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2019

Interim Condensed Report
January to June 2019

Report on the Six Months Ended June 30, 2019, and Interim Condensed Consolidated Financial Statements

Contents

Santhera Announces Financial Results for the First Half-Year 2019

Santhera Pharmaceuticals (SIX: SANN) reports first half-year results as of June 30, 2019, and provides an update on its pipeline and strategic focus.

Thomas Meier, PhD, Chief Executive Officer of Santhera, said: "Following a series of transactions which strategically reshaped the direction of the Company, we are well positioned to advance our long-term growth strategy of focusing on medicines to treat neuromuscular and pulmonary diseases. For 2019, we are on track to achieve our strategic objectives, having filed for conditional marketing authorization of Puldysa® (idebenone) in Europe for the preservation of respiratory function in Duchenne muscular dystrophy (DMD) patients. We expect an opinion from the EMA's Committee for Medicinal Products for Human Use (CHMP) mid-2020. Our goal is to help all DMD patients, irrespective of causative mutations, disease stage or age. We were therefore very pleased about the recent publication by ReveraGen of positive Phase IIa-extension study results with vamorolone in the journal *Neurology*. These data together with previous publications provide proof-of-concept that vamorolone can improve gross muscle function outcomes in young patients with DMD and indicate a better tolerability profile than standard corticosteroids."

"During the period, we announced an exclusive agreement with Chiesi Group to out-license Raxone® for the treatment of Leber's hereditary optic neuropathy (LHON) for a total consideration of up to EUR 93 million with an upfront payment of EUR 44 million. In monetizing this commercial product, we have strengthened our financial position to focus on upcoming regulatory milestones and commercialization activities for our neuromuscular and pulmonary product candidates, which are core to our long-term growth strategy."

Financial highlights:

- 1H-2019 sales on track with CHF 18.3 million, an increase of 14% compared to 1H-2018
- Operating expenses of CHF 38.2 million (1H-2018: CHF 39.9 million)
- Operating result of CHF –22.4 million (1H-2018: CHF –26.3 million) leading to a net result of CHF –26.9 million (1H-2018: CHF –27.4 million)
- Cash and cash equivalents of CHF 43.7 million (August 31, 2019)
- Full-year sales guidance of CHF 25-27 million

First half-year overview:

Regulatory momentum for Puldysa in DMD

In June, the European Medicines Agency (EMA) validated Santhera's application for conditional marketing authorization (CMA) for Puldysa (idebenone) in the treatment of respiratory dysfunction in patients with Duchenne muscular dystrophy (DMD) who are not using glucocorticoids. The review process by the EMA's CHMP has begun and the Company expects an opinion by the CHMP around mid-2020. In addition, the UK's Medicines and Healthcare products Regulatory Agency (MHRA) renewed its positive scientific opinion for idebenone for patients with DMD in respiratory function decline who are not taking glucocorticoids, under the Early Access to Medicines Scheme (EAMS).

Long-term data with Puldysa

In February, Santhera announced the results of its SYROS study, which investigated long-term efficacy with idebenone in slowing respiratory function loss in patients with DMD. The study demonstrated that long-term treatment with idebenone consistently contributed to the preservation of respiratory function for up to 6 years in a real-world setting. This long-term data further supports the potential for idebenone to positively modify the course of respiratory function decline and delay the time to clinically relevant milestones.

Strong Raxone performance underscores Santhera's commercial expertise

Net sales of Raxone in Europe amounted to CHF 18.3 million (1H-2018: CHF 16.0 million) which corresponds to a 14% increase year-on-year, in line with the previous full-year product sales guidance. In May, the Company announced that it had entered into an exclusive license agreement with Chiesi Group for Raxone for the treatment of LHON, for a total consideration of up to EUR 93 million. With the closing at the end of July, Santhera has now transferred all rights to Chiesi Group for the development, commercialization and distribution of Raxone for the treatment of LHON and any other potential ophthalmological indications for all territories worldwide except the US and Canada. After the closing and for an interim period, Santhera will provide support services to Chiesi Group to enable a seamless handover of the business and will continue to commercialize Raxone for LHON in France.

Positive study data with vamorolone published– pivotal study enrolling

Last month, positive data from 6-month Phase IIa-extension study (VBP15-003) with vamorolone in DMD were published by ReveraGen in *Neurology* [1]. The data demonstrated dose-related improvement of gross muscle function in patients with DMD treated with vamorolone. Vamorolone was reported to be safe and well tolerated up to the highest dose tested (6.0 mg/kg/day). Biomarker data indicated reduced occurrence of side effects typical for traditional corticosteroid drugs. Based on these data, vamorolone has potential to replace standard corticosteroids currently used in patients with DMD.

ReveraGen is presently enrolling the Phase IIb VISION-DMD study [2] (VBP15-004; clinicaltrials.gov: NCT03439670), designed as a pivotal efficacy and safety trial. The study is expected to be fully enrolled by the end of 2019. Accordingly, the 6-month randomized placebo-controlled treatment period would end by mid-2020, followed by data analysis. NDA submission could be towards year end 2020.

Collaboration to advance gene therapy research for rare neuromuscular disease

Santhera initiated a collaboration with the Biozentrum of the University of Basel to advance gene therapy research for the treatment of LAMA2-deficient congenital muscular dystrophy (LAMA2 MD or MDC1A). The preclinical research collaboration builds on previous work with omigapil, which was recently studied in a Phase I clinical trial and could act complementary. The program is supported by public funding for innovation in Switzerland through a grant from Innosuisse – the Suisse Innovation Agency.

Continued investment in clinical development

Santhera is running several late-stage clinical trials, among them SIDEROS, the largest ever conducted study in patients with DMD. The SIDEROS study is 84% recruited and positive outcome of the study will form the basis for NDA submission for patients with DMD irrespective of glucocorticoid status, currently expected in 2H-2021. In addition, the preparation of the MAA for Puldysa, remaining post-approval clinical work for Raxone in LHON and increased clinical development work with POL6014 entailed slightly higher development expenses of CHF 19.3 million (+2% year-on-year). Overall, operating expenses showed a small decline (–4%) driven by lower expenses for commercial activities (–10%).

Liquidity base allows for the continuation of the strategy as planned

In April, Santhera raised CHF 7.1 million by the placement of 500,000 shares. As of the end of June 2019, Santhera had cash and cash equivalents of CHF 12.7 million (December 31, 2018: CHF 22.0 million). Together with the net proceeds from the initial payment from Chiesi Group following closing of the licensing transaction, liquid funds amounted to CHF 43.7 million (August 31, 2019), allowing the Company to proceed with its clinical trial program and regulatory filings as planned.

Outlook and Guidance

Based on the performance in the first half-year of 2019, the Company expects to achieve annual net sales with Raxone in the currently approved indication LHON of CHF 25-27 million in 2019, taking into account that Chiesi Group has taken over commercial sales for Raxone from August in all European countries except France. The operational priorities for 2019 are the preparation of European market entry with Puldysa in DMD in 2020, completing enrollment into the DMD SIDEROS trial to support the planned US-submission of Puldysa in DMD, and advancing the other clinical stage candidates in the pipeline, particularly vamorolone and POL6014.

References:

[1] Hoffman EP et al. (2019). Vamorolone trial in Duchenne muscular dystrophy shows dose-related improvement of muscle function. Neurology 2019. https://n.neurology.org/lookup/doi/10.1212/WNL.0000000000008168

[2] https://vision-dmd.info/

Interim Condensed Consolidated Financial Statements

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Interim Consolidated Balance Sheet

In CHF thousands	Notes	**June 30, 2019 (reviewed)**	Dec. 31, 2018 (audited)
Assets			
Tangible assets	9	5,927	2,269
Intangible assets	9	59,914	61,467
Financial assets long-term		712	690
Restricted cash long-term	6	0	1,500
Deferred tax assets		1,067	1,285
Noncurrent assets		**67,620**	**67,211**
Prepaid expenses and accrued income		1,183	969
Inventories	5	9,029	9,282
Trade and other receivables		12,562	7,861
Restricted cash short-term	6	3,000	3,000
Cash and cash equivalents	6	12,698	21,971
Current assets		**38,472**	**43,083**
Total assets		**106,092**	**110,294**
Equity and liabilities			
Share capital	7	11,165	10,665
Capital reserves and share premium		445,112	435,795
Retained earnings		–441,167	–414,267
Employee benefit reserve		–4,303	–2,675
Treasury shares		–726	–904
Other components of equity		–827	–785
Total equity		**9,254**	**27,829**
Convertible bonds	8	55,345	54,569
Derivative financial instruments	8	1,394	204
Noncurrent lease liabilities	8	2,924	0
Pension liabilities		9,946	7,983
Total noncurrent liabilities		**69,609**	**62,756**
Trade and other payables		9,143	8,306
Accrued expenses		11,823	11,041
Accruals for income taxes		475	362
Current lease liabilities	8	1,017	0
Current loan	8	4,771	0
Total current liabilities		**27,229**	**19,709**
Total liabilities		**96,838**	**82,465**
Total equity and liabilities		**106,092**	**110,294**

Interim Consolidated Income Statement (Reviewed)

For the half-year ended June 30, in CHF thousands	Notes	2019	2018
Net sales	**9**	**18,315**	**16,027**
Cost of goods sold		–2,557	–2,441
Of which amortization intangible asset		*–1,519*	*–1,519*
Other operating income		16	0
Development	10	–19,325	–18,854
Marketing and sales	10	–11,611	–12,921
General and administrative	10	–7,206	–8,051
Other operating expenses	10	–66	–57
Operating expenses	**10**	**–38,208**	**–39,883**
Operating result		**–22,434**	**–26,297**
Financial income		859	2,512
Financial expenses		–4,924	–3,473
Result before taxes		**–26,499**	**–27,258**
Income taxes	11	–401	–93
Net result		**–26,900**	**–27,351**
Basic and diluted loss per share (in CHF)		–2.47	–4.25

Interim Consolidated Statement of Comprehensive Income (Reviewed)

For the half-year ended June 30, in CHF thousands	2019	2018
Net result	**–26,900**	**–27,351**
Items never to be reclassified subsequently to net income in subsequent periods:		
Net actuarial gains/(losses) from defined benefit plans	–1,628	1,155
Items to be reclassified subsequently to net income in subsequent periods:		
Currency translation differences	–42	–11
Other comprehensive result	**–1,670**	**1,144**
Total comprehensive result	**–28,570**	**–26,207**

Interim Consolidated Statement of Cash Flows (Reviewed)

For the half-year ended June 30, in CHF thousands	Notes	2019	2018
Result before taxes		**–26,499**	**–27,258**
Depreciation of tangible assets		827	311
Amortization of intangible assets		1,553	1,579
Expenses for equity rights plans		3,229	3,735
Change in fair value of derivatives	8	1,190	–1,687
Change in fair value of financial assets short-term		0	269
Other non-cash items (Polyphor clinical material)		0	290
Change in pension liabilities		335	302
Taxes paid		–183	–298
Change in net working capital		–3,173	1,166
Total financial result		4,065	961
Interest received		8	1
Interest paid		–1,571	–1,525
Cash flow from operating activities		**–20,219**	**–22,154**
Investments in tangible assets		–24	–1,261
Investments in intangible assets		–28	–33
Investments in other financial assets long-term		0	–69
Change in restricted cash		1,500	1,500
Cash flow from investing activities		**1,448**	**137**
Capital increase		7,125	0
Proceeds from sale of treasury shares		1,222	1,476
Purchase of treasury shares		–1,155	–2,583
Proceeds from current loan	8	4,732	0
Payment of lease liabilities		–523	0
Cost of issuance share capital		–1,936	0
Cash flow from financing activities		**9,465**	**–1,107**
Effects of exchange rate changes on cash and cash equivalents		33	11
Net increase/(decrease) in cash and cash equivalents		**–9,273**	**–23,113**
Cash and cash equivalents at January 1		21,971	45,195
Cash and cash equivalents at June 30		**12,698**	**22,082**

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, 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	10	0	3,735	0	0	0	0	**3,735**
Capital increase Polyphor		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**
Balance at January 1, 2019		**10,665**	**435,795**	**–414,267**	**–2,675**	**–904**	**–785**	**27,829**
Net result		0	0	–26,900	0	0	0	**–26,900**
Other comprehensive income		0	0	0	–1,628	0	–42	**–1,670**
Total comprehensive result for the period		**0**	**0**	**–26,900**	**–1,628**	**0**	**–42**	**–28,570**
Share-based payment transactions	10	0	3,229	0	0	0	0	**3,229**
Capital increase	7	500	6,625	0	0	0	0	**7,125**
Cost of issuance share capital		0	–426	0	0	0	0	**–426**
Change in treasury shares		0	–111	0	0	178	0	**67**
Balance at June 30, 2019		**11,165**	**445,112**	**–441,167**	**–4,303**	**–726**	**–827**	**9,254**

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 2, 2019.

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, 2018, except for the adoption of new standards and interpretations as of January 1, 2019, 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, 2018.

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's cash and cash equivalents amounted to CHF 12.7 million as of June 30, 2019. In August 2019, Santhera received the initial payment of EUR 44 million upon closing of the transaction (see note 14 "Subsequent Events") with Chiesi Farmaceutici S.p.A., Parma, Italy (**Chiesi**).

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.

In May 2019, Santhera filed an application for conditional marketing authorization (**CMA**) for Puldysa® (idebenone) in Europe for the treatment of respiratory dysfunction in patients with Duchenne muscular dystrophy (**DMD**) to the European Medicines Agency (**EMA**). A decision by the EMA is expected around mid 2020 and upon marketing authorization Santhera intends to commercialize Puldysa in Europe.

Santhera is considering various options including a capital increase in order to generate further financing of the Group. Shareholders should note that whilst the Management and Board of Directors continue to apply best

efforts to evaluate available options, there is no guarantee that any transaction can be realized or that such transaction would generate sufficient funds to finance further operations. This may cast significant doubts about the going concern of the Company.

The Management and Board of Directors believe that the Company is prepared to secure additional funds needed (i.e., through further equity financing) 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.

Changes in accounting policies

The Group has not early adopted any other standard, interpretation or amendment that had been issued but is not yet effective.

IFRS standard effective with January 1, 2019

IFRS 16 was issued in January 2016 and it replaces IAS 17 *Leases*, IFRIC 4 *Determining whether an Arrangement Contains a Lease*, SIC-15 *Operating Leases - Incentives* and SIC-27 *Evaluating the Substance of Transactions Involving the Legal Form of a Lease*. IFRS 16 sets out the principles for the recognition, measurement, presentation and disclosure of leases and requires lessees to account for leases under a single on-balance sheet model similar to the accounting for finance leases under IAS 17.

Santhera introduced IFRS 16 using the modified retrospective method of adoption with the date of initial application of January 1, 2019. Under this method, the standard is applied retrospectively with the cumulative effect of initially applying the standard recognized at the date of initial application. The Group has opted to apply the transition practical expedient according to which the standard may be applied only to contracts that were previously identified as leases in accordance with IAS 17 and IFRIC 4 at the date of initial application. The Group has also elected to use the recognition exemptions for lease contracts that, at the commencement date, have a lease term of 12 months or less and do not contain a purchase option ("short-term leases"), and lease contracts for which the underlying asset is of low value ("low-value assets").

a) Nature of the effect of adoption of IFRS 16

The Group has lease contracts for offices and equipment (vehicles). Before the adoption of IFRS 16, all the Group leases were classified as operating leases.

Upon adoption of IFRS 16, the Group applied a single recognition and measurement approach for all leases, except for short-term leases and leases of low-value assets. The standard provides specific transition requirements and practical expedients, which have been applied by Santhera. The Group has recognized right-of-use (ROU) assets and lease liabilities for its leases previously classified as operating leases, except for short-term leases and leases of low-value assets. The right-of-use assets were recognized based on the amount equal to the lease liabilities. Lease liabilities were recognized based on the present value of the remaining lease payments, discounted using the incremental borrowing rate at the date of initial application.

The Group also applied the available practical expedients; this means that it:

- used a single discount rate to a portfolio of leases with reasonably similar characteristics.
- relied on its assessment of whether leases are onerous immediately before the date of initial application.
- applied the short-term leases exemptions to leases with lease terms that end within 12 months at the date of initial application.
- used subsequent consideration in determining the lease term where the contract contains options to extend or terminate the lease.

Based on the foregoing, as at January 1, 2019:

- Right-of-use (ROU) assets of CHF 4.4 million were recognized within tangible assets.
- Lease liabilities of CHF 4.4 million were recognized (current and noncurrent).

The lease liabilities as at January 1, 2019 can be reconciled to the operating lease commitments as at December 31, 2018 as follows:

	In CHF thousands
Operating lease commitments as at December 31, 2018	**2,626**
Weighted average incremental borrowing rate as at January 1, 2019	2.81%
Operating lease commitments at January 1, 2019 (discounted)	2,567
Less:	
Commitments relating to short-term leases	-62
Add:	
Commitments relating to early cancellation right not exercised	1,872
Lease liabilities as at January 1, 2019	**4,377**

b) Summary of new accounting policies

Set out below are the new accounting policies of the Group upon adoption of IFRS 16, which have been applied from the date of initial application:

Right-of-use assets

The Group recognizes right-of-use assets at the commencement date of the lease. Right-of-use assets are measured at cost, less any accumulated depreciation and impairment losses, and adjusted for any remeasurements of lease liabilities. The cost of right-of-use assets includes the amount of lease liabilities recognized, initial direct costs incurred, and lease payments made at or before the commencement date. Unless the Group is reasonably certain to obtain ownership of the leased asset at the end of the lease term, the recognized right-of-use assets are depreciated on a straight-line basis over the shorter of its estimated useful life and the lease term. Right-of-use assets are subject to impairment.

Lease liabilities

At the commencement date of the lease, the Group recognizes lease liabilities measured at the present value of lease payments to be made over the lease term. The lease payments include fixed payments less any lease incentives receivable, variable lease payments that depend on an index or a rate, and amounts expected to be paid under residual value guarantees. The lease payments also include the exercise price of a purchase option reasonably certain to be exercised by the Group and payments of penalties for terminating a lease, if the lease term reflects the Group exercising the option to terminate. The variable lease payments that do not depend on an index or a rate are recognized as expense in the period during which the event or condition that triggers the payment occurs.

In calculating the present value of lease payments, the Group uses the incremental borrowing rate at the lease commencement date if the interest rate implicit in the lease is not readily determinable. After the commencement date, the amount of lease liabilities is increased to reflect the accumulation of interest and reduced by the lease payments made. In addition, the carrying amount of lease liabilities is remeasured if there is a modification, a change in the lease term, a change in the in-substance fixed lease payments or a change in the assessment to purchase the underlying asset.

Short-term leases and leases of low-value assets

The Group applies the short-term lease recognition exemption to its short-term leases. It also applies the lease of low-value assets recognition exemption to leases that are considered of low value (i.e. below CHF 5,000). Lease payments on short-term leases and leases of low-value assets are recognized as expense over the lease term.

c) Amounts recognized in the Balance Sheet and Income Statement

In CHF thousands	**Right-of-use assets**		**Total**	**Lease liabilities**
	Vehicles	Buildings		
At December 31, 2018	**0**	**0**	**0**	**0**
IFRS 16 first time adoption January 1, 2019	612	3,765	4,377	4,377
Additions	78	28	106	106
Depreciation	–153	–397	–550	0
Exchange differences	–9	–10	–19	–19
Interest expense	0	0	0	58
Payments	0	0	0	–581
At June 30, 2019	**528**	**3,386**	**3,914**	**3,941**

The following new, revised or amended standards became effective January 1, 2019, but did not have an impact on these Interim Condensed 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)

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, 2019	June 30, 2018	**June 30, 2019**	Dec. 31, 2018
1 Euro (**EUR**)	**1.1294**	1.1696	**1.1093**	1.1265
1 US dollar (**USD**)	**0.9996**	0.9661	**0.9740**	0.9850
1 British pound (**GBP**)	**1.2935**	1.3293	**1.2349**	1.2546
1 Canadian dollar (**CAD**)	**0.7497**	0.7562	**0.7437**	0.7236

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, 2019**	Dec. 31, 2018
Cash at banks and on hand		
in CHF	8,452	9,111
in EUR	3,510	10,149
in USD	306	2,036
in GBP	306	493
other currencies	124	182
Total at period end	**12,698**	**21,971**

6.2 Restricted cash

In CHF thousands	**June 30, 2019**	Dec. 31, 2018
Long-term	0	1,500
Short-term	3,000	3,000
Total at period end	**3,000**	**4,500**

Restricted cash is designated for interest payments due related to the convertible bonds during the first 3 years (starting 2017, next payments are in 2019 and 2020). 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, 2019, 500,000 Shares were issued out of the authorized share capital. As a result, as of June 30, 2019, the issued nominal share capital amounted to CHF 11,164,563, divided into 11,164,563 Shares at a nominal value of CHF 1 each.

7.2 Authorized share capital

In April 2019, 500,000 Shares were issued out of the authorized share capital in connection with a private placement. On the occasion of the Annual General Meeting (**AGM**) on May 28, 2019, 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 May 27, 2021, through the issuance of up to 3,000,000 Shares with a nominal value of CHF 1 each.

7.3 Conditional share capital

As of June 30, 2019, the Company had conditional share capital, pursuant to which the share capital may be increased by

(i) a maximum amount of CHF 687,552 (2018: CHF 691,302) through the issuance of up to 687,552 (2018: 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 13 "Equity Rights Plans").

(ii) a maximum amount of CHF 2,500,000 (2018: CHF 930,000) by issuing up to 2,500,000 (2018: 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 Financial Liabilities

8.1 Financial current liabilities – Credit line

Early April 2019, the Company secured a credit line facility of up to CHF 15.0 million from a group of private lenders. The credit line served as a short-term loan due for full repayment at December 31, 2019, or any closing of an asset monetization transaction taking place before that date. By June 30, 2019, Santhera had drawn an amount of CHF 5.0 million under the facility. Transaction expenses of CHF 0.3 million were deducted from the drawn amount and were expensed over the duration of the loan. As per June 30, 2019, the undrawn amount of the credit line amounted to CHF 10 million.

The loan bore monthly rising interest rates and was subject to a minimum interest payment of 15% going up to 20% if the line was fully utilized. For the provision of the credit line, Santhera had to pay a commitment fee on the undrawn amount computed at the rate of 40% of the respective interest. Interest and commitment fees were due at repayment and termination of the credit line. The credit line has been repaid in August 2019 (see note 14 "Subsequent Events").

8.2 Financial noncurrent liabilities – Bonds

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, 2019, amounts to CHF 43.2 million (December 31, 2018: CHF 41.7 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, 2019, was at 90% (June 30, 2018: 66%).

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 amount of equity instruments 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, 2018, the value was CHF 0.2 million and at period end CHF 1.4 million (June 30, 2019). The change in the fair value (CHF 1.2 million) was recognized in financial expenses.

Sensitivity analysis:

	June 30, 2019		June 30, 2018	
	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%	–87	+5%	–112
	–5%	70	–5%	146

Changes in liabilities from convertible bonds and derivative financial instruments:

In CHF thousands	Convertible bonds	Derivative financial instruments
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**
Change in fair value of derivative financial instruments	0	–901
Effective interest/amortized cost calculation	744	0
December 31, 2018	54,569	204
Change in fair value of derivative financial instruments	0	1,190
Effective interest/amortized cost calculation	776	0
June 30, 2019	**55,345**	**1,394**

9 Segment and Geographic Information

9.1 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.

9.2 Geographic information

Net sales

Half-year ended June 30, in CHF thousands	**2019**	2018
Europe	18,144	15,956
Rest of the world	171	71
Total	**18,315**	**16,027**

In the reporting period 2019, net sales amounted to CHF 18.3 million (2018: 16.0 million). Raxone was sold in 23 European countries, with the majority of sales generated in France and Germany (in the reporting period 2018, sales went into 20 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, 2019**	Dec. 31, 2018
Switzerland	64,757	63,504
Rest of Europe	969	164
North America	115	68
Total	**65,841**	**63,736**

10 Operating Expenses by Nature

Half-year ended June 30, in CHF thousands	**2019**	2018
External development expenses	–11,731	–12,243
Patent and license expenses	–264	–184
Marketing expenses	–4,257	–5,145
Employee expenses	–18,306	–18,196
Of which non-cash-relevant expenses for share-based payments	*–3,229*	*–3,735*
General and administrative expenses	–2,564	–2,966
Depreciation and amortization	–861	–370
Facility related and lease expenses	–159	–723
Other operating expenses	–66	–56
Total operating expenses	**–38,208**	**–39,883**

11 Income Taxes

Half-year ended June 30, in CHF thousands	**2019**	2018
Current income taxes	–183	–298
Deferred taxes	–218	205
Total	**–401**	**–93**

Movements on deferred taxes relate to temporary differences on inventory.

12 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 2019, the same range of valuation parameters as disclosed in the financial statements as of December 31, 2018, was applied, except for the exercise prices (equal to the Share prices at grant) which were between CHF 6.61 and CHF 14.50. The non-cash-relevant expenses for all unvested SAR and stock options in the reporting period 2019 amounted to CHF 3.2 million (2018: CHF 3.7 million).

12.1 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, the ESARP 2017 and the ESARP 2018, 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 since 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 2019, Santhera has introduced ESARP 2019 in order to set the date of the annual grant to the day after the annual general meeting (instead of January 1 of each year). All other terms remained the same.

In the reporting period ended June 30, 2019, a total of 880,476 SAR with exercise prices between CHF 6.61 and CHF 14.50 were granted. In the half-year period ending June 30, 2018, a total of 617,282 SAR with exercise prices between CHF 18.90 and CHF 36.70 were granted.

Number of SAR outstanding

Half-year ended June 30, number of SAR	**2019**	2018
Outstanding at January 1	**730,388**	**360,110**
Granted[1]	880,476	617,282
Exercised	0	0
Forfeited	–20,717	–129,869
Expired	0	0
Outstanding at June 30	**1,590,147**	**847,523**

[1] The weighted average fair value of the SAR granted during the reporting period in 2019 was CHF 5.71 (in the comparative reporting period 2018 the weighted average fair value of SAR granted was CHF 12.16).

12.2 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.

Since July 1, 2016, no more stock options are available for grants under ESOP 2015 and/or BSOP 2015. Stock option plans were replaced by share appreciation rights (**SAR**).

Number of stock options outstanding

Half-year ended June 30, number of stock options	**2019**	2018
Outstanding at January 1	**262,837**	**288,442**
Granted	0	0
Forfeited	–3,527	–11,556
Expired	0	0
Exercised	0	0
Outstanding at June 30	**259,310**	**276,886**

13 Related Party Transactions

During the reporting period 2019, a total of 78,944 SAR were granted to members of the Board and 175,273 SAR were granted to members of the Executive Management. In the same period in 2018, a total of 62,659 SAR were granted to members of the Board and 134,194 SAR to members of the Executive Management.

14 Subsequent Events

On August 2, 2019, Santhera announced the closing of the licensing transaction with Chiesi Farmaceutici S.p.A., Parma, Italy (Chiesi), whereby Chiesi has in-licensed Raxone for the treatment of Leber's hereditary optic neuropathy (LHON). In connection with this transaction, Santhera received an initial payment of EUR 44 million (approximately CHF 48 million). Additional consideration of up to EUR 49 million becomes payable contingent on the achievement of certain milestones. With the closing, Santhera has licensed its rights for the development, commercialization and distribution of Raxone for the treatment of LHON and any other potential ophthalmological indications to Chiesi for all territories worldwide except the US and Canada. In an interim phase, Santhera will provide support services to Chiesi to enable a seamless handover and will continue to commercialize Raxone for LHON in France.

In August 2019, upon closing of the transaction with Chiesi, the amounts drawn in connection with the credit line facility, the related accrued interest and the commitment fees were repaid (see note 8 "Financial Liabilities").

Building a better working world

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

Basle, September 2, 2019

Report on the review of interim condensed consolidated financial statements

Introduction

We have reviewed the accompanying interim condensed consolidated financial statements (pages 7 to 21) of Santhera Pharmaceuticals Holding AG for the period from January 1, 2019 to June 30, 2019. 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 of the interim condensed consolidated financial statements, which indicates that the Group’s ability to continue operations as planned for the next twelve months depends on cash flows from ongoing product sales, the results of its development activities and the capability to raise additional funds through an ordinary capital increase. This fact together with other factors 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 review conclusion is not modified in respect of this matter.

Ernst & Young Ltd

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

/s/ Karina Gawron
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 neuromuscular and pulmonary diseases with high unmet medical need. Santhera is building a leading Duchenne muscular dystrophy (DMD) franchise. A marketing authorization application for Puldysa® (idebenone) is currently under review by the European Medicines Agency. Santhera has an option to license vamorolone, a first-in-class dissociative steroid currently investigated in a pivotal study in patients with DMD to replace standard corticosteroids. The clinical stage pipeline also includes POL6014 to treat cystic fibrosis (CF) and other neutrophilic pulmonary diseases, as well as omigapil and an exploratory gene therapy approach targeting congenital muscular dystrophies. Santhera out-licensed ex-North American rights to its first approved product, Raxone® (idebenone), for the treatment of Leber's hereditary optic neuropathy (LHON) to Chiesi Group. For further information, please visit www.santhera.com.

Trademarks

Raxone® and Puldysa® are trademarks 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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