santhera

THEIR FUTURE – OUR FOCUS

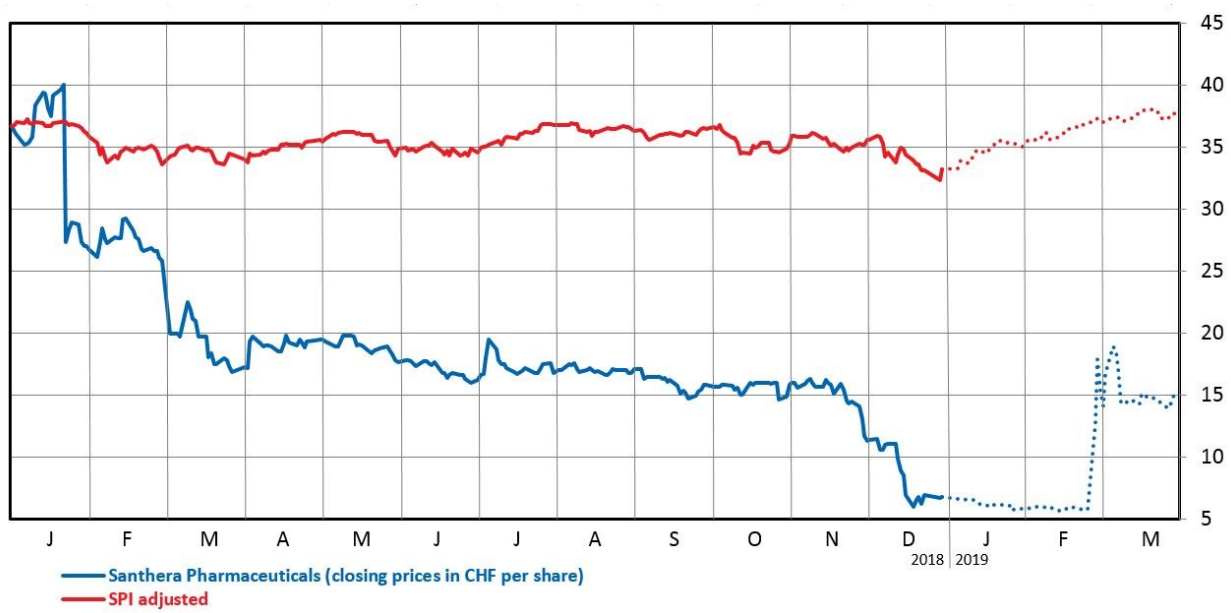
Annual Report 2018

Financial Key Figures

IFRS consolidated, in CHF thousands	**2018**	2017
Net sales	**31,657**	22,943
Operating expenses	**–78,687**	–69,563
Operating result	**–51,420**	–50,454
Net result	**–54,186**	–51,532
Basic and diluted net result per share (in CHF)	**–7.86**	–8.22
Freely available liquid funds at December 31 *	**21,971**	58,206
Net change in cash and cash equivalents	**–23,224**	–4,620

* Cash and cash equivalents and short-term financial assets

Share Price Development in 2018



High	CHF 40.10 (January 23, 2018)
Low	CHF 5.99 (December 17, 2018)
Share price performance in 2018	–81.6%
Share price at year-end	CHF 6.74
Market capitalization at year-end	CHF 72 million
Average trading volume	29,106 shares/day

(based on closing share prices)

Contents

Letter to Our Shareholders

Dear Shareholders,

We are proud to have come through a challenging year and achieved defining milestones which will lead our Company onto a path of sustainable growth. It is therefore right to begin with expressing our thanks to our employees for their dedication and endurance, and to our shareholders for their continued confidence and support.

Top-line growth in 2018 was very strong at 38%, driven by sales of Raxone® (idebenone) for the treatment of Leber's hereditary optic neuropathy (**LHON**) which reached CHF 31.7 million, the upper end of our guidance. We anticipate continued double-digit growth for Raxone as we are expanding our reach, to include territories of the EU and outside Europe, and additional reimbursement schemes.

Broadening our development pipeline was a priority strategic goal in 2018, with the aim to diversify the pipeline with clinical stage drug candidates for rare diseases. By securing the rights to **POL6014** and analogs, a novel compound class with the potential to treat cystic fibrosis (**CF**) and other neutrophilic pulmonary diseases, and ***vamorolone***, a first-in-class dissociative steroid as a potential treatment for Duchenne muscular dystrophy (**DMD**) and other inflammatory diseases, we broadened our pipeline with two highly promising drug candidates in areas with high unmet medical need. These acquisitions also underscore our ability to in-license high-quality, clinical-stage rare disease assets, which leverage our existing capabilities and expertise.

The option to sub-license *vamorolone* from Idorsia has brought us a big step forward in achieving our vision of becoming a leader in making innovative drugs available for patients with neuromuscular diseases, especially DMD. *Vamorolone*, currently being developed by US-based ReveraGen, is a highly promising drug candidate for the treatment of patients with DMD and a perfect strategic fit alongside our own compound *idebenone*. With these two drug candidates, our late-stage DMD drug portfolio now covers DMD patients at all disease stages from young children to adults and irrespective of the underlying genetic background. We look forward to working with ReveraGen in the development of *vamorolone*, which has the potential to replace standard glucocorticoids as a treatment for DMD. Alongside this agreement, we were also delighted to welcome Idorsia as our largest shareholder and strategic partner.

With *idebenone*, we are approaching an important milestone as we plan to file for Conditional Marketing Authorization (**CMA**) in Europe in the second quarter of 2019 under a new tradename, Puldysa®. In the past 12 months, we have focused our work on substantially strengthening our data package to allow for a new submission. We have collected additional clinical data from patients treated with *idebenone*, conducted new analyses of previously submitted data as requested by regulators and provided new natural history data and analyses, collectively addressing the queries of regulatory authorities. In its entirety, these new data demonstrate clinically relevant patient benefits and sustained therapeutic efficacy during treatment with *idebenone* for up to six years in patients with DMD. The new data package and filing strategy have been discussed in several pre-submission meetings with national regulatory authorities.

The goal of our activities and decisions is to develop treatments for rare diseases, often life-altering conditions with a wide variety of symptoms and for which no adequate treatment is available. Working together with patient organizations and clinical experts in the field, we continue to embed the patient perspective in all our development activities. Our initial disease awareness campaigns on respiratory function in patients with DMD were met with great enthusiasm by the patient and caregiver community and encouraged us to continue on this path.

Securing financing to fund Santhera's activities in the longer term remains a priority goal for the current year. The share capital increase in December 2018 – in connection with the acquisition of the option to license *vamorolone* – in unforeseen challenging market conditions provided us with CHF 23.5 million, much lower than the anticipated target amount. In April 2019, in order to close the funding gap, Santhera took out a newly established credit line which, subject to certain conditions, amounts up to CHF 15.0 million, and placed the remaining authorized share capital adding an additional CHF 7.1 million. Total proceeds of these near-term financing initiatives provided new liquid funds of up to CHF 22.1 million.

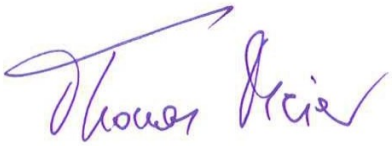
The Company expects major inflection points over the next 12 – 18 months including the start of the regulatory review of our Puldysa filing by the CHMP, topline results of the pivotal trial with *vamorolone* and ultimately the launch of Puldysa in the EU. Additional financial resources are needed to reach these inflection points and we are currently exploring options, including non-dilutive measures such as the monetization of assets, to ensure such financing over the course of this year.

Outlook and Guidance

Based on the performance in the first quarter of the current year, the Company expects continued strong growth and projects net sales of Raxone in the currently approved indication LHON to reach CHF 35-37 million in 2019. The priorities for 2019 are the submission of the marketing authorization application for Puldysa to treat DMD in Europe, advancing the other clinical stage candidates in our pipeline, particularly *vamorolone* and POL6014, and securing the necessary financial resources of Santhera to achieve these objectives.

We again thank all of our employees for their hard work and commitment to Santhera during 2018 and the start of 2019. Our thanks also go to our scientific and clinical partners and advisors for their continued support, all our shareholders for their unwavering confidence in Santhera's ambition in very challenging times and, most importantly, the patients who are involved in the clinical development of our products.

Sincerely,



Elmar Schnee
Chairman

Thomas Meier
Chief Executive Officer

FINANCIAL HIGHLIGHTS

Santhera Delivers Strong Top-line Growth in 2018

Santhera has achieved excellent sales performance and robust revenue growth in 2018 and expanded its rare disease pipeline with clinical-stage drug candidates.

Strong double-digit sales growth from Raxone® for LHON

In 2018, net revenues from product sales of Raxone for Leber's hereditary optic neuropathy (LHON) reached CHF 31.7 million which corresponds to a high double-digit growth of 38% year-on-year (2017: CHF 22.9 million). Growth was primarily driven by higher market penetration and additional reimbursement schemes coming into effect. The roll-out of Raxone in the approved indication is progressing as planned with reimbursement and launches in additional countries expected in 2019.

Raxone® sales up 38% year-on-year

Operating and net results reflect higher investment in clinical development and for regulatory submissions

The increase in development expenses to CHF 38.2 million (2017: CHF 26.6 million) reflects the additional efforts for the preparation of the regulatory filing for Puldysa for DMD in Europe, the ongoing late stage clinical studies such as the Phase III SIDEROS trial in DMD, ongoing post-authorization studies for LHON and clinical work with POL6014, which was acquired early in 2018. The Company reports combined marketing and sales expenses of CHF 24.9 million (2017: CHF 28.5 million) and general and administrative expenses of CHF 15.4 million (2017: CHF 14.4 million), generally in line with previous year. In summary, total operating expenses reached CHF 78.7 million (2017: CHF 69.6 million) whereas the operating result remained almost unchanged at CHF –51.4 million (2017: CHF –50.5 million). For the full-year 2018, Santhera reported a net result of CHF –54.2 million (2017: CHF –51.5 million), similar to the previous year's level.

Increased expenses to fund late-stage pipeline and filing preparations

Cash and cash flows primarily impacted by business development success in enlarging the product pipeline

Santhera expanded its development pipeline in two transactions, acquiring licenses to POL6014 from Polyphor and *vamorolone* from Idorsia, which the company paid for with a mix of shares and cash. In December 2018, the Company completed an ordinary capital increase with the placement of 3,133,334 new shares and raised gross proceeds of CHF 23.5 million. The proceeds were used in part for the cash payment of the latter transaction, namely to complete the acquisition of the option for the exclusive sub-license to *vamorolone*.

As a consequence of these acquisitions, as of December 31, 2018, freely available liquid funds (cash and cash equivalents and short-term financial assets) amounted to CHF 22.0 million (December 31, 2017: CHF 58.2 million). In addition, the Company reported CHF 4.5 million (December 31, 2017: CHF 7.5 million) of restricted cash designated for the interest payments related to the convertible bonds during the first three years.

Significant cash needs for pipeline expansion

FINANCIAL HIGHLIGHTS

Near-term corporate financing initiatives completed

In April 2019, after the reporting date, Santhera took out a syndicated credit line, which, subject to certain conditions, will provide up to CHF 15.0 million over a period of nine months. Alongside this, Santhera placed all of the remaining 500,000 registered shares from its existing authorized share capital in a private placement, resulting in aggregate gross proceeds of CHF 7.1 million. Taken together, the proceeds of these near-term financing initiatives provided new liquid funds of up to CHF 22.1 million. In addition, and to fund operations up to the achievement of key milestones expected in 2020, Santhera is considering various options including a capital increase and/or the monetization of certain assets.

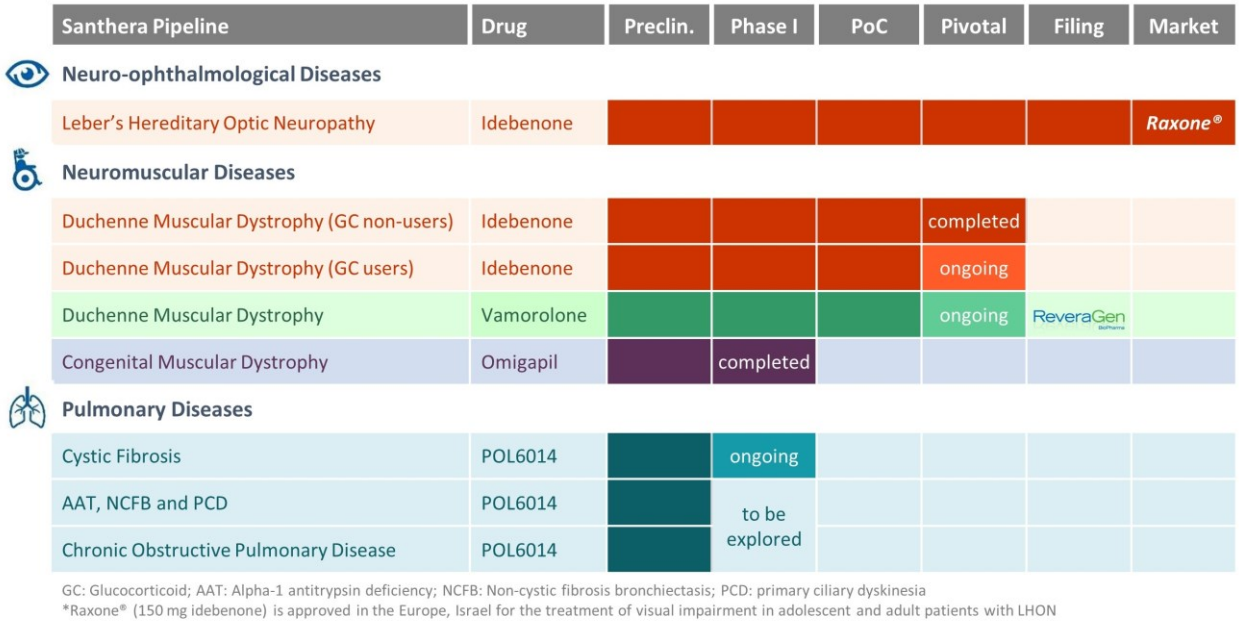
Near-term funding needs secured

Santhera plans to submit a marketing authorization application for Puldysa for the treatment of DMD in Europe in the second quarter of 2019. With the objective to secure financing of the Company until the expected market entry of Puldysa in Europe, the Board is proposing to the forthcoming Annual General Meeting on May 28, 2019, an increase of conditional capital and the creation of authorized capital. Subject to approval by the Company's shareholders, this increase would enable Santhera to raise new capital in a flexible way.

OUR RARE DISEASE APPROACH

Our Rare Disease Pipeline

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



GC: Glucocorticoid; AAT: Alpha-1 antitrypsin deficiency; NCFB: Non-cystic fibrosis bronchiectasis; PCD: primary ciliary dyskinesia
*Raxone® (150 mg idebenone) is approved in the Europe, Israel for the treatment of visual impairment in adolescent and adult patients with LHON

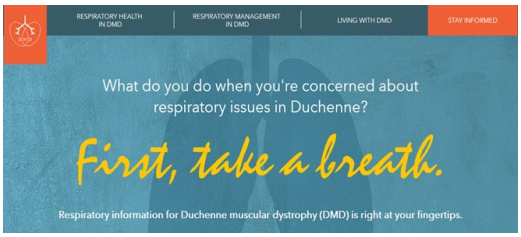
Santhera's rare disease portfolio comprises clinical stage and marketed medicines:

Santhera's Raxone® (idebenone) is authorized in the European Union, Norway, Iceland, Liechtenstein, Israel and Serbia for the treatment of Leber's hereditary optic neuropathy (**LHON**) and is currently commercialized in more than 20 countries. Puldysa® (idebenone) is in regulatory pre-submission in Europe for the treatment of Duchenne muscular dystrophy (**DMD**). A large clinical trial (SIDEROS) in DMD-patients not using glucocorticoids is progressing. *Vamorolone*, to which Santhera acquired an exclusive in-licensing option, is in clinical trials for the lead indication DMD. POL6014, in-licensed from Polyphor in February 2018, is a clinical stage selective inhibitor of human neutrophil elastase in early clinical trials as a potential treatment of cystic fibrosis (**CF**) with additional indications under evaluation. In addition, Santhera's pipeline includes *omigapil*, an investigational drug with anti-apoptotic properties, in development to address unmet medical needs for patients with congenital muscular dystrophy (**CMD**).

OUR RARE DISEASE APPROACH

Be Aware, Show You Care

Rare diseases present unique challenges and initiatives to raise awareness about disease diagnosis, progression and management to enhance therapy outcomes. Reassured by the high acceptance of our educational websites www.breatheduchenne.com and www.takeabreath.com, which provide comprehensive information on breathing in DMD, we continued along this path to heighten awareness for both the rare conditions as well as the needs of affected patients.



Raising awareness for respiratory function in DMD

Together with Parent Project Muscular Dystrophy, Santhera has developed educational videos which explain the importance of breathing to patients and caregivers and can serve as an educational resource for physicians. [1]

Giving patients a voice

We are sharing stories of patients with neuromuscular diseases in conjunction with *Living in the Light*™, a patient advocacy initiative telling rare disease stories to educate about the realities of rare diseases and their unprecedented effect on families and daily life.

The *Limitless* collection of real-life patient stories provides a platform for patients to become ambassadors thereby fostering awareness and deepening understanding of their condition. [2]

[1] www.parentprojectmd.org/care/care-guidelines/by-area/care-for-lung-muscles/pulmonary-video-series/

[2] www.santhera.com/patients/patient-stories

LEBER'S HEREDITARY OPTIC NEUROPATHY

Living with LHON

Giving patients a voice – CHAZ

Chaz was born with perfect eyesight. One morning in his freshman year of college, his vision suddenly changed. "I woke up one day and my right eye couldn't see at all," Chaz says. "My vision was distorted. Light was coming through with some peripheral vision, but my central vision was gone." The first round of medical tests revealed no answers. It wasn't until a few months later, when the vision in his left eye deteriorated, that Chaz met with experts at Tufts University. A neuro-ophthalmologist told Chaz he had LHON. "I was told that I would be legally blind for the rest of my life. That summer, I began drinking. I went into my room and turned the lights off. I didn't want to do anything."

"I want to educate the larger community. I want people to see visually impaired people as assets, not as helpless." – Chaz

A few months into his self-described period of "self-pity", Chaz woke up and thought I'm not doing this anymore. "Somehow, I got my spark back," he says. Newly blind, he returned to school. Although Chaz was succeeding academically, he missed competitive running. He longed to be a part of a team again, and to feel once more the physicality that was his natural born gift. "That fall, I tried to run the preseason trials with my team, but I fell down a bunch of times, tripping on rocks and sticks." Chaz's coach suggested he give indoor track a shot because of the smooth running surface. "With my peripheral vision, I could discern the lines of the track. I didn't fall at all. I could innately sense other runners around me." Running track gave Chaz a boost in confidence. He increased the intensity of his training and soon became the fastest runner on the team.

The 2016 Paralympics in Rio de Janeiro were on the horizon. Three years after forcing himself out of the darkness of his bedroom, he set an American record. Coming off the high of the Paralympics, Chaz says it was a challenge to adjust to everyday life. "I kind of realized then… I'm actually blind. The world around me is a sighted world. It's not set up for blind people to succeed. I needed more [life] training." He moved to Denver and enrolled in a masters in social work program at the University of Denver, where he is pursuing clinical social work within the blind community. He also recently made his marathon debut at the California International Marathon running it at 2:31:48, setting an American record in the visual impairment category.

Chaz insists his accomplishments are more than just personal successes. "I want to educate the larger community. I want people to see visually impaired people as assets, not as helpless. I am a representative of the blind community whenever I'm in public. If I can inspire someone just by being myself, then that's good."

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Extract from "LIMITLESS - Stories of Defiance, Opportunity, and Ability". Created by Living in the Light in partnership with Santhera. Read the full article at www.santhera.com/patients/patient-stories

LEBER'S HEREDITARY OPTIC NEUROPATHY

RAXONE® (idebenone) in LHON

LHON cause. Leber's hereditary optic neuropathy (LHON) is a heritable genetic disease causing profound vision loss and blindness. The disease is caused by genetic mutations leading to deficiencies in the small cell structures called mitochondria which are no longer able to produce sufficient energy to work properly (retinal ganglion cell dysfunction), leading to the characteristic loss of vision.

LHON symptoms. LHON affects approximately two in every 100,000 people in Europe and presents in young adulthood, more commonly in males, as rapid, painless loss of central vision, usually leading to permanent bilateral vision loss within a few months of the onset of symptoms. Without treatment, vision loss will be permanent in the majority of people with the condition.

Raxone in LHON. Santhera's Raxone (idebenone), the first pharmaceutical product for a mitochondrial disorder, is the only medicine approved for LHON. It is authorized in the European Union, Norway, Iceland, Liechtenstein, Israel and Serbia and is currently commercialized in more than 20 countries. Raxone has been shown to promote recovery of visual acuity and current data demonstrate that around 50% of patients may benefit from treatment, either by preventing from progression of visual acuity loss or by experiencing a clinically relevant recovery of visual acuity. Raxone for the treatment of LHON has orphan drug status in the EU, US, Switzerland and South Korea.

RAXONE® idebenone 150mg tablets

Highlights 2018/2019 (to date):

- In June 2018, Santhera submitted a new drug application for Raxone for the treatment of LHON to the Korean Ministry of Food and Drug Safety (MFDS). The authorities have accepted the dossier for review and a response is expected by mid 2019.
- Early in 2019, Raxone was approved for LHON by the Medicines and Medical Devices Agency of Serbia. Ewopharma, Santhera's marketing partner for Eastern Europe, will commercialize Raxone in Serbia.
- Recruitment into LEROS trial completed with 197 patients enrolled. LEROS is a Phase IV, external natural history controlled, open-label intervention study to further assess the efficacy and safety of long-term treatment with Raxone in patients with LHON (ClinicalTrials.gov Identifier: NCT02774005). The primary objective of the trial is to assess the efficacy of Raxone to improve or stabilize visual acuity (VA) in patients starting treatment up to one year after the onset of vision loss, compared to an external natural history control group. Santhera is conducting the LEROS trial in 31 study sites across nine European countries and the USA and expects to complete 24-month study by the second quarter 2021.
- Expanded Access Program completed with 111 patients receiving Raxone treatment, as well as completion of a retrospective natural history data collection with data from 592 patients.
- Progress with PAROS, a multicenter, prospective, non-interventional post-authorization safety study (PASS) for patients with LHON treated with Raxone (ClinicalTrials.gov Identifier: NCT02771379).

DUCHENNE MUSCULAR DYSTROPHY

Living with DMD

Giving patients a voice – TAYJUS

At the foundation of what makes Tayjus' life function is his dedication to keeping himself healthy. As is typical in men with DMD, his lungs are a fragile ecosystem, susceptible to complications from the common cold. Respiratory infection can be fatal. During his second semester of freshman year, Tayjus contracted pneumonia. With fluid-filled lungs and a raging fever, he was bedridden for two weeks and required three rounds of antibiotics. "The doctor said my lung function was a lot lower than it used to be. He said I had to use a cough-assist machine twice a day, even when I was not sick." To demonstrate, Tayjus asks for the mask to be placed on his nose and mouth.

The machine sounds like a vacuum, forcing him to inhale first, and then suction out a powerful cough, to dislodge any phlegm or mucus built up in his lungs. "It's hard to cough on your own with Duchenne. I do this once in the morning and once at night. If I get sick—the minute I get a scratchy throat—I use it aggressively."

He admits he get scared when he sees older men with Duchenne who are tracheostomized or wear a BiPap (or other breathing machine), full-time. "I do whatever I can to preserve my lung function."

"I want to be as independent as possible, while I still can." — Tayjus

Part of Tayjus' advocacy involves speaking with newly diagnosed DMD families. "When a child is first diagnosed with Duchenne, the parents think about their kid not walking or playing sports. They are worried about how long their child will live." He reminds them that this is the best time for a kid to be born with Duchenne because there are so many options. He tells people about the dozens of companies that are working to improve people's lives through innovative therapies, and the creation of devices to improve health and slow down muscle degeneration. "It gives them hope," he says.

The ideals of biotech and pharmaceutical companies resonate with Tayjus' professional goals: to extend and improve the lives of boys and men living with DMD. He plans to work in public policy, lobbying for access to care for all individuals with Duchenne. Already, he has worked closely with Parent Project Muscular Dystrophy, having served on several advisory boards and advocating on Capitol Hill. He also has brought a patient perspective to a number of pharmaceutical developers, working as an intern for two companies who are researching drugs for DMD. When asked if he'd ever consider running for office, his eyes hinted at a sparkle. "I don't know. Maybe." With his education, work as an advocate and his personal experience living with DMD, he is uniquely poised to unlock each one in due time.

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Extract from "LIMITLESS - Stories of Defiance, Opportunity, and Ability". Created by Living in the Light in partnership with Santhera. Read the full article at www.santhera.com/patients/patient-stories

DUCHENNE MUSCULAR DYSTROPHY

PULDYSA® (idebenone) and *Vamorolone* in DMD

DMD cause. DMD is a genetic, degenerative disease characterized by a loss of the protein dystrophin, leading to cell damage, impaired calcium homeostasis, elevated oxidative stress, reduced energy production in muscle cell and progressive muscle cell loss and weakness.

DMD symptoms. DMD is one of the most common and devastating types of progressive muscle weakness and degeneration starting at an early age and leading to early morbidity and mortality due to respiratory failure. It occurs almost exclusively in males with an incidence of up to 1 in 3,500 live male births worldwide. With age, often only after the loss of mobility, progressive respiratory muscle weakness affecting thoracic accessory muscles and the diaphragm causes respiratory disease, impaired clearance of airway secretions, recurrent pulmonary infections due to ineffective cough, and eventually respiratory failure. Therapy at earlier stages focuses primarily on the preservation of skeletal muscle strength and patients' mobility. There is currently no approved treatment of respiratory dysfunction in patients with DMD.

Our pipeline in DMD. With Puldysa (idebenone) and *vamorolone*, Santhera has two clinical stage development compounds which in combination address the medical need of DMD patients at all disease stages and irrespective of their underlying mutation. Therapy at earlier stages focuses primarily on the preservation of skeletal muscle strength and patients' mobility while preservation of respiratory function emerges as a therapeutic priority target as the disease progresses.

Idebenone has demonstrated benefits in clinical trials for the treatment of respiratory dysfunction in patients with DMD. In conjunction with the extrapolation of natural history data, these findings indicated a potential of *idebenone* to delay the time to clinically relevant milestones of disease progression, such as assisted ventilation, by up to three years. During long-term therapy the beneficial effects of *idebenone* were sustained year on year for up to six years. Furthermore, the preservation of respiratory functions shown with *idebenone* was shown to reduce the risk of important patient-relevant outcomes, including bronchopulmonary adverse events and hospitalizations due to respiratory causes.

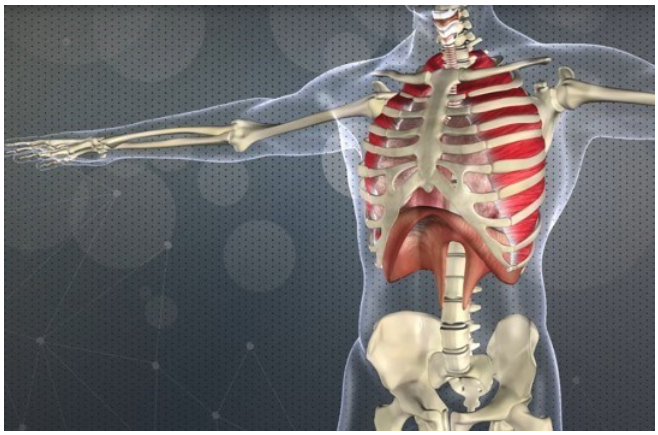
Vamorolone is a first-in-class drug candidate that binds to the same receptors as glucocorticoids but modifies the downstream activity of the receptors. This has the potential to 'dissociate' efficacy from typical steroid safety concerns and therefore could replace existing glucocorticoids as the current standard of care such as prednisone in children and adolescent patients with DMD. There is significant unmet medical need in this patient group as high dose glucocorticoids have severe systemic side effects, which limit long-term usage. *Vamorolone* was discovered by US-based ReveraGen BioPharma Inc. and is currently being developed with participation in funding and design of studies by several international non-profit foundations. Santhera has obtained an option to an exclusive license to *vamorolone* worldwide except Japan and South Korea.

DUCHENNE MUSCULAR DYSTROPHY

Puldysa (idebenone) highlights 2018/2019 (to date):

- In February 2018, Santhera Pharmaceuticals (SIX: SANN) launched a U.S. Expanded Access Program (EAP) referred to as BreatheDMD with *idebenone* for patients with DMD. Through the BreatheDMD program, eligible patients in the U.S. with DMD who are 10 years and older and in respiratory function decline, can obtain access to investigational *idebenone*.

- In June 2018, the UK's Medicines and Healthcare products Regulatory Agency (MHRA) renewed the Early Access to Medicines Scheme (EAMS) scientific opinion for Raxone for patients with DMD in respiratory function decline who are not taking glucocorticoids. Under EAMS, patients with life threatening or seriously debilitating conditions obtain access to medicines that do not yet have a marketing authorization when there is a clear unmet medical need.



- In July 2018, Santhera presented novel data linking measurements of functional parameters to patient benefits. The data demonstrate that a decline in peak expiratory flow is a direct indicator of time to initiation of assisted ventilation and risk of death and can serve as a measure for disease progression.

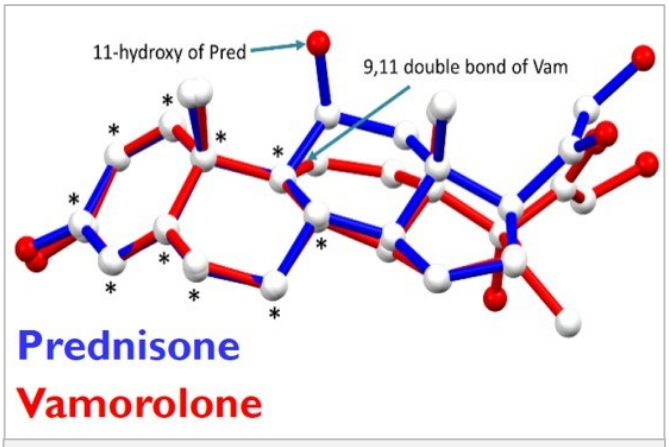
- In January 2019, Santhera announced SYROS study results demonstrating that long-term treatment with *idebenone* consistently reduced the rate of respiratory function loss in patients with DMD for up to 6 years in a real-world setting. This long-term data further supports the potential for *idebenone* to modify the course of respiratory function decline and delay the time to clinically relevant milestones.

- In Q2 2019, Santhera plans to file an application for Conditional Marketing Authorization (CMA) for Puldysa® (idebenone) for the treatment of respiratory dysfunction in Duchenne muscular dystrophy (DMD) with the European Medicines Agency (EMA). Following an earlier rejection, the regulatory dossier has been expanded and substantiated with additional clinical data from patients treated with *idebenone*, new analyses of previously submitted data and new comprehensive natural history data, addressing requests from regulatory authorities. In its entirety, these new data demonstrate clinically relevant patient benefits and sustained therapeutic efficacy during treatment with *idebenone* for up to six years in patients with DMD.

DUCHENNE MUSCULAR DYSTROPHY

***Vamorolone* highlights 2018/2019 (to date)**

- In November 2018, Santhera Pharmaceuticals and Idorsia Ltd entered into an agreement under which Santhera acquired the option to exclusively in-license, by way of sub-license, the first-in-class dissociative steroid *vamorolone* in all indications and all countries worldwide except Japan and South Korea. ReveraGen, inventor and developer of *vamorolone*, remains fully responsible to conduct and complete the ongoing pivotal Phase IIb VISION-DMD trial as planned. Entering the sub-license agreement with Idorsia has positioned Santhera as a leading company in the DMD space.



Comparison of the experimentally determined crystal structures of vamorolone (red) and the glucocorticoid prednisone (blue). Taken from Hoffman et al. (2018) Steroids 134:43–52

- At present, ReveraGen is conducting the pivotal Phase IIb VISION-DMD study (**VBP15-004**, ClinicalTrials.gov Identifier: NCT03439670). The study builds on the available promising preliminary safety and efficacy data from Phase IIa trial and is designed to bridge exploratory biomarker data to clinical outcomes. This pivotal study will enroll approximately 120 boys aged 4 to <7 with DMD that have not yet been treated with glucocorticoids, randomized to *vamorolone* (two dosage strengths), prednisone or placebo. The study is being conducted at approximately 30 sites across North America, Europe, Israel and Australia. Top-line results for the primary endpoint after 6 months of treatment are expected in second half of 2020. *Vamorolone* has received Orphan Drug Designation in the US and in Europe and fast-track status in the US.

For additional information consult www.reveragen.com and www.vision-dmd.info

CONGENITAL MUSCULAR DYSTROPHY

Omigapil in CMD

CMD cause. CMD refers to a variety of inherited neuromuscular conditions characterized by different forms of progressive loss of muscle tissue. The most common forms of CMD are the MDC1A- or LAMA2-type (resulting from laminin alpha2-deficiency) and COL6-type (resulting from collagen VI-deficiency) dystrophies.

CMD symptoms. CMDs are characterized by congenital-onset weakness and hypotonia and have associated dystrophic findings on muscle biopsy. 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 for CMD.

Omigapil in CMD. Omigapil is a deprenyl-analog with anti-apoptotic properties. Santhera obtained an exclusive license for omigapil from Novartis for the development in CMDs. 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. Omigapil has orphan drug designations for CMD in the US and Europe and was granted Fast Track Designation by the FDA.

Highlights 2018/2019 (to date):

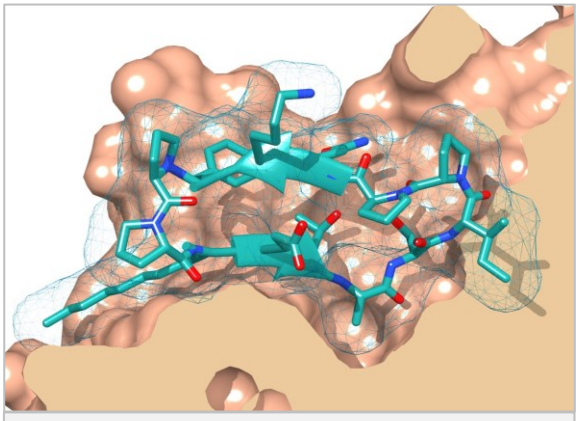
- In April 2018, Santhera reported the successful completion of the first clinical trial with omigapil in patients with two forms of CMD conducted in the US at the National Institutes of Health (**NIH**). The ascending multiple dose cohort study (**CALLISTO**, ClinicalTrials.gov identifier NCT01805024) met its primary objective to establish a favorable pharmacokinetic profile of omigapil and demonstrated that the study drug was safe and well tolerated in children and adolescents with CMD.
- Following further data analysis, the Company will further discuss these results with clinical experts and regulatory authorities to assess a potential development path forward in patients with CMD.

CYSTIC FIBROSIS

POL6014 in CF

CF cause. CF is a rare, hereditary, life-threatening, progressive disease. Activated or necrotic neutrophils liberate human neutrophil elastase (**hNE**) in the lung that causes damage to structural, cellular and soluble components of the pulmonary microenvironment. High levels of hNE play a central role in the pathophysiology of CF and correlate with disease severity.

CF symptoms. CF affects 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. 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. There is still a high unmet medical need in treating the chronic inflammation in patients with cystic fibrosis.



POL6014 in complex with human neutrophil elastase shows binding of POL6014, occupying a large part of the enzyme pocket

POL6014 in CF. POL6014 is a highly potent and selective inhibitor of hNE and was shown to reach high concentrations in the lung when administered by inhalation via an optimized eFlow® nebulizer (PARI Pharma GmbH). In Phase I studies, the drug candidate was well tolerated and safe with a favorable pharmacokinetic profile and strong elastase inhibition as previously shown in animal models.

Highlights 2018/2019 (to date):

- In February, Santhera entered into a license agreement with Polyphor for POL6014. Under the agreement, Santhera obtained the worldwide, exclusive rights to develop and commercialize POL6014 as a potential treatment for cystic fibrosis and other pulmonary diseases.
- In October 2018, the Committee for Orphan Medicinal Products (**COMP**) of the European Medicines Agency (EMA) issued a positive opinion on orphan drug designation for POL6014 in the treatment of CF, thereby acknowledging the needs of patients with CF, and for a novel treatment approach with POL6014 to support these patients. The orphan drug designation will provide Santhera with regulatory and financial incentives to develop POL6014 in the treatment of CF.
- Also in October 2018, Santhera started a Phase Ib/IIa multiple ascending dose (**MAD**, ClinicalTrials.gov Identifier: NCT03748199) trial with POL6014 in patients with CF. The study is designed to evaluate the safety, tolerability, pharmacokinetics and pharmacodynamics of orally inhaled multiple doses of POL6014 in up to 40 patients with CF. Patients will be treated for 15 days with either placebo or one of three ascending doses of POL6014 given once or twice daily. The trial, which is taking place in sites in Germany and Poland, is expected to complete in 2H 2019.

Consolidated Financial Statements

Contents

Consolidated Balance Sheet

As of December 31, in CHF thousands	Notes	2018	2017
Assets			
Tangible assets	5	2,269	2,157
Intangible assets	6	61,467	23,560
Financial assets long-term		690	713
Restricted cash long-term	11	1,500	4,500
Deferred tax assets	16	1,285	1,242
Noncurrent assets		**67,211**	**32,172**
Prepaid expenses	8	969	853
Inventories	9	9,282	10,147
Trade and other receivables	10	7,861	5,402
Financial assets short-term	15	0	13,011
Restricted cash short-term	11	3,000	3,000
Cash and cash equivalents	11	21,971	45,195
Current assets		**43,083**	**77,608**
Total assets		**110,294**	**109,780**
Equity and liabilities			
Share capital	12	10,665	6,289
Capital reserves and share premium		435,795	392,002
Retained earnings		–414,267	–360,081
Employee benefit reserve		–2,675	–4,905
Treasury shares	12	–904	–335
Translation differences		–785	–714
Total equity		**27,829**	**32,256**
Convertible bonds	15	54,569	53,111
Derivative financial instruments	15	204	2,792
Pension liabilities	24	7,983	8,375
Total noncurrent liabilities		**62,756**	**64,278**
Trade and other payables	17	8,306	4,734
Accrued expenses	18	11,041	8,080
Accruals for income taxes		362	432
Total current liabilities		**19,709**	**13,246**
Total liabilities		**82,465**	**77,524**
Total equity and liabilities		**110,294**	**109,780**

Consolidated Income Statement

For the year ended December 31, in CHF thousands	Notes	**2018**	2017
Net sales	21	31,657	22,943
Cost of goods sold		–4,702	–4,104
Of which amortization intangible assets		*–3,039*	*–3,039*
Other operating income	22	312	270
Development	23	–38,240	–26,561
Marketing and sales	23	–24,884	–28,522
General and administrative	23	–15,365	–14,416
Other operating expenses	23	–198	–64
Operating expenses	**23**	**–78,687**	**–69,563**
Operating result		**–51,420**	**–50,454**
Financial income	25	4,371	4,134
Financial expenses	25	–6,815	–4,955
Result before taxes		**–53,864**	**–51,275**
Income taxes	26	–322	–257
Net result		**–54,186**	**–51,532**
Basic and diluted earnings/loss per share (in CHF)	27	–7.86	–8.22

Consolidated Statement of Comprehensive Income

For the year ended December 31, in CHF thousands	Notes	**2018**	2017
Net result		**–54,186**	**–51,532**
Items never to be reclassified to net income in subsequent periods:			
Actuarial gains/losses on defined benefit plans	24	2,230	–171
Items to be reclassified to net income in subsequent periods:			
Currency translation differences		–71	82
Other comprehensive result		**2,159**	**–89**
Total comprehensive result		**–52,027**	**–51,621**

Consolidated Cash Flow Statement

For the year ended December 31, in CHF thousands	Notes	**2018**	2017
Result before taxes		**–53,864**	**–51,275**
Depreciation of tangible assets	5	605	257
Amortization of intangible assets	6	3,136	3,125
Expenses for equity rights plans	20, 23	7,426	9,687
Change in fair value of derivatives		–2,588	–2,540
Change in fair value of financial assets short-term		293	–96
Other non-cash items (Polyphor clinical material)	13	290	0
Change in pension liabilities	24	1,838	2,021
Taxes paid		–365	–392
Change in net working capital		5,959	315
Total financial result	25	2,445	821
Interest received	25	1	5
Interest paid	25	–3,041	–1,561
Cash flow from operating activities		**–37,865**	**–39,633**
Investments in tangible assets	5	–1,348	–1,261
Investments in intangible assets	6, 14	–20,294	–136
Investments in financial assets short-term	15	0	–12,915
Disposal of other financial assets short-term	15	12,718	0
Change in investments in other long-term financial assets		15	–427
Change in restricted cash	11	3,000	–7,500
Cash flow from investing activities		**–5,909**	**–22,239**
Capital increase	12	23,500	0
Capital increases from options exercised	12	17	34
Proceeds from sale of treasury shares	12	2,278	9,372
Purchase of treasury shares	12	–3,413	–9,567
Cost of issuance share capital	12	–1,738	0
Proceeds from convertible bonds	15	0	57,269
Cash flow from financing activities		**20,644**	**57,108**
Effects of exchange rate changes on cash and cash equivalents		–94	144
Net increase/decrease in cash and cash equivalents		**–23,224**	**–4,620**
Cash and cash equivalents at January 1		45,195	49,815
Cash and cash equivalents at December 31		**21,971**	**45,195**

For a discussion of significant non-cash transactions under investments in intangibles assets (POL6014, sub-license vamorolone) see note 13 "*Transaction with Polyphor*" and note 14 "*Transaction with Idorsia*"

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, 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	24	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	20, 23	0	9,687	0	0	0	0	**9,687**
Capital increase from options exercise	12	9	25	0	0	0	0	**34**
Change in treasury shares	12	0	–32	0	0	–163	0	**–195**
Balance at December 31, 2017		**6,289**	**392,002**	**–360,081**	**–4,905**	**–335**	**–714**	**32,256**
Balance at January 1, 2018		**6,289**	**392,002**	**–360,081**	**–4,905**	**–335**	**–714**	**32,256**
Net result		0	0	–54,186	0	0	0	**–54,186**
Other comprehensive result	24	0	0	0	2,230	0	–71	**2,159**
Total comprehensive result for the period		**0**	**0**	**–54,186**	**2,230**	**0**	**–71**	**–52,027**
Transactions for equity rights plans	20, 23	0	7,426	0	0	0	0	**7,426**
Capital increase from options exercise	12	4	13	0	0	0	0	**17**
Capital increase Polyphor	12	239	6,261	0	0	0	0	**6,500**
Capital increase Idorsia	12	1,000	13,540	0	0	0	0	**14,540**
Capital increase	12	3,133	20,367	0	0	0	0	**23,500**
Cost of issuance share capital	12	0	–3,248	0	0	0	0	**–3,248**
Change in treasury shares	12	0	–566	0	0	–569	0	**–1,135**
Balance at December 31, 2018		**10,665**	**435,795**	**–414,267**	**–2,675**	**–904**	**–785**	**27,829**

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 registered offices at Hohenrainstrasse 24 in 4133 Pratteln, Switzerland.

The consolidated financial statements were approved for publication by the Board of Directors (**Board**) on April 26, 2019. They are subject to approval by the Annual General Meeting of Shareholders (**AGM**) on May 28, 2019.

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 revaluation 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.

Material uncertainties and ability to continue operations

Santhera's cash and cash equivalents amounted to CHF 22.0 million as of December 31, 2018. Material uncertainties remain as to whether the Company's current funding is sufficient to support the going concern assumption for the next twelve months. The ability to continue as a going concern and to execute the Company's strategy depends on further funding to ensure the continuation of its operations through the next twelve months.

Santhera plans to file an application for Conditional Marketing Authorization (CMA) in Europe with additional clinical data from patients treated with idebenone for the treatment of DMD in the second quarter of 2019.

Furthermore, the Company raised CHF 7.1 million by a placement of 500,000 Shares from authorized capital in April 2019 and also established a new credit line in April 2019. The credit line can be drawn on, up to a maximum

amount of CHF 15 million, with principal and interest payment due at the end of 2019. Additionally, Santhera is considering various options including a capital increase and/or the monetization of certain assets.

Shareholders should note that whilst the Management and Board of Directors continue to apply best efforts to evaluate available options and take the steps described, there is no guarantee that any transaction can be realized or that such transaction would generate sufficient funds to finance further operations.

The Management and Board of Directors believe that the Company is prepared to secure additional funds needed (i.e., through further equity financing and/or monetizing certain assets) in order to operate its business as planned with the objective to meet all of its obligations for the next twelve months. Hence, the consolidated financial statements have been prepared on a going concern basis.

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, Pratteln, 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; Santhera (UK) Limited, London, United Kingdom; and Santhera Pharmaceuticals (Spain), S.L.U, Irun, Spain.

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.

IFRS 9 requires Santhera to measure all financial assets at amortized cost at the expected credit loss (ECL) on a 12-month or lifetime basis.

Changes in accounting policies

The adopted accounting policies are consistent with the previous year except for those described below.

IFRS standards effective with January 1, 2018

IFRS 9 Financial Instruments
This standard brings together all three aspects of the accounting for financial instruments: classification and measurement, impairment and hedge accounting. The application of the classification and measurement requirements of IFRS 9 had no material impact on the Group's equity and profit or loss. All assets and liabilities measures at amortized cost under IAS 39 continue to be measured at amortized cost under IFRS 9 and those liabilities classified as at fair value through profit or loss remain in the same measurement category under IFRS 9. Furthermore, the Group does not apply hedge accounting. IFRS 9 requires Santhera to record expected credit losses (**ECL**) on all of

its financial assets at amortized cost on a 12-month or lifetime basis. The Group applied the simplified approach and records lifetime expected losses on its trade receivables. Based on the nature of its receivables, the application of the impairment model under IFRS 9 had no material impact on the Group's balance sheet or equity. Consequently, no impact in equity was recorded on January 1, 2018. Comparative information has not been restated and continues to be measured and presented in accordance with IAS 39 and IFRS 7 effective until December 31, 2017.

IFRS 15 Revenue from Contracts with Customers

The Group applied, for the first time in 2018, IFRS 15 Revenue from Contracts with Customers. Under IFRS 15, revenue is recognized at an amount that reflects the consideration to which an entity expects to be entitled in exchange for transferring goods or services to a customer. The Group adopted IFRS 15 applying the modified retrospective approach and adopted IFRS 15 on January 1, 2018 without restating comparatives. Revenue from sales of products is recognized at the point in time when the customer obtains control of the goods or services, which for the Group is generally upon delivery at the customer. The adoption of IFRS 15 had no material impact on the Group's revenue and profit or loss. If Santhera had applied the previous accounting policies for revenue in 2018, the consolidated income statement and balance sheet would not be materially different.

Due to the limited impact of IFRS 15 and IFRS 9 Santhera has not reproduced the accounting policies for revenue and financial instruments applied until December 31, 2017. These are disclosed in the annual report 2017.

New, revised or amended IFRS standards and interpretations as from 2019 or later

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 (**ROU**) 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. Santhera has decided to apply the modified retrospective adoption method and that, as per January 1, 2019, it will only recognize leases with a term of more than 12 months on its balance sheet. Based on this date ROUs will be calculated by reference to the measurement of the lease liability. The Group will not recognize ROU assets of low value; payments for such leases continue to be recognized in the income statement.

The most significant impact of the new standard results from a recognition of a right-of-use asset and a lease liability for office space and leased cars currently treated as operating leases (see note 19 "*Commitments and Contingent Liabilities"* for a disclosure of non-cancellable operating lease commitments). The amounts to be capitalized under IFRS 16 as right-of-use asset and lease liability for the payment obligations will be measured by reference to the discounted future lease payments over the lease period, including reasonably certain to be exercised lease options. Furthermore, interest expense are recognized separately from operating expenses in profit or loss, which is the main impact on Santhera's income statement.

Santhera will apply the standard from its mandatory adoption date of January 1, 2019, using the modified retrospective method and will not restate comparative figures for the year prior to first adoption. During 2018, Santhera continued its assessment of potential impact on the balance sheet as of the transition date and expects to recognize right-of-use assets of approximately CHF 4.4 million and lease liabilities of approximately the same amount as of January 1, 2019.

The following new, revised or amended standards have been published but are not yet effective and have not been early adopted by the Group. They are not expected to have a significant impact on the consolidated financial statements.

- IFRIC 23 Uncertainty over Income Tax Positions (effective January 1, 2019)
- IFRS 9 Amendments to IFRS 9, Prepayment Features with negative Compensation (effective January 1, 2019)
- IAS 28 Amendments to IAS 28, Long-term Interests in Associates and Joint Ventures (effective January 1, 2019)
- IAS 19 Amendments to IAS 19, Plan Amendment, Curtailment or Settlement (effective January 1, 2019)
- Various Annual Improvements to IFRS Standards 2015–2017 Cycle. (effective January 1, 2019)
- Various Amendments to References to Conceptual Framework in IFRS Standards. (effective January 1, 2020)
- Amendment to IFRS 3: Definition of a Business (effective January 1, 2020)
- Amendments to IAS 1 and IAS 8: Definition of Material (effective January 1, 2020)
- IFRS 17 Insurance Contracts (effective January 1, 2021)

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, sub-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 life-time 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. The 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 based on expected credit losses.

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 subsequently measured at amortized cost

These are financial assets held to collect contractual cash flows representing principal and interest only. With the exception of trade receivables, they are initially measured at fair value plus transaction costs. Trade receivables are measured at the transaction price established in accordance with IFRS 15. Subsequent to initial recognition these financial assets are measured at amortized cost using the effective interest rate and are subject to impairment using the expected credit loss model.

Until December 31, 2017 Santhera applied an incurred loss model to determine its bad debt allowance.

They are included in current assets, except for maturities longer than 12 months after the balance sheet date. These are classified as noncurrent assets.

Financial assets at fair value through profit or loss

Santhera classifies all other financial assets at fair value through profit or loss. Principally, these are instruments held for trading. 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. 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.

Interest income

Interest income is recognized on a pro rata temporis basis 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 less transaction costs 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, Santhera reassesses unrecognized deferred tax assets and the carrying amount of deferred tax assets. Santhera 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 from contracts with customers is recognized at an amount that reflects the consideration to which Santhera expects to be entitled in exchange for transferring goods or services to a customer.

Net sales from the sale of products is recognized at the point in time when the customer obtains control of those products which is generally upon delivery at the customer. Revenue is net of value-added tax, rebates, discounts, returns and after eliminating intercompany sales.

Where revenue arrangements include variable consideration, such amounts are not included in the estimated transaction price unless it is highly probable that a significant reversal of the cumulative revenues recognized will not occur in future periods once the uncertainty related to the variable consideration is resolved. Payment terms usually range between 30 and 60 days for the sale of goods. Customer returns and variable consideration are not material.

Revenue from out-licensing, incl. revenue from royalties

Out-licensing agreements are concluded, where the counterparty has to pay license fees which are usually in the form of upfront and milestone payments as well as royalty payments. Santhera determines its performance obligations under such arrangements and in case of multiple deliverables allocates the transaction price to each distinct performance obligation on a relative stand-alone selling price basis. Typically, these arrangements include obligations such as maintenance of patents, research and development support and services, memberships in joint steering committees and other involvement in the arrangement, in which case the upfront and milestone payments may represent advance payments for future services and/or the right to access the underlying intellectual property of the Group.

Sales-based or usage-based royalties received in exchange for licenses of intellectual property are recognized as revenue at the later of when: (1) the subsequent sale or usage occurs; or (2) the performance obligation to which some or all of the sales-based or usage-based royalty has been allocated is satisfied (in whole or in part) where the license is the only or predominant item to which the royalty relates.

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 not yet available for use, see note 7 *"Impairment Test for Intangible Assets"*.
- Measurement and testing for net realizable value of inventory, see note 9 *"Inventories"*.
- Valuation of derivative financial instruments in connection with the convertible bonds, see note 15 "*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 20 "*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 24 *"Employee Expenses and Benefits"*.

4 Exchange Rates of Principal Currencies

	Income statement in CHF average rates		**Balance sheet in CHF** year-end rates	
	2018	**2017**	**2018**	**2017**
1 Euro (**EUR**)	1.1547	1.1114	1.1265	1.1691
1 US dollar (**USD**)	0.9779	0.9847	0.9850	0.9753
1 British pound (**GBP**)	1.3052	1.2683	1.2546	1.3173
1 Canadian dollar (**CAD**)	0.7547	0.7590	0.7236	0.7777

5 Tangible Assets

In CHF thousands	Equipment	IT hardware	Leasehold improvements	**2018**
Cost				
At January 1	**362**	**1,106**	**1,469**	**2,937**
Additions	538	38	147	**723**
Disposals	–15	–81	0	**–96**
Exchange differences	–6	–3	–1	**–10**
At December 31	**879**	**1,060**	**1,615**	**3,554**
Accumulated depreciation				
At January 1	**212**	**514**	**54**	**780**
Additions	106	251	248	**605**
Disposals	–15	–81	0	**–96**
Exchange differences	–3	–2	1	**–4**
At December 31	**300**	**682**	**303**	**1,285**
Net book value	**579**	**378**	**1,312**	**2,269**

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**

6 Intangible Assets

In CHF thousands	Option to Vamorolone sub-license (in process R&D) 2	POL6014 (in process R&D) [1]	Idebenone	Fipamezole	IT software/ patents	**2018**
Cost						
At January 1	**0**	**0**	**30,387**	**3,918**	**654**	**34,959**
Additions	34,780	6,210	0	0	54	**41,044**
Disposals	0	0	0	0	–42	**–42**
At December 31	**34,780**	**6,210**	**30,387**	**3,918**	**666**	**75,961**
Accumulated amortization						
At January 1	**0**	**0**	**7,091**	**3,918**	**390**	**11,399**
Additions	0	0	3,039	0	98	**3,137**
Disposals	0	0	0	0	–42	**–42**
At December 31	**0**	**0**	**10,130**	**3,918**	**446**	**14,494**
Net book value	**34,780**	**6,210**	**20,257**	**0**	**220**	**61,467**

[1] see note 13 "*Transaction with Polyphor*"

[2] see note 14 "*Transaction with Idorsia*"

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**

During 2018 there was no trigger for impairment of intangible assets in use (see to note 7 "*Impairment Test for Intangible Assets*" for a discussion on impairment testing performed on assets not yet available for use).. "Idebenone" represents the main intangible asset currently in use of Santhera. It has become available for use in September 2015 and has an estimated useful life of 10 years.

7 Impairment Test for Intangible Assets

Idebenone

During 2018 there was no trigger for impairment of intangible assets. "Idebenone" represents the intangible asset of Santhera which has become available for use in September 2015 and has an estimated useful life of 10 years.

POL6014 and Option to Vamorolone sub-license

"POL6014" and "Option to Vamorolone sub-license" are intangible assets which were added in 2018 (see note 13 "*Transaction with Polyphor*" and note 14 "*Transaction with Idorsia*"). They are not yet available for use. Therefore they are subject to an impairment test at least once annually or in case of triggers for impairment.
Management used a risk-adjusted Net Present Value (**rNPV**) model which contains several assumptions in order to verify the recoverable amount. This is a customary way for the valuation of pharmaceutical intangibles. The rNPV model considers mainly the period over the net cash flows of the development and use patent period of the products; hence the models contain data for the periods from 2019 through 2032 (for option to vamorolone sub-license) and 2037 (for POL6014). No terminal value was calculated since it is probable that after the exclusivity period the sales could decrease. For the purpose of estimating these cash flows as per December 31, 2018, Santhera made general estimates for:

WACC	10.4%
Tax rate	9.3%

Considering the markets and respective risk-profile for both assets, Santhera used the same WACC and tax rate assumptions for the impairment testing of POL6014 and the option to vamorolone sub-license. Other input elements for the calculation of the rNPV are based on the individual agreement with Polyphor or Idorsia such as the expected revenues based on estimated market size and patient numbers, expected market penetration rates, product pricing and project- or product-related costs. Probability of reaching the market lies between 22% (POL6014) and 27% (option to vamorolone sub-license), reflecting the uncertainty as to whether a final and successful market registration can be achieved for POL6014 and the sub-license vamorolone.

The impairment test of the recoverable amount of the intangible assets performed, as of December 31, 2018, did not result in the requirement to recognize an impairment. Santhera performed a sensitivity analysis taking into account reasonable changes in the assumptions used, such as changes in number of patients on drug, sales price and probability of reaching the market. The sensitivity analysis for 2018 did not reveal any indicators of impairment as at the reporting date.

8 Prepaid Expenses

In CHF thousands	**2018**	**2017**
Prepayments	969	853
Total at December 31	**969**	**853**

9 Inventories

In CHF thousands	2018	2017
Raw material (active pharmaceutical ingredients)	7,488	7,488
Semi-finished goods	628	2,335
Finished goods	1,166	324
Total at December 31	**9,282**	**10,147**

10 Trade and Other Receivables

In CHF thousands	2018	2017
Trade receivables (gross)	5,536	4,249
Other receivables	2,402	1,208
Expected credit losses / allowance for doubtful debts	**–77**	**–55**
Total at December 31	**7,861**	**5,402**

Trade receivables in 2018 result from product sales, see note 21 *"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, 2018, an allowance for expected credit losses (ECL) of TCHF 77 was recognized on the trade receivables (as of December 31, 2017: TCHF 55).

In CHF thousands	2018	2017
Expected credit losses January 1	**–55**	**0**
Allowance for doubtful debts	0	–55
Reversal of allowance for doubtful debts	55	0
Allowance for expected credit losses (ECL)	–77	0
Expired	0	0
Outstanding at December 31	**–77**	**–55**

11 Cash and Cash Equivalents and Restricted Cash

11.1 Cash and cash equivalents

In CHF thousands	**2018**	**2017**
Cash at banks and on hand		
In CHF	9,111	34,730
In EUR	10,149	8,152
In GBP	493	697
In USD	2,036	1,496
In CAD	138	120
Other currencies	44	0
Total at December 31	**21,971**	**45,195**
Of which: Short-term deposits		
In CHF	0	21,007

11.2 Restricted cash

in CHF thousands	**Dec. 31, 2018**	Dec. 31, 2017
Long-term	1,500	4,500
Short-term	3,000	3,000
Total at period end	**4,500**	**7,500**

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.

12 Share Capital

Ordinary share capital

As of January 1, 2017, the share capital amounted to CHF 6,279,857, divided into 6,279,857 shares ("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.

In February 2018, 238,924 Shares were issued in out of the authorized share capital in connection with the agreement with Polyphor (see note 13 *"Transaction with Polyphor"*). In November 2018, 1,000,000 Shares were issued out of the authorized share capital in connection with the first step of the agreement with Idorsia (see note 14 *"Transaction with Idorsia"*). In December 2018, 3,133,334 Shares were issued in connection with the second step of the agreement with Idorsia (see note 14 *"Transaction with Idorsia"*). During 2018 3,750 Shares were issued from

conditional capital upon the exercise of stock options. As a result, as of December 31, 2018, the share capital amounted to CHF 10,664,563, divided into 10,664,563 Shares at a nominal value of CHF 1 each.

On occasion of the Extraordinary General Meeting, held December 11, 2018, the shareholders approved the increase of the share capital by up to 5,000,000 Shares. On December 21, 2018, 3,133,334 Shares were used to increase the share capital accordingly.

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, 2018, Santhera held 53,290 treasury Shares (2017: 9,921 treasury Shares).

Authorized share capital

In February 2018, 238,924 Shares were issued out of the authorized share capital in connection with the agreement with Polyphor (see note 13 *"Transaction with Polyphor"*). On the occasion of the AGM on April 12, 2018, 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 April 11, 2020, through the issuance of up to 1,500,000 Shares with a nominal value of CHF 1 each. On November 21, 2018, 1,000,000 Shares were issued out of the authorized share capital in connection with the first step of the agreement with Idorsia (see note 14 *"Transaction with Idorsia"*). As a result, as of December 31, 2018, the Board is authorized to increase the share capital at any time until April 11, 2020, through the issuance of up to 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

As of December 31, 2018, the Company had a conditional share capital, pursuant to which the share capital may be increased by

- a maximum amount of CHF 687,552 (2017: CHF 691,302) through the issuance of up to 687,552 (2017: 691,302) Shares, under the exclusion of shareholders' pre-emptive rights, for equity rights being exercised under the Company's equity rights plans, see note 20 *"Equity Rights Plans"*, and
- a maximum amount of CHF 930,000 (2017: CHF 930,000) by issuing up to 930,000 (2017: 930,000) 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.

13 Transaction with Polyphor

On February 15, 2018, Santhera announced that it had 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. Under the terms of the agreement, Santhera may be required to make cash payments due to future development, regulatory and sales milestones of up to CHF 121 million (i.e. contingent payments). Consistent with existing licensing agreements, such contingent payments have not been capitalized.

The consideration for the acquisition of the license and the clinical material was paid by issuing 238,924 shares of Santhera's authorized share capital for a total amount of CHF 6.5 million (CHF 27.2053 per share; see note 12 *"Share Capital"*). Santhera acquired a license (POL6014) in the amount of CHF 6.2 million, which was recognized as an addition to the intangible assets (see note 6 *"Intangible Assets"*). The intangible asset is being developed and hence not yet available for use and not amortized. In addition to the license, the Group purchased clinical material in the amount of CHF 0.3 million, which was accounted for as a development expense. The amounts of the two parts were based on their relative fair values.

14 Transaction with Idorsia

On November 21, 2018, Santhera announced that it had entered into an agreement to acquire an option from Idorsia Ltd, Allschwil, Switzerland, for an exclusive sub-license of dissociative steroid vamorolone. Under the terms of the agreement, Idorsia will grant Santhera the option to obtain an exclusive sub-license for vamorolone in all indications and all territories except Japan and South Korea. In a first step Idorsia received as consideration for entering into the agreement 1,000,000 new registered shares from Santhera's existing authorized share capital. In a second step Idorsia received an upfront cash component of USD 20 million (CHF 20.2 million). Santhera may exercise the option upon receipt of data from the Phase IIb VISION-DMD study (VBP15-004) and following a one-time consideration to Idorsia of USD 30 million.

Following the exercise of the worldwide vamorolone license option by Idorsia and exercise of the vamorolone sub-license option for all territories worldwide except Japan and South Korea by Santhera, Santhera will pay to Idorsia regulatory and commercial milestone payments of up to USD 80 million in the DMD indication and four one-time sales milestone payments of up to USD 130 million in aggregate. Regulatory milestone payments by Santhera to Idorsia for three additional indications amount to up to USD 205 million in aggregate. Upon commercialization of vamorolone, Santhera has committed to pay tiered royalties ranging from a single-digit percentage to low double-digit percentage on the annual net sales of vamorolone to Idorsia.

The first step of the consideration for the agreement was paid by issuing shares of Santhera Pharmaceuticals Holding AG, while the second step was paid in cash. The valuation of the 1,000,000 shares amounts to CHF 14.6 million and the payment of USD 20 million converts into CHF 20.2 million. Therefore the consideration for the option to the sub-license is valued at CHF 34.8 million and recognized as an addition to intangible assets (see note 6 *"Intangible Assets"*). The intangible asset is being developed and hence not yet available for use and not amortized.

15 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.

15.1 Financial assets short-term

Financial assets (units in a fund) were held for trading and measured at fair value based on quoted prices (Level 1) through profit or loss. The instruments were sold in 2018, generating proceeds of CHF 12.7 million. A loss of TCHF 293 (financial expenses) resulted during the reporting period (2017: gain of TCHF 96).

15.2 Financial liabilities

On February 17, 2017, Santhera issued senior unsecured convertible bonds in the nominal amount of CHF 60 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 was fixed at CHF 86.4006 and was reset in accordance with the terms of the bond in February 2018 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. 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 fair value of the bond (Level 1) at December 31, 2018, amounts to CHF 41.7 million (2017: CHF 51.6 million).
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, 2018, was 71% (December 31, 2017: 87%).
The embedded conversion right and the reset mechanism are directly related and have the same risk exposure. Therefore, these two derivatives are accounted for as a single instrument (i.e. a compound derivative). Due to the reset mechanism, the compound derivative is not settled for a fixed number of equity and hence classifies as a financial liability.

The value of the derivatives, initially amounted to CHF 5.3 million (February 17, 2017). At December 31, 2017, the value was CHF 2.8 million and at December 31, 2018 CHF 0.2 million. The change in the fair value was recognized in financial income and amounted in 2018 to TCHF 2,588 (2017: TCHF 2,540).

Sensitivity analysis:

	December 31, 2018		December 31, 2017	
	Increase/decrease in volatility assumption	Effect on result before taxes in CHF thousands	Increase/decrease in volatility assumption	Effect on result before taxes in CHF thousands
Change in volatility	+5%	–55	+5%	175
	–5%	84	–5%	–181

Changes in liabilities arising from financing activities

In CHF thousands	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**
Change in fair value of derivative financial instruments	0	–2,588
Effective interest/amortized cost calculation	1,458	0
December 31, 2018	**54,569**	**204**

16 Deferred Taxes

Net deferred taxes recorded

In CHF thousands	2018	2017
Temporary differences on inventory	1,285	1,242
Deferred tax assets recognized	**1,285**	**1,242**
Temporary differences on intangible assets, net	1,657	1,905
Temporary differences on intercompany loans	13,449	13,449
Temporary differences on convertible bonds	409	321
Tax loss carryforwards	–15,515	–15,675
Deferred tax liabilities recognized	**0**	**0**
Tax loss carryforwards	343,389	339,492
Of which recorded	–187,824	–197,008
Of which unrecorded	**155,565**	**142,484**
Expiring in		
1 year	39,311	22,671
2 years	4,223	30,569
3 years	0	4,223
4 years	0	0
5 years	41,104	0
More than 5 years	41,624	54,552
Without expiration	29,303	30,469
Total unrecorded tax loss carryforwards	**155,565**	**142,484**

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 7,983 at December 31, 2018, and TCHF 8,375 at December 31, 2017, respectively).

17 Trade and Other Payables

In CHF thousands	2018	2017
Trade payables	7,194	3,585
Other payables (nonfinancial)	1,112	1,149
Total at December 31	**8,306**	**4,734**

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

18 Accrued Expenses

In CHF thousands	2018	2017
Development programs	4,046	1,547
Liabilities to employees	3,665	3,429
Accruals for pricing and reimbursement	501	839
Accrued marketing and sales expenses	505	469
Accruals for audit, consulting and other	1,207	688
Accruals for interest expenses	1,117	1,108
Total at December 31	**11,041**	**8,080**

19 Commitments and Contingent Liabilities

Commitments

Commitments for operating lease (noncancellable)

In CHF thousands	2018	2017
Within 1 year	1,200	1,176
After 1 year through to 5 years	1,426	1,177
After 5 years	0	34
Total at December 31	**2,626**	**2,387**

Commitments to future payments under license agreements

Option to sub-license with Idorsia

On November 21, 2018, Santhera announced that it had entered into an agreement to acquire an option from Idorsia Ltd, Allschwil, Switzerland, for an exclusive sub-license of dissociative steroid vamorolone. Under the terms of the agreement, Idorsia will grant Santhera the option to obtain an exclusive sub-license for vamorolone in all indications and all territories except Japan and South Korea. Santhera may exercise the option upon receipt of data from the Phase IIb VISION-DMD study (VBP15-004) and following a one-time consideration to Idorsia of USD 30 million.
Following the exercise of the worldwide vamorolone license option by Idorsia and exercise of the vamorolone sub-license option for all territories worldwide except Japan and South Korea by Santhera, Santhera will pay to Idorsia regulatory and commercial milestone payments of up to USD 80 million in the DMD indication and four one-time sales milestone payments of up to USD 130 million in aggregate. Regulatory milestone payments by Santhera to Idorsia for three additional indications amount to up to USD 205 million in aggregate. Upon commercialization of vamorolone, Santhera has committed to pay tiered royalties ranging from a single-digit percentage to low double-digit percentage on the annual net sales of vamorolone to Idorsia.

License agreement with Polyphor

On February 15, 2018, Santhera announced that it had 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. Under the terms of the agreement, Santhera may be required to make cash payments due to future development, regulatory and sales milestones of up to CHF 121 million (i.e. contingent payments). Consistent with existing licensing agreements, such contingent payments have not been capitalized.

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 in-licensed 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 sub-licenses 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 and technical development services. Commitments are within current market prices and can be terminated at the Company's discretion.
In connection with its clinical studies, Santhera entered into commitments for the purchase of material in the amount of up to CHF 3.3 million (to be delivered in 2019).

Contingent liabilities

Santhera believes that the accruals (see note 18 *"Accrued Expenses"*) are adequately 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.

20 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.

20.1 Stock Option Plans

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. 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						**2018**
	At January 1	Exercised	Granted	Forfeited	Expired	At December 31
ESOP 2010	29,551	–3,750	0	0	0	25,801
ESOP 2015	245,329	0	0	–21,855	0	223,474
Total	**274,880**	**–3,750**	**0**	**–21,855**	**0**	**249,275**

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**

Board Stock Option Plans

The Company adopted the BSOP 2015 (collectively the **BSOP**) to provide incentives to members of the Board. The BSOP contains 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 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						**2018**
	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						**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**

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 20.2 *"Share Appreciation Rights Plans".*

Number of stock options outstanding and exercisable

Number of options	**2018**	**2017**
Outstanding at January 1	**288,442**	**313,365**
Granted	0	0
Exercised[1]	–3,750	–8,698
Forfeited	–21,855	–15,472
Expired	0	–753
Outstanding at December 31	**262,837**	**288,442**
Exercisable at December 31	**190,188**	**102,642**

[1] The average closing share price of options exercised during the reporting period 2018 was CHF 7.30 (2017: CHF 39.05).

The value of stock options granted is recognized as personnel expense over the period Santhera receives services. In 2018, previously granted stock options resulted in personnel expenses of TCHF 1,077 (TCHF 127 related to Development, TCHF 594 related to Marketing and sales (**M&S**) and TCHF 356 to General and administrative (**G&A**)) and in 2017, such grants resulted in personnel expenses of TCHF 2,778 (TCHF 374 related to Development, TCHF 1,525 related to M&S and TCHF 879 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)	**2018 Number exercisable**	Number outstanding	Weighted average remaining contractual life (years)	**2017 Number exercisable**
from 3.85 to 4.53	21,251	4.20	21,251	25,001	5.13	25,001
at 22.25	4,550	5.50	4,550	4,550	6.50	3,275
at 69.30	13,150	7.25	7,400	14,800	8.25	0
from 82.10 to 114.50	223,886	6.61	156,987	244,091	7.73	74,366
Total	**262,837**	**6.37**	**190,188**	**288,442**	**7.61**	**102,642**

20.2 Share Appreciation Rights Plans

Starting with July 1, 2016, Santhera switched from stock option plans to Share Appreciation Rights Plans (**SARP**). It introduced Board Share Appreciation Plans (**BSARP**), the BSARP 2016, the BSARP 2017, for the members of its Board and Employee Share Appreciation Rights Plans (**ESARP**), the ESARP 2016 and the ESARP 2017, for the Executive Management, employees and consultants. Share appreciation rights (**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). In January 2018, Santhera has introduced ESARP 2018 in order to provide a special grant for the Executive Management and employees. Besides the usual terms, this grant contains an additional vesting condition, which is based on Santhera obtaining a positive opinion of the Committee for Medicinal Products for Human Use (**CHMP**) with respect to the marketing authorization of idebenone for the treatment of patients with DMD in the European Union (**EU**). 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						**2018**
	At January 1	Exercised	Granted	Forfeited	Expired	At December 31
ESARP 2016	85,589	0	0	–42,277	0	43,312
BSARP 2017	15,120	0	62,659	0	0	77,779
ESARP 2017	259,401	0	479,751	–149,907	0	589,245
ESARP 2018	0	0	79,872	–59,820	0	20,052
Total	**360,110**	**0**	**622,282**	**–252,004**	**0**	**730,388**

Number of SARs						**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**

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:

	2018	**2017**
Market price of stock	CHF 5.80 to 41.15	CHF 29.00 to 82.00
Exercise prices	CHF 16.20 to 36.70	CHF 38.60 to 77.80
Weighted average fair value of SAR granted	CHF 12.11	CHF 25.94
Expected volatility[1]	37%	38%
CHF risk-free interest rate	0.0 to 0.05% 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	2018	2017
Outstanding at January 1	**360,110**	**56,581**
Granted	622,282	350,243
Exercised	0	0
Forfeited	–252,004	–46,714
Expired	0	0
Outstanding at December 31	**730,388**	**360,110**
Exercisable at December 31	**155,424**	**0**

The value of SAR granted is recognized as personnel expense over the period Santhera receives services. In 2018, SAR grants resulted in personnel expenses of TCHF 5,641 (TCHF 2,178 related to Development, TCHF 1,957 related to M&S and TCHF 1,506 to G&A) and in 2017, such 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). The above expenses of TCHF 5,641 are net of a reversal for SAR forfeited under ESARP 2018 in the amount of TCHF 525 (TCHF 180 related to Development, TCHF 185 related to M&S and TCHF 160 to G&A).

In the course of allocating SAR annually to its employees as part of the long-term incentive (**LTI**), Santhera plans to grant up to 768,000 SAR after the Annual General Meeting (**AGM**) which is to be held in May 2019. These SAR form part of the long-term incentive (**LTI**) award to employees for the year ended December 31, 2018. Although these SAR were not legally granted in 2018, Executive Management considers it appropriate to recognize expenses in 2018 as employees have been rendering services in 2018 in expectation of the annual LTI allocation. Personnel expenses in 2018 for this amounted to TCHF 708 (TCHF 298 related to Development, TCHF 207 related to M&S and TCHF 203 related to G&A) based on an estimate of fair value (in 2017 personnel expenses 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)). 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 May 28, 2019. After the AGM the grant date fair value of the SAR will be determined and the cumulative expense and number of SAR to be granted adjusted.

Terms of SAR outstanding at December 31

Exercise price range for SAR (in CHF)	Number outstanding	Weighted average remaining contractual life (years)	**2018 Number exercisable**	Number outstanding	Weighted average remaining contractual life (years)	**2017 Number exercisable**
from 16.20 to 18.90	67,659	9.32	0	0	0	0
from 36.70 to 38.70	402,999	9.00	11,087	33,257	9.76	0
from 51.75 to 54.85	232,458	8.00	132,921	283,435	9.01	0
from 76.50 to 77.80	27,272	8.08	11,416	43,418	9.08	0
Total	**730,388**	**9.04**	**155,424**	**360,110**	**9.04**	**0**

21 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	2018	2017
EU	31,544	22,859
Rest of the world	113	84
Total	**31,657**	**22,943**

In 2018, net sales amounted to CHF 31.7 million with its product Raxone only (2017: CHF 22.9 million). Raxone was sold in 20 European countries, with the majority of sales reached in France and Germany (2017: 20 European countries).

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

In CHF thousands	2018	2017
Switzerland	63,504	25,451
EU	164	171
North America	68	95
Total	**63,736**	**25,717**

22 Other Operating Income

This position consists primarily of reimbursements and income from scientific programs.

23 Operating Expenses by Nature

In CHF thousands	2018	2017
External Development expenses	–24,208	–14,762
Patent and license expenses	–621	–381
Marketing expenses	–10,113	–13,018
Employee expenses	–36,125	–35,488
Of which non-cash-relevant expenses for equity rights plans	*–7,426*	*–9,687*
Other administrative expenses	–5,415	–4,727
Depreciation and amortization	–703	–343
Lease expenses	–1,304	–780
Other operating expenses	–198	–64
Total operating expenses	**–78,687**	**–69,563**

24 Employee Expenses and Benefits

Employee expenses

In CHF thousands	2018	2017
Wages and salaries	–20,926	–18,372
Social security and other personnel-related expenses[1]	–7,773	–7,429
Of which non-cash-relevant adjustments of pension fund	*–1,838*	*–2,021*
Expenses for equity rights plans	–7,426	–9,687
Total employee costs	**–36,125**	**–35,488**

Average number of full-time equivalents[2]	**109.6**	**92.9**
Full-time equivalents at year-end	**114.9**	**106.2**
Total headcount at year-end	**119**	**112**

1 Thereof TCHF 458 were expensed for defined contribution plans in North America and some European countries (2017: TCHF 306).

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, Pratteln, 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	**2018**	**2017**
Present value of obligation, January 1	**24,152**	**21,279**
Current employer service cost	2,302	1,614
Past service cost[1]	702	1,240
Interest cost	181	138
Employee contributions	824	514
Benefits paid / transfer payments	–2,375	–620
Insurance premiums	–179	–266
Remeasurements[2]	–2,332	253
Present value of obligation, December 31	**23,275**	**24,152**

1 Increase of obligation due to higher savings contributions and changes in the conversion rates for the over-mandatory part of the retirement capital.

2 Details of remeasurements:

In CHF thousands	**2018**	**2017**
Effect of changes in demographic assumptions	0	0
Actuarial gain/loss due to changes in financial assumptions	–1,206	–400
Actuarial gain/loss due to experience adjustments	–1,126	653
Subtotal gain/loss	**–2,332**	**253**
Return/loss on plan assets (excluding interest income)	102	–82
Total remeasurements in other comprehensive income gain/loss	**–2,230**	**171**

Changes in plan assets

In CHF thousands	2018	2017
Fair value of assets, January 1	**15,777**	**15,096**
Interest income on assets	114	101
Employer contributions	1,233	870
Employee contributions	824	514
Benefits paid / transfer payments	–2,375	–620
Insurance premiums	–179	–266
Remeasurements (return/loss on plan assets (excluding interest income))	–102	82
Fair value of assets, December 31	**15,292**	**15,777**

Net defined benefit asset/obligation

In CHF thousands	2018	2017
Present value of obligation, December 31	23,275	24,152
Fair value of assets, December 31	15,292	15,777
Net defined asset/obligation	**–7,983**	**–8,375**

Asset allocation

In CHF thousands	2018	2017
Cash	107	0
Debt instruments	6,881	0
Equity instruments	4,557	0
Property	2,906	0
Assets from insurance contracts	0	15,777
Others	841	0
Total value of assets	**15,292**	**15,777**

For 2017 an asset breakdown is not available since AXA fully insured them as an insurance contract.

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

In %	2018	2017
Discount rate	1.00	0.70
Expected future salary increases	1.50	1.50

Sensitivity analysis for 2018:

In CHF thousands	Defined benefit obligation		Gross (net) service cost	
	Increase assumption	Decrease assumption	Increase assumption	Decrease assumption
Discount rate +/–0.25%	–941	1,017	–154	165
Salary increase +0.25%	223	–	0	–
Life expectancy +1 year	394	–	40	–

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	–

Mortality rate:

Life expectancy at age 65 (in years)	**2018**	**2017**
Male	22.6	22.5
Female	24.7	24.5

The expected employer contributions for fiscal year 2019 amount to approximately TCHF 1,193 (2018: TCHF 936). Benefit obligations of pensioners amounted to TCHF 2,988 at December 31, 2018 (2017: TCHF 3,103). The duration of the plan liabilities calculated is 20.9 years as of December 31, 2018 (2017: 20.3 years).

25 Financial Income/Expenses

Financial income

In CHF thousands	**2018**	**2017**
Interests on cash and cash equivalents	1	5
Change in fair value of financial derivative instruments	2,588	2,540
Income from financial assets	92	267
Realized and unrealized foreign exchange gains	1,690	1,322
Total	**4,371**	**4,134**

Financial expenses

In CHF thousands	2018	2017
Interest expenses	–4,501	–3,843
Expenses from financial assets	–429	–197
Realized and unrealized foreign exchange losses	–1,885	–915
Total	**–6,815**	**–4,955**

26 Income Taxes

In CHF thousands	2018	2017
Current income tax income/expense	–365	–390
Deferred tax income/expense	43	133
Total	**–322**	**–257**

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

In CHF thousands	2018	2017
Result before taxes	–53,864	–51,275
Tax expense/income at expected group tax rate of 9.3%	5,009	4,769
Effect of tax rate difference group versus local	–417	–422
Effect of nondeductible expenses	–1,382	–784
Prior year DTA (deferred tax assets) decrease	0	–289
Utilization of previously unrecognized tax losses	19	24
Recognition of previously unrecognized DTL (deferred tax liabilities)	0	0
Recognition of DTA on previously unrecognized tax losses	0	0
Unrecognized deferred taxes	–3,551	–3,555
Effective tax income/expense	**–322**	**–257**

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.

27 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).

	2018	2017
Net result attributable to shareholders (in TCHF)	–54,186	–51,532
Weighted average number of shares issued and outstanding	6,891,610	6,269,813
Basic and diluted net result per share (in CHF)	**–7.86**	**–8.22**

For the years ended December 31, 2018 and 2017, 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.

28 Related Party Transactions

Board and Executive Management compensation

Total compensation of Board and Executive Management

In CHF thousands	2018	2017
Compensation, wages and salaries	2,859	3,344
Post-employment benefits (pension fund and defined benefit contributions)	353	272
Share-based payment expenses (fair value according to IFRS 2)	2,333	3,826
Total	**5,545**	**7,442**

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 2018, no stock options were exercised by members of the Board (2017: no stock options exercised). During 2018, 3,750 stock options were exercised by the Executive Management (2017: 5,000 stock options exercised).

29 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, 2018 and 2017.

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		
2018	+5%	+488
	–5%	–488
2017	+5%	+411
	–5%	–411

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 2018, variances of +/–50 basis points were calculated, resulting in fluctuations of +/–TCHF 132 before tax (end of 2017: +/–50 basis points resulting in fluctuations of +/–TCHF 263 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. Santhera estimates its expected credit losses (ECL) based on default probabilities and the ageing of outstanding invoices.

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 15 *"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, 2018 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	67,500	70,500	54,569
Trade payables	0	7,194	0	0	7,194	7,194
Accrued expenses	0	7,738	0	0	7,738	7,738
Total	**0**	**16,432**	**1,500**	**67,500**	**85,432**	**69,501**

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**

Categories of financial instruments

Year ended December 31, 2018 (IFRS 9 measurement categories) In CHF thousands	Book value	Financial assets at amortized cost	Other liabilities at amortized cost	At fair value through profit or loss
Assets				
Financial assets long-term	690	690	0	0
Restricted cash long-term	1,500	1,500	0	0
Trade receivables	5,459	5,459	0	0
Other receivables	98	98	0	0
Financial assets short-term	0	0	0	0
Restricted cash short-term	3,000	3,000	0	0
Cash and cash equivalents	21,971	21,971	0	0
Total	**32,718**	**32,718**	**0**	**0**
Liabilities				
Convertible bonds	54,569	0	54,569	0
Derivative financial instruments	204	0	0	204
Trade payables	7,194	0	7,194	0
Accrued expenses	7,738	0	7,738	0
Total	**69,705**	**0**	**69'501**	**204**

Year ended December 31, 2017 (IAS 39 measurement categories) 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**

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, 2015 and 2018, 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.

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

30 Events after the Reporting Date

On April 3, 2019, Santhera announced that it has taken out a syndicated credit line which, subject to certain conditions, will provide up to CHF 15.0 million over a time period of nine months.

On April 4, 2019, Santhera announced that it has placed all of the remaining 500,000 registered shares from its existing authorized share capital in a private placement, resulting in aggregate gross proceeds of CHF 7.1 million. Following completion of the placement, Santhera's share capital amounts to CHF 11,164,563, divided into 11,164,563 registered shares with a nominal value of CHF 1 each.

Ernst & Young Ltd
Aeschengraben 9
P.O. Box
CH-4002 Basle

Phone +41 58 286 86 86
Fax +41 58 286 86 00
www.ey.com/ch

To the General Meeting of
Santhera Pharmaceuticals Holding Ltd, Pratteln

Basle, April 26, 2019

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, 2018 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 20 to 63) give a true and fair view of the consolidated financial position of the Group as at December 31, 2018, 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.

Material uncertainty related to going concern

We draw attention to note 2 of the consolidated financial statements, which indicates that the Group's ability to continue operations as planned for the next twelve months depends on further funding. This fact together with other matters disclosed in note 2 indicates that a material uncertainty exists that may cast significant doubt about the Group's ability to continue as a going concern. Our opinion is not modified in respect of this matter.

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. In addition to the matters described in the *Material uncertainty related to going concern* section of our report, we have determined the matters described below to be the key audit matters to be communicated in our report. 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.

Accounting for board and employee share-based payment arrangements

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 2018 expenses recorded for equity rights plans amounted to CHF 7.4 million compared to CHF 9.7 million in the same period in 2017. *Refer to note 20 related to equity rights plans.*
Our audit response	We tested the determination of fair value for all grants made in 2018 and assessed the accuracy of the share-based payment expenses recognized. We obtained and read documentation related to new share appreciation rights plans established during financial year 2018 to understand the terms of the grants. Furthermore, we inspected new grants on a sample basis and referenced the grants to supporting documentation such as the communication to the employees. We reconciled the number of awards granted to the calculation of the expenses and recalculated the amounts to be recognized over the vesting period. Additionally, 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. Lastly, we also considered the adequacy of the disclosures made in relation to share-based payments. Our audit procedures did not lead to any reservations regarding the accounting for board and employee share-based payment arrangements.

Impairment assessment of intangible assets not yet available for use

Areas of focus	As a result of the transactions with Polyphor and Idorsia in 2018, the Group capitalized intangible assets not yet available for use in the amount of CHF 41.0 million. Based on the requirements of IAS 36, such intangible assets need to be tested for impairment at least annually. The impairment assessment of the intangible assets not yet available for use is a key audit matter based on the magnitude of the balances and the inherent judgement in the respective model and assumptions used as part of management's impairment assessment, specifically those related to timing and magnitude of future cash flows and to the determination of the respective discount rate. *Refer to note 2 "Summary of Significant Accounting Policies", note 3 "Critical Accounting Estimates, Assumptions and Judgments", and note 7 "Impairment Test for Intangible Assets".*

Our audit response

We evaluated the Company's valuation model for the intangible assets not yet available for use and analyzed the underlying key assumptions and discount rates, including risk adjustments for the probabilities of development success. We assessed the assumptions regarding future revenues and margins and we evaluated sensitivity in the valuation resulting from changes to the key assumptions applied. With respect to the discount rates applied, we evaluated the reasonableness of the discount rates determined by management by assessing the cost of capital for the Company and comparable organizations, as well as considering territory specific factors.
Our audit procedures did not lead to any reservations regarding the measurement of intangible assets not yet available for use.

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	2018	2017
Assets			
Cash and cash equivalents		7,348	30,738
Financial assets short-term		0	13,011
Other receivables from third parties		77	54
Prepaid expenses and accrued income		141	153
Restricted cash short-term		3,000	3,000
Current assets		**10,566**	**46,956**
Loans to shareholdings	3.1	118,451	63,293
Investments in shareholdings	3.2	246	198
Restricted cash long-term		1,500	4,500
Noncurrent assets		**120,197**	**67,991**
Total assets		**130,763**	**114,947**
Liabilities and equity			
Trade accounts payable to third parties		1,386	311
Other accounts payable to third parties		230	35
Accrued expenses		1,768	1,538
Current liabilities		**3,384**	**1,884**
Senior unsecured convertible bonds[1]	2	60,000	60,000
Noncurrent liabilities		**60,000**	**60,000**
Total liabilities		**63,384**	**61,884**
Share capital	3.3	10,665	6,289
Reserves from capital contributions[2]		*17,581*	*7,450*
Other capital reserves		*6,165*	*2,916*
Statutory capital reserves		23,746	10,366
Accumulated result		*–23,623*	*–13,752*
Results carried forward		*–13,752*	*–6,451*
Net result for the period		*–9,871*	*–7,301*
Other voluntary reserves (free reserves)		*57,495*	*50,495*
Voluntary accumulated result and other reserves		33,872	36,743
Treasury shares	3.4	–904	–335
Total equity		**67,379**	**53,063**
Total liabilities and equity		**130,763**	**114,947**

1 Interest bearing

2 Value as per December 31, 2018, to be confirmed by Swiss Federal Tax Administration (SFTA).

Income Statement

For the year ended December 31, in CHF thousands	Notes	2018	2017
Income from shareholdings	3.5	1,218	1,325
Other operating income		0	11
Total operating income		**1,218**	**1,336**
General and administrative expenses	3.6	–6,243	–5,407
Employee expenses		–1,139	–1,122
Other operating expenses		–20	–12
Total operating expenses		**–7,402**	**–6,541**
Operating result		**–6,184**	**–5,205**
Financial income		370	1,011
Financial expenses		–4,105	–3,189
Financial result		**–3,735**	**–2,178**
Reversal on allowance of investment		49	82
Result before and after taxes / net result		**–9,870**	**–7,301**
Direct taxes		0	0
Net result		**–9,870**	**–7,301**

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 registered offices at Hohenrainstrasse 24 in 4133 Pratteln, Switzerland.

Material uncertainties and ability to continue operations

Cash and cash equivalents of Santhera Group amounted to CHF 22.0 million as of December 31, 2018. Material uncertainties remain as to whether the Company's current funding is sufficient to support the going concern assumption for the next twelve months. The ability to continue as a going concern and to execute the Company's strategy depends on further funding to ensure the continuation of its operations through the next twelve months.

Santhera plans to file an application for Conditional Marketing Authorization (CMA) in Europe with additional clinical data from patients treated with idebenone for the treatment of DMD in the second quarter of 2019.

Furthermore, the Company raised CHF 7.1 million by a placement of 500,000 Shares from authorized capital in April 2019 and also established a new credit line in April 2019. The credit line can be drawn on, up to a maximum amount of CHF 15 million, with principal and interest payment due at the end of 2019. Additionally, Santhera is considering various options including a capital increase and/or the monetization of certain assets.

Shareholders should note that whilst the Management and Board of Directors continue to apply best efforts to evaluate available options and take the steps described, there is no guarantee that any transaction can be realized or that such transaction would generate sufficient funds to finance further operations.

The Management and Board of Directors believe that the Company is prepared to secure additional funds needed (i.e., through further equity financing and/or monetizing certain assets) in order to operate its business as planned with the objective to meet all of its obligations for the next twelve months. Hence, the financial statements have been prepared on a going concern basis.

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, 2018: CHF 290.8 million; December 31, 2017: CHF 235.6 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, 2018, Executive Management concluded that approximately 41% 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 2018 and 2017, the following companies are direct subsidiaries of Santhera Pharmaceuticals Holding AG (100% ownership and 100% voting rights):

	Share capital at December 31	**2018**	**2017**
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	2018	2017
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
Santhera Pharmaceuticals (Spain), S.L.U Irun, Spain	EUR	50,000	n/a

3.3 Share capital

During 2018, the share capital was increased by a total amount of CHF 4,376,008 to CHF 10,664,563 as of December 31, 2018 (2017: CHF 6,288,555): The increase consisted of three parts: 1) an increase through the exercise of 3,750 employee stock options (from conditional capital); 2) an increase through the issuance of 238,924 Shares (from authorized capital) in connection with Polyphor; 3.1) an increase through the issuance of 1,000,000 Shares (from authorized capital) in connection with Idorsia (step 1) and 3.2) an increase through the issuance of 3,133,334 Shares (from the Extraordinary General Meeting) in connection with Idorsia (step 2).
On occasion of the Extraordinary General Meeting, held December 11, 2018, the shareholders approved the increase of the share capital by up to 5,000,000 Shares. On December 21, 2018, 3,133,334 Shares were used to increase the share capital accordingly.

3.4 Treasury shares

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

	No of Shares	TCHF
January 1, 2017	**3,616**	**172**
Purchase	180,083	9,567
Sale	–173,778	–9,404
December 31, 2017	**9,921**	**335**
Purchase	157,436	3,413
Sale	–114,067	–2,844
December 31, 2018	**53,290**	**904**

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	2018	2017
Administrative expenses	1,268	1,220
Consulting expenses	1,727	1,456
Expenses in connection with convertible bonds	0	2,731
Expenses in connection with capital increase	3,248	0
Total	**6,243**	**5,407**

4 Other Information

4.1 Full-time equivalents

The number of full-time equivalents at period end was not above 10 in 2018 and 2017.

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, 2018: 10,660,813 shares (December 31, 2017: 6,279,857 shares):

	2018 Shares	2018 %	2017 Shares	2017 %
Idorsia Pharmaceuticals Ltd., Switzerland	1,333,333	12.5	n/a	n/a
Bertarelli Ernesto, Guichard-Bertarelli Donata and Bertarelli Maria-Iris, Switzerland	759,371	7.1	545,777	8.7
Iglu Group, Switzerland	422,500	4.0	557,350	8.9
JPMorgan Chase & Co., US[1]	278,620	2.6	n/a	n/a
Roderick Wong (RTW Master Fund, LTD, US)	209,841	2.0	315,339	5.0
Norges Bank (the Central Bank of Norway)[2]	128,398	1.2	214,258	3.4
Polyphor AG, Switzerland	110,200	1.0	n/a	n/a
The Goldman Sachs Group, Inc., Corporation Trust Centre[3]	10	0	457,309	7.3

[1] 278,620 shares and rights convertible into 229,089 shares

[2] Purchase positions in connection with securities lending transactions

[3] Purchase positions in connection with securities lending transactions

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, 2018:

	Number of Shares	Number of vested equity rights	Number of un-vested equity rights	Total number of equity rights
Board of Directors				
Elmar Schnee, Chairman	12,000	2,244	20,833	23,077
Martin Gertsch, Vice-Chairman	38,109	5,970	21,679	27,649
Philipp Gutzwiller, Director	7,100	1,581	14,659	16,240
Thomas Meier, Director		--- See below ---		
Patrick Vink, Director	1,000	1,662	18,225	19,887
Executive Management				
Thomas Meier, CEO	79,402	20,670	38,366	59,036
Günther Metz, Head Business Development	0	23,201	23,956	47,157
Christoph Rentsch, Chief Financial Officer	0	22,979	33,920	56,899
Kristina Sjöblom Nygren, Chief Medical Officer & Head Development	0	0	36,937	36,937
Giovanni Stropoli, Chief Commercial Officer Europe and Rest of World, until September 30, 2018 [1]	250	20,751	0	20,751
Oliver Strub, General Counsel and Secretary to the Board	0	15,478	24,492	39,970

[1] Number of Shares as of March 31, 2018

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	75,652	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 & 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

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

	2018 Quantity	**2018** Value (in TCHF)[1]	**2017** Quantity	**2017** Value (in TCHF)[1]
Board of Directors	62,659	463	17,913	524
Executive Management	139,194	1,266	104,033	3,686
Employees of Santhera Group	420,429	5,809	228,297	4,876
Total	**622,282**	**7,538**	**350,243**	**9,086**

[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 20 *"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.

After the Annual General Meeting to be held in May 2019, it is planned to grant up to 768,000 share appreciation rights (**SAR**s) to employees of Santhera. These SARs are part of the bonus award for the year 2018 to employees of the Group. These SARs will be granted under ESARP 2016 (see note 20 *"Equity Rights Plans"*).

	Quantity	Value (in TCHF)[1]
Executive Management	157,600	437
Employees of Santhera Group	610,400	1,584
Total	**768,000**	**2,021**

[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 20 *"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 2020.

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 April 3, 2019, Santhera announced that it has taken out a syndicated credit line which, subject to certain conditions, will provide up to CHF15.0 million over a time period of nine months.

On April 4, 2019, Santhera announced that it has placed all of the remaining 500,000 registered shares from its existing authorized share capital in a private placement, resulting in aggregate gross proceeds of CHF 7.1million. After completion of the placement, Santhera's share capital will amount to CHF 11,164,563, divided into 11,164,563 registered shares with a nominal value of CHF 1 each.

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	2018	2017
Result carried forward	**–13,751,583**	–6,451,188
Net result of the year	–9,870,826	–7,300,395
Accumulated result	**–23,622,409**	**–13,751,583**
Result to be carried forward	**–23,622,409**	**–13,751,583**

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 after Annual General Meeting (April 12th, 2018)	449,947
Share premium from option exercise during 2018	13,069
Share premium of capital increase December 2018	17,118,176
Reserves from capital contribution	**17,581,192**
Transfer from reserves from capital contribution to other voluntary reserves (free reserves)	–17,500,000
Reserves from capital contribution	**81,192**

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

	In CHF
Other capital reserves at December 31, 2018	6,165,107
Transfer from reserves from capital contribution to other voluntary reserves (free reserves)	–3,000,000
Other capital reserves	**3,165,107**

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

	In CHF
Other voluntary reserves (free reserves) after Annual General Meeting (April 12th, 2018)	**57,494,714**
Transfer from reserves from capital contribution	17,500,000
Transfer from other capital reserves	3,000,000
Free reserves	**77,994,714**

Ernst & Young Ltd
Aeschengraben 9
P.O. Box
CH-4002 Basle

Phone +41 58 286 86 86
Fax +41 58 286 86 00
www.ey.com/ch

To the General Meeting of
Santhera Pharmaceuticals Holding Ltd, Pratteln

Basle, April 26, 2019

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 69 to 78), for the year ended December 31, 2018.

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, 2018 comply with Swiss law and the company's articles of incorporation.

Emphasis of matter

We draw attention to note 1 of the financial statements, which indicates that the Company's ability to continue operations as planned for the next twelve months depends on further funding. This fact together with other matters disclosed in note 1 indicates that a material uncertainty exists that may cast significant doubt about the Company's ability to continue as a going concern. Our opinion is not modified in respect of this matter. Should the going concern assumption no longer be appropriate, the financial statements would have to be prepared based on liquidation values. In this case, a serious concern of over-indebtedness in the sense of article 725 para. 2 CO would exist and the relevant provisions would have to be complied with.

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. In addition to the matters described in the *Emphasis of matter* section of our report, we have determined the matters described below to be the key audit matters to be communicated in our report. 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.

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 related to the investment in and the long-term receivables from shareholdings.*
Our audit response	We evaluated management's impairment assessment, which is based on an income approach, and analyzed 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. Our audit procedures did not lead to any reservations regarding the valuation of investments and long-term receivables from shareholdings.

Building a better working world

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 2018, as well as shareholdings of the members of the Board and EM members.

Compensation Governance

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 is to assist the Board with the:

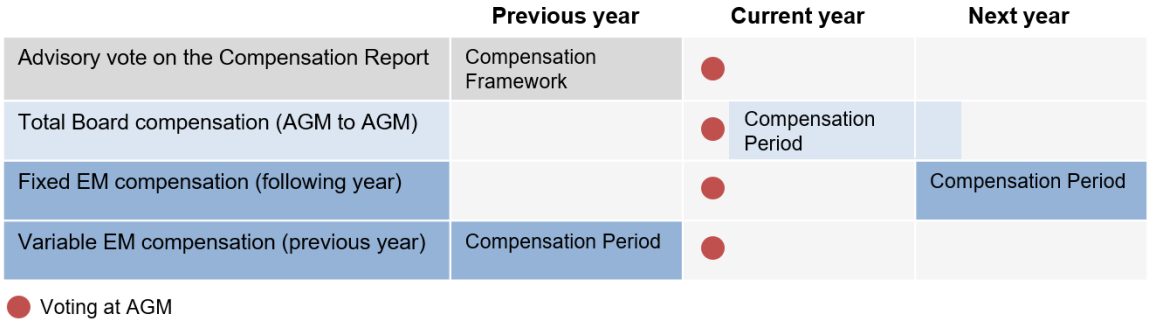
- Determination and review of remuneration policies and guidelines.
- Determination and review of performance objectives.
- Proposals to the AGM concerning the compensation of the Board and of EM.
- Resolution of other compensation related matters.

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 AGM 2019

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

1. Consultative vote on the Compensation Report 2018.
2. Board

   The maximum total amount of the fixed compensation for the period between the AGM 2019 and the AGM 2020.
3. EM

   3.1. The maximum total amount of the fixed compensation for the period from January 1, 2020 to December 31, 2020.

   3.2. The maximum total amount of the variable compensation for the period from January 1, 2018 to December 31, 2018.

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

Compensation Principles & Elements

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 market 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. The compensation system shall ensure compliance and best practice. In addition, compensation elements are focused on rewarding the delivery of outstanding and sustainable results without inappropriate risk-taking.

Market competitiveness

The compensation structure and level of the EM members is reviewed locally on a regular basis in order to ensure market competitiveness. Such review takes into consideration comparable functional and financial responsibilities. 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 more than 20 Swiss and international Biotech and Pharma companies[1] from which most of our talents are likely to join from. With respect to total compensation (base salary, annual cash bonus & share appreciation rights (**SAR**) entitlement), we position ourselves at the market median at target in our industry.

The benchmark analysis served as a basis for the CC to review the compensation structure and level of the EC members. No further benchmark analysis was conducted in 2018.

[1] Defined national and international peer group:
AC Immune, Alexion, Amgen, Basilea, Bayer, Cassiopea, Celgene, Cosmo, GSK, Jazz Pharmaceuticals, Lonza, Merck, Molecular Partners, Newron, Novartis, Pfizer, Roche, Sanofi, Siegfried, Shire, Straumann, Ypsomed, Vifor Pharma

Board Compensation Elements

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.

For more information about the underlying Plan, see note 20 *"Equity Rights Plans"* in the consolidated financial statements.

Executive Management Compensation Elements

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

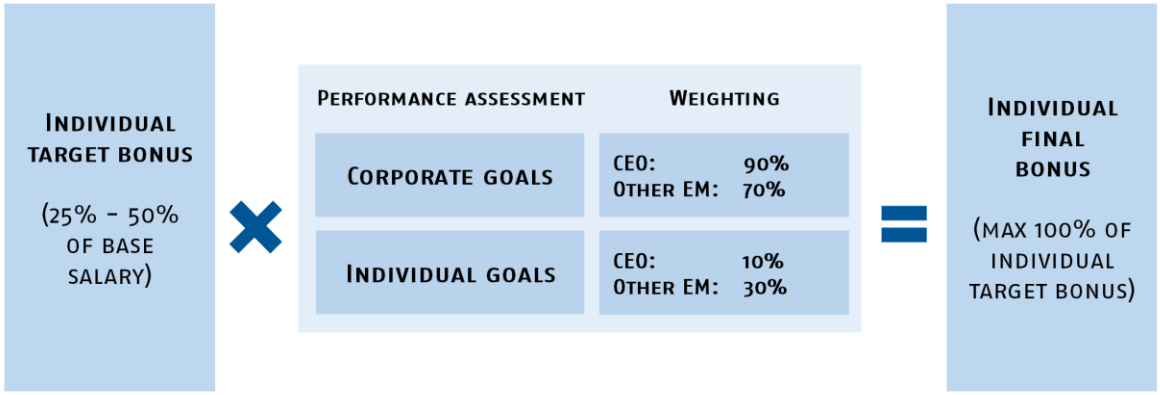
The fixed compensation for Executives includes base salary, social security contributions and payments to the pension fund by the Company. The base salary takes into account the position, responsibilities, experience and skills of an individual EM member. Base salaries are reviewed annually by the CC, taking into account individual performance and the results of the external benchmarking.

Annual cash bonus

The annual cash bonus is based on the achievement of Company and individual goals and will be paid in the month following the AGM, subject to the shareholders' approval. 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



Share Appreciation rights (SAR)

Under the Employee Share Appreciation Rights Plan (**ESARP**), members of the EM receive an annual individually allocated amount of Share Appreciation Rights (SAR) which is calculated as follows:

The Board defines for each EM function a certain percentage of the base salary which is the basis for the SAR calculation. The individual SAR amount is based on the achievement of corporate and individual goals and the same weightings apply as for the annual cash bonus.

The final calculated amount is then divided by the fair market value of a SAR resulting in a maximum number of SAR. The fair market value is calculated by applying the option model "Hull-White".

Starting from the financial year 2019, the grant date for SAR will be one business day after the AGM of a particular year.

For more information about the underlying Plan, see note 20 *"Equity Rights Plans"* in the consolidated financial statements.

Compensation awarded to the Board of Directors in 2018

Annual cash fees

At the AGM 2018, the shareholders approved a total cash compensation for the entire Board of a maximum of CHF 500,500. For the period between the 2018 and the AGM 2019, 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 AGM 2018, the shareholders approved a total maximum amount of CHF 500,500 to be granted in SAR for the period until the AGM 2019. In accordance with the Board Share Appreciation Rights Plan (**BSARP 2017**), 62,659 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 13, 2018. The exercise price was the closing price of Santhera's share on April 13, 2018 and amounted to CHF 18.90 (2017: CHF 77.00).

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.

Disclosure of compensation of members of the Board for the financial years 2018 and 2017 (audited)

In CHF	Annual cash fees	Stock options[1] SAR	Social security[1, 2]	Total compensation	Number of stock options/SAR granted
2018					
Elmar Schnee	148,500	137,499	10,312	296,311	18,591
Martin Gertsch	127,315	127,315	19,651	274,281	17,214
Philipp Gutzwiller	96,759	96,762	14,935	208,456	13,083
Thomas Meier[3]	0	0	0	0	0
Patrick Vink	101,852	101,850	15,721	219,423	13,771
Total	**474,426**	**463,426**	**60,619**	**998,471**	**62,659**
2017					
Elmar Schnee	111,375	137,501	10,313	259,189	4,486
Martin Gertsch	144,236	127,325	20,994	292,555	4,154
Philipp Gutzwiller	72,569	96,766	13,015	182,350	3,157
Thomas Meier[3]	0	0	0	0	0
Patrick Vink	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**

[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 market value of allocated options. For all SARs held by Board members as of December 31, 2018, 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 2018 is CHF 0 (2017: CHF 0).

[3] 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 AGM 2018, the shareholders approved a maximum total amount of fixed compensation for the Board of CHF 500,500 for the period from the AGM 2018 to the AGM 2019. In addition, the shareholders approved the allocation of SAR with a fair market value of a maximum of CHF 500,500.

The table below represents the approved maximum compensation for the Board, the actual amounts paid in 2018 and those still payable until AGM 2019.

	Approved AGM 2018 – AGM 2019	Paid/payable AGM 2018 – AGM 2019
Board fees (CHF)	500,500	500,288
SAR[1] (CHF)	500,500	498,183
Total (CHF)	**1,001,000**	998,471
SAR (number)	n/a	62,659

[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 AGM 2018 (CHF 7.3960).

Compensation awarded to the members of the Executive Management in 2018

Comparison of the approved and paid EM fixed compensation

At the AGM 2017, shareholders approved a maximum total compensation for the EM for 2018 as follows: CHF 3,200,000 for the fixed compensation in cash.

In CHF	Approved 2018	Paid 2018
Base salary	3,200,000	2,420,707

Annual cash bonus

The annual cash bonus for 2018 is based on the achievement of Company and individual goals and will be paid in June 2019, subject to the shareholders' approval. The Company goals included the achievement of sales targets, the successful completion of a financing, certain business development activities (in-licensing) and the completed enrolment of the Company's SIDEROS study. As the Company did not succeed in completing the SIDEROS study enrolment and achieved other goals only partially, the corporate target achievement was determined by the Board to amount to 57.5%.

The proposal to the shareholders at the AGM 2019 is for a maximum cash bonus payment of CHF 550,000 (of which a maximum of CHF 121,000 for social security contributions).

SAR entitlement in CHF

The annual SAR entitlement for 2018 is based on the achievement of Company and individual goals as described in section above "Annual cash bonus".

The proposal to the shareholders at the AGM 2019 is for a maximum SAR entitlement of CHF 1,286,000 (of which a maximum of CHF 96,000 for social security contributions).

To calculate the number of SAR to be allocated, the total SAR amount would be divided by the fair market value of the SAR on the date of their grant (one business day after the AGM). The fair market 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).

It was decided to cap the distributable group-wide SAR allocation numbers at 7.2% of the outstanding share capital to account for the shareholder dilution, the currently low share price, and the upside potential. Therefore, across the company, the calculation of SARs will be adapted pro rata to ensure that the 7.2% threshold is not exceeded. Last year, the distributed SARs were representing 7.2% of the outstanding share capital.

Disclosure of compensation of members of the Executive Management for the years 2018 and 2017 (audited)

In CHF	Base salary	Cash bonus[1]	SAR[1, 2, 5]	Social security and pension[2,3]	Total compensation	Number of stock options/ SAR granted[4]
2018						
Meier Thomas	400,000	121,500	291,600	147,749	960,849	
Other five members of EM	1,557,504	306,148	893,968	517,851	3,275,471	
Total	**1,957,504**	**427,648**	**1,185,568**	**665,600**	**4,236,320**	
2017[6]						
Kristina Nygren	330,000	58,163	232,644	107,017	727,824	16,633
Kristina Nygren Retention award	45,600[7]	0[8]	399,992[8]	34,407	479,999	18,617
Kristina Nygren Total	**375,600**	**58,163**	**632,636**	**141,424**	**1,207,823**	**35,250**
Other seven 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**

1 Proposal for approval by the AGM 2019.

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; the 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 market value of allocated SAR.

4 Starting from the financial year 2018, the grant date for SAR will be one business day after the AGM of a particular year.

5 The distributable group-wide SAR allocation numbers will be capped at 7.2% of the outstanding share capital to account for the shareholder dilution, the currently low share price and the upside potential.

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 revaluation, the value of his SAR amounted to CHF 201,852 and for the other five members of Executive Management, to CHF 3,084,101.

7 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.

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

Changes in the Executive Management in 2018

Giovanni Stropoli, Chief Commercial Officer Europe & Rest of World and Member of the EM, left the Company effective September 30, 2018. As a consequence, all his unvested SAR forfeited.

Event	Date	Number of Executives
Giovanni Stropoli's leaving	September 30, 2018	5

Executive Contracts

The employment contracts with the EM members are compliant with the OaEC and the Company's Articles of Incorporation. Any noncompeting 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.

Indirect Benefits

The Company contributes to pension plans which are based on defined contributions, for old age pension, disability and death. The risk portion provides benefits for widows (spouse), orphans and long term disability in case of sickness. In addition, there will be paid a lump sum in case of death due to accident or sickness. The amount of pension benefits depends on the employee's age and insured compensation. Both employee and employer contribute to the aforementioned pension plans.

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 2018, 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 no such payments in 2018 (2017: none).

Disclosure of compensation of former Board members for the years 2018 and 2017 (audited)

In CHF	Total payment
2018	
n/a	–
Total	**0**
2017	
n/a	–
Total	**0**

With regard to the former EM members, Santhera made no such payments in 2018 (2017: none).

Disclosure of compensation of former EM members for the years 2018 and 2017 (audited)

In CHF	Total payment
2018	
n/a	0
Total	0
2017	
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 2018 and 2017 (audited)

December 31, 2018	Number of shares	Number of stock options (vested)	Number of stock options (unvested)	Number of SAR (vested)	Number of SAR (unvested)
Elmar Schnee	12,000	0	0	2,244	20,833
Martin Gertsch	38,109	3,891	2,390	2,079	19,289
Philipp Gutzwiller	7,100	0	0	1,581	14,659
Thomas Meier[1]	0	0	0	0	0
Patrick Vink	1,000	0	0	1,662	18,225
Total	**58,209**	**3,891**	**2,390**	**7,566**	**73,006**

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[1]	0	0	0	0	0
Patrick Vink	1,000	0	0	0	6,116
Jürg Ambühl	30,000	7,281	0	0	0
Total without Jürg Ambühl	**71,609**	**8,781**	**4,781**	**0**	**17,913**

[1] Thomas Meier's shareholdings are listed in the table below.

Disclosure of shareholdings in the Company of Executive Management members for the years 2018 and 2017 (audited)

December 31, 2018	Number of shares	Number of stock options (vested)	Number of stock options (unvested)	Number of SAR (vested)	Number of SAR (unvested)
Thomas Meier	79,402	9,563	5,312	11,107	33,054
Günther Metz	0	16,560	2,560	6,641	21,396
Christoph Rentsch	0	14,750	7,250	8,229	26,670
Kristina Sjöblom Nygren	0	0	0	0	36,937
Giovanni Stropoli[1]	250	14,033	0	6,718	0
Oliver Strub	0	8,621	2,620	6,857	21,872
Total	**79,652**	**63,527**	**17,742**	**39,552**	**139,929**

[1] Giovanni Stropoli forfeited all his unvested Options/SAR on September 30, 2018, the effective date of his resignation.

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	75,652	13,313	5,312	0	19,038
Todd Bazemore	0	0	0	0	0
Nicholas Coppard	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	**75,902**	**56,189**	**50,612**	**3,489**	**95,297**

Outlook for Board compensation

At the AGM 2019, 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.

The Board will continue with an Audit Committee (**AC**) and a Compensation Committee (**CC**). For rules and responsibilities of these two committees, see section *"Work methods of the Board and its Committees"* (DCG 3.5.3) in the Corporate Governance report.

Both committee chairmanships as well as memberships of the Board and its committees are proposed to be remunerated as per the table under section "*Compensation awarded to the Board of Directors in 2018*" of this Compensation Report. The proposal does not foresee an increase of the Board compensation.

The total maximum compensation of CHF 1,001,000 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 market value of the SAR on the date of their grant. The fair market 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).

Outlook for EM compensation

The AGM 2018 has already approved the fix compensation for 2019 in the amount of CHF 3,200,000. Giovanni Stropoli's tasks have been allocated to Executives and employees of the European commercial operations. The Company is currently considering filling the vacancy and to increase the number of Executives.

For the fix compensation for 2020, the Board will propose an amount of CHF 3,000,000 to the AGM 2019 which would be based on the planned six Executives, including a reserve amount which would allow increasing the fix compensation of the EM if deemed appropriate by the Board.

Ernst & Young Ltd
Aeschengraben 9
P.O. Box
CH-4002 Basle

Phone +41 58 286 86 86
Fax +41 58 286 86 00
www.ey.com/ch

To the General Meeting of
Santhera Pharmaceuticals Holding Ltd, Pratteln

Basle, April 26, 2019

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, 2018. 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 84 to 96 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, 2018 of Santhera Pharmaceuticals Holding 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.

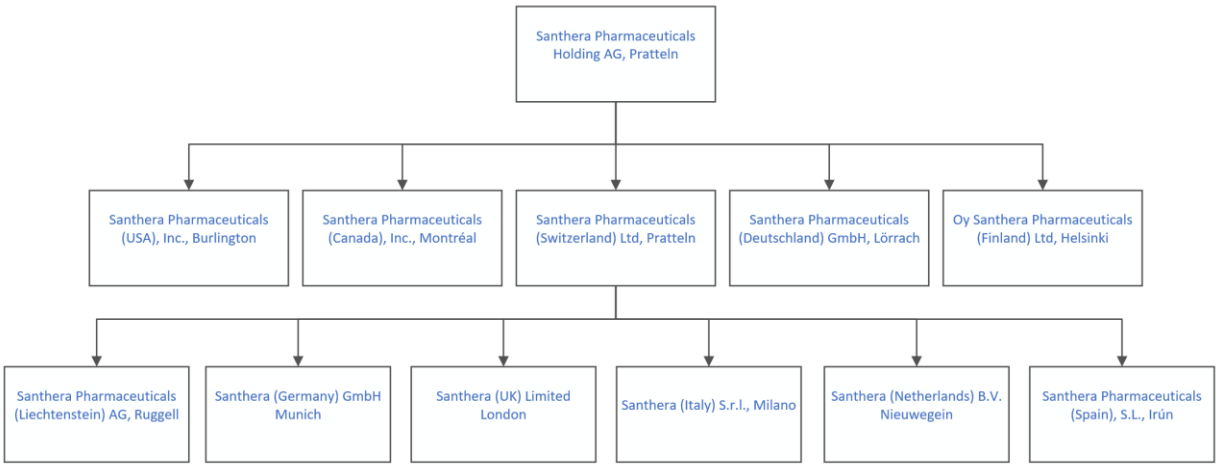
Group Structure and Shareholders (DCG 1)

Group structure (DCG 1.1)

Listed company

Name	Santhera Pharmaceuticals Holding AG (**Company**, together with its affiliates, **Santhera**)
Legal Domicile	Hohenrainstrasse 24, 4133 Pratteln, Switzerland.
Register number	CHE-105.388.338
Listing	SIX Swiss Exchange
Symbol	SANN
Security ID	2714864
ISIN	CH0027148649
Market capitalization	CHF 72 million (December 28, 2018)
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 (Spain), S.L.U	EUR 50,000	Irún, ES	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.

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, 2018, 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	10,660,813	100.0	10,664,563	100.0		3
Authorized capital	500,000	4.7	500,000	4.7	April 11, 2020	3a
Conditional capital for warrants/option rights granted in connection with debt instruments	930,000	8.7	930,000	8.7	For conversion rights: 10 years from issue date. For options: 7 years from issue date.	3c
Conditional capital for ESOP/BSOP/EIP	691,302	6.5	687,552	6.5		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 20 "*Equity Rights Plans*" to the consolidated financial statements.

Changes in share capital (DCG 2.3)

For changes in capital that occurred in 2016 and 2017, see the Company's Annual Report 2017, which can be downloaded from http://www.santhera.com/investors-and-media/investor-toolbox/financial-reports. For changes that took place in 2018, see note 12 *"Share Capital"* to the consolidated financial statements of the Company.

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

As of December 31, 2018, 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 20 "*Equity Rights Plans*" to the consolidated financial statements.

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	○		●

● = 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.

Elmar Schnee

Elmar Schnee is Board Secretary of MindMaze SA, 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 Board Member and Financial Advisor in the life sciences industry. Up to January 2014, he served as Chief Financial Officer of Acino Holding. Prior to this he was Vice President Head of Finance EMEA at Synthes and held Chief Financial Officer 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, Martin was an Audit Engagement Manager at PricewaterhouseCoopers, Basel, Switzerland. Martin is a Swiss-certified fiduciary and Swiss-certified public accountant. He has also completed several executive-level development programs at IMD (the lnternational Institute for Management Development) in Lausanne, Switzerland. He serves as a Board Member of Evolva Holding, and the University Center of Dentistry, Basel (**UZB**). He is also Chairman of a privately-held diagnostic start-up company.

Philipp Gutzwiller

Philipp Gutzwiller is a Managing Director 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 nonexecutive 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 28 "*Related Party Transactions*" to the consolidated financial statements.

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 2019 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 2018

In 2018, the Board held five meetings in person which on average lasted about seven and a half hours. In addition, the Board held sixteen teleconferences which on average lasted somewhat more than 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 discussed the Company's major risks during the Board meetings. Among the key risks identified were the financial situation of the Company, 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**), 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, also in connection with Brexit. 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. In addition, the Company had a consultant conduct select internal audits during the review period.

Executive Management (DCG 4 and 3.6)

In the beginning of the reporting period, the Executive Management consisted of six Executives, and after Giovanni Stropoli's resignation of five.

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	
Christoph Rentsch	Chief Financial Officer, EVP	CH	1959	
Giovanni Stropoli	Chief Commercial Officer Europe & Rest of World, EVP	IT	1960	Until September 30, 2018
Oliver Strub	General Counsel & Secretary to the Board, EVP	CH	1963	

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).

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

Christoph Rentsch joined Santhera as Chief Financial Officer (CFO) and member of Santhera's Executive Management effective July 1, 2015. With a background in finance and long-standing experience in the pharmaceutical industry, Mr 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. He joined F. Hoffmann La Roche in 1994 and became Head of Group Funding and Capital Markets. For almost 10 years he was responsible for all of Roche's finance transactions on group level. In 2003, Mr. Rentsch became partner of Caperis Ltd, an investment management and life science advisory firm, before joining then still privately held Polyphor as CFO, where he supported the company in key stages of its development. Mr. Rentsch holds a Master 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.

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 BSARPs and the ESARPs 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, 2018, 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 (audit-related fees have been incurred in connection with capital increases and related comfort letters and review procedures):

In CHF thousands	2018	2017
Audit services	325	409
Audit-related services	524	11

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 2019

The 2019 Annual General Meeting will be held on Tuesday, May 28, 2019. See also www.santhera.com/investors-and-media/corporate-calendar.

Contact

Eva Kalias, Head External Communications
Phone +41 61 906 89 26
eva.kalias@santhera.com

About Santhera

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

Raxone® and Puldysa® are trademarks 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…

Santhera Pharmaceuticals Holding AG
Hohenrainstrasse 24
4133 Pratteln
Switzerland
Phone +41 61 906 89 50
Fax +41 61 906 89 51
www.santhera.com