santhera
THEIR FUTURE - OUR FOCUS

2018

Interim Condensed Report
January to June 2018

Report on the Six Months Ended June 30, 2018, and Interim Condensed Consolidated Financial Statements

Contents

Santhera Announces Financial Results for the First Half-Year 2018 and Updates on Operational Progress and Growth Strategy

Santhera Pharmaceuticals (SIX: SANN) reports first half-year results as of June 30, 2018, outlines the Company's vision and strategy as it moves through the second half of 2018, and positions itself for future growth.

Thomas Meier, PhD, Chief Executive Officer of Santhera, said: "Our vision is to be a leader in the development and commercialization of rare disease therapies for neuro-ophthalmology, neuromuscular and pulmonary indications. Our strategy to achieve this vision focuses on three distinct pillars: One, we continue to expand our commercial reach and grow sales of our revenue generating product Raxone® for the treatment of LHON. Turnover during the first half-year has been above expectation and we are on track to exceed our 2018 guidance. Two, we are progressing our pipeline assets towards regulatory approval in the EU and the U.S. and, with the inclusion of new data, intend to submit marketing authorization applications for idebenone in DMD in 2019. Three, we are pursuing an active in-licensing strategy for high quality, late-stage rare disease assets with a short time to market."

"We see multiple business development opportunities to leverage our existing development, regulatory and commercial capabilities and our recent in-licensing for POL6014 to treat cystic fibrosis is the first example of Santhera advancing this strategy. With this vision and strategy in mind, we believe Santhera is optimally positioned to create value with its existing and future product portfolio opportunities."

Financial highlights:

- 1H 2018 sales of CHF 16.0 million, increase of 48% compared to 1H 2017
- Operating expenses of CHF 39.9 million (1H 2017: CHF 30.5 million)
- Operating result of CHF –26.3 million (1H 2017: CHF –21.4 million) leading to a net result of CHF –27.4 million (1H 2017: CHF –22.7 million)
- Cash, cash equivalents and short-term financial assets of CHF 34.8 million (June 30, 2018)
- Full year sales guidance raised to CHF 30-32 million

Operational highlights:

- Acquisition of worldwide exclusive license to develop and commercialize clinical stage candidate POL6014 for cystic fibrosis (CF) and other pulmonary diseases
- Renewal of the Early Access to Medicines Scheme (EAMS) Scientific Opinion by UK's Medicines and Healthcare products Regulatory Agency (MHRA) for idebenone for patients with Duchenne muscular dystrophy (DMD) in the UK
- Launch of Expanded Access Program with idebenone for patients with DMD in the U.S.
- Submissions of regulatory dossiers for Raxone in Leber's hereditary optic neuropathy (LHON) in South Korea and Serbia
- Analysis of new data linking study findings with idebenone in DMD to clinically relevant patient benefits for inclusion in regulatory submissions in Europe and the U.S. (planned for 2019)
- Progress with clinical development candidates having successfully completed first clinical trial with omigapil in patients with congenital muscular dystrophy (CMD) and advanced preparations for multiple-ascending dose trial for POL6014 in CF

First half-year overview

- **Strong Raxone sales in 1H 2018**

  Net sales of Raxone in Europe amounted to CHF 16.0 million (1H 2017: CHF 10.9 million) which corresponds to a strong 48% increase year-on-year. Turnover was mainly driven by increased number of patients receiving the drug in existing markets and new launches in additional EU countries. Santhera's goal is to provide treatment to LHON patients worldwide and the Company has submitted a new drug application for LHON in South Korea, one of the major markets in Asia. A decision from the South Korean drug regulatory authorities who granted orphan drug designation for Raxone in LHON can be expected by summer 2019. At the end of the first half of 2018, Santhera was marketing Raxone in more than 20 countries.

- **Broadened product pipeline with licensing agreement**

  In February, Santhera completed the first step in its strategy to in-license pipeline strengthening, clinical stage product candidates in neuro-ophthalmology, neuromuscular and pulmonary diseases by entering into a license agreement with Polyphor for POL6014. Under the agreement, Santhera obtained the worldwide, exclusive rights to develop and commercialize POL6014, a clinical stage selective inhibitor of human neutrophil elastase with the potential to treat cystic fibrosis and other pulmonary diseases.

- **UK's MHRA renewed EAMS positive scientific opinion for Raxone in DMD**

  In June, the UK's MHRA renewed the EAMS scientific opinion for Raxone for a further year for patients with DMD in respiratory function decline who are not taking glucocorticoids. Inclusion in EAMS allows eligible patients with DMD, who meet criteria defined under this scheme, to gain free of charge access to Raxone in the UK.

- **Launch of U.S. Expanded Access Program with idebenone for patients with DMD**

  Santhera has successfully launched and enrolled the first patients in a U.S. Expanded Access Program (EAP), called BreatheDMD, with idebenone. 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, at no cost, through a growing network of research centers across the U.S.

- **New supporting data to be included in submissions for marketing authorization applications for DMD in 2019**

  In July, Santhera announced results of a comparative analysis of the Phase III DELOS trial outcome with new data from natural history studies. This analysis showed that the treatment effect with idebenone observed in the DELOS trial can be linked to a delay in the initiation of assisted ventilation by three years, which is of high clinical relevance. In coming months, Santhera and its academic partners will prepare for the publication of additional clinical data that demonstrate long-term efficacy of idebenone on respiratory function outcomes in patients with DMD, thereby supporting the positive data from the successful Phase III DELOS trial. The findings will be discussed with regulators in the coming months and will be included in the regulatory dossier in preparation of marketing authorization applications for idebenone in DMD in Europe and the U.S. in 2019.

- **Omigapil safe and well tolerated in patients with congenital muscular dystrophy (CMD)**

  The single-center interventional trial to establish the pharmacokinetic profile and to evaluate the safety and tolerability of omigapil in pediatric and adolescent patients with CMD was successfully completed. Santhera plans to seek advice on the clinical development program of omigapil by the TREAT-NMD Advisory Committee for Therapeutics (TACT).

- **Liquidity base allows for the continuation of the strategy as planned**

  As of the end of June 2018, Santhera had cash, cash equivalents and short-term financial assets of CHF 34.8 million (December 31, 2017: CHF 58.2 million). These funds will allow the Company to proceed with its clinical trial program and regulatory filings as foreseen.

Revenue Guidance

Santhera will continue to grow its international business, advance its pipeline programs and proceed business development initiatives to expand its late stage product portfolio. Based on its sales performance in the first six months of the current year and the positive outlook, the Company expects to exceed its guidance of CHF 28-30 million and anticipates reaching a higher turnover of CHF 30-32 million in 2018.

Interim Condensed Consolidated Financial Statements

Contents

Interim Consolidated Balance Sheet

in CHF thousands	Notes	**June 30, 2018 (reviewed)**	Dec. 31, 2017 (audited)
Assets			
Tangible assets	10	2,481	2,157
Intangible assets	8, 10	28,224	23,560
Financial assets long-term		782	713
Deferred tax assets		1,447	1,242
Restricted cash long-term	6	3,000	4,500
Noncurrent assets		**35,934**	**32,172**
Prepaid expenses and accrued income		807	853
Inventories	5	9,443	10,147
Trade and other receivables		8,144	5,402
Financial assets short-term	9	12,742	13,011
Restricted cash short-term	6	3,000	3,000
Cash and cash equivalents	6	22,082	45,195
Current assets		**56,218**	**77,608**
Total assets		**92,152**	**109,780**
Equity and liabilities			
Share capital	7	6,528	6,289
Capital reserves and share premium		401,772	392,002
Retained earnings		–387,432	–360,081
Employee benefit reserve		–3,750	–4,905
Treasury shares		–1,216	–335
Other components of equity		–725	–714
Total equity		**15,177**	**32,256**
Senior unsecured convertible bonds	9	53,825	53,111
Derivative financial instruments	9	1,105	2,792
Pension liabilities		7,522	8,375
Total noncurrent liabilities		**62,452**	**64,278**
Trade and other payables		5,580	4,734
Accrued expenses		8,943	8,512
Total current liabilities		**14,523**	**13,246**
Total liabilities		**76,975**	**77,524**
Total equity and liabilities		**92,152**	**109,780**

Interim Consolidated Income Statement (Reviewed)

for the half-year ended June 30, in CHF thousands	Notes	2018	2017
Net sales	**10**	**16,027**	**10,859**
Cost of goods sold		–2,441	–1,954
Of which amortization intangible asset		*–1,519*	*–1,519*
Other operating income		0	242
Development	11	–18,854	–11,703
Marketing and sales	11	–12,921	–12,622
General and administrative	11	–8,051	–6,113
Other operating expenses	11	–57	–75
Operating expenses	**11**	**–39,883**	**–30,513**
Operating result		**–26,297**	**–21,366**
Financial income		2,512	846
Financial expenses		–3,473	–2,135
Result before taxes		**–27,258**	**–22,655**
Income taxes	12	–93	–57
Net result		**–27,351**	**–22,712**
Basic and diluted loss per share (in CHF)		–4.25	–3.62

Interim Consolidated Statement of Comprehensive Income (Reviewed)

for the half-year ended June 30, in CHF thousands	2018	2017
Net result	**–27,351**	**–22,712**
Items never to be reclassified subsequently to net income in subsequent periods:		
Net actuarial gains/(losses) from defined benefit plans	1,155	133
Items to be reclassified subsequently to net income in subsequent periods:		
Currency translation differences	–11	3
Other comprehensive result	**1,144**	**136**
Total comprehensive result	**–26,207**	**–22,576**

Interim Consolidated Statement of Cash Flows (Reviewed)

for the half-year ended June 30, in CHF thousands	Notes	2018	2017
Result before taxes		**–27,258**	**–22,655**
Depreciation of tangible assets		311	110
Amortization of intangible assets		1,579	1,557
Expenses for equity rights plans		3,735	3,692
Change in fair value of derivatives	9	–1,687	–549
Change in fair value of financial assets short-term	9	269	0
Other non-cash items (Polyphor clinical material)	8	290	0
Change in pension liabilities		302	389
Taxes paid		–298	–152
Change in net working capital		1,166	–3,087
Total financial result		961	1,289
Interest received		1	0
Interest paid		–1,525	–25
Cash flow from operating activities		**–22,154**	**–19,431**
Investments in tangible assets		–1,261	–180
Investments in intangible assets		–33	–45
Investments in other financial assets short-term		0	–5,984
Investments in other financial assets long-term		–69	–143
Change in restricted cash		1,500	–9,000
Cash flow from investing activities		**137**	**–15,352**
Proceeds from sale of treasury shares		1,476	4,640
Purchase of treasury shares		–2,583	–4,908
Proceeds from convertible bonds		0	57,269
Cash flow from financing activities		**–1,107**	**57,001**
Effects of exchange rate changes on cash and cash equivalents		11	–47
Net increase/(decrease) in cash and cash equivalents		**–23,113**	**22,171**
Cash and cash equivalents at January 1		45,195	49,815
Cash and cash equivalents at June 30		**22,082**	**71,986**

For a disclosure of the non-cash transaction with Polyphor and the related acquisition of intangible assets and clinical material, see note 8 *"Transaction with Polyphor"*.

Interim Consolidated Statement of Changes in Equity (Reviewed)

in CHF thousands	Notes	Share capital	Capital reserves and share premium	Retained earnings	Employee benefit reserve	Treasury shares	Trans-lation differen ces	**Total**
Balance at January 1, 2017		**6,280**	**382,322**	**–308,549**	**–4,734**	**–172**	**–796**	**74,351**
Net result		0	0	–22,712	0	0	0	**–22,712**
Other comprehensive income		0	0	0	133	0	3	**136**
Total comprehensive result for the period		**0**	**0**	**–22,712**	**133**	**0**	**3**	**–22,576**
Share-based payment transactions	11	0	3,692	0	0	0	0	**3,692**
Change in treasury shares		0	28	0	0	–296	0	**–268**
Balance at June 30, 2017		**6,280**	**386,042**	**–331,261**	**–4,601**	**–468**	**–793**	**55,199**
Balance at January 1, 2018		**6,289**	**392,002**	**–360,081**	**–4,905**	**–335**	**–714**	**32,256**
Net result		0	0	–27,351	0	0	0	**–27,351**
Other comprehensive income		0	0	0	1,155	0	–11	**1,144**
Total comprehensive result for the period		**0**	**0**	**–27,351**	**1,155**	**0**	**–11**	**–26,207**
Share-based payment transactions	11	0	3,735	0	0	0	0	**3,735**
Capital increase Polyphor	8	239	6,261	0	0	0	0	**6,500**
Change in treasury shares		0	–226	0	0	–881	0	**–1,107**
Balance at June 30, 2018		**6,528**	**401,772**	**–387,432**	**–3,750**	**–1,216**	**–725**	**15,177**

Notes to the Interim Condensed Consolidated Financial Statements (Reviewed)

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 rare indications with high unmet medical needs.

The Company, having the 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 interim financial statements were approved for publication by the Board of Directors (**Board**) on September 3, 2018.

2 Summary of Significant Accounting Policies

The accounting policies used in the preparation of the interim financial statements are consistent with those used in the preparation of the Group's annual financial statements for the year ended December 31, 2017, except for the adoption of new standards and interpretations as of January 1, 2018, as noted below.

Basis of preparation

These unaudited consolidated interim financial statements were prepared in accordance with IAS 34, Interim Financial Reporting, of the International Financial Reporting Standards (**IFRS**) and should be read in conjunction with the annual financial statements for the year ended December 31, 2017.

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

Material uncertainties and going concern

Santhera is subject to different risks and uncertainties, including but not limited to the uncertainty of the development of its clinical studies, regulatory approval and marketing activities in order to achieve profitability. The Group's ability to continue operations as planned for the next 12 months depends on cash flows from ongoing product sales, the results of its development activities and the capability to raise additional funds (equity, debt financing).

Santhera continues to generate increasing income from product sales for the indication Leber's hereditary optic neuropathy (**LHON**). Moreover, the Company has collected additional data in patients with Duchenne muscular dystrophy (**DMD**) needed for submission of a Marketing Authorization Application for this indication in the European Union (**EU**) in early 2019. Santhera also prepares for submission of a New Drug Application for patients with DMD in the United States.

Santhera's cash, cash equivalents and short-term financial assets amounted to CHF 34.8 million as of June 30, 2018. A material uncertainty remains as to whether Santhera's current funding is sufficient to support its going concern for another twelve months. Santhera seeks to secure additional financing in due course. Shareholders should note that whilst the Board and Executive Management continue to apply best efforts to raise additional funds, there is no guarantee that such funds can be raised. The availability of sufficient funds is crucial for Santhera and its ability to continue and grow its operations. Based on the Board's and the Executive Management's plan as discussed above the Board of Directors is confident to ensure business continuation and meet its obligations for a further twelve months. Hence, the interim consolidated financial statements have been prepared on a going concern basis.

Changes in accounting policies

The accounting policies adopted in the preparation of the interim consolidated financial statements are consistent with those followed in the preparation of the Group's annual consolidated financial statements for the year ended December 31, 2017, except for the adoption of new standards effective as of January 1, 2018. The Group has not early adopted any other standard, interpretation or amendment that has been issued but is not yet effective.

The Group applies, for the first time, IFRS 15 Revenue from Contracts with Customers and IFRS 9 Financial Instruments. Several other amendments and interpretations apply for the first time in 2018, but do not have an impact on the interim consolidated financial statements of Santhera. IFRS 15 supersedes IAS 11 Construction Contracts, IAS 18 Revenue and related interpretations and it applies to all revenue arising from contracts with customers, unless those contracts are in the scope of other standards. 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. Revenue from sales of products is recognized at the point in time when the customer obtains control of the goods or services, which 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.

IFRS 9 Financial Instruments replaces IAS 39 Financial Instruments: Recognition and Measurement for annual periods beginning on or after January 1, 2018, bringing 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. Furthermore, the Group does not apply hedge accounting. IFRS 9 requires Santhera to record expected credit losses (**ECL**) on all of its trade receivables, either on a 12-month or lifetime basis. The Group applied the simplified approach and records lifetime expected losses. 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.

3 Seasonality

The operating result is not subject to significant seasonal variations during the financial year.

4 Exchange Rates of Principal Currencies

	Income statement in CHF average rates for half-year ended		Balance sheet in CHF as of period end	
	June 30, 2018	June 30, 2017	**June 30, 2018**	Dec. 31, 2017
1 Euro (**EUR**)	**1.1696**	1.0762	**1.1571**	1.1691
1 US dollar (**USD**)	**0.9661**	0.9947	**0.9933**	0.9753
1 British pound (**GBP**)	**1.3293**	1.2516	**1.3080**	1.3173
1 Canadian dollar (**CAD**)	**0.7562**	0.7459	**0.7514**	0.7777

5 Inventories

This position consists mainly of active pharmaceutical ingredients and semi-finished products which are kept by Santhera as stock for market supply, development and inventory risk management purposes (security stock) for Raxone.

6 Cash and Cash Equivalents and Restricted Cash

6.1 Cash and cash equivalents

in CHF thousands	**June 30, 2018**	Dec. 31, 2017
Cash at banks and on hand		
in CHF	13,385	34,730
in EUR	6,191	8,152
in USD	1,709	1,496
in GBP	582	697
in CAD	121	120
other currencies	94	0
Total at period end	**22,082**	**45,195**

6.2 Restricted cash

in CHF thousands	**June 30, 2018**	Dec. 31, 2017
Long-term	3,000	4,500
Short-term	3,000	3,000
Total at period end	**6,000**	**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.

7 Share Capital

7.1 Ordinary share capital

During the reporting period ending June 30, 2018, 238,924 Shares were issued out of the authorized share capital. With these Shares Santhera obtained from Polyphor, Allschwil, Switzerland, the worldwide, exclusive rights to develop and commercialize POL6014, an innovative macrocycle elastase inhibitor (see note 8 *"Transaction with Polyphor"*).

As a result, as of June 30, 2018, the issued nominal share capital amounted to CHF 6,527,479, divided into 6,527,479 Shares at a nominal value of CHF 1 each.

7.2 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 8 *"Transaction with Polyphor"*). On the occasion of the Annual Shareholders' Meeting (**ASM**) on April 12, 2018, Santhera's shareholders approved the increase of the authorized share capital of the Company.

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.

7.3 Conditional share capital

As of June 30, 2018, the Company had conditional share capital, pursuant to which the share capital may be increased by

(i) a maximum amount of CHF 691,302 (2017: CHF 700,000) through the issuance of up to 691,302 (2017: 700,000) Shares, under the exclusion of shareholders' pre-emptive rights, for equity rights being exercised under the Company's equity rights plans (see note 13 *"Equity Rights Plans"*).

(ii) 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.

8 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.

Significant non-cash transaction

The consideration for the acquisition of the license and the clinical material was paid by issuing shares of Santhera Pharmaceuticals Holding AG for a total amount of CHF 6.5 million (CHF 27.2053 per share; see note 7 *"Share Capital"*). Santhera acquired on one hand a license (POL6014) in the amount of CHF 6.2 million, which was recognized as an addition to the intangible assets. The intangible asset is being developed and hence not yet available for use and not amortized. On the other hand, the Group purchased clinical material in the amount of CHF 0.3 million, which was booked as a development expense. The amounts of the two parts were based on their relative fair values.

9 Financial Assets and Liabilities

9.1 Financial assets short-term

Financial assets (units in a fund) are classified as held for trading. They are measured at fair value through profit or loss and based on quoted prices (level 1). A loss of TCHF 269 (financial expenses) resulted during the reporting period (2017: TCHF 16).

9.2 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 has been 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. The fair value of the bonds (Level 1) at June 30, 2018, amounts to CHF 43.8 million (December 31, 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 June 30, 2018, was at 65.7% (June 30, 2017: 88.0%).

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 period end CHF 1.1 million (June 30, 2018). The change in the fair value (CHF 1.7 million) was recognized in financial income.

Sensitivity analysis:

	June 30, 2018		June 30, 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%	–112	+5%	343
	–5%	146	–5%	–273

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 in 2017	**57,269**	**0**
Non-cash changes		
Initial recognition derivative financial instruments	–5,332	5,332
Change in fair value of derivative financial instruments	0	–549
Effective interest/amortized cost calculation	488	0
June 30, 2017	**52,425**	**4,783**
Change in fair value of derivative financial instruments	0	–1,991
Effective interest/amortized cost calculation	686	0
December 31, 2017	**53,111**	**2,792**
Change in fair value of derivative financial instruments	0	–1,687
Effective interest/amortized cost calculation	714	0
June 30, 2018	**53,825**	**1,105**

10 Segment and Geographic Information

10.1 Segment information

Santhera operates in one business segment, namely 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, assess the reporting data and allocate resources as one segment on an aggregated 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.

10.2 Geographic information

Net sales

half-year ended June 30, in CHF thousands	**2018**	**2017**
Europe	15,956	10,802
Rest of the world	71	57
Total	**16,027**	**10,859**

In the reporting period 2018, net sales amounted to CHF 16.0 million. Raxone was sold in more than 20 European countries, with the majority of sales generated in France and Germany (in the reporting period 2017, sales went into 17 European countries, with a majority of sales in France and Germany).

Noncurrent assets (excluding financial instruments and deferred tax assets)

in CHF thousands	**June 30, 2018**	Dec. 31, 2017
Switzerland	30,474	25,451
Rest of Europe	149	171
North America	82	95
Total	**30,705**	**25,717**

11 Operating Expenses by Nature

half-year ended June 30, in CHF thousands	**2018**	**2017**
External development expenses	–12,243	–6,812
Patent and license expenses	–184	–147
Marketing expenses	–5,145	–5,547
Employee expenses	–18,196	–15,609
Of which non-cash-relevant expenses for share-based payments	*–3,735*	*–3,692*
General and administrative expenses	–2,966	–1,833
Depreciation and amortization	–370	–147
Lease expenses	–723	–343
Other operating expenses	–56	–75
Total operating expenses	**–39,883**	**–30,513**

Increased expenses for the reporting period in 2018 mainly resulted from additional development activities (e.g. DMD and POL6014) and staff hired for development activities.

12 Income Taxes

half-year ended June 30, in CHF thousands	**2018**	**2017**
Current income taxes	–298	–152
Deferred taxes	205	95
Total	**–93**	**–57**

Movements on deferred taxes relate to temporary differences on inventory.

13 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. New grants are only possible under Share Appreciation Rights Plans (**SARP**).

The fair value of equity rights (share appreciation rights (**SAR**) or stock options) is determined at each grant date by using the Hull-White pricing model. For the calculation of the fair value of SAR granted during the reporting period in 2018 the same range of valuation parameters as disclosed in the financial statements as of December 31, 2017, was applied, except for the exercise prices (equal to the Share prices at grant) which were between CHF 18.90 and CHF 36.70. The non-cash-relevant expenses for all unvested SAR and stock options in the reporting period 2018 amounted to CHF 3.7 million (2017: CHF 3.7 million).

13.1 Share Appreciation Rights Plans

Santhera has established a 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. SAR grants are made mainly periodically at the full discretion of the Board or as contractually agreed with employees. SARP introduced in 2017 foresee vesting of 1/3 of the SAR on the first anniversary; the remaining 2/3 vest 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**).

In the reporting period ended June 30, 2018, a total of 617,282 SARs with exercise prices between CHF 18.90 and CHF 36.70 were granted. In the half-year period ending June 30, 2017, a total of 316,986 SAR with exercise prices between CHF 54.85 and CHF 77.80 were granted.

Number of SAR outstanding

half-year ended June 30, number of SAR	**2018**	**2017**
Outstanding at January 1	**360,110**	**56,581**
Granted[1]	617,282	316,986
Exercised	0	0
Forfeited	–129,869	0
Expired	0	0
Outstanding at June 30	**847,523**	**373,567**

[1] The weighted average fair value of the SAR granted during the reporting period in 2018 was CHF 12.16 (in the comparative reporting period 2017 the weighted average fair value of SAR granted was CHF 26.75).

13.2 Stock Option Plans

Santhera has established Employee Stock Option Plans (**ESOP**), the ESOP 2010, the ESOP 2015, and Board Stock Option Plans (**BSOP**), the BSOP 2015, to align the long-term interests of the Board, the Executive Management and employees. Options granted under the stock option plans are equity-settled. No grants are made under ESOP and BSOP anymore.

In the reporting period ended June 30, 2018, no stock options were granted. In the half-year period ending June 30, 2017, no stock options were granted.

Number of stock options outstanding

half-year ended June 30, number of stock options	**2018**	**2017**
Outstanding at January 1	**288,442**	**313,365**
Granted	0	0
Forfeited	–11,556	–9,687
Expired	0	–240
Exercised	0	0
Outstanding at June 30	**276,886**	**303,438**

14 Related Party Transactions

During the reporting period 2018, a total of 62,659 SAR were granted to members of the Board and 134,194 SAR were granted to members of the Executive Management. In the same period in 2017, a total of 15,120 SAR were granted to members of the Board and 104,033 SAR to members of the Executive Management.

15 Subsequent Events

None.

Building a better working world

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To the Board of Directors of
Santhera Pharmaceuticals Holding Ltd, Pratteln

Basle, September 3, 2018

Report on the review of interim condensed consolidated financial statements

Introduction

We have reviewed the accompanying interim condensed consolidated financial statements (pages 7 to 20) of Santhera Pharmaceuticals Holding AG for the period from January 1, 2018 to June 30, 2018. The Board of Directors is responsible for the preparation and presentation of these interim condensed consolidated financial statements in accordance with International Financial Reporting Standard IAS 34 “Interim Financial Reporting”. Our responsibility is to express a conclusion on these interim condensed consolidated financial statements based on our review.

Scope of Review

We conducted our review in accordance with International Standard on Review Engagements 2410 “Review of Interim Financial Information Performed by the Independent Auditor of the Entity”. A review of interim financial information consists of making inquiries, primarily of persons responsible for financial and accounting matters, and applying analytical and other review procedures. A review is substantially less in scope than an audit conducted in accordance with International Standards on Auditing and consequently does not enable us to obtain assurance that we would become aware of all significant matters that might be identified in an audit. Accordingly, we do not express an audit opinion.

Conclusion

Based on our review, nothing has come to our attention that causes us to believe that the interim condensed consolidated financial statements are not prepared, in all material respects, in accordance with International Financial Reporting Standard IAS 34 “Interim Financial Reporting”.

Material Uncertainty Related to Going Concern

We draw attention to note 2 in the interim condensed consolidated financial statements, which indicates the existence of a material uncertainty which casts significant doubt about the Group’s ability to continue as a going concern in connection with the ability to raise additional funds (equity, debt financing). Our conclusion is not modified in respect of this matter.

Ernst & Young Ltd

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

/s/ Jan Meyer
Licensed audit expert

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 and Israel for the treatment of Leber's hereditary optic neuropathy (LHON) and currently commercialized in more than 20 countries. For further information, please visit www.santhera.com.

Trademarks

Raxone® is a trademark of Santhera Pharmaceuticals.

Forward-Looking Statements

This Interim 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 Interim 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.

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