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2020

Interim Condensed Report
January to June 2020

Report on the Six Months Ended June 30, 2020, and Interim Condensed Consolidated Financial Statements

Contents

Santhera Announces Financial Results for the First Half-Year 2020 and Updates on Corporate Progress

- **Net revenues of CHF 7.8 million, operating expenses reduced by 16%, net result of CHF -31.8 million**
- **Closing of financing arrangements in April and July to provide in total up to CHF 32 million with the option to increase to up to CHF 44 million**
- **Puldysa® (idebenone) expecting CHMP opinion in Q4-2020 following inclusion of data from SIDEROS trial interim analysis**
- **Vamorolone VISION-DMD study nearing full enrollment and 6-month topline results expected in Q2-2021**
- **Worldwide rights to vamorolone in Duchenne muscular dystrophy and all other indications obtained in September 2020**

Pratteln, Switzerland, September 8, 2020 – Santhera Pharmaceuticals (SIX: SANN) announces the Company's financial results for the first half-year 2020 and provides an update on progress made in advancing its lead compounds for the treatment of Duchenne muscular dystrophy (DMD).

"Our primary focus continues to be on advancing our business towards several significant inflection points with our DMD drug candidates, Puldysa and vamorolone, and securing sufficient funds to enable the ongoing implementation of our growth plans," said **Dario Eklund, Chief Executive Officer of Santhera**. "After having successfully secured all rights to vamorolone, we now have a DMD franchise that has the potential to address the unmet medical needs of a broad patient population. For Puldysa, we intend to present the data from the planned interim analysis of the SIDEROS study to the European regulators and, subject to a positive CHMP opinion in the fourth quarter 2020, aim at a launch in Europe in the first quarter 2021. Whilst for vamorolone, our second DMD drug candidate, we now expect read-out of topline data from the pivotal VISION-DMD trial in the second quarter 2021 due to the Covid-19-related delays."

He added: "Our sincere thanks go to all clinical trial participants, their families and carers, for their commitment to advancing our drug candidates in minimizing the delays under the severe constraints imposed by the Covid-19 pandemic. Likewise, we are also appreciative of the investor support which allowed us to obtain additional financing under challenging market conditions. I am confident that the upcoming value-enhancing inflection points will trigger further interest in Santhera and facilitate the raising of additional finance to support ongoing development and commercialization activities."

KEY FINANCIALS

- Net revenues from product sales of CHF 7.8 million
- Operating expenses of CHF -31.9 million, reduced by 16%
- Net result of CHF -31.8 million (1H-2019: CHF -26.9 million)
- Cash and cash equivalents of CHF 19.4 million (June 30, 2020)
- Operating cash flow CHF -19.8 million

Sales of Raxone phasing out after transfer of business to Chiesi Group

In the first six months 2020, Santhera reported revenue from contracts with customers of CHF 7.8 million (1H-2019: CHF 18.3 million). The majority of this revenue reflects sales of Raxone for the treatment of Leber's hereditary optic neuropathy (LHON) in France, where Santhera continues to market the product following the out-licensing and transfer to Chiesi Group. As previously announced, Chiesi Group has in-licensed Raxone for LHON and all other ophthalmologic indications for all territories worldwide, except the US and Canada, for a total consideration of up to CHF 105 million of which CHF 46.4 million was recognized as revenue in 2019.

Operating cost reductions of 16%

As a result of rigorous cost saving measures, total operating expenses for the first half 2020 decreased by 16% year-on-year to CHF 31.9 million (1H-2019: CHF 38.2 million). Development expenses were down 8% to CHF 17.7 million (1H-2019: CHF 19.3 million) and reflect primarily costs for ongoing late stage clinical studies, including the Phase 3 SIDEROS trial in DMD, and efforts associated with the pending marketing authorization application for Puldysa for DMD in Europe. Marketing and sales expenditures declined by 42% to CHF 6.8 million (1H-2019: CHF 11.6 million) as commercial activities were rescheduled and aligned with the expected market entry of Puldysa in the first quarter 2021. General and administrative expenses of CHF 7.2 million were unchanged year-on-year (1H-2019: CHF 7.2 million). Overall, the Company reported an operating result of CHF -25.9 million (1H-2019: CHF -22.4 million). Decreased revenues following the out-licensing were partially compensated by cost reduction measures, resulting in a net result of CHF -31.8 million (1H-2019: CHF -26.9 million).

Recent financings provide additional liquidity

As of June 30, 2020, cash and cash equivalents amounted to CHF 19.4 million (December 31, 2019: CHF 31.4 million). In recent months, Santhera successfully secured additional funds which provide a runway to advance value-enhancing developments and pre-commercialization activities for the neuromuscular compounds Puldysa and vamorolone.

In April, Santhera entered into a financing arrangement with IRIS (France) in the initial gross amount of up to CHF 12 million over 12 months, with the extension option for another CHF 12 million over the following 12 months. As of June 30, 2020, the Company had received gross proceeds of CHF 8 million from the arrangement with IRIS. Post period end, on July 14, 2020, Santhera closed an up to CHF 20 million financing facility with a fund managed by Highbridge Capital Management, LLC, an existing investor in the Company. As of September 7, 2020, the Company has received the first tranche of CHF 7.5 million from this arrangement with Highbridge Capital Partners, LLC, CHF 5 million of which has been repaid via the exchange for Company shares. Additional tranches of up to CHF 12.5 million may be drawn contingent on the achievement of milestones. The conditional and authorized capital approved in April 2020 by the Annual General Meeting was in part used to serve these equity-linked financing arrangements.

In parallel, the Company is evaluating a restructuring of the CHF 60 million Senior Unsecured Convertible Bonds with a February 2022 maturity.

For ongoing development activities, the preparation for commercial launch of Puldysa and the payments due following the exercise of the sub-license option for vamorolone, Santhera will require further additional funding.

From September 21, 2020, Santhera will be included in both of the SIX Swiss Exchange healthcare indices: SXI Life Sciences®, which includes pharmaceutical, biotechnology and medical technology companies, and its more narrowly defined sub-index SXI Bio+Medtech®, focused on biotech and medtech companies, which is expected to further enhance investor visibility.

PIPELINE MILESTONES AND PROGRESS

With Puldysa and vamorolone, Santhera is building a complementary DMD product portfolio. The Company expects the availability of both vamorolone and Puldysa to address the medical needs of DMD patients, from early to late disease stages, irrespective of age, underlying dystrophin mutation or ambulatory status. Santhera's pipeline priorities for the remainder of 2020 continue to focus on advancing its neuromuscular franchise in DMD, with Puldysa and vamorolone, towards value-enhancing inflection points:

- September 2, 2020: Santhera obtains worldwide rights to vamorolone in all indications
- Q4-2020: Interim analysis of SIDEROS study and inclusion of data into regulatory dossier
- Q4-2020: CHMP opinion on marketing authorization application for Puldysa in DMD in Europe
- Q1-2021: Launch of Puldysa in first European markets
- Q2-2021: Read-out of topline data of pivotal Phase 2b trial for vamorolone in DMD
- Q4-2021: NDA (new drug application) filing in the US for vamorolone in DMD

Puldysa—ahead of DMD SIDEROS interim analysis and CHMP opinion

Puldysa highlights in the first half-year were the renewal of the Early Access to Medicines Scheme (EAMS) scientific opinion in the UK and the completion of enrollment into the Phase 3 SIDEROS study in DMD. With the EAMS renewal for another year, the UK's Medicines and Healthcare products Regulatory Agency (MHRA) has confirmed its positive scientific opinion for idebenone enabling continued pre-approval access to idebenone for patients with DMD in respiratory function decline who are not taking glucocorticoids. In May, Santhera announced full recruitment into the SIDEROS study and its intention of conducting an interim analysis by the independent Data and Safety Monitoring Board (DSMB), subject to approval of the necessary study protocol amendment. If positive results are shown, the additional clinical data will be included into the European conditional marketing authorization application for which the Company now expects a CHMP opinion in the fourth quarter of 2020. Simultaneously, preparations for market entry will be advanced to allow for a launch in Europe in the first quarter 2021, subject to timely product approval. In the US, a positive outcome of the interim analysis, followed by an early completion of the SIDEROS study, could allow acceleration of a regulatory filing with the FDA by approximately one year. Santhera estimates the peak sales potential for Puldysa to be in excess of USD 500 million in the US and the largest five EU countries.

Vamorolone—encouraging new long-term clinical data and VISION-DMD nearly fully enrolled

In June, Santhera's partner ReveraGen completed a long-term, open-label 24-month extension study (VBP15-LTE) in patients with DMD. Enrolled in this study were patients who had previously completed a 6-month dose escalation study (VBP15-003) which demonstrated dose-dependent improvement in timed function tests and good

tolerability. Taken together, ReveraGen has now obtained safety and efficacy data with vamorolone over a period of 2.5 years in 41 boys with DMD. Currently ongoing is the pivotal VISION-DMD study which compares the efficacy and tolerability profile of vamorolone versus placebo and prednisolone to determine whether vamorolone improves muscle strength and function compared to placebo and whether it has less side effects compared to prednisolone, thereby potentially making it a valuable alternative to standard corticoid treatments. The next study milestone will be the soon expected full enrollment into the study followed by topline 6-month data readout anticipated in the second quarter of 2021 which, if positive, could allow for an NDA filing in the fourth quarter of 2021.

Vamorolone—worldwide rights for all indication obtained

On September 2, 2020, Santhera announced the signing of agreements with Idorsia (SIX: IDIA) and ReveraGen BioPharma Inc., making Santhera a direct license holder of vamorolone [1]. Under the agreements, Santhera has obtained an exclusive license from ReveraGen, the originator of vamorolone, for all indications worldwide. The agreements create further value for Santhera through the transfer of rights for the previously excluded markets Japan and South Korea, the right to grant sublicenses and a share in the expected Priority Review Voucher. Under the amended terms, Santhera expects a reduction in cash outflow in the range of USD 18-24 million in the next 12-18 months. Santhera estimates the peak sales potential for vamorolone for the DMD indication alone to be in excess of USD 500 million in the US and the largest five EU countries.

Progress made with earlier pipeline projects

Santhera is advancing its Phase 1b study with clinical stage candidate lonodelestat for cystic fibrosis. The compound's potential as a therapeutic intervention for COVID-19-related acute respiratory distress syndrome (ARDS) is investigated in preclinical research by Cold Spring Harbor Laboratory (CSHL) under a collaboration agreement. The pipeline also includes a discovery-stage gene therapy approach for which Santhera signed agreements with University Basel and Rutgers University as part of its program to advance gene therapy research for the treatment of LAMA2-deficient congenital muscular dystrophy (LAMA2 MD or MDC1A). As an ongoing business development activity, Santhera is evaluating further diversification of its platform-type pipeline products vamorolone and lonodelestat into additional indications which may include partnering with other companies.

Outlook

The operational priorities for Santhera in the second half of 2020 are the preparation for European market entry of Puldysa in DMD in early 2021, advancing vamorolone towards the VISION-DMD top-line data readout and securing additional funding to allow the Company to pursue its operations as planned.

Reference:

[1] Press release "Santhera Exercises Option to Obtain Worldwide Rights to Vamorolone in Duchenne Muscular Dystrophy and All Other Indications", September 2, 2020, accessible here.

Interim Condensed Consolidated Financial Statements

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Interim Consolidated Balance Sheet

In CHF thousands	Notes	**June 30, 2020 (reviewed)**	Dec. 31, 2019 (audited)
Assets			
Tangible assets		5,084	5,604
Intangible assets		56,918	58,479
Financial assets long-term		621	664
Deferred tax assets		897	1,049
Noncurrent assets		**63,520**	**65,796**
Prepaid expenses		2,174	637
Inventories	5	6,906	6,859
Trade and other receivables		8,121	8,901
Restricted cash short-term	6	0	1,500
Cash and cash equivalents	6	19,353	31,358
Current assets		**36,554**	**49,255**
Total assets		**100,074**	**115,051**
Equity and liabilities			
Share capital	7	13,185	11,165
Capital reserves and share premium		457,692	448,084
Retained earnings		-465,067	-433,240
Employee benefit reserve		-3,028	-3,160
Treasury shares		-1,625	-745
Other components of equity		-947	-857
Total equity		**210**	**21,247**
Convertible bonds	8	56,997	56,154
Noncurrent derivative financial instruments	8	278	617
Noncurrent contract liabilities		563	1,126
Noncurrent lease liabilities		2,507	2,827
Pension liabilities		9,549	9,116
Total noncurrent liabilities		**69,894**	**69,840**
Trade and other payables		10,615	9,532
Accrued expenses		14,441	11,427
Income tax payables		348	395
Current contract liabilities		1,569	1,597
Current lease liabilities		1,038	1,013
Current convertible notes (IRIS)	8	1,100	0
Current derivative financial instruments (IRIS)	8	859	0
Total current liabilities		**29,970**	**23,964**
Total liabilities		**99,864**	**93,804**
Total equity and liabilities		**100,074**	**115,051**

Interim Consolidated Income Statement (Reviewed)

For the half-year ended June 30, in CHF thousands	Notes	2020	2019
Net sales	9	6,133	18,315
Net sales to licensing partner	9	1,642	0
Revenue from contracts with customers		**7,775**	**18,315**
Cost of goods sold		-2,114	-2,557
Of which amortization intangible asset		*-1,519*	*-1,519*
Other operating income		357	16
Development	10	-17,688	-19,325
Marketing and sales	10	-6,766	-11,611
General and administrative	10	-7,209	-7,206
Other operating expenses	10	-248	-66
Operating expenses	**10**	**-31,911**	**-38,208**
Operating result		**-25,893**	**-22,434**
Financial income	11	547	859
Financial expenses	11	-6,120	-4,924
Result before taxes		**-31,466**	**-26,499**
Income taxes	12	-361	-401
Net result		**-31,827**	**-26,900**
Basic and diluted loss per share (in CHF)		-2.78	-2.47

Interim Consolidated Statement of Comprehensive Income (Reviewed)

For the half-year ended June 30, in CHF thousands	2020	2019
Net result	**-31,827**	**-26,900**
Items never to be reclassified subsequently to net income in subsequent periods:		
Net actuarial gains/(losses) from defined benefit plans	132	-1,628
Items to be reclassified subsequently to net income in subsequent periods:		
Currency translation differences	-90	-42
Other comprehensive result	**42**	**-1,670**
Total comprehensive result	**-31,785**	**-28,570**

Interim Consolidated Statement of Cash Flows (Reviewed)

For the half-year ended June 30, in CHF thousands	Notes	2020	2019
Result before taxes		**-31,466**	**-26,499**
Depreciation of tangible assets		789	827
Amortization of intangible assets		1,567	1,553
Expenses for equity rights plans		1,749	3,229
Change in fair value of derivatives	8	-339	1,190
Change in pension liabilities		565	335
Taxes paid		-208	-183
Change in net working capital		3,562	-3,173
Total financial result	11	5,574	4,065
Interest received		1	8
Interest paid		-1,589	-1,571
Cash flow from operating activities		**-19,795**	**-20,219**
Investments in tangible assets		-24	-24
Investments in intangible assets		-5	-28
Change in investments in other financial assets long-term		35	0
Change in restricted cash		1,500	1,500
Cash flow from investing activities		**1,506**	**1,448**
Capital increase		0	7,125
Proceeds from sale of treasury shares		598	1,222
Purchase of treasury shares		-633	-1,155
Proceeds from convertible notes (IRIS)	8	6,983	0
Proceeds from current loan	8	0	4,732
Payment of lease liabilities		-543	-523
Cost of issuance share capital		0	-1,936
Cash flow from financing activities		**6,405**	**9,465**
Effects of exchange rate changes on cash and cash equivalents		-121	33
Net increase/(decrease) in cash and cash equivalents		**-12,005**	**-9,273**
Cash and cash equivalents at January 1		31,358	21,971
Cash and cash equivalents at June 30		**19,353**	**12,698**

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, 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		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**
Balance at January 1, 2020		**11,165**	**448,084**	**-433,240**	**-3,160**	**-745**	**-857**	**21,247**
Net result		0	0	-31,827	0	0	0	**-31,827**
Other comprehensive income		0	0	0	132	0	-90	**42**
Total comprehensive result for the period		**0**	**0**	**-31,827**	**132**	**0**	**-90**	**-31,785**
Share-based payment transactions	10	0	1,749	0	0	0	0	**1,749**
Capital increase from conversions of loans (IRIS)	8	1,945	7,484	0	0	-1,036	0	**8,393**
Capital increase	7	75	566	0	0	0	0	**641**
Change in treasury shares		0	-191	0	0	156	0	**-35**
Balance at June 30, 2020		**13,185**	**457,692**	**-465,067**	**-3,028**	**-1,625**	**-947**	**210**

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 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 7, 2020.

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, 2019, except for the changes in accounting policies as noted further 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, 2019.

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 ability to continue operations

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**) with the European Medicines Agency (**EMA**). The opinion by the Committee for Medicinal Products for Human Use (**CHMP**) for this CMA filing is expected in Q4 2020 and, upon receiving marketing authorization (**MA**), Santhera intends to launch Puldysa in Europe in the first quarter 2021. The Company is also expecting the topline results of the pivotal Phase 2b study with vamorolone in ambulant patients with DMD, being conducted by ReveraGen in Q2-2021.

During April 2020, the Company entered into an agreement with IRIS, Paris, France, for the issuance of and subscription to warrants in the initial gross amount of CHF 12 million giving access to notes which are convertible into shares over a period of 12 months. The Company has the option to extend the financing in the aggregate gross amount of up to additional CHF 12 million over a further period of up to 12 months after the initial period, under similar terms and conditions. Refer to note 8 "Financial Liabilities" for further details of this financing arrangement.

In addition, during July 2020 the Company entered into an agreement with Highbridge Capital Partners LLC, to provide funding of up to CHF 20 million, of which the first tranche of CHF 7.5 million has been received as of September 7, 2020, the date of these financial statements. Additional tranches of up to CHF 12.5 million may be drawn on the achievement of certain milestones. Refer to note 15 “Subsequent Events” for further details.

On September 2, 2020 Santhera announced that Idorsia has assigned its original agreement with ReveraGen to Santhera. Santhera has thus become a direct contracting party with ReveraGen and with a signed early option exercise this allows Santhera to gain exclusive and immediate access to vamorolone and defers some early milestone-related payments until after study readout. Under the amended terms, Santhera expects a reduction in cash outflow in the range of USD 18-24 million in the next 12-18 months. Refer to note 15 “Subsequent Events” for further details.

Santhera’s cash and cash equivalents amounted to CHF 19.4 million as of June 30, 2020. Due to cost saving measures already implemented, these funds together with available funds available from existing financing agreements, are expected to fund the operations of the Company to the CHMP opinion on Puldysa, a significant value inflection point, expected in the fourth quarter 2020, following inclusion of data from SIDEROS trial interim analysis. In the event that the CHMP decision is positive, the Company plans to raise additional financing in the fourth quarter of 2020 mainly to fund the preparation for the commercial launch of Puldysa in early 2021.

In case of a negative CHMP opinion, the Management and the Board would immediately initiate restructuring measures to defer Puldysa commercialization activities and further significantly reduce its cost base. In such scenario of a negative CHMP decision and restructuring measures, the Management and the Board in order to reach the vamorolone pivotal study 6-month topline readout expected in the second quarter 2021 would also seek additional funding that may include renegotiation of certain existing financing arrangements. However, because executing the Company’s strategy depends on further funding to ensure the continuation of its operations, material uncertainties remain as to the Company’s ability to continue as a going concern until June 30, 2021.

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 operations through June 30, 2021. This material uncertainty may cast significant doubts about the going concern of the Company.

However, the Management and the Board of Directors believe that the Company is prepared to secure additional funds needed in order to operate its business as planned with the objective to meet all of its obligations until June 30, 2021. 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.

The following new, revised or amended standards became effective January 1, 2020, but did not have an impact on these Interim Condensed Consolidated Financial Statements.

- Amendments 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)
- Various Amendments to References to Conceptual Framework in IFRS Standards (effective January 1, 2020)

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, 2020	June 30, 2019	**June 30, 2020**	Dec. 31, 2019
1 Euro (**EUR**)	**1.0639**	1.1294	**1.0679**	1.0858
1 US dollar (**USD**)	**0.9660**	0.9996	**0.9518**	0.9683
1 British pound (**GBP**)	**1.2179**	1.2935	**1.1684**	1.2739
1 Canadian dollar (**CAD**)	**0.7085**	0.7497	**0.6955**	0.7430

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 and Puldysa.

6 Cash and Cash Equivalents and Restricted Cash

6.1 Cash and cash equivalents

In CHF thousands	**June 30, 2020**	Dec. 31, 2019
Cash at banks and on hand		
in CHF	12,667	19,285
in EUR	5,363	7,875
in USD	734	1,243
in GBP	431	2,769
other currencies	158	186
Total at period end	**19,353**	**31,358**
Of which: Short-term deposits		
In CHF	117	127

6.2 Restricted cash

In CHF thousands	**June 30, 2020**	Dec. 31, 2019
Short-term	0	1,500
Total at period end	**0**	**1,500**

Restricted cash was kept in escrow with the bond agent and was designated for interest payments due related to the convertible bonds during the first 3 years (starting 2017, last payment in February 2020).

7 Share Capital

7.1 Ordinary share capital

During the reporting period ending June 30, 2020, 1,945,000 Shares were issued out of the authorized share capital for financing arrangements in connection with IRIS (see note 8 "Financial Liabilities"); 75,000 Shares were issued in connection the exchangeable notes with Highbridge Capital Management, LLC (**Highbridge**) (see note 15 "Subsequent Events"). As a result, as of June 30, 2020, the issued nominal share capital amounted to CHF 13,185,063, divided into 13,185,063 Shares at a nominal value of CHF 1 each.

7.2 Authorized share capital

In April and June 2020, the aggregate amount of 2,020,000 Shares was issued out of the authorized share capital in connection with financing arrangements. On the occasion of the Annual General Meeting (**AGM**) on April 22, 2020, 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 21, 2022, through the issuance of up to 4,630,000 Shares with a nominal value of CHF 1 each.

7.3 Conditional share capital

As of June 30, 2020, the Company had conditional share capital, pursuant to which the share capital may be increased by

(i) a maximum amount of CHF 687,052 (2019: CHF 687,552) through the issuance of up to 687,052 (2019: 687,552) 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 4,800,000 (2019: CHF 2,500,000) by issuing up to 4,800,000 (2019: 2,500,000) 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 noncurrent liabilities – Convertible 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 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 (volume weighted average price) of the Shares is at least 160% of the conversion price. The convertible bonds are measured at amortized costs applying the effective interest method. The fair value of the bond (Level 1) at June 30, 2020, amounts to CHF 23.1 million (December 31, 2019: CHF 39.5 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, 2020, was 91% (June 30, 2019: 90%).

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, 2019, the value was CHF 0.6 million and at period end CHF 0.3 million (June 30, 2020). The change in the fair value (CHF 0.3 million) was recognized in financial income.

Sensitivity analysis:

	June 30, 2020		June 30, 2019	
	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%	-32	+5%	-87
	-5%	60	-5%	70

8.2 Financial current liabilities – Equity-linked Financing with IRIS

On April 8, 2020, Santhera entered into an agreement with IRIS, Paris, France, for the issuance of and subscription to warrants in the initial gross amount of CHF 12 million giving access to notes which are convertible into shares over a period of 12 months. The financing has been established to provide additional liquidity in support of the Company's ongoing regulatory and development programs. Santhera has the option to extend the financing in the aggregate gross amount of up to additional CHF 12 million over a further period of up to 12 months after the initial period, under similar terms and conditions.

As a result of the implementation of the contractual agreements Santhera issued for free 4,800 warrants in favor of IRIS. These warrants entitle IRIS to subscribe to convertible notes under certain conditions. Subscriptions are made in bundles with the exercise of at least 400 warrants which is equal to CHF 1 million. One convertible note's principal amount is CHF 2,500 and the note does not bear interest. The subscription price for a convertible note for IRIS is 97% of the principal amount. Each note is convertible at the discretion of its holder into a number of shares of Santhera. If a note is not converted optionally, it is mandatorily convertible 18 months after issuance of the relevant note. The pricing of the conversion of a note is at market, whereby during the period of five consecutive trading days, preceding a notice date for conversion, the lowest VWAP (volume weighted average price) is identified, providing that such value may not be lower than CHF 5. A relevant percentage of 94% or 95% is applied to this lowest VWAP, which is then the divisor for the number of shares eligible for IRIS in the event of a conversion of notes. IRIS is selling the shares on the market or in block trades. The shares used for conversion are taken from Santhera's treasury shares and shall immediately bear the same rights of all other existing shares and can be traded at the SIX.

The warrants issued under the agreement are measured at fair value through profit or loss considering the 3% discount on exercise as well as the discount and terms of the conversion feature of the convertible notes to which the warrants give right to. As of June 30, 2020, 3,200 of the 4,800 warrants issued were exercised, the fair value of the 1,600 outstanding warrants amounted to TCHF 657 and is included in current derivative financial instruments. Based on certain conditions it is at the discretion of IRIS to decide on the timing for the consecutive exercise of warrants and hence subscription of convertible notes.

The convertible notes are classified as a hybrid contract containing a host that is a financial liability and embedded derivatives separated from the host and measured at fair value with all changes in fair value recognized in profit or loss.

Both, the warrants and the embedded financial derivatives (e.g. discounts of 3% (warrants) and 5% or 6% (conversion right) respectively, conversion right based on lowest VWAP of five preceding trading days) are valued by an independent consultant initially and at period end at fair value, applying a simulation-based valuation approach. The valuation of the warrants and embedded financial 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 the historical volatility of the past five years which is longer than the remaining life of the convertible notes. This approach was chosen due to the high historical volatility of Santhera shares. The volatility used as per June 30, 2020, was 76%.

The embedded derivatives of the convertible notes are closely related to each other and are therefore accounted for as a single instrument (i.e. a compound derivative). Due to the conversion based on market share price, the conversion right triggers a variable number of shares and the embedded derivatives are therefore classified as a financial liability.

The carrying amount of the host contract at initial recognition is the difference between the carrying amount of the hybrid contract and the fair value of the embedded derivatives. The host is then subsequently measured at amortized cost, using the effective interest rate method. However, as the conversions of the convertible notes are soon after the warrant exercise date, Santhera has expensed the transaction costs in profit or loss.

As of June 30, 2020, the carrying amount of the host for notes issued but not yet converted amounted to TCHF 1,100 and is included in the balance sheet under current liabilities. The fair value of the embedded derivatives of the outstanding notes amounted to TCHF 202 and is included in current derivative financial instruments.

Expenses related to fair value measurement of warrants and embedded derivatives of TCHF 2,591 as well as transaction cost of TCHF 777 were recorded as financial expenses in profit or loss.

Sensitivity analysis:

	June 30, 2020	
	Increase/decrease in volatility assumption	Effect on result before taxes in CHF thousands
Change in volatility	+5%	-52
	-5%	52

Changes in liabilities from convertible bonds, current loan, convertible notes and their derivative financial instruments:

In CHF thousands	Convertible bonds	Noncurrent derivative financial instruments	Current loan	Convertible notes	Current derivative financial instruments
December 31, 2018	**54,569**	**204**	**0**	**0**	**0**
Proceeds from current loan	0	0	4,732	0	0
Change in fair value of derivative financial instruments	0	1,190	0	0	0
Effective interest/amortized cost calculation	776	0	0	0	0
June 30, 2019	**55,345**	**1,394**	**4,732**	**0**	**0**
Change in fair value of derivative financial instruments	0	-777	0	0	0
Effective interest/amortized cost calculation	809	0	0	0	0
Repayment of current loan	0	0	-4,732	0	0
December 31, 2019	**56,154**	**617**	**0**	**0**	**0**
Proceeds from convertible notes	0	0	0	6,983	0
Cash flows in 2020				**6,983**	**0**
Non-cash changes					
Initial recognition of derivative financial instruments	0	0	0	0	2,579
Derecognition of derivative financial instruments on warrants exercise	0	0	0	240	-240
Nominal value of convertible notes converted into shares	0	0	0	-6,900	0
Derecognition of derivative financial instruments on conversion of notes	0	0	0	0	-1,492
Change in fair value of derivative financial instruments	0	-339	0	0	12
Effective interest/amortized cost calculation/amortization of transaction cost	843	0	0	777	0
June 30, 2020	**56,997**	**278**	**0**	**1,100**	**859**

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 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 (Leber's hereditary optic neuropathy) and sales to licensing partner. Geographic revenue information is based on location of the customer.

9.2 Geographic information

Revenue from contracts with customers

Half-year ended June 30, in CHF thousands	**2020**	2019
Net sales		
EU	6,133	18,144
Rest of the world	0	171
Subtotal net sales	6,133	18,315
Net sales to licensing partner		
EU	1,642	0
Total	**7,775**	**18,315**

In the reporting period 2020, net sales amounted to CHF 6.1 million (2019: 18.3 million). Raxone was sold in 5 European countries, with the majority of sales generated in France (in the reporting period 2019, prior to the execution of the outlicensing agreement with Chiesi, sales went into 23 European countries, with a majority of the sales in France and Germany).

Noncurrent assets (excluding financial instruments and deferred tax assets)

In CHF thousands	**June 30, 2020**	Dec. 31, 2019
Switzerland	60,826	62,874
EU	980	949
North America	196	260
Total	**62,002**	**64,083**

10 Operating Expenses by Nature

Half-year ended June 30, in CHF thousands	**2020**	2019
External development expenses	-10,989	-11,731
Patent and license expenses	-266	-264
Marketing expenses	-1,692	-4,257
Employee expenses	-15,201	-18,306
Of which non-cash-relevant expenses for share-based payments	*-1,749*	*-3,229*
General and administrative expenses	-2,562	-2,564
Depreciation and amortization	-851	-861
Facility related and lease expenses	-102	-159
Other operating expenses	-248	-66
Total operating expenses	**-31,911**	**-38,208**

11 Financial Income/Expenses

Financial income

Half-year ended June 30, in CHF thousands	**2020**	2019
Interests on cash and cash equivalents	1	8
Change in fair value of financial derivative instruments	339	0
Realized and unrealized foreign exchange gains	207	851
Total	**547**	**859**

Financial expenses

Half-year ended June 30, in CHF thousands	**2020**	2019
Interest expenses	-2,381	-2,674
Interest expenses on lease liabilities	-51	-58
Change in fair value of financial derivative instruments	0	-1,190
Initial recognition of financial derivative instruments (IRIS)	-2,591	0
Transaction cost financial instruments (IRIS)	-777	0
Realized and unrealized foreign exchange losses	-320	-1,002
Total	**-6,120**	**-4,924**

12 Income Taxes

Half-year ended June 30, in CHF thousands	**2020**	2019
Current income taxes	-208	-183
Deferred taxes	-153	-218
Total	**-361**	**-401**

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 2020, the similar range of valuation parameters as disclosed in the financial statements as of December 31, 2019, was applied, except for the exercise prices (equal to the Share prices at grant) which were between CHF 7.22 and CHF 7.96. The non-cash-relevant expenses for all unvested SAR and stock options in the reporting period 2020 amounted to CHF 1.7 million (2019: CHF 3.2 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, the ESARP 2017, the ESARP 2018 and the ESARP 2019, 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 generally 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 the reporting period ended June 30, 2020, a total of 1,112,467 SAR with exercise prices between CHF 7.22 and CHF 7.96 were granted. In the half-year period ending June 30, 2019, a total of 880,476 SAR with exercise prices between CHF 6.61 and CHF 14.50 were granted.

Number of SAR outstanding

Half-year ended June 30, number of SAR	**2020**	2019
Outstanding at January 1	**1,757,514**	**730,388**
Granted[1]	1,112,467	880,476
Exercised	0	0
Forfeited	-65,948	-20,717
Expired	0	0
Outstanding at June 30	**2,804,033**	**1,590,147**

[1] The weighted average fair value of the SAR granted during the reporting period in 2020 was CHF 3.28 (in the comparative reporting period 2019 the weighted average fair value of SAR granted was CHF 5.71).

13.2 Stock Option Plans

No stock option grants are made anymore. Stock option plans were replaced by equity plans with share appreciation rights.

Number of stock options outstanding

Half-year ended June 30, number of stock options	**2020**	2019
Outstanding at January 1	**257,604**	**262,837**
Granted	0	0
Forfeited	-131	-3,527
Expired	0	0
Exercised	0	0
Outstanding at June 30	**257,473**	**259,310**

14 Related Party Transactions

During the reporting period 2020, a total of 165,332 SAR were granted to members of the Board and 283,127 SAR were granted to members of the Executive Management. In the same period in 2019, a total of 78,944 SAR were granted to members of the Board and 175,273 SAR to members of the Executive Management.

15 Subsequent Events

On July 14, 2020, Santhera announced the closing of the first tranche of a financing transaction in the amount of CHF 7.5 million nominal with a fund managed by Highbridge Capital Management, LLC (**Highbridge**), as previously announced on June 4, 2020. Under the financing instrument, Santhera may borrow up to an aggregate amount of CHF 20 million through the issuance of senior secured exchangeable notes. In connection with this agreement, Santhera agreed to issue 300,000 shares of the Company, as consideration for the commitment and utilization of the financing (with 75,000 of such shares being issued upon signing of the commitment in June 2020). The exchangeable notes have a maximum term of 18 months and will pay a fixed interest, which Santhera can pay in cash at a rate of 12% per annum or in kind at a rate of 13% per annum. Subject to certain restrictions, Highbridge may elect to exchange notes for Company shares. Also, Santhera has an option to redeem the notes in shares under certain circumstances. As of September 7, 2020, CHF 5 million of the initial tranche of CHF 7.5 million has been repaid via the exchange for Company shares.

On September 2, 2020 Santhera announced that Idorsia has assigned its original agreement with ReveraGen to Santhera. Santhera has thus become a direct contracting party with ReveraGen and with a signed early option exercise this allows Santhera to gain exclusive and immediate access to vamorolone and defers some early milestone-related payments until after study readout. Under the terms of the agreements now signed with ReveraGen and Idorsia, Santhera has obtained an exclusive license, including sublicensing rights, for vamorolone in all indications and all territories worldwide, now also including Japan and South Korea. Additionally, ReveraGen is the holder of a Rare Pediatric Disease designation, which may result in receipt of a Priority Review Voucher upon approval of vamorolone for DMD. Santhera will have a share in any revenues of a potential sale of such a voucher.

The 2018 license agreements were revised and include the following term amendments: As consideration for the assignment of its licensing option for vamorolone to Santhera, Idorsia received 366,667 Santhera shares, increasing Idorsia's equity position in Santhera to close to 12%. Milestone payments by Santhera up to and including potential FDA approval will be reduced by USD 18 million to USD 72 million (previously USD 90 million). Santhera's obligations are a payment of up to USD 7 million, payable in monthly instalments of up to USD 500,000 to ReveraGen, to fund development including the Phase 2b VISION-DMD study; USD 5 million to ReveraGen at the time when FDA supports an NDA filing with Phase 2b 6-month data; a non-interest bearing exchangeable note to Idorsia in the amount of CHF 10 million; and USD 50 million (previously USD 60 million) in total for FDA approval. Since the exchangeable note is payable up to 65% in Santhera shares, at Santhera's discretion, this could potentially reduce cash outlay by an additional USD 6.5 million. Furthermore, Santhera will receive 10% of any potential proceeds that could arise from the monetization of the expected Priority Review Voucher. Upon achievement of the first USD 100 million annual revenue, an additional USD 5 million milestone payment is due to Idorsia.

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

Basle, September 7, 2020

Report on the review of interim condensed consolidated financial statements

Introduction

We have reviewed the interim condensed consolidated financial statements (consolidated balance sheet, consolidated income statement, consolidated statement of comprehensive income, consolidated statement of cash flows, consolidated statement of changes in equity and notes, pages 8 to 24) of Santhera Pharmaceuticals Holding AG for the period from January 1, 2020 to June 30, 2020. 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 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 review conclusion is not modified in respect of this matter.

Ernst & Young Ltd

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

/s/ Diana Vejina
ACCA

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 Duchenne muscular dystrophy (DMD) product portfolio to treat patients from early to late disease stages, irrespective of causative mutations, ambulatory status or age. A marketing authorization application for Puldysa® (idebenone) is currently under review by the European Medicines Agency. Santhera has an exclusive license for all indications worldwide to vamorolone, a first-in-class anti-inflammatory drug candidate with novel mode of action, currently investigated in a pivotal study in patients with DMD as an alternative to standard corticosteroids. The clinical stage pipeline also includes lonodelestat (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. Further information at www.santhera.com.

Trademarks

Puldysa® and Raxone® 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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