to certain conditions as defined by the ESARP.

The term of the SAR grant is 10 years. For more information about the underlying Plan, see note 17 *"Equity Rights Plans"* in the consolidated financial statements.

Disclosure of compensation of members of the Executive Management for the years 2017 and 2016 (audited)

In CHF	Base salary	Cash bonus[1]	SAR[1, 2, 5]	Social security and pension[2,3]	Total compen-sation	Number of stock options/ SAR granted
2017						
Kristina Nygren	330,000	58,163	232,644	107,017	727,824	16,633
Kristina Nygren Retention award	45,600[8]	0[8]	399,992[9]	34,407	479,999	18,617
Kristina Nygren Total	**375,600**	**58,163**	**632,636**	**141,424**	**1,207,823**	**35,250**
Other 7 members of EM	2,071,520	389,969	1,231,938	685,411	4,378,838	88,078
Total	2,447,120	448,132	1,864,574	826,835	5,586,661	123,328
2016[6]						
Todd Bazemore	118,940	44,275	116,915[7]	20,008	300,138	5,247
Todd Bazemore Retention award	0	0	726,222[4, 7]	10,530	735,752	34,881[4]
Todd Bazemore Total	**118,940**	**44,275**	**843,137**	**30,538**	**1,036,890**	**40,128**
Other 5 members of EM	1,888,644	343,300	1,786,350	630,012	4,648,306	80,169
Total	**2,007,584**	**387,575**	**2,629,487**	**660,550**	**5,685,196**	**120,297**

[1] Proposal for approval by the 2018 AGM.

[2] Reflects value of share-based payments in accordance with IFRS 2 at grant, i.e. the value of unvested stock options/SAR attributable at grant; tax value of such stock options/SAR is CHF 0 until stock options/SAR are exercised. Such stock option/SAR values are theoretical values and do not reflect income tax values and do also take into consideration certain vesting provisions.

[3] Included in the amounts are social security payments on the fair value of allocated options/SAR. For all options/SAR granted to EM members for 2017, the social security contributions have been calculated to amount to CHF 139,842 (2016: CHF 132,099). The total amount of social security payments on options exercised by members of EM during 2017 is CHF 15,798 (2016: CHF 20,541). The award of SAR requires retrospective approval by the 2018 AGM. Following such approval, a revised fair value will be determined for accounting purposes only. The fair value will be re-valuated for accounting purposes only following the shareholders' approval; the change in value will be disclosed in the consolidated financial statements.

[4] Todd Bazemore had joined the Company on September 6, 2016. In addition to the proposed amount of CHF 116,915 for the grant of SAR, in 2016, he received SAR with a fair market value of CHF 726,222 as a retention award. This amount had been calculated on the contractually agreed amount of USD 750,000 based on the FX rate of USD 1.0000 = CHF 0.9683. The retention award consisted of a grant that was subject to reimbursement by Todd Bazemore to the Company if before September 6, 2017, Todd Bazemore would resign his employment without good reason or if he is terminated by the Company for cause. As a consequence of Todd's resignation effective November 17, 2017, all of his 40,128 SAR (5,247 annual SAR plus 34,881 retention award SAR) were cancelled.

[5] In the year under review, only SAR have been granted; in 2016 SAR and stock options have been granted.

[6] In 2017, the AGM approved a variable compensation amount of a maximum of CHF 1,905,000 for the allocation of SAR to Executives for their merits in 2016. The number of such SAR was based on the fair market value (**FMV**) of each SAR as of January 1, 2017 of CHF 22.28, calculated in accordance with the Company's consistently applied Hull-White model and resulted in an allocation of 85,416 SAR (as disclosed in the invitation to the 2017 AGM). The said FMV had to be re-valuated at the date of the AGM and then amounted to CHF 38.47 per SAR, an increase of 73.1% over the January 1 FMV. As a consequence, the total amount of the values of the SAR in the table above (CHF 3,285,954) are 73.1% higher than the mentioned maximum amount of CHF 1,905,000. For Todd Bazemore, after such re-valuation, the value of his SAR amounted to CHF 201,852 and for the other 5 members of Executive Management, to CHF 3,084,101.

[7] As a consequence of Todd Bazemore's resignation effective as of November 17, 2017, he forfeited all his SAR.

[8] Consisting of a payment of CHF 42,000 for a forfeited bonus of Kristina Nygren's former employment and CHF 3,600 housing allowance for two months.

[9] The number of SAR granted to Kristina Nygren has been agreed in connection with her joining the Company.

Comparison of the approved and paid EM compensation

At the 2016 AGM, shareholders approved a maximum total compensation for the EM for 2017 as follows: CHF 2,600,000 for the fixed compensation in cash.

In CHF	Approved 2017	Paid 2017[1]
Base salary	2,600,000	2,600,000
Additional amount	1,300,000	838,282
Total maximum amount	3,900,000	3,438,626

Changes in the Executive Management in 2017

Effective January 1, 2017, Kristina Sjöblom Nygren joined the Company as Chief Medical Officer (**CMO**), Head of Development and Member of the Executive Management, while Nicholas Coppard retired on January 31, 2017.

As the maximum total compensation approved at the 2016 AGM for 2017 was not sufficient to compensate the newly appointed CMO, her compensation was based on the additional amount as provided for in art. 26 of the Articles of Incorporation to the extent that the approved amount for the fix compensation (CHF 2,600,000) was not sufficient to compensate Kristina Sjöblom Nygren. The actual payments (including social security payments) to EM other than Kristina Sjöblom Nygren amounted to CHF 2,555,344, leaving an amount of CHF 44,656 usable to pay part of Kristina Sjöblom Nygren's total fix compensation of CHF 883,282 (consisting of base salary, retention award, social security contributions thereon and retention award SAR). As a result, the balance compensation of CHF 838,282 had to be paid out of the additional amount (which is capped at 50% of the fix compensation of all EM members as approved by the 2016 AGM, i.e. at CHF 1,300,000).

Todd Bazemore, COO Santhera Pharmaceuticals (USA) Inc. and Member of the Executive Management, left the Company effective November 17, 2017. As a consequence, he forfeited all his SAR.

Event	Date	Number of Executives
Kristina Sjöblom Nygren joined	January 1, 2017	8
Nicholas Coppard retired	January 31, 2017	7
Todd Bazemore resigned	November 17, 2017	6

Outlook for EM compensation

The AGM 2017 has already approved the fix compensation for 2018 in the amount of CHF 3,200,000. Todd Bazemore's tasks have been allocated to Executives and employees of the Company's US operations. The Company is currently considering filling the vacancy and to increase the number of Executives.

For the fix compensation for 2019, the Board will propose an amount of CHF 3,200,000 to the 2018 AGM which would be based on the planned seven Executives, including a reserve amount which would allow to increase the fix compensation of the EM if deemed appropriate by the Board.

Executive Contracts

The employment contracts with the EM members are compliant with the OaEC and the Company's Articles of Incorporation. Any noncompete clauses for the period after termination of an employment agreement shall not exceed one year with the maximum compensation for such period of the last total annual compensation of an EM member in question.

Loans and Credits

In accordance with the Articles of Incorporation, loans to members of the Board and EM may only be on market terms and may only be made by the Company or by any of its directly or indirectly controlled companies, whereas the total sum of total outstanding loans to a particular member, including the amount to be granted, shall not exceed twice the most recent annual compensation to such member. In 2017, no loans or credits were made to the members of the Board, EM or to their related parties.

Compensation of Former Members of the Board and Executive Management

In connection with option exercises by several former members of the Board and EM, Santhera had to contribute to the proceeds from options, as these are subject to social security payments in accordance with applicable laws. With regard to the former Board members, Santhera made a total of CHF 0 (2016: CHF 10,037) for such payments in 2017.

Disclosure of compensation of former Board members for the years 2017 and 2016 (audited)

In CHF	Total payment
2017	
n/a	–
Total	**0**
2016	
Klaus Schollmeier	5,287
Bernd Seizinger	4,750
Total	**10,037**

With regard to the former EM members, Santhera made payments of CHF 0 in 2017 (2016: CHF 0).

Disclosure of compensation of former EM members for the years 2017 and 2016 (audited)

In CHF	Total payment
2017	
n/a	0
Total	0
2016	
n/a	0
Total	0

Shareholdings of Members of the Board and Executive Management

Disclosure of shareholdings in the Company of Board members for the years 2017 and 2016 (audited)

December 31, 2017	Number of shares	Number of stock options (vested)	Number of stock options (unvested)	Number of SAR (vested)	Number of SAR (unvested)
Elmar Schnee	2,000	0	0	0	4,486
Martin Gertsch	38,109	1,500	4,781	0	4,154
Philipp Gutzwiller	500	0	0	0	3,157
Thomas Meier	75,562	13,313	5,312	0	19,038
Patrick Vink	1,000	0	0	0	6,116
Jürg Ambühl	30,000[1]	7'281	0	0	0
Total without Jürg Ambühl	117,171	14,813	10,093	0	36,951
December 31, 2016					
Martin Gertsch	38,109	0	6,281	0	6,281
Jürg Ambühl	30,000	0	7,281	0	7,281
Total	**68,109**	**0**	**13,562**	**0**	**13,562**

[1] As of April 4, 2017. Jürg Ambühl was member of the Board until April 4, 2017, the day of the AGM 2017 when he no longer stood for re-election. In accordance with the terms of the BSOP, all his unvested options vested immediately after the AGM.

Disclosure of shareholdings in the Company of Executive Management members for the years 2017 and 2016 (audited)

December 31, 2017	Number of shares	Number of stock options (vested)	Number of stock options (unvested)	Number of SAR (vested)	Number of SAR (unvested)
Thomas Meier	75,562	13,313	5,312	0	19,038
Todd Bazemore[1]	0	0	0	0	0
Nicholas Coppard[2]	0	7,875	7,375	3,489	6,977
Günther Metz	0	14,000	5,120	0	11,380
Christoph Rentsch	0	7,500	14,500	0	14,101
Kristina Sjöblom Nygren	0	0	0	0	18,617
Giovanni Stropoli	250	7,500	13,065	0	13,434
Oliver Strub	0	6,001	5,240	0	11,750
Total	75,812	56,189	50,612	3,489	95'297

[1] Todd Bazemore forfeited all his SAR on November 17, 2017, the effective date of his resignation.
[2] As of January 31, 2017, the effective date of his retirement.

December 31, 2016	Number of shares	Number of stock options (vested)	Number of stock options (unvested)
Thomas Meier	72,902	3,750	14,875
Todd Bazemore	0	0	34,881[1]
Nicholas Coppard	0	0	12,250
Günther Metz	0	12,000	7,120
Christoph Rentsch	0	0	22,000
Giovanni Stropoli	600	0	20,565
Oliver Strub	0	9,001	7,240
Total	**73,502**	**24,751**	**118,931**

[1] Todd Bazemore forfeited all his SAR on November 17, 2017, the effective date of his resignation.

Building a better working world

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To the General Meeting of
Santhera Pharmaceuticals Holding Ltd, Liestal

Basle, March 19, 2018

Report of the statutory auditor on the compensation report

We have audited the compensation report of Santhera Pharmaceuticals Holding Ltd for the year ended December 31, 2017. The audit was limited to the information according to articles 14–16 of the Ordinance against Excessive Compensation in Stock Exchange Listed Companies (Ordinance) contained in the tables labeled “audited” on pages 79 to 91 of the compensation report.

Board of Directors’ responsibility
The Board of Directors is responsible for the preparation and overall fair presentation of the compensation report in accordance with Swiss law and the Ordinance. The Board of Directors is also responsible for designing the compensation system and defining individual compensation packages.

Auditor’s responsibility
Our responsibility is to express an opinion on the compensation report. We conducted our audit in accordance with Swiss Auditing Standards. Those standards require that we comply with ethical requirements and plan and perform the audit to obtain reasonable assurance about whether the compensation report complies with Swiss law and articles 14 – 16 of the Ordinance.

An audit involves performing procedures to obtain audit evidence on the disclosures made in the compensation report with regard to compensation, loans and credits in accordance with articles 14 – 16 of the Ordinance. The procedures selected depend on the auditor’s judgment, including the assessment of the risks of material misstatements in the compensation report, whether due to fraud or error. This audit also includes evaluating the reasonableness of the methods applied to value components of compensation, as well as assessing the overall presentation of the compensation report.

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

Opinion
In our opinion, the compensation report for the year ended December 31, 2017 of Santhera Pharmaceuticals Ltd complies with Swiss law and articles 14 – 16 of the Ordinance.

Ernst & Young Ltd

/s/ Frederik Schmachtenberg
Licensed audit expert
(Auditor in charge)

/s/ Jan Meyer
Licensed audit expert

Corporate Governance Report

Contents

General Information

The Company's corporate governance principles are laid out in its articles of incorporation (**Articles**), the organizational rules (**Organizational Rules**; *Organisationsreglement*), by-laws of the Company's Audit and Compensation Committees and of executive management (**Executive Management**) adopted by the Board of Directors (**Board**) and a comprehensive set of Group directives, including a Code of Conduct and insider trading rules that require a trading preclearance for the Board and the Company's officers and employees, as well as an internal control system, and a risk management process.

The information published below conforms to the Directive Corporate Governance (**DCG**) of the SIX Swiss Exchange (**SIX**). In order to avoid redundancies, references are inserted to other parts of the financial report. Santhera's website www.santhera.com provides more detailed information.

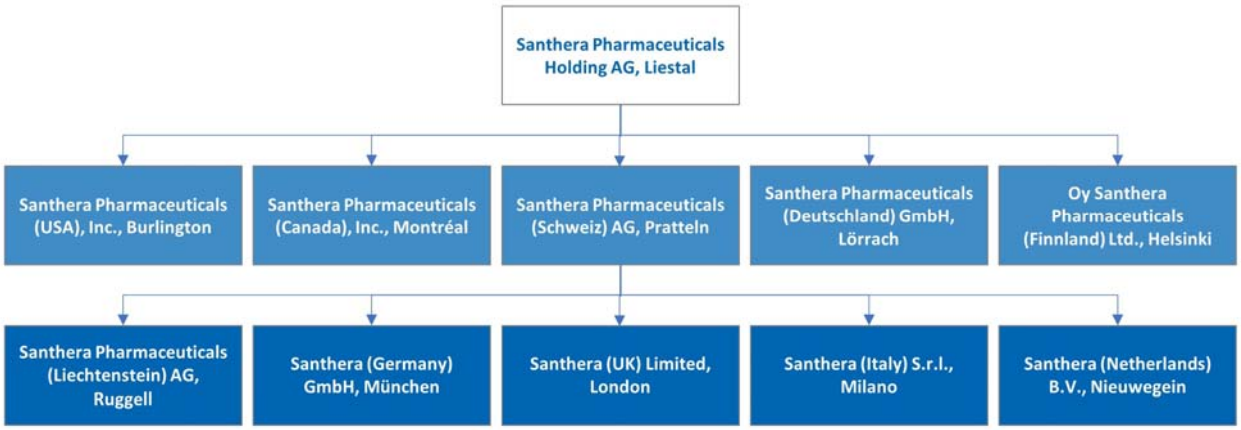
Group Structure and Shareholders (DCG 1)

Group structure (DCG 1.1)

Listed company

Name	Santhera Pharmaceuticals Holding AG (**Company**, together with its affiliates, **Santhera**)
Legal Domicile	Hammerstrasse 49, 4410 Liestal, Switzerland. At the 2018 Annual General Meeting (**AGM**), the shareholders will vote on a change of the legal domicile to the address of the office location mentioned below. This change requires an amendment of the Articles of Incorporation.
Office location	Hohenrainstrasse 24, 4132 Pratteln, Switzerland
Register number	CHE-105.388.338
Listing	SIX Swiss Exchange
Symbol	SANN
Security ID	2714864
ISIN	CH0027148649
Market capitalization	CHF 226 million (December 29, 2017)
Website	www.santhera.com
Duration of company	Not limited
Subsidiaries	See following section as well as note 3.2 *"Investments in shareholdings"* to the statutory financial statements of the Company.

Santhera operates through its wholly owned subsidiaries (DCG 1.1.3):



Company	Share Capital	Domicile	Activities
Santhera Pharmaceuticals (Schweiz) AG	CHF 125,000	Pratteln, CH	Headquarters; development of pharmaceutical drugs, administrative functions
Santhera Pharmaceuticals (Liechtenstein) AG	CHF 50,000	Ruggell, LI	Logistics/distribution
Santhera (Germany) GmbH	EUR 50,000	München, DE	Medical information
Santhera (Netherlands) B.V.	EUR 50,000	Nieuwegein, NL	Medical information
Santhera (UK) Limited	GBP 50,000	London, GB	Medical information
Santhera (Italy) S.r.l.	EUR 50,000	Milano, IT	Medical information
Santhera Pharmaceuticals (Canada), Inc.	CAD 1,000	Montréal, CA	Development of pharmaceutical drugs
Santhera Pharmaceuticals (USA), Inc.	USD 1,000	Burlington, Massachusetts, US	Advocacy/patient liaison
Santhera Pharmaceuticals (Deutschland) GmbH	EUR 25,000	Lörrach, DE	Regulatory and development in the EU
Oy Santhera Pharmaceuticals (Finland) Ltd	EUR 2,500	Helsinki, FI	Administrative

None of these subsidiaries is listed on a stock exchange (DCG 1.1.2). The development activities are managed by Santhera Pharmaceuticals (Schweiz) AG and are mostly performed in Switzerland, the EU and the US (DCG 1.1.1).

Significant shareholders (DCG 1.2)

See note 4.2 *"Significant Shareholders"* to the statutory financial statements of the Company on page 69.

Cross-shareholdings (DCG 1.3)

There are no cross-shareholdings.

Capital Structure (DCG 2)

Ordinary, conditional and authorized capital (DCG 2.1/2.2)

The Company has one class of registered shares with a nominal value of CHF 1 each (**Shares**). As of December 31, 2017, it had the following ordinary, authorized and conditional share capital:

Type of capital	Capital as per commercial register		Effectively outstanding capital			
	Amount in CHF	As % of ordinary capital	Amount in CHF	As % of ordinary capital	Expiry	Section in Articles
Ordinary capital	6,279,857	100.0	6,288,555	100.0		3
Authorized capital	1,500,000	23.9	1,500,000	23.9	May 10, 2018	3a
Conditional capital for warrants/option rights granted in connection with debt instruments	930,000	14.8	930,000	14.8	For conversion rights: 10 years from issue date. For options: 7 years from issue date.	3c
Conditional capital for ESOP/BSOP/EIP	700,000	11.1	691,302	11.0		3b

For details with regard to terms and conditions of potential share issues under the Company's authorized and conditional share capital, see sections 3a, 3b and 3c of the Company's Articles, which can be downloaded from http://www.santhera.com/investors-and-media/investor-toolbox/articles-of-incorporation, and the section on DCG 2.7 below.

For details with regard to the Company's ESOP, BSOP, ESARP and BSARP and EIP, see note 17 "*Equity Rights Plans*" to the consolidated financial statements on page 40.

Changes in share capital (DCG 2.3)

For changes in capital that occurred in 2015 and 2016, see the Company's Annual Report 2016, which can be downloaded from http://www.santhera.com/investors-and-media/investor-toolbox/financial-reports. For changes that took place in 2017, see note 12 *"Share Capital"* to the consolidated financial statements of the Company on page 34.

Shares, participation and dividend right certificates (DCG 2.4/2.5)

As of December 31, 2017, the Company had one single class of registered Shares with a nominal value of CHF 1 each. All Shares were fully paid in and are nonassessable. The Company has not issued any participation certificates or any profit-sharing certificates.

As a consequence of the Swiss Federal Intermediated Securities Act (**FISA**) that entered into force on January 1, 2010, the Company may issue its Shares in the form of uncertificated securities, single certificates or global certificates. The shareholder has no right to demand the printing and delivery of share certificates. However, a registered shareholder may, at any time, request the Company to confirm in writing its shareholding as entered into the share register. The transfer of the Shares is effected via electronic book entry only by the intermediary holding the securities account, usually a bank. The transferability of the Shares is not affected by the changes required by FISA.

Subject to section 5 in the Company's Articles on share register, transfer restrictions and nominees, each Share carries one vote (see section on DCG 2.6) and is entitled to dividends if the AGM resolves in favor of a dividend payment.

Limitations on transferability and nominee registrations (DCG 2.6)

The Company's Shares are freely transferable, provided that the acquirers declare that they acquired the Shares in their own name and for their own account. There is no percentage limitation (DCG 2.6.1), and accordingly, the Company did not grant any exception (DCG 2.6.2).

The Board may register individual nominees (**Nominees**) with the right to vote in the share register up to 2% of the share capital as set forth in the commercial register. Shares in excess of 2% of the total share capital are entered without voting rights, unless the Nominee discloses the names, addresses and number of Shares of persons for whose account it holds such excess Shares. Nominees are persons who do not explicitly declare to hold Shares for their own account. Groups of persons who are interrelated or otherwise act in concert to circumvent the Nominee provisions are treated as a Nominee (DCG 2.6.3). In the year under review, the Company granted no exception.

The Board delegated the administration of the share register to the Group General Counsel (**GC**) who may cancel registration of shareholders if such registration was based on false information and if the GC has previously heard such shareholder or Nominee. No statutory privileges of limitations on transferability exist (DCG 2.6.4).

Convertible bonds and warrants/options (DCG 2.7)

Convertible bonds

On February 10, 2017, the Company placed CHF 60 million senior unsecured convertible bonds (**Convertible Bonds**) due 2022. The Convertible Bonds have a 5-year maturity and a coupon of 5.00% per annum. The Conversion Price was fixed at CHF 86.4006, representing a premium of 20% over the volume weighted average price (**VWAP**) of the Santhera shares between the launch and pricing of the Convertible Bonds (**Reference Share Price**; CHF 71.9969). The Convertible Bonds were issued at 100% of their principal amount and, unless previously redeemed, converted or repurchased and cancelled, will mature on February 17, 2022, at 100% of their principal amount. Each bond (with a denomination of CHF 5,000) was initially convertible into 57.87 Shares. With an issue volume of CHF 60 million, this would have required a maximum of 694,440 Shares to be issued at conversion.

Santhera may call the Convertible Bonds at any time on or after the second anniversary of the issue date at par, plus accrued interest, if any, if the VWAP of the shares is at least 160% of the Conversion Price.

Since the average VWAP of the Santhera Share was below the Reference Share Price on 20 trading days within one year from the launch of the Bond, the Conversion Price was adjusted to CHF 64.80. Each bond is now convertible into 77.16 Shares. With an issue volume of CHF 60 million, this would require a maximum of 925,919 Shares to be issued at conversion. These shares would be issued from the Company's conditional capital of CHF 930,000, allowing for an issue of a maximum of 930,000 Shares with a nominal value of CHF 1 each.

Options, warrants

See the statutory financial statements of the Company and note 17 "*Equity Rights Plans*" to the consolidated financial statements on page 40.

Board of Directors (DCG 3)

Board and committee memberships (DCG 3.1/3.2/3.3/3.4 and 3.5.2)

Composition of the Board of Directors (**BoD**), the Audit Committee (**AC**) and the Compensation Committee (**CC**):

	Year of birth	Nationality	First elected	BoD	AC	CC
Elmar Schnee[1]	1959	CH	2017	●		○
Martin Gertsch	1965	CH	2006	⊙	●	
Philipp Gutzwiller[1]	1968	CH	2017	○	○	
Thomas Meier[1, 2]	1962	DE	2017	○		
Patrick Vink[1, 3]	1963	NL	2017	○		●
Jürg Ambühl[4]	1949	CH	2009			

● = Chairman ⊙ = Vice Chairman ○ = Member

1 Elected for the first time at the 2017 AGM on April 4, 2017.

2 Thomas Meier is also Delegate of the Board and CEO of Santhera

3 In the time between September 2016 and the 2017 AGM, Patrick Vink served as an advisor to the Board.

4 Jürg Ambühl was member of the Board and of the Compensation Committee until the 2017 AGM (April 4, 2017), when he did not stand for re-election.

Elmar Schnee

Elmar Schnee is advisor to management of MindMaze, a neuro-technology company spun off from the Swiss Federal Institute of Technology in Lausanne (**EPFL**). Prior to that, he was chairman, CEO and board member of Cardiorentis in Zug, Switzerland. Previously, he was a general partner and member of the executive board of Merck KGaA, responsible for its worldwide pharmaceutical business. He also led the major restructuring of the business including the acquisition and integration of Serono. Prior to Merck, Mr. Schnee held senior roles as managing director and in marketing, licensing, strategy and business development with UCB Pharma, Sanofi-Synthelabo, Migliara Kaplan and Fisons. He currently serves on the board of directors of listed Jazz Pharmaceuticals and Stallergenes Greer as well as of several privately held life science companies.

Martin Gertsch

Martin Gertsch is an experienced chief financial officer in the life science industry. Until January 2014, he served as chief financial officer of Acino Holding. Before, he was vice president head of finance EMEA at Synthes and held chief financial and chief operating officer positions at Delenex Therapeutics and ESBATech, two privately held biotech companies. From 2002 to the beginning of 2006, he was chief financial officer of Straumann, which he had joined in 1997 as head of group controlling and reporting. Between 1986 and 1997, Mr. Gertsch was an audit engagement manager at PricewaterhouseCoopers, Basel, Switzerland. Mr. Gertsch is a Swiss certified fiduciary and Swiss certified public accountant. He has also completed several executive-level development programs at IMD (International Institute for Management Development) in Lausanne, Switzerland. Mr. Gertsch serves as a board member of Evolva Holding, the University Center of Dentistry, Basel (**UZB**), and several privately held life-science companies.

Philippe Gutzwiller

Philipp Gutzwiller is Global Head Healthcare at Lloyds Banking Group plc in London. He has accumulated over 15 years of experience as a banker to the broader healthcare industry, advising corporate and private equity clients on the assessment, financing and execution of acquisitions and capital market transactions. He started his career at Roche as a financial controller and later worked as an executive in Roche's corporate mergers and acquisitions team.

Thomas Meier

Thomas Meier was appointed CEO of Santhera, effective October 1, 2011, after having served for seven years as Chief Scientific Officer (**CSO**) for the Company. Mr. Meier was the founder and CEO of MyoContract, a Basel/Switzerland-based research company focused on orphan neuromuscular diseases, which he merged in 2004 with Graffinity of Heidelberg, Germany, to form today's Santhera. In 1999, Mr. Meier became an independent research group leader and lecturer in the Department of Pharmacology and Neurobiology at the University of Basel, Switzerland, where he established MyoContract as first start-up of the Biozentrum. Mr. Meier received his PhD in biology from the University of Basel, Switzerland, in 1992 and subsequently joined the University of Colorado Health Sciences Center, Denver, Colorado, US. He has a distinguished scientific track record in the field of neuromuscular research. Before joining the industry, Mr. Meier was awarded the International Research Fellowship Award from the US National Institutes of Health and a long-term fellowship from the Human Frontier Science Foundation. In 2007, he received the BioValley Basel Award for his outstanding contributions to the life sciences in the area. For additional information, see Section on Executive Management below.

Patrick Vink

Patrick Vink, MD, has been an advisor to Santhera's Board since 2016. He advises the life sciences industry and has over 25 years of global industry experience. In his latest assignment, he was employed as Chief Operating Officer at Cubist Pharmaceuticals, overseeing all worldwide commercial and technical operations as well as global alliance management and managing the company's P&L. Previously, Mr. Vink held several senior management positions with Mylan Inc., Novartis Generics/Sandoz, Biogen and Sanofi-Synthelabo. He currently is chairman of the board of listed Targovax ASA and privately-held NMD Pharma and Acacia Pharma and a member of the board of Concordia International Corp. and Spero Therapeutics Inc., both listed, and several privately held life science companies.

Independence of Board members (DCG 3.1.b and c)

With the exception of Thomas Meier, Santhera's CEO, all other four Board members (80%) are non-executive and none has ever been a member of the Executive Management of the Company or any of its subsidiaries. Thomas Meier does not receive any separate compensation as a Board member. He is also not a member of any Board Committees.

Business connections between Board members and the Company (DCG 3.1.c)

See note 25 "*Related Party Transactions*" to the consolidated financial statements on page 52.

Other activities and vested interests (DCG 3.2)

Other than described above, none of the members of the Board has any position in governing or supervisory bodies of any major organization, institution or foundation under private or public law, permanent management or consultancy function for major interest groups, official function or political mandate.

Permitted mandates in other companies (DCG 3.3)

See table in section on DCG 4.3.

Elections and terms of office (DCG 3.4)

According to the Company's Articles, the Board consists of no more than eight members. All members of the Board, including the Chairman in his function as a chairman, are appointed or removed exclusively by a resolution of the shareholders. The Board members are elected on an individual basis for a term of office which must not exceed one year, whereby a year means the period between two AGMs. The terms of the Board members end at the 2018 AGM.

Organizational structure/areas of responsibility and information flow (DCG 3.5)

Allocation of tasks within the Board (DCG 3.5.1)

In accordance with the Organizational Rules of the Company, the Chairman convenes and presides over the Board meetings. After consultation with the CEO, the CFO and the GC, who also acts as the Secretary to the Board, he decides on agenda items and motions. The other Board member may request that items be placed on the agenda. In case of urgency, the Chairman may approve transactions and measures on behalf of the full Board. The Board also approves the Company's news releases.

The Board committees (DCG 3.5.2)

The Compensation Committee consists of two Board members, Patrick Vink (Chairman) and Elmar Schnee (member). The members of the Compensation Committee are elected individually by the AGM for a term of office until the end of the next AGM. The CC's Chairman is elected the Board.

The Audit Committee consists of two Board members, Martin Gertsch (Chairman) and Philipp Gutzwiller (member). Chairman and member of the AC are elected by the Board.

Board – organizational structure and areas of responsibility (DCG 3.5/3.6)

Core tasks of the Board

The Board is entrusted with the ultimate direction of the Company and supervision of the Executive Management. The Board's nontransferable and inalienable duties include the following:

- The ultimate management of the Company, by determining the strategy of the Company based on discussions with Executive Management, e.g. whether to evaluate, pursue or execute a financing, M&A or licensing transaction or a regulatory strategy before regulatory authorities such as the European Medicines Agency (**EMA**) and the US Food and Drug Administration (**FDA**).
- The determination of the organizational structure of the Company, in terms of both organization by departments and organization through the legal structure of the Group, specifically in a tax effective way.
- The oversight of the accounting system, financial control (including the Company's internal control system, risk management as well as financial planning), through structured processes of budgeting (both bottom up and top down), variance analyses, regular latest estimates.
- The appointment, recall and supervision of the Executive Management, the determination of their areas of responsibility and their signing authorities.

The Board is also responsible for the preparation of the Annual Report and the AGM, carrying out shareholders' resolutions, and notification to the judge in case of overindebtedness of the Company.

The Board has delegated the execution of the strategies defined by it and the day-to-day management of the Company to the CEO and Delegate of the Board who relies on a management team where the main functional areas of the Company are represented.

Work methods of the Board and its Committees (DCG 3.5.3)

Board

The adoption of resolutions and elections by the Board requires a majority of the votes cast. To validly pass a resolution, more than half of the members of the Board must be present at the meeting. In case of an impasse, the Chairman has a casting vote. In the period under review, all resolutions by the Board were taken unanimously. Meetings may also be held by teleconference.

Audit Committee

The Audit Committee (**AC**) monitors the integrity of the financial statements of the Company, assesses the independent audit firm's and its representatives' qualifications, the performance of the Company's internal audit function and independent public accountants, and the compliance of the Company with legal and regulatory requirements. The AC reviews the Company's financial statements and budgets on an ongoing basis. It also assesses the Company's internal control system and is responsible for the Company's risk management, accounting principles and policies as well as tax structures. The AC, together with the Board, communicates with the Company's external auditors concerning the results of their interim audits, audits of the annual and reviews of the interim financial statements and assesses important or critical accounting topics with the Executive Management and the external auditors.

Compensation Committee

The tasks of the Compensation Committee are described in the Compensation Report.

Meetings in 2017

In 2017, the Board held five meetings in person which on average lasted more than seven hours. In addition, the Board held ten teleconferences which on average lasted three quarters of an hour, not counting additional calls as required.

Information and control instruments vis-à-vis the Executive Management (DCG 3.7)

As a rule, the CEO, the CFO and the Board's Secretary, who is also the Company's GC, participate in all Board meetings and report to the Board on the current course of business and all significant issues and transactions. Other members of Executive Management are invited to attend discussions of their areas of responsibility (commercial operations, development and business development). Other members of senior management are present when human resources (**HR**), financial, and supply chain topics are discussed. In addition, other employees are invited for certain agenda items covering their area of expertise, for example, to discuss results and progress of clinical studies and submissions to regulatory authorities. From time to time, the Board also invites the Company's auditors and tax advisors to its meetings.

For the year under review, the Board had a risk report prepared by management. Among the key risks identified were the regulatory risk in the EU and the US with respect to the marketing authorization application of idebenone for the treatment of patients with Duchenne muscular dystrophy (**DMD**), sales of Raxone in Leber's hereditary optic neuropathy (**LHON**), the financial situation of the Company, potential loss of key personnel, compliance (**GxP** compliance and compliance with respect to interactions with health care professionals and qualification and validation of computerized systems) and an out of stock risk. For all these risks, mitigation strategies have been and are being implemented.

On a monthly basis, the CFO sends the Management Report to the Board members. Such report contains – inter alia – monthly income statement, balance sheet, overview of cash flows and liquid funds, including sales figures, operating expenses, net operating cash flow, COGS, gross profit, EBIT and EBT, headcount and FTE numbers.

Extraordinary transactions and issues must be reported by the CEO to the Board immediately. The CEO is in regular contact with the Board. Each member of the Board is entitled to request and receive information on all matters of the Company and has access to the Company's and the Company's subsidiaries' property, records and personnel.

Due to its size, Santhera does not have an internal audit function, but parts of this function have been allocated to its finance department and the manager of quality assurance.

Executive Management (DCG 4 and 3.6)

In the beginning of the reporting period, the Executive Management consisted of eight Executives, after Nicholas Coppard's retirement of seven and after Todd Bazemore's resignation of six.

Executive	Function	Nationality	Year of Birth	Remarks
Thomas Meier	Chief Executive Officer, Board Member and Delegate of the Board	DE	1962	
Günther Metz	Head Business Development, EVP	DE	1958	
Kristina Sjöblom Nygren	Chief Medical Officer & Head Development, EVP	SE	1961	Joined effective January 1, 2017
Christoph Rentsch	Chief Financial Officer	CH	1959	
Giovanni Stropoli	Chief Commercial Officer Europe & Rest of World, EVP	IT	1960	
Oliver Strub	General Counsel & Secretary to the Board, EVP	CH	1963	
Todd Bazemore	COO Santhera Pharmaceuticals (USA) Inc	US	1970	Resigned effective November 17, 2017
Nicholas Coppard	Senior Vice President, Development	GB	1959	Retired effective January 31, 2017

The current Members of Executive Management are listed in **bold** font

Members of the Executive Management are appointed by the Board upon proposal by the CEO with the exception of the CEO himself who is appointed upon proposal by the Chairman of the Board.

During the Board and Board committee meetings the CEO reports to the Board as well as whenever required on an ad hoc basis.

The CEO, together with Executive Management, is responsible for implementation of the strategy and the decisions taken by the Board and its Committees within the approved budget. With the support of the management team – consisting of the members of Executive Management, the Senior Vice President (**SVP**) Head Human Resources and the VP Technical Development & Operations – he prepares the business strategy and business plan for decision by the Board. The CEO approves material contracts, decides on the Company's intellectual property rights and the handling of lawsuits. He also allocates financial, personnel and other resources within Santhera and supervises the members of the management team. The management team has regular meetings that usually cover the following topics: product revenues, development programs and clinical studies, regulatory strategies, resource allocation, business development, competitive situation, risk management and internal control system, corporate affairs including important contracts, supply chain and information on subsidiaries, financing situation and strategies, internal and external financial reporting, financial controlling, public and investor relations, human resources, taxes, legal and compliance.

Thomas Meier

See section on Board of Directors (DCG 3) on page 98.

Günther Metz

Günther Metz spent more than 20 years in the life science industry and has been working for Santhera since its inception in 2004. Mr. Metz began his career in drug discovery at the French company Fournier Pharma, and thereafter joined the German start-up Graffinity, which in 2004 merged with MyoContract to form Santhera. Mr. Metz held various research management positions in cross-functional teams and while working at Santhera gained broad experience across the preclinical and clinical pharmaceutical value chain in diverse indications. In 2008, he transitioned to a new area of responsibilities in business development and licensing, taking up the role of VP Business Development at Santhera. Mr. Metz received his PhD in biophysics from the University Freiburg, Germany, in 1992 and subsequently held a postdoctoral research position at Yale University, New Haven, Connecticut, US, supported by a fellowship from the Alexander von Humboldt Foundation.

Christoph Rentsch

With a background in finance, and long-standing experience in the pharmaceutical industry, Christoph Rentsch brings a profound knowledge of the international public and private funding markets to Santhera. Mr. Rentsch started his career in investment banking at Credit Suisse. Subsequently, he worked in various senior management functions for the Alusuisse-Lonza Group both in Switzerland and in the USA. As Head of Group Funding and Capital Markets at Roche, he was responsible for all finance transactions on group level for more than 8 years. In 2003 he became partner of Caperis Ltd, an investment advisory and management firm, before joining privately held Polyphor as CFO, where he supported the company in key stages of its development. Mr. Rentsch joined Santhera in 2015. He holds a degree in Economics and Business Administration from the University of Applied Sciences, Basel.

Kristina Sjöblom Nygren

Kristina Sjöblom Nygren joined Santhera as Chief Medical Officer (CMO) and Head of Development and member of Santhera's Executive Management effective January 1, 2017. Ms. Sjöblom Nygren studied chemistry and biochemistry and graduated as a medical doctor from the Karolinska Institute, Sweden. She brings over 18 years of experience as biopharmaceutical executive in drug development across multiple therapeutic areas, including orphan diseases. During her career she has worked in clinical development roles at Wyeth, AstraZeneca and Biovitrum. Prior to joining Santhera Ms. Sjöblom Nygren served as VP and Head of Clinical Development at Sobi where she was leading the clinical development of all programs from first in man to commercialization and life cycle management.

Giovanni Stropoli

Giovanni Stropoli is an experienced commercial manager in the pharmaceutical industry. Until January 2015, he served as SVP for the region called Mid-Sized Countries at InterMune, Switzerland, an orphan drug company acquired by Roche in 2014. For InterMune, Mr. Stropoli successfully launched Esbriet in 11 countries. Before this assignment he was holding several roles at Eisai, Tokyo, Japan, including country manager in Italy, regional manager for Mid-Sized Countries and finally SVP, New Markets, in London, UK, from 2005 until 2011. Before joining Eisai, Mr. Stropoli was country manager Italy for ALK-Abelló, Copenhagen, Denmark, a market leader in vaccine therapy for allergy. Earlier he held several positions in marketing and sales at Eli Lilly, Indianapolis, Indiana, USA, with assignments in Italy, the US and Spain. Mr. Stropoli started his professional career in 1998 as sales representative with Alfa-Wassermann, Bologna, Italy. Mr. Stropoli holds a degree in veterinary medicine from Sassari University, Sardinia, Italy.

Oliver Strub

Oliver Strub is an experienced commercial lawyer, also responsible for the Company's general legal affairs, insurances, trademarks, IT and facility management. Mr. Strub joined Santhera in 2006 as General Counsel, shortly before the Company listed its shares on the SIX. From 1995 to 2006, he was with Ciba-Geigy, then Ciba Specialty Chemicals (now part of BASF), both Basel, Switzerland, where he was Head Corporate Law and Chief Compliance Officer. Mr. Strub holds a degree in law from Basel University, Basel, Switzerland.

Other activities and vested interests (DCG 4.2)

No member of Executive Management has any position in governing or supervisory bodies of any major organization, institution or foundation under private or public law, permanent management or consultancy function for major interest groups, official function or political post.

Permitted mandates in other companies (DCG 3.3 and 4.3)

Body	Maximum of mandates on board of listed companies	Maximum of mandates on board of privately held companies
Board members	4	8
Members of Executive Management	2	4

Management contracts (DCG 4.4)

There are no management contracts between the Company and third parties.

Compensation, Shareholdings and Loans (DCG 5)

An extensive description of the compensation system and the amounts paid in the year under review are available in the separate Compensation Report.

Shareholders' Participation (DCG 6)

Voting rights restrictions and representation (DCG 6.1)

There are no voting rights restrictions, no statutory group clauses and hence no rules on making exceptions. As a consequence, there is neither a procedure nor a condition for their cancellation.

A shareholder may be represented by his legal representative, the independent proxy or by another shareholder. Shareholders can instruct the independent proxy by completing an instruction form. There are no provisions in the Company's Articles of Incorporation that differ from statutory provisions where the participation of shareholders in the AGM is concerned (DCG 6.1.5)

Statutory quora (DCG 6.2)

There are no statutory quora which differ from the applicable legal provisions.

Convocation of the Shareholders' Meeting (DCG 6.3)

There are no statutory rules on the convocation of the Shareholders' Meeting that differ from the applicable legal provisions.

Agenda rules (DCG 6.4)

The Board decides on agenda items and motions of the AGM. Shareholders with voting rights whose combined holdings represent Shares with a nominal value of at least CHF 1 million or 10% of the Company's share capital may, up to 60 days before the date of the meeting, demand that items be included in the agenda. Such a request must be in writing and must specify the items and the motions to be submitted.

Registrations in the share register (DCG 6.5)

Shareholders entered into the share register as shareholders on a specific qualifying day designated by the Board (record date), which is usually less than five business days before the AGM, are entitled to attend such AGM and to exercise their votes.

Changes of Control and Defense Measures (DCG 7)

Duty to make an offer (DCG 7.1)

At the 2013 AGM, shareholders approved an "opting out" clause in the Articles by which it completely excluded the obligation of a shareholder to submit a public takeover offer for all outstanding Shares if he had acquired 33⅓% of all the Company's voting rights (art. 125 para. 4 FMIA in conjunction with art. 135 para. 1 FMIA).

Clauses on changes of control (DCG 7.2)

The ESOP 2004, 2008, 2010, 2015, the BSOP 2011 and 2015, the BSARP and the ESARP under which most options and all share appreciation rights to receive Shares have been granted, contain clauses according to which all options granted under these plans vest immediately upon a sale of more than 50% of the Shares.

Other than that, as of December 31, 2017, agreements and plans from which members of the Board and/or the Executive Management or other members of senior management benefit or may benefit contain no clauses on changes of control.

Auditors (DCG 8)

Duration of the mandate and term of office of the lead auditor (DCG 8.1)

Ernst & Young, Basel, assumed the existing auditing engagement for Santhera's predecessor company MyoContract in 2002 (DCG 8.1.1). The Shareholders' Meeting elects the Company's auditors for a term of office of one year. The auditor in charge is Frederik Schmachtenberg. He assumed his responsibility in 2017 (DCG 8.1.2).

Auditing fees and additional fees (DCG 8.2/8.3)

The following fees were charged for professional services rendered by Ernst & Young, for the 12-month period ended December 31:

In CHF thousands	2017	2016
Audit services	409	335
Audit-related services	11	0

Audit services are defined as the standard audit work that needs to be performed each year in order to issue an opinion on the consolidated financial statements of Santhera and to issue reports on the local statutory financial statements. It also includes services that can only be provided by the Group auditor and includes the verification of the implementation of new or revised accounting policies and from reporting periods 2007 onwards the audit of the Company's internal control system and risk management. Audit-related services include those other services provided by auditors but not restricted to those that can only be provided by the auditor signing the audit report. They comprise services in relation to general accounting matters. For reasons of good corporate governance, Santhera contracted the provision of tax and internal control system/risk management services to a company other than Ernst & Young.

Supervisory and control instruments pertaining to the audit (DCG 8.4)

The Board performs its supervisory and control functions towards the external auditors. In particular, the Board meets with the auditors at the end of an audit or review to discuss in depth the audit procedures, any findings made and recommendations proposed. The auditor's reports to the Board are also extensively discussed.

Information Policy (DCG 9)

Santhera reports to its shareholders, employees, business partners and other public stakeholders in an open, transparent and timely manner. Equal treatment of all stakeholders is the guiding principle behind its partnership-based approach. In doing so, Santhera is able to promote an understanding of its objectives, strategy and business activities, and to ensure an increasing degree of awareness about Santhera. The Company has adopted a comprehensive disclosure policy to protect Santhera's interests and assets, to release material information in a timely and controlled manner, to observe the legal requirements and rules and in particular to also distinguish competencies and responsibilities of corporate and strategic disclosure and those applicable in marketing and sales or development.

The most important information tools are news releases, the AGMs, the Annual Report, the Interim Reports and the website www.santhera.com.

Investors and other parties interested in subscribing to the Company's news service may do so by registering themselves on www.santhera.com/investors-and-media/news-and-media-center/news-subscriptions. For contact details, see www.santhera.com/contact.

Corporate events 2017

The 2017 Annual General Meeting will be held on Tuesday, April 12, 2018, in Basel, Switzerland. See also www.santhera.com/investors-and-media/corporate-calendar.

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About Santhera

Santhera Pharmaceuticals (SIX: SANN) is a Swiss specialty pharmaceutical company focused on the development and commercialization of innovative medicines for orphan and other diseases with high unmet medical needs. The portfolio comprises clinical stage and marketed treatments for neuro-ophthalmologic, neuromuscular and pulmonary diseases. The most advanced pipeline product, idebenone, is in clinical Phase III for the treatment of Duchenne muscular dystrophy (DMD). Santhera's Raxone® (idebenone) is authorized in the European Union, Norway, Iceland, Liechtenstein and Israel for the treatment of Leber's hereditary optic neuropathy (LHON) and currently commercialized in 20 countries. For further information, please visit www.santhera.com.

Raxone® is a trademark of Santhera Pharmaceuticals.

Forward-Looking Statements

This Annual Report expressly or implicitly contains certain forward-looking statements concerning Santhera Pharmaceuticals Holding AG and its business. Such statements involve certain known and unknown risks, uncertainties and other factors, which could cause the actual results, financial condition, performance or achievements of Santhera Pharmaceuticals Holding AG to be materially different from any future results, performance or achievements expressed or implied by such forward-looking statements. There can be no guarantee that any of the development projects described will succeed or that any new products or indications will be brought to market. Similarly, there can be no guarantee that Santhera Pharmaceuticals Holding AG or any future product or indication will achieve any particular level of revenue. In particular, management's expectations could be affected by, among other things, uncertainties involved in the development of new pharmaceutical products, including unexpected clinical trial results; unexpected regulatory actions or delays or government regulation generally; the Company's ability to obtain or maintain patent or other proprietary intellectual property protection; competition in general; government, industry, and general public pricing and other political pressures. Santhera Pharmaceuticals Holding AG is providing the information in this Annual Report as of the date of the publication and does not undertake any obligation to update any forward-looking statements contained herein as a result of new information, future events or otherwise.

let's go further...

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