2022

Interim Condensed Report
January to June 2022

Report on the Six Months Ended June 30, 2022 and Interim Consolidated Financial Statements

Contents

Santhera Announces Half-Year 2022 Financial Results and Provides Corporate Update

- Revenue from contracts with customers of CHF 6.3 million (H1-2021: CHF 4.5 million)
- Operating result of CHF -25.5 million (H1-2021: CHF -19.5 million) and net result of CHF -29.7 million (H1-2021: CHF -20.5 million)
- Cash and cash equivalents of CHF 12.7 million (June 30, 2022), together with existing facilities enabling cash reach into Q1-2023
- Key milestones reached with U.S. and EU regulatory submissions (NDA, MAA) for vamorolone in Duchenne muscular dystrophy (DMD)
- Financing initiatives ongoing to support vamorolone launch and other ongoing activities

Pratteln, Switzerland, October 31, 2022 – Santhera Pharmaceuticals (SIX: SANN) announces the Company's financial results for the six months ended June 30, 2022, reports on the regulatory and clinical progress with its lead drug candidate vamorolone for the treatment of DMD in the U.S. and Europe, and provides updates on its financing initiatives.

"The year 2022 to date was fully geared towards registration and approval of vamorolone in Duchenne muscular dystrophy (DMD) and I am delighted that, over the last month, we submitted and received validation for a marketing authorization application (MAA) in the EU followed by completion of the rolling submission of a new drug application (NDA) in the U.S. This represents a tremendous achievement for Santhera and a major step towards our goal of bringing this investigational therapy to patients living with DMD," said Dario Eklund, CEO of Santhera. "With equally high priority, we are pursuing additional near-term financing, primarily to allow us to fund market entry preparations for vamorolone. We are evaluating various non-dilutive options including but not limited to licensing agreements and monetization of assets in addition to debt and royalty financing and, depending on market conditions, may also consider equity-based funding options."

PIPELINE MILESTONES AND PROGRESS REVIEW

Half-year key events and post-period updates

- New drug application (NDA) submitted to the U.S. Food and Drug Administration (FDA) for vamorolone in DMD
- Marketing authorization application (MAA) submitted to and validated by the European Medicines Agency (EMA) for vamorolone in DMD
- Efficacy, safety and bone health data with vamorolone published in JAMA Neurology and presented at scientific conferences
- Activities advanced to establish launch readiness for vamorolone in the U.S.
- Exclusive license agreement concluded with Sperogenix for vamorolone in rare diseases in the Greater China Region
- Phase 2 trial started with vamorolone in boys aged 2 to <4 years and 7 to <18 years with DMD to assess drug effects in wider age range
- First participant dosed in FDA-funded study with vamorolone in Becker muscular dystrophy
- Lonodelestat development program deprioritized owing to limited resources

The primary operational focus of Santhera in 2022 continues to be the advancement of the regulatory submissions for vamorolone in the U.S. and the EU towards approval. In parallel, the Company plans to advance operational preparations for launches anticipated for 2023 in both regions.

NDA submission to the U.S. FDA for vamorolone in DMD completed

In October, Santhera completed the rolling submission of an NDA to the U.S. FDA, seeking priority review for vamorolone for the treatment of DMD. Typically, within 60 days of the receipt of the dossier, the FDA will inform if a priority review will be granted. A priority review designation indicates FDA's goal to take action on an application within six months (compared to ten months under standard review) which would set an anticipated approval date for as early as mid-2023. Subject to approval, vamorolone is set to become available to patients in the U.S. in H2-2023.

European MAA for vamorolone in DMD submitted – review by the EMA has started

In September, Santhera submitted an MAA for vamorolone for the treatment of DMD to the EMA. Validation, received in October, confirms that the submission is complete and that the review by the EMA's Committee for Medicinal Products for Human Use (CHMP) has begun. Santhera expects the CHMP to complete the review and issue an opinion regarding approval to EMA's European Commission (EC) in late Q3-2023. Subject to EC approval later in 2023, vamorolone will receive marketing authorization in all member states of the European Union, as well as in Norway, Liechtenstein and Iceland.

Findings on bone health published for vamorolone alongside efficacy and safety data

Vamorolone is under joint development by ReveraGen and Santhera for DMD patients with the objective to provide an anti-inflammatory and muscle preserving treatment with a favorable safety and tolerability profile as an alternative to the current standard of care. In addition to long-term efficacy and safety data with vamorolone, recent publications and presentations further characterized vamorolone's differentiated profile especially with regard to bone health [1-5].

In July 2022, data assessing the impact of long-term treatment with vamorolone on bone health were presented at the *10th International Conference on Children's Bone Health* [1]. After 2.5 years of treatment with vamorolone, bone turnover markers were not suppressed, bone age delay was minimal, and the vertebral fracture burden was lower compared with published data on daily prednisolone. Efficacy data showed that vamorolone can maintain muscle function in boys with DMD, similar to standard of care glucocorticoid treatment.

In August 2022, *JAMA Neurology* published the positive 24-week results from the pivotal VISION-DMD study evaluating vamorolone in patients with DMD compared to placebo and prednisone [2]. Vamorolone met its primary endpoint by demonstrating statistically significant and clinically relevant improvement in time to stand from floor compared to placebo and showed consistent results across multiple secondary endpoints. The relative efficacy of vamorolone 6 mg/kg/day was comparable to that seen with prednisone 0.75 mg/kg/day across primary and secondary efficacy endpoints. Importantly, no negative impact on biomarkers of bone health and no loss of linear growth were observed with vamorolone. Patients treated with prednisone experienced reductions in serum biomarkers of bone formation, which promptly recovered to baseline values when subjects were switched from prednisone to vamorolone.

In October 2022, key opinion leaders further highlighted the bone-related profile of vamorolone in different presentations at the *World Muscular Society Congress 2022* which can be viewed on Santhera's website here.

Vamorolone, an investigational drug, was generally safe and well tolerated. The most commonly reported adverse events versus placebo were cushingoid features, vomiting and vitamin D deficiency. Adverse events were generally of mild to moderate severity.

U.S. pre-commercialization measures advanced

The U.S. subsidiary made further progress in establishing launch readiness with the hiring into critical roles and a focus on long lead-time priority projects. These include medical and market access activities, working closely with key clinical opinion leaders to facilitate presentations and papers as well as engaging with patient advocacy groups.

Started Phase 2 study evaluating vamorolone in a wider age range of patients with DMD

Health care professionals routinely prescribe glucocorticoid steroids in DMD to preserve muscle strength and function in ambulant boys, starting at an early stage. In most cases treatment is continued until deleterious side effects prevent further therapy and lead to early discontinuation. The clinical development program for vamorolone included patients 4-7 years old and this new study aims at collecting information on vamorolone outside the original age range. The ongoing Phase 2 VBP-006 study (ClinicalTrials.gov ID: NCT05185622) is an open-label, multiple dose study to evaluate the clinical efficacy, safety and tolerability of vamorolone 2 or 6 mg/kg/day over a treatment period of 12 weeks in 44 steroid-naïve boys ages 2 to <4 years, and glucocorticoid-treated and currently untreated boys ages 7 to <18 years with DMD. The estimated study completion date is end of 2023.

First patient dosed in FDA-funded study with vamorolone in Becker muscular dystrophy (BMD)

This Phase 2 pilot study is a randomized, double-blind, placebo-controlled study to evaluate the safety, tolerability and exploratory clinical efficacy on motor function outcomes of vamorolone compared to placebo over a treatment period of 24 weeks in 39 males (aged ≥18 and <65 years) with BMD (ClinicalTrials.gov ID: NCT05166109). The study is funded by a USD 1.2 million grant from the FDA under their "Clinical Studies of Orphan Products Addressing Unmet Needs of Rare Diseases (R01)" grants program. Vamorolone has shown efficacy in the pivotal VISION-DMD study in DMD, a more severe but related disease, and, based on these findings and its mechanisms of action, this developmental compound may show a benefit in BMD.

Lonodelestat development paused owing to resource constraints

Vamorolone is the main strategic focus and in the near-term will consume all financial and human resources. Santhera's focus for months to come will be on advancing vamorolone through the regulatory process towards approval and on preparations for market entry. As a consequence, Santhera has put the development program for lonodelestat, its second clinical development candidate targeting pulmonary indications, on hold. Preparations for a Phase 2 study in an acute pulmonary indication are far advanced, however, continuation of the program will be subject to funding. Santhera explores various opportunities via collaboration and/or partnerships to resume the project as quickly as possible.

Post-authorization measures (PAMs) with Raxone in LHON completed

In July 2022, the last part of the PAMs for Raxone (idebenone) was completed. Cornerstone of the PAMs were the long-term Phase 4 studies LEROS and PAROS with Raxone for the treatment of Leber's hereditary optic neuropathy (LHON). As previously reported, LEROS met the primary endpoint, the proportion of eyes with clinically relevant benefit after 12 months treatment with Raxone, with high statistical significance (p=0.002). PAROS, a prospective non-interventional study in routine clinical settings in LHON patients treated with Raxone®, suggested a maintenance of treatment effect and showed a similar safety profile observed to that from the LEROS study. This clinically robust evidence of long-term effectiveness and safety confirms and extends previous findings and is expected to facilitate market access, allowing patients to benefit where currently no effective treatments alternatives are available. Santhera intends to discuss a path forward towards U.S. approvability with drug regulators.

In 2019, Santhera out-licensed rights for Raxone (idebenone) for the treatment of LHON outside North America and France to Chiesi Group while remaining the EU marketing authorization holder for the product.

Strategic licensing agreements to exploit pipeline potential and tap non-dilutive funding

Santhera continues to pursue out-licensing agreements in the rare disease space to further exploit the potential of its pipeline products, and for securing additional funding. As previously announced, two agreements were closed in H1-2022:

In January 2022, the Company entered into an exclusive license agreement for the Greater China area with Sperogenix Therapeutics, a China-based company specializing in orphan diseases. Under this agreement, Sperogenix has in-licensed vamorolone for rare disease indications for a total consideration of up to USD 124 million and plans to initiate a regulatory filing for vamorolone for DMD in China upon U.S. FDA approval, which could lead approval in China as early as 2024.

In February 2022, Santhera signed an agreement with SEAL Therapeutics to further develop a gene therapy approach intended for the treatment of congenital muscular dystrophy in exchange for payments based on future proceeds of SEAL Therapeutics.

FINANCIAL HALF-YEAR PERFORMANCE & FINANCING OUTLOOK

- Net revenue from contracts with customers of CHF 6.3 million (H1-2021: CHF 4.5 million)
- Operating result of CHF -25.5 million (H1-2021: CHF -19.5 million)
- Net result of CHF -29.7 million (H1-2021: CHF -20.5 million)
- Cash flow from operating activities CHF -12.0 million (H1-2021: CHF -18.6 million)
- Cash and cash equivalents of CHF 12.7 million (June 30, 2022)
- Additional accrual due to uncertainties over Raxone reimbursement in France impacted H1-2022 results
- Full repayment of 2017/22 Bonds and further reduction of outstanding convertible bonds to CHF 31.5 million
- Financing agreement with Highbridge amended to meet immediate liquidity requirements
- Renegotiated the timing of vamorolone approval milestone payment reducing near-term financial obligations of the Company by CHF 20 million

Update on reimbursement negotiations for Raxone in France

Since its launch in 2018, Raxone was reimbursed in France for the treatment of patients with LHON under a temporary financing scheme. From August 2021, Santhera has supplied Raxone free of charge based on an agreement reached with the authorities in France after the temporary pricing was challenged and Raxone removed from the list of reimbursed drugs. Reimbursement discussions are still ongoing. Due to ongoing uncertainties with the status of pricing reimbursement negotiations, the Company has accrued an additional CHF 8.1 million towards a settlement, of which CHF 6.0 million was recognized against sales, and CHF 2.1 million recognized as marketing and sales expenses. As of June 30, 2022, Santhera has recognized a total accrual amount of CHF 25.0 million included within noncurrent liabilities. Once an agreement is reached, including for the final pricing for Raxone, Santhera expects to be able to settle the liability from future sales of Raxone in France.

Santhera continues to supply the product in the French market following the out-licensing and transfer of Raxone outside North America and France to Chiesi Group in 2019.

Net revenue

In the first half-year 2022, Santhera reported net revenue from contracts with customers of CHF 6.3 million (H1-2021: CHF 4.5 million).

Net sales amounted to CHF -5.9 million (H1-2021: CHF 2.9 million). The negative sales are attributable to an additional CHF 6.0 million that has been accrued and offset against sales in the context of ongoing reimbursement negotiations in France, as described above. Until an agreement is reached with the Comité économique des produits de santé (CEPS) on the future pricing of Raxone for LHON, the Company continues to provide Raxone to patients in France free of charge. Outside of France and North America, Santhera has out-licensed Raxone to Chiesi Group.

During the six months ended June 30, 2022, Santhera recognized revenue from out-licensing transactions in the amount of CHF 11.2 million (H1-2021: CHF 0 million). This largely reflects an initial income from the out-licensing transaction of vamorolone for the Greater China Region with Sperogenix.

Cost of goods

Cost of goods sold amounted to CHF 1.9 million (H1-2021: CHF 2.0 million) and represents continuing supply of Raxone and amortization of intangibles.

Operating expenses and result

Operating expenses of CHF 30.0 million (H1-2021: CHF 21.9 million) were higher, primarily due to higher external development expenses related to vamorolone.

Development expenses amounted to CHF 16.9 million (H1-2021: CHF 13.6 million). The increase was primarily due to higher expenses for third-party clinical and regulatory service providers for finalizing data analysis and the assembly of the regulatory dossiers for vamorolone in DMD to U.S. and European authorities.

Marketing and sales expenses were CHF 5.9 million (H1-2021: CHF 2.0 million). The increase was a result of the additional accrual of CHF 2.1 million in relation to ongoing reimbursement negotiations in France, as described above, as well as ongoing pre-commercialization activities for vamorolone.

General and administrative expenses of CHF 7.1 million (H1-2021: CHF 6.3 million), for which the increase year-on-year reflects the addition of personnel in key functions in view of market readiness preparations for vamorolone in the U.S.

The operating result amounted to CHF -25.5 million (H1-2021: CHF -19.5 million).

Financial income and expenses

The net financial expense amounted to CHF 3.6 million (H1-2021: CHF 0.4 million). The change from prior year same period is largely a reflection of a recognized gain on exchange of the 2017/22 Bonds in H1-2021, which had been partially offset by the costs associated with financing transactions.

Net result

The net result for the half-year ended June 30, 2022, was a loss of CHF 29.7 million or CHF -0.52 per share, compared to a net loss of CHF 20.5 million or CHF -0.92 per share for the half-year ended June 30, 2021.

Cash flow and cash balance

As of June 30, 2022, the Company had cash and cash equivalents of CHF 12.7 million compared to CHF 21.2 million as of December 31, 2021.

Net cash outflow for operating activities was lower year-on-year and amounted to CHF 12.0 million (H1-2021: CHF 18.6 million).

Net cash inflow from financing activities was lower year-on-year and amounted to CHF 3.5 million (H1-2021: CHF 14.3 million) as the net additional proceeds from exchangeable notes was largely offset by the repayment of convertible bonds.

Shareholders' equity

Total consolidated net equity deficit as of June 30, 2022, amounted to CHF -13.8 million compared to total equity of CHF 1.3 million as of December 31, 2021, as a result of the net loss incurred for the period.

Year-to-date debt and equity-linked financing

In February 2022, Santhera fully repaid its senior unsecured convertible bonds (2017/22 Bonds) with a remainder amount of CHF 13.9 million. Of the senior unsecured convertible bonds (2021/24 Bonds) maturing in August 2024, an aggregate amount of CHF 19.6 million was still outstanding on June 30, 2022, unchanged from December 31, 2021 as no repayment or conversion into shares took place during the period. Of the private convertible bonds (2021/24 Private Bonds) in the amount of CHF 15 million issued to Highbridge, CHF 3 million were converted into shares during the period, leaving a remainder of CHF 12 million at June 30, 2022. In summary, this significantly reduced total and short-term convertible debt from an original amount of CHF 60 million maturing in February 2022 to approximately CHF 31.5 million maturing in August 2024.

During the six months ending June 30, 2022, in order to provide additional fundraising flexibility, Santhera issued new shares to be held as treasury shares totaling 19,107,892 with a nominal value of CHF 1 each. As a result, Santhera's issued share capital amounts to CHF 73,725,702 as of June 30, 2022. Santhera expects to hold its treasury shares until market conditions allow for a favorable financing transaction.

Concurrently with the ordinary capital increase, and as approved by the Extraordinary General Meeting (EGM) on December 15, 2021 and the Annual General Meeting (AGM) on June 30, 2022, Santhera's authorized capital has increased during the six months ending June 30, 2022, from CHF 27,303,905 to CHF 36,860,687 and its conditional capital for financing has increased from CHF 21,878,228 to CHF 31,370,336. Together with the 19,485,946 treasury shares held as of June 30, 2022, the Company plans to use these shares for financing activities if required.

With the shareholders' approval at the AGM held on June 30, 2022, Santhera reduced the nominal value of the shares from CHF 1.00 to CHF 0.01 per share with effective date September 6, 2022.

On June 2, 2022, the Company entered into an amendment to the timing of an upcoming milestone payment to partner ReveraGen, resulting in a reduction of the milestone payment (expected in the second half of 2023) due upon FDA approval in exchange for an increase of the sales milestone in the same amount. Thereby near-term financial obligations of the Company were reduced by CHF 20 million.

In addition, on June 2, 2022, the Company upsized its existing financing arrangement with certain funds managed by Highbridge Capital Management, LLC (Highbridge) by up to an additional CHF 40 million. An initial unconditional drawdown of CHF 20 million was received on June 3, 2022. The remaining balance of CHF 20 million is divided into two tranches of CHF 10 million each, available for drawdown subject to Highbridge Capital Management's consent.

Amendment of Highbridge facility to satisfy immediate cash requirements

On September 28, 2022, Santhera and Highbridge agreed to amend the existing financing arrangement that has been announced on June 2, 2022, and again mentioned above, to provide for the immediate drawdown of a CHF 10 million tranche in senior secured Exchangeable Notes and amend certain provisions. Of this amount, approximately CHF 5 million was used to repurchase part of the outstanding convertible bonds issued to Highbridge in 2021 and due in 2024 at a 25 percent discount to its nominal value plus interest. The Exchangeable Notes can be exchanged by Highbridge for shares at a discount to VWAP, subject to a reduced floor price. As part of this new money financing and further commitments, Santhera has agreed on a new conversion price of CHF 1.20 for the remaining outstanding private convertible bond and a new exercise price of CHF 0.80 per share for the existing warrants held by Highbridge. A further tranche of CHF 10 million available for drawdown is conditional on management achieving certain milestones and other conditions.

Funding outlook

Santhera still has treasury shares, conditional and authorized capital from past EGM and AGM authorizations which are available for future placement, subject to market conditions. This, in combination with cash balances of CHF 12.7 million (at June 30, 2022), the recent drawdown from the Highbridge facility and remaining facilities, is expected to provide a liquidity runway into Q1-2023.

In order to ensure the execution of the Company's operating plans to mid-2023, when approval of vamorolone for DMD in the U.S. is expected at the earliest, Santhera will need to secure additional funds. Santhera is pursuing strategic options including but not limited to non-dilutive funding in the form of out-licensing agreements and/or the monetization of assets and, in parallel, is also evaluating debt financing, royalty financing, standby equity distribution agreement or, depending on market conditions, equity-based funding.

References:
[1] Guglieri M et al (2022). JAMA Neurol. Published online August 29, 2022. doi:10.1001/jamaneurol.2022.2480. Link.
[2] Mah JK et al (2022). JAMA Netw Open. 2022;5(1):e2144178. doi:10.1001/jamanetworkopen.2021.44178. Link.
[3] Guglieri, et al (2022) JAMA. doi:10.1001/jama.2022.4315
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[5] Liu X, et al (2020). Proc Natl Acad Sci USA 117:24285-24293

Half-year Report

The Santhera Half-year Report 2022 is available for download on the Company's website at www.santhera.com/financial-reports.

Interim Consolidated Financial Statements

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Interim Consolidated Balance Sheet

In CHF thousands	Notes	**Jun 30, 2022** (unaudited)	**Dec 31, 2021** (audited)
Assets			
Tangible assets		1,202	1,324
Intangible assets		63,055	64,596
Financial assets long-term		477	468
Deferred tax assets		10	88
Pension assets		475	-
Noncurrent assets		**65,219**	**66,476**
Prepaid expenses		708	1,069
Inventories		213	428
Trade and other receivables		945	1,936
Cash and cash equivalents	5	12,697	21,208
Current assets		**14,563**	**24,641**
Total assets		**79,782**	**91,117**
Equity and liabilities			
Share capital	6	73,726	54,608
Capital reserves and share premium		514,015	509,513
Retained earnings/(deficit)		(586,149)	(556,425)
Employee benefit reserve		4,939	(437)
Treasury shares		(19,486)	(5,020)
Currency translation differences		(890)	(911)
Total equity		**(13,845)**	**1,328**
Noncurrent convertible bonds	7.2	24,899	25,796
Noncurrent derivative financial instruments	7	2,001	3,683
Noncurrent warrant liabilities	7	2,293	4,723
Noncurrent lease liabilities		913	1,203
Noncurrent accrued expenses	10.2	24,945	16,808
Pension liabilities		-	4,794
Total noncurrent liabilities		**55,051**	**57,007**
Trade and other payables		4,868	4,585
Accrued expenses		13,557	9,710
Income tax payable		449	266
Current lease liabilities		672	609
Current Exchangeable Notes	7.1	14,157	1,488
Current convertible bonds	7.2	-	13,880
Current derivative financial instruments	7.1	3,929	402
Current warrant liabilities	7	800	1,650
Current provisions	9	144	192
Total current liabilities		**38,576**	**32,782**
Total liabilities		**93,627**	**89,789**
Total equity and liabilities		**79,782**	**91,117**

Interim Consolidated Income Statement

In CHF thousands (except per share data)	Notes	**Six months ended** (unaudited)	
		June 30, 2022	**June 30, 2021**
Net sales	10	(5,873)	2,853
Revenue from out-licensing transactions	10	11,190	-
Net sales to licensing partner	10	933	1,639
Revenue from contracts with customers		**6,250**	**4,492**
Cost of goods sold		(1,875)	(2,031)
Of which amortization of intangible asset		*(1,519)*	*(1,519)*
Other operating income		79	-
Development	12	(16,870)	(13,592)
Marketing and sales	12	(5,917)	(2,008)
General and administrative	12	(7,133)	(6,307)
Other operating expenses	12	(70)	(31)
Operating expenses		**(29,990)**	**(21,938)**
Operating result		**(25,536)**	**(19,477)**
Financial income	13	5,291	13,957
Financial expense	13	(8,887)	(14,346)
Result before taxes		**(29,132)**	**(19,866)**
Income tax expense	14	(592)	(653)
Net result		**(29,724)**	**(20,519)**
Basic and diluted loss per share	16	(0.52)	(0.92)

Interim Consolidated Statement of Comprehensive Income

In CHF thousands	**Six months ended** (unaudited)	
	June 30, 2022	**June 30, 2021**
Net result	**(29,724)**	**(20,519)**
Items that will not be reclassified to profit or loss in subsequent periods:		
Net actuarial gains/(losses) from defined benefit plans	5,376	1,143
Items that may be reclassified to profit or loss in subsequent periods:		
Currency translation differences	21	41
Other comprehensive result	**5,397**	**1,184**
Total comprehensive result	**(24,327)**	**(19,335)**

Interim Consolidated Statement of Cash Flows

In CHF thousands	Notes	**Six months ended** (unaudited)	
		June 30, 2022	**June 30, 2021**
Result before taxes		**(29,132)**	**(19,866)**
Depreciation of tangible assets		297	342
Amortization of intangible assets		1,519	1,551
Share-based compensation	15	1,451	1,152
Change in fair value of financial instruments, net		(4,968)	(312)
Change in pension liabilities		106	(342)
Reversal of current provisions		-	(589)
Change in noncurrent accruals	10.2	8,137	-
Taxes paid		(58)	(159)
Change in net working capital		2,550	760
Total financial result		8,559	389
Interest paid		(418)	(1,533)
Net cash flow from/(used in) operating activities		**(11,957)**	**(18,607)**
Purchase of tangible assets		-	(44)
Change in investments in other long-term financial assets		-	(31)
Net cash flow from/(used in) investing activities		**-**	**(75)**
Proceeds from sale of treasury shares		-	81
Purchase of treasury shares		-	(56)
Proceeds from Exchangeable Notes	7.1	18,000	17,000
Repayment of convertible bonds	7.2	(13,935)	-
Payment of lease liabilities		(365)	(392)
Financing transaction costs		(5)	(2,357)
Cost of issuance of capital		(207)	-
Net cash flow from/(used in) financing activities		**3,488**	**14,276**
Effects of exchange rate changes on cash and cash equivalents		(42)	(14)
Net increase/(decrease) in cash and cash equivalents		**(8,511)**	**(4,420)**
Cash and cash equivalents at January 1		21,208	12,411
Cash and cash equivalents at June 30		**12,697**	**7,991**

Interim Consolidated Statement of Changes in Equity

In CHF thousands	Notes	Share capital	Capital reserves and share premium	Retained earnings/(deficit)	Employee benefit reserve	Treasury shares	Trans-lation differ-ences	**Total**
Balance, January 1, 2021		**19,430**	**480,005**	**(500,899)**	**(2,320)**	**(1,580)**	**(990)**	**(6,354)**
Net result		-	-	(20,519)	-	-	-	**(20,519)**
Other comprehensive income		-	-	-	1,143	-	41	**1,184**
Total comprehensive result		**-**	**-**	**(20,519)**	**1,143**	**-**	**41**	**(19,335)**
Share-based compensation		-	1,152	-	-	-	-	**1,152**
Shares issued		11,874	-	-	-	(11,695)	-	**179**
Delivery of Shares on conversion of Exchangeable Notes into Shares		-	14,154	-	-	6,394	-	**20,458**
Delivery of Shares on conversion of convertible bonds into Shares		-	7,954	-	-	4,249	-	**12,203**
Delivery of Shares for financing facility		-	-	-	-	233	-	**233**
Cost of issuance of capital		-	(545)	-	-	-	-	**(545)**
Change in treasury shares		-	(252)	-	-	524	-	**272**
Balance, June 30, 2021 (unaudited)		**31,304**	**502,468**	**(521,418)**	**(1,177)**	**(1,875)**	**(949)**	**8,353**
Balance, January 1, 2022		**54,608**	**509,513**	**(556,425)**	**(437)**	**(5,020)**	**(911)**	**1,328**
Net result		-	-	(29,724)	-	-	-	**(29,724)**
Other comprehensive income		-	-	-	5,376	-	21	**5,397**
Total comprehensive result		**-**	**-**	**(29,724)**	**5,376**	**-**	**21**	**(24,327)**
Share-based compensation	15	-	1,451	-	-	-	-	**1,451**
Shares issued	6	19,118	-	-	-	(19,108)	-	**10**
Delivery of Shares on conversion of Exchangeable Notes into Shares	7.1	-	437	-	-	1,712	-	**2,149**
Delivery of Shares on conversion of convertible bonds into Shares	7.2	-	2,592	-	-	2,193	-	**4,785**
Delivery of Shares on settlement of convertible bonds interest expense	7.2	-	229	-	-	659	-	**888**
Delivery of Shares for financing transactions		-	-	-	-	78	-	**78**
Cost of issuance of capital		-	(207)	-	-	-	-	**(207)**
Balance, June 30, 2022 (unaudited)		**73,726**	**514,015**	**(586,149)**	**4,939**	**(19,486)**	**(890)**	**(13,845)**

Notes to the Interim Consolidated Financial Statements

1 General Information

Santhera Pharmaceuticals Holding AG (herein 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 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 interim consolidated financial statements were approved for publication by the Board of Directors (**Board**) on October 30, 2022.

2 Summary of Significant Accounting Policies

2.1 Basis of presentation

The Company's interim consolidated financial statements are prepared in accordance with International Financial Reporting Standards (**IFRS**), IAS 34 *Interim Financial Reporting*. Accordingly, the interim financial information does not include all the information and notes required under IFRS for annual consolidated financial statements. Therefore, such information should be read in conjunction with the Group's audited consolidated financial statements for the year ended December 31, 2021.

Except as described in 2.2 below, the accounting policies applied in these unaudited interim consolidated financial statements are consistent with those applied in the audited consolidated financial statements for the year ended December 31, 2021.

The presentation currency is Swiss francs (**CHF**). Amounts shown are rounded to the nearest CHF 1,000 unless otherwise indicated.

2.2 Changes in accounting policies

In 2022 the Group adopted various amendments to existing standards and interpretations. Adoption of these amendments had no material impact on the Group's overall results and financial position.

2.3 Material uncertainties and ability to continue operations

The interim consolidated financial statements have been prepared under the going concern assumption despite several material uncertainties present as of June 30, 2022, that may perceived to be contrary to this assumption. In order to support ongoing activities including preparation for launch of vamorolone in H2-2023, the Company requires additional funds to reach the next inflection point, namely the regulatory approvals of vamorolone in the U.S., as early as mid-2023, and the EU in late 2023.

Cash at hand and additional funds available as of June 30, 2022, and as of the date of issuance of these interim consolidated financial statements are insufficient to allow the Company to reach the value inflection points of approval during 2023 after the completion of vamorolone regulatory filing during the remainder of 2022. Hence, material uncertainties remain as to the Company's ability to continue as a going concern until June 30, 2023.

Executing the Company's strategy significantly depends on the following:

- The acceptance by the U.S. FDA of the NDA submission for vamorolone expected in Q4-2022
- Additional funding to ensure the continuation of its operations through June 30, 2023
- Ability to settle current debt obligations

The European and U.S. NDA submissions for vamorolone in ambulant patients with DMD are in progress, with the acceptance of the submissions expected in Q4-2022. In the event of acceptance, the Management and the Board plan to raise additional funds to finance further development activities and to support pre-commercialization activities. Should sufficient further funding not be available, the Company may review further organizational restructuring measures and reduction in business activities as well as consider the monetization of assets (e.g., out-licensing rights of lonodelestat or out-licensing rights in certain geographic markets of vamorolone).

As previously announced, on June 2, 2022, the Company increased its financing facility with Highbridge Capital Management, by an additional CHF 40 million that provides liquidity through Q1-2023. This agreement provides for a tranche of CHF 20 million, which is unconditional and available for immediate drawdown. The remaining balance of CHF 20 million is divided into two tranches, each amounting to CHF 10 million, and each drawdown subject to Highbridge Capital Management's consent. See Note 7.1 and Note 18 for more information about this financing facility with Highbridge Capital Management.

In addition, on June 2, 2022, the Company announced an amendment to the agreement with ReveraGen, resulting in a reduction of the USD 40 million milestone payment due upon FDA approval (expected in the second half of 2023) by USD 20 million in exchange for an increase of the sales milestone by USD 20 million (due when vamorolone annual revenue reaches USD 100 million).

As of June 30, 2022, the Company had cash and cash equivalents in the amount of CHF 12.7 million, providing for liquidity runway into Q1-2023. However, in addition to the extension of the financing facility described above (arranged in June 2022), to ensure the execution of the Company's operating plan through to June 30, 2023, and beyond, additional funding will be needed. If the Company is unable to obtain the required funding to run its operations and to develop and commercialize its product candidates, the Company could be forced to delay, reduce or stop some or all of its research and development programs with the objective to ensure it remains solvent. The Company may seek additional funding through public or private financings or licensing agreements. The sale of additional equity may dilute existing shareholders.

Shareholders should note that whilst the Management and the Board consistently continue to apply best efforts to evaluate and execute available options, there is no guarantee that the development studies will be successful, regulatory approvals obtained, and that any transaction can be realized or that such transaction would generate sufficient funds to finance operations through to June 30, 2023. These material uncertainties may cast significant doubts about the ability of the Company to continue as a going concern.

However, the Management and the Board are of the view that it is more likely than not that the Group will continue 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, 2023. Hence, the interim consolidated financial statements have been prepared on a going concern basis.

2.4 Financial liabilities

Financial liabilities at fair value through profit or loss

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

Derivatives may be embedded in other contractual arrangements. The Company accounts for an embedded derivative separately from the host contract when:

- the host contract is not an asset in the scope of IFRS 9 *Financial Instruments*
- the host contract is not itself carried at fair value through profit or loss
- the terms of the embedded derivative would meet the definition of a derivative if they were contained in a separate contract
- the economic characteristics and risks of the embedded derivative are not closely related to the economic characteristics and risks of the host

Separated embedded derivatives are initially and subsequently measured at fair value, with all changes in fair value recognized in profit or loss.

Other financial liabilities measured at amortized cost

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

3 Seasonality

The Group's operating result is not subject to significant seasonal variations.

4 Principal Currencies Translation Rates

	average rates for six months ended		period-end rates	
	Jun 30, 2022	**Jun 30, 2021**	**Jun 30, 2022**	**Dec 31, 2021**
1 Euro (**EUR**)	1.0250	1.0945	0.9978	1.0339
1 US dollar (**USD**)	0.9696	0.9091	0.9557	0.9127
1 British pound (**GBP**)	1.1949	1.2564	1.1605	1.2330
1 Canadian dollar (**CAD**)	0.7565	0.7230	0.7401	0.7176

5 Cash and Cash Equivalents

In CHF thousands	**Jun 30, 2022**	**Dec 31, 2021**
Cash at banks and on hand		
in CHF	11,185	20,301
in EUR	847	288
in USD	470	468
in GBP	47	61
in CAD	20	24
other currencies	128	66
Total cash and cash equivalents	**12,697**	**21,208**

6 Share Capital

6.1 Ordinary share capital

During the six months ending June 30, 2022, a total of 19,117,892 new Shares were issued for financing transactions, share-based compensation, and for treasury shares. As of June 30, 2022, issued share capital totals CHF 73,725,702, consisting of 73,725,702 Shares with a nominal value of CHF 1 each. As of December 31, 2021, issued share capital totaled CHF 54,607,810, consisting of 54,607,810 Shares with a nominal value of CHF 1 each.

At the Annual General Meeting (**AGM**) held on June 30, 2022, the shareholders approved a reduction of the nominal value of the Shares from CHF 1.00 to CHF 0.01 per share, with allocation of the amount by which the Company's share capital has been reduced to the reserves from capital contribution. The legal process for this change was concluded in September 2022.

6.2 Treasury shares

During the six months ending June 30, 2022, a total of 19,107,892 new Shares were issued to be held as treasury shares intended to be used for financing transactions and share-based compensation, and of which 4,641,825 treasury shares were used for the same.

As of June 30, 2022, the Company holds 19,485,946 treasury shares with a nominal value of CHF 1 each for a total value of CHF 19,485,946. As of December 31, 2021, the Company held 5,019,879 treasury shares with a nominal value of CHF 1 each for a total value of CHF 5,019,879.

6.3 Authorized shares

At the Extraordinary General Meeting (**EGM**) held on December 15, 2021, the shareholders approved an increase of authorized shares to 27,303,905 and in a second step, by an additional 10,000,000 within three months from December 15, 2021, the date of the EGM. At the AGM held on June 30, 2022, the shareholders approved an increase of an additional 2,656,782 authorized shares (and its extension until June 29, 2024). Accordingly, during the six months ending June 30, 2022, authorized shares increased by 12,656,782. In March 2022, 3,100,000 shares were issued out of the authorized capital for new Share issuances intended to be held as treasury shares.

As of June 30, 2022, authorized share capital totals CHF 36,860,687, consisting of 36,860,687 shares with a nominal value of CHF 1 each. As of December 31, 2021, authorized share capital totaled CHF 27,303,905, consisting of 27,303,905 shares with a nominal value of CHF 1 each.

6.4 Conditional shares

Pursuant to Article 3b and Article 3c of the Company's Articles of Incorporation, the Company has conditional shares. The conditional shares represent conditional capital authorized for issuance for share-based compensation, under the exclusion of shareholders' pre-emptive rights, and financing transactions, respectively.

During the six months ending June 30, 2022, a total of 10,000 shares were issued out of Article 3b conditional capital for new Share issuances for share-based compensation. As of June 30, 2022, Article 3b conditional capital totals CHF 5,415,677, consisting of 5,415,677 shares with a nominal value of CHF 1 each. As of December 31, 2021, the total was CHF 5,425,677, consisting of 5,425,677 shares with a nominal value of CHF 1 each.

At the EGM held on December 15, 2021, the shareholders approved an increase of Article 3c conditional shares to 21,878,228 and in a second step, by an additional 10,000,000 within three months from December 15, 2021, the date of the EGM. Accordingly, during the six months ending June 30, 2022, Article 3c conditional shares increased by 10,000,000 shares. In February 2022, 507,892 shares were issued out of the Article 3c conditional shares for new Share issuances for financing transactions. As of June 30, 2022, Article 3c conditional capital totals CHF 31,370,336, consisting of 31,370,336 shares with a nominal value of CHF 1 each. As of December 31, 2021, the total was CHF 21,878,228, consisting of 21,878,228 shares with a nominal value of CHF 1 each.

7 Financial Liabilities

7.1 Equity-linked financing arrangements

Exchangeable Notes – Highbridge Capital Management

In July 2020, the Company and its subsidiary Santhera Pharmaceuticals (Schweiz) AG (**Santhera Schweiz**), entered into a subscription agreement with a fund managed by Highbridge Capital Management LLC (any such entity, **Highbridge**), providing for the issuance of senior secured Exchangeable Notes (**Exchangeable Notes**), subject to certain conditions and available in tranches, and exchangeable for Shares. The Exchangeable Notes are guaranteed by the Company and certain of its subsidiaries and secured by a comprehensive security package, including security over all shares of Santhera Schweiz and other subsidiaries of the Company, as well as over the Group's material intellectual property and other assets.

The Highbridge Exchangeable Notes are considered hybrid contracts containing a host that is a financial liability and different embedded derivatives. Since the economic characteristics and risks of the host and the embedded derivatives are not closely related, the embedded derivatives are separated from the host. The compound embedded derivative includes different features like interest rate choices, a compound interest rate calculation based on the interest rate choice, discounts based on Share prices, a floor for Share prices and different exchange rights. There is an interdependence between the mentioned features, which is why they are recognized as one compound embedded derivative with their fair value. The Exchangeable Notes are recognized as financial liabilities measured at amortized cost using the effective interest method and the embedded derivatives are recognized as financial liabilities measured at fair value through profit or loss.

As of December 31, 2021, the carrying value of the Exchangeable Notes totaled CHF 1.5 million and the fair value of the embedded derivatives totaled CHF 0.4 million.

In June 2022, the Company and Highbridge agreed on an amendment to increase the commitment principal amount by an additional CHF 40 million. For the six months ended June 30, 2022, proceeds from Exchangeable Notes totaled CHF 18 million. As of June 30, 2022, the carrying value of the Exchangeable Notes total CHF 14.2 million and the fair value of the embedded derivatives totaled CHF 3.9 million.

Warrants – Highbridge

In connection with the Highbridge Exchangeable Notes amendments of 2021, warrants were granted in two tranches equal to 15% of the total aggregate amount of the remaining existing facility, and new money tranches. In May 2021, a total of 984,769 warrants with a fair value of CHF 1.58 per warrant were granted. Each of the May warrants is exercisable at any time from the date of grant until March 15, 2026, for one Share at an exercise price of CHF 2.74. In September 2021, an additional 1,000,000 warrants with a fair value of CHF 1.05 per warrant were granted. Each of the September warrants is exercisable at any time until September 22, 2026, for one Share at an exercise price of CHF 2.00. The warrants are initially and subsequently recognized at fair value through profit or loss and are classified as financial liabilities until exercised by the holder.

The combined fair value of the warrants granted in 2021 was recognized as prepaid financing transaction costs. Once the Exchangeable Notes are issued, the prepaid financing transaction cost is expensed on a pro rata basis. As of June 30, 2022, prepaid financing transaction costs totaled CHF 0.3 million. As of December 31, 2021, prepaid financing transaction costs totaled CHF 0.8 million.

As of June 30, 2022, the May 2021 and September 2021 warrants granted to Highbridge have a combined fair value of CHF 0.6 million and nil have been exercised. As of December 31, 2021, the combined fair value of the warrants totaled CHF 1.2 million and nil had been exercised.

Warrants – equity raise

In September 2021, an equity offering transaction totaling CHF 20 million was entered into wherein investors subscribed (based on different subscription agreements) to a total of 12,670,078 Shares. As part of the equity raise transaction, Santhera issued one warrant for every two shares, for a total of 6,335,039 warrants granted with a fair value of CHF 1.05 per warrant at the grant date and an exercise price of CHF 2.00. The warrants are initially and subsequently recognized at fair value through profit or loss and are classified as financial liabilities until exercised by the holder.

As of June 30, 2022, the warrants granted in the September 2021 equity raise have a combined fair value of CHF 2 million and nil have been exercised. As of December 31, 2021, the combined fair value of the warrants totaled CHF 4.2 million and nil had been exercised.

Exchangeable Notes – Idorsia

In September 2020, as part consideration for the assignment of the license for vamorolone, Santhera issued non-interest-bearing Exchangeable Notes in the amount of CHF 10 million with a maturity date of September 1, 2021, to Idorsia, a biopharmaceutical company headquartered in Allschwil, Switzerland. These Exchangeable Notes entitled Santhera to varying redemption options including settling the nominal value fully in cash or by delivering a combination of cash and Santhera shares with differing discounts on the share price, depending on the portion of Santhera shares delivered. The Exchangeable Notes issued to Idorsia were considered compound financial instruments, including a host contract which classifies as a financial liability and different embedded derivatives that have been valued as one compound derivative. The value of the embedded derivatives was solely based on entity specific information and was insignificant.

In September 2021, Santhera repaid the Idorsia Exchangeable Notes in full by transferring cash in the amount of CHF 3.5 million and 3,594,759 Shares with a value of CHF 6.5 million.

The table below summarizes the changes in financial liabilities arising from equity-linked financing arrangements and their financial instruments during the six months ending June 30, 2022, and June 30, 2021:

In CHF thousands	Exchangeable Notes Highbridge	Exchangeable Notes Highbridge derivatives	Warrants Highbridge	Warrants equity raise	Exchangeable Notes Idorsia
Balance, December 31, 2020	**642**	**125**	**-**	**-**	**9,953**
Proceeds from Exchangeable Notes	17,000	-	-	-	-
Non-cash changes:					
Initial recognition of financial instruments at fair value	(7,111)	5,496	1,556	-	-
Nominal value of Exchangeable Notes converted into Shares	(15,000)	(4,944)	-	-	-
Effective interest/amortized cost/fair value adjustments	5,842	-	-	-	35
Balance, June 30, 2021	**1,372**	**677**	**1,556**	**-**	**9,988**
Proceeds from Exchangeable Notes	5,000	-	-	-	-
Repayment of Exchangeable Notes	-	-	-	-	(3,500)
Non-cash changes:					
Initial recognition of financial instruments at fair value	(1,351)	1,290	1,050	6,651	-
Nominal value of Exchangeable Notes converted into Shares	(5,750)	-	-	-	(6,500)
Derecognition of financial instruments on conversion of Exchangeable Notes into Shares	-	(1,565)	-	-	-
Effective interest/amortized cost/fair value adjustments	2,216	-	(1,404)	(2,470)	12
Balance, December 31, 2021	**1,488**	**402**	**1,202**	**4,181**	**-**
Proceeds from Exchangeable Notes	18,000	-	-	-	-
Non-cash changes:					
Initial recognition of financial instruments at fair value	(5,693)	5,693	-	-	-
Nominal value of Exchangeable Notes converted into Shares	(1,500)	-	-	-	-
Derecognition of financial instruments on conversion of Exchangeable Notes into Shares	-	(1,497)	-	-	-
Effective interest/amortized cost/fair value adjustments	1,862	(669)	(616)	(2,154)	-
Balance, June 30, 2022	**14,157**	**3,929**	**586**[(a)]	**2,027**[(a)]	**-**

[(a)] The Highbridge warrants and equity raise warrants have a combined fair value of CHF 2.6 million, of which CHF 2.3 million is classified as noncurrent warrant liability and CHF 0.3 million is classified as current warrant liability.

Equity-linked financial instruments valuation and sensitivity analysis

The equity-linked financing arrangements' financial instruments includes the embedded derivatives and warrants. The financial instruments valuations are based on Level 3 unobservable input parameters applying a simulation-based approach. The implied volatility, a significant valuation input, is determined by reference to the annualized daily trading volatility of Santhera's Shares for a historical lookback period equal to the expected remaining life of the conversion right as of the valuation date. By construction, the compound financial instrument issued to Highbridge is assumed will be exercised before maturity. For valuation purposes, it is therefore assumed that the expected exercise date is between the investing date and the maturity date.

The table below shows the implied volatility as of the valuation date:

Financial instruments	**Jun 30, 2022**	**Dec 31, 2021**
Equity-linked financing arrangements – derivatives	57%	73%
Equity-linked financing arrangements – warrants:		
Granted in May 2021	79%	78%
Granted in September 2021	75%	81%

The table below shows the impact that a 5% increase/decrease in volatility has on the fair value for each category of financial instrument and its effect on result before taxes as of the valuation date.

In CHF thousands		**Jun 30, 2022**	**Dec 31, 2021**
Financial instruments	Increase/decrease in volatility assumption	Effect on result before taxes	Effect on result before taxes
Equity-linked financing arrangements – derivatives			
Change in volatility	+5%	(11)	(5)
	-5%	15	4
Equity-linked financing arrangements – warrants			
Change in volatility	+5%	(22)	(40)
	-5%	29	50

7.2 Financing arrangements – convertible bonds

2017/22 Bonds

On February 17, 2017, Santhera issued senior unsecured convertible bonds with a maturity date of February 17, 2022 in the nominal value of CHF 60 million (**2017/22 Bonds**). The bonds were listed on the SIX and had interest bearing (5%) with a maximum term of five years and were convertible into Shares 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 could call the 2017/22 Bonds at any time on or after the second anniversary of the issue date at par, plus accrued interest, if any, if the volume weighted average price (**VWAP**) of the Shares was at least 160% of the conversion price.

On March 25, 2021, Santhera announced an exchange offer for the 2017/22 Bonds. The holders of the 2017/22 Bonds who accepted the exchange offer would receive for each of their 2017/22 Bonds, one new bond issued in 2021 with a maturity in 2024 and 26 Shares on exchange.

With effective date May 4, 2021, upon settlement of the 2017/22 Bonds' restructuring, the aggregate nominal value of the 2017/22 Bonds was reduced from the original CHF 60 million to CHF 15.2 million. As consideration for

the exchange, new bonds with a nominal value of CHF 30.3 million were issued (see below the "2021/24 Bonds").

As of December 31, 2021, the 2017/22 Bonds had a remaining aggregate nominal value of CHF 13.9 million and a carrying value of CHF 13.9 million.

During the six months ended June 30, 2022, the remaining 2017/22 Bonds outstanding were fully repaid with effective date February 17, 2022 and delisted from the SIX Swiss Exchange.

2021/24 Bonds

On May 4, 2021, Santhera issued senior unsecured convertible bonds with a maturity date of August 17, 2024 in the nominal value of CHF 30.3 million (**2021/24 Bonds**). The bonds, listed on the SIX, have interest bearing (7.5%) with a maximum term of 39 months, and are convertible into Shares with a nominal value of CHF 1 each. The initial conversion price is fixed at CHF 3.0029. In addition, Santhera could call the 2021/24 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 150% of the conversion price.

The 2021/24 Bonds were offered as consideration for the 2017/22 Bonds. Accordingly, Santhera did not receive any cash proceeds upon issuance of the 2021/24 Bonds.

During 2021, 2021/24 Bonds with a total aggregate nominal value of CHF 10.7 million were converted into Shares. As of December 31, 2021, the 2021/24 Bonds had a remaining aggregate nominal value of CHF 19.6 million and a carrying value of CHF 15.4 million and the fair value of the derivatives totaled CHF 1.1 million.

During the six months ended June 30, 2022, nil 2021/24 Bonds were converted into Shares. As of June 30, 2022, the 2021/24 Bonds have a remaining aggregate nominal value of CHF 19.6 million and a carrying value of CHF 16 million and the fair value of the derivatives totals CHF 0.4 million.

2021/24 Private Bonds

On October 14, 2021, in a private offering, Santhera issued senior unsecured convertible bonds to Highbridge with an aggregate nominal value of CHF 15 million (**2021/24 Private Bonds**). The terms of the 2021/24 Private Bonds are substantially similar to those of the 2021/24 Bonds, except for the conversion price fixed at CHF 1.76 and the floor price for purposes of interest payments fixed at CHF 1.25.

The proceeds from issuance of the 2021/24 Private Bonds was used for the repayment of the outstanding 2017/22 Bonds due on February 17, 2022.

As consideration for its commitment to subscribe for the 2021/24 Private Bonds, Highbridge received 1.5 million warrants with a fair value of CHF 1.05 per warrant at the date of issuance. Each warrant is exercisable at any time until September 22, 2026, for one Share at an exercise price of CHF 2.00. The warrants are initially and subsequently recognized at fair value through profit or loss and are classified as financial liabilities until exercised by the holder.

During 2021, nil 2021/24 Private Bonds and warrants were converted to Shares. As of December 31, 2021, the 2021/24 Private Bonds had an aggregate nominal value of CHF 15 million and a carrying value of CHF 10.4 million. As of December 31, 2021, the fair value of the warrants totaled approx. CHF 1 million and the fair value of the derivatives totaled CHF 2.6 million.

During the six months ended June 30, 2022, 2021/24 Private Bonds with a total aggregate nominal value of CHF 3 million were converted into Shares. Nil warrants were exercised and converted into Shares. As of June 30, 2022, the 2021/24 Private Bonds have an aggregate nominal value of CHF 12 million and a carrying value of CHF 8.9 million. As of June 30, 2022, the fair value of the warrants totaled approx. CHF 0.5 million and the fair value of the derivatives totaled CHF 1.6 million.

The following table summarizes the nominal and carrying values of the convertible bonds as of June 30, 2022, and December 31, 2021:

In CHF thousands					**Jun 30, 2022**		**Dec 31, 2021**	
	Offering	Currency	Interest	Maturity	Nominal value	Carrying value	Nominal value	Carrying value
2017/22 Bonds (ISIN: CH0353955195)	Public	CHF	5%	2022	-	-	13,945	13,880
2021/24 Bonds (ISIN: CH0563348744)	Public	CHF	7.5%	2024	19,568	16,043	19,568	15,387
2021/24 Private Bonds	Private	CHF	7.5%	2024	11,971	8,856	15,002	10,409
Total convertible bonds					**31,539**	**24,899**	**48,515**	**39,676**
Less current portion of convertible bonds with short-term maturities						-		(13,880)
Noncurrent portion of convertible bonds with long-term maturities						24,899		25,796

The table below summarizes the changes in financial liabilities arising from convertible bond issuances and their financial instruments during the six months ending June 30, 2022, and June 30, 2021:

In CHF thousands	2017/22 Bonds	2021/24 Bonds	2021/24 Bonds derivatives	2021/24 Private Bonds	2021/24 Private Bonds derivatives	2021/24 Private Bonds warrants
Balance, December 31, 2020	**57,875**	**-**	**-**	**-**	**-**	**-**
Redemption on exchange	(44,845)	-	-	-	-	-
Issue on exchange	-	30,270	-	-	-	-
Initial recognition of financial instruments at fair value	-	(7,693)	7,693	-	-	-
Nominal value of bonds converted into Shares	-	(10,702)	-	-	-	-
Derecognition of financial instruments on conversion of bonds into Shares	-	-	(2,408)	-	-	-
Effective interest/amortized cost/fair value adjustments	1,819	2,905	(312)	-	-	-
Balance, June 30, 2021	**14,849**	**14,780**	**4,973**	**-**	**-**	**-**
Proceeds from new bonds	-	-	-	15,002	-	-
Repurchased bonds	(1,210)	-	-	-	-	-
Initial recognition of financial instruments at fair value	-	-	-	(4,849)	3,274	1,575
Nominal value of bonds converted into Shares	-	(7)	-	-	-	-
Effective interest/amortized cost/fair value adjustments	241	614	(3,847)	256	(717)	(585)
Balance, December 31, 2021	**13,880**	**15,387**	**1,126**	**10,409**	**2,557**	**990**

(continued)	2017/22 Bonds	2021/24 Bonds	2021/24 Bonds derivatives	2021/24 Private Bonds	2021/24 Private Bonds derivatives	2021/24 Private Bonds warrants
Repayment of bonds	(13,935)	-	-	-	-	-
Nominal value of bonds converted into Shares	(10)	-	-	(3,031)	-	-
Derecognition of financial instruments on conversion of bonds into Shares	-	-	-	-	(1,117)	-
Effective interest/amortized cost/fair value adjustments	65	656	(774)	1,478	209	(510)
Balance, June 30, 2022	**-**	**16,043**	**352**	**8,856**	**1,649**	**480**

Convertible bonds financial instruments valuation and sensitivity analysis

The convertible bonds conversion rights, reset mechanisms, and early redemption options are considered embedded financial derivatives and requires initial recognition and subsequent measurement at fair value through profit or loss. The valuation of the embedded derivatives is based on Level 3 unobservable input parameters applying a simulation-based valuation approach. The implied volatility is determined by reference to the annualized daily trading volatility of Santhera's Shares for a historical lookback period equal to the expected remaining life of the conversion right as of the valuation date.

The embedded conversion rights and reset mechanisms are directly related and have the same risk exposure. Therefore, these two derivatives are accounted for as a single financial instrument (i.e., a compound derivative). Due to the reset mechanisms, the compound derivative is not settled for a fixed number of Shares and hence classifies as a financial liability. The convertible bonds are recognized as financial liabilities measured at amortized cost using the effective interest method and the embedded derivatives are recognized as financial liabilities measured at fair value through profit or loss.

A key input to determine the valuation of the financial instruments, the identified volatility, is calculated based on the historical returns of the Company's Shares over a period commensurate to the duration of the instrument.

The table below shows the implied volatility as of the valuation date:

Financial instruments	**Jun 30, 2022**	**Dec 31, 2021**
Derivatives:		
2017/22 Bonds	-	34%
2021/24 Bonds	68%	81%
2021/24 Private Bonds	68%	64%
Warrants:		
2021/24 Private Bonds	79%	81%

The table below shows the impact that a 5% increase/decrease in volatility has on the fair value for each category of financial instrument and its effect on result before taxes as of the valuation date:

In CHF thousands		**Jun 30, 2022**	**Dec 31, 2021**
Financial instruments	Increase/decrease in volatility assumption	Effect on result before taxes	Effect on result before taxes
2021/24 Bonds – derivatives			
Change in volatility	+5%	(80)	(56)
	-5%	57	219
2021/24 Private Bonds – derivatives			
Change in volatility	+5%	(90)	(132)
	-5%	105	107
2021/24 Private Bonds – warrants			
Change in volatility	+5%	(45)	(60)
	-5%	60	75

8 Fair Value of Financial Liabilities Arising from Financing Activities

The table below summarizes the fair value hierarchy of financial liabilities measured at amortized cost and measured at fair value through profit or loss as of June 30, 2022, and December 31, 2021. During the six months ended June 30, 2022, there have been no transfers between the different hierarchy levels.

In CHF thousands	**June 30, 2022**				
	Carrying value	Level 1	Level 2	Level 3	Total
Exchangeable Notes	14,157	-	14,157	-	**14,157**
2021/24 Bonds	16,043	16,765	-	-	**16,765**
2021/24 Private Bonds	8,856	-	8,827	-	**8,827**
Total financial liabilities at amortized cost	**39,056**	**16,765**	**22,984**	**-**	**39,749**
Derivative financial instruments	5,930	-	-	5,930	**5,930**
Warrant financial instruments	3,093	-	-	3,093	**3,093**
Total financial liabilities at fair value through profit or loss	**9,023**	**-**	**-**	**9,023**	**9,023**

In CHF thousands	December 31, 2021				
	Carrying value	Level 1	Level 2	Level 3	Total
Exchangeable Notes	1,488	-	1,488	-	**1,488**
2017/22 Bonds	13,880	14,000	-	-	**14,000**
2021/24 Bonds	15,387	14,300	-	-	**14,300**
2021/24 Private Bonds	10,409	-	10,209	-	**10,209**
Total financial liabilities at amortized cost	**41,164**	**28,300**	**11,697**	**-**	**39,997**
Derivative financial instruments	4,085	-	-	4,085	**4,085**
Warrant financial instruments	6,373	-	-	6,373	**6,373**
Total financial liabilities at fair value through profit or loss	**10,458**	**-**	**-**	**10,458**	**10,458**

The Group applies the following assumptions in estimating fair values of financial liabilities carried on an amortized cost basis:

- The carrying amounts of short-term debt and current maturities of long-term debt, excluding finance lease obligations, are deemed a reasonable approximation of fair values
- Long-term debt, excluding finance lease obligations: Fair values of the Company's publicly traded convertible bonds are determined using quoted market prices (Level 1 inputs). For convertible bonds and Exchangeable Notes without available quoted market prices, the fair values are determined by reference to the present value of future contractual cash flows discounted at observable market interest rates for instruments with similar characteristics to those held by the Company (Level 2 inputs)

9 Current Provisions

Current provisions mainly consist of restructuring liabilities. In 2020, the Group initiated a restructuring plan in response to the discontinuation of Puldysa development and mainly represent employee-related costs. The changes in restructuring liabilities for the six months ended June 30, 2022 are as follows:

In CHF thousands	
Balance, December 31, 2021	192
Utilization of provision	(48)
Balance, June 30, 2022	**144**

10 Segment and Geographic Information

10.1 Segment information

The Group operates as 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. The Group currently generates revenue from sales of Raxone for the treatment of Leber's hereditary optic neuropathy (**LHON**) and revenues from licensing agreements. Geographic revenue information is based on location of the customer.

10.2 Geographic information – revenues

The following table presents the Company's revenues from contracts with customers disaggregated by region.

In CHF thousands	Six months ended					
	June 30, 2022			**June 30, 2021**		
	Europe	Asia	**Total**	Europe	Asia	**Total**
Net sales	(5,873)	-	**(5,873)**	2,853	-	**2,853**
Revenue from out-licensing transactions	-	11,190	**11,190**	-	-	**-**
Net sales to licensing partner	933	-	**933**	1,639	-	**1,639**
Total revenue from contracts with customers	**(4,940)**	**11,190**	**6,250**	**4,492**	**-**	**4,492**

During the six months ending June 30, 2022, there have been new developments in relation to the ongoing negotiations around pricing reimbursement for sales of Raxone in France as described in Note 18 in the Group's audited consolidated financial statements for the year ended December 31, 2021.

In the Company's opinion, based on the most recent communications with the Comité économique des produits de santé (**CEPS**), its experience to date and discussions with external counsel, it is probable and reasonably estimated for the need to accrue an additional CHF 8.1 million in settlement of the pricing reimbursement. Accordingly, as of June 30, 2022, the Company has recognized an aggregate of CHF 24.9 million in noncurrent accrued expenses. As of December 31, 2021, the aggregate accrual was CHF 16.8 million in noncurrent accrued expenses. This estimate could change substantially over time as new facts emerge and the legal case progresses. Should Santhera be required to make a significant cash payment within the next twelve months, the Group's financial condition, results of operations, and future prospects could be materially adversely affected.

Of the CHF 8.1 million additionally accrued during the six months ending June 30, 2022, CHF 6 million was recognized against net sales and CHF 2.1 million was recognized as marketing and sales expenses. Excluding this adjustment, net sales total approx. CHF 0.1 million and marketing and sales expenses total CHF 3.8 million.

Until a settlement is reached with the CEPS on the future pricing of Raxone for LHON in France, the Company continues to provide Raxone to patients free of charge.

During the six months ending June 30, 2022, Raxone direct sales are to Italy, France, and Switzerland. During the six months ending June 30, 2021, Raxone was sold in five European countries, excluding France, with the majority of sales generated in Germany.

10.3 Geographic information – noncurrent assets

The following table presents the Company's noncurrent assets (excluding financial instruments, deferred tax assets, and pension assets) disaggregated by country.

In CHF thousands	**Jun 30, 2022**	**Dec 31, 2021**
Switzerland	64,139	65,884
United States and Canada	118	36
Total	**64,257**	**65,920**

11 Out-licensing Agreement with Sperogenix Therapeutics Limited

On January 4, 2022, Santhera entered into an exclusive licensing agreement with Sperogenix Therapeutics Limited (**Sperogenix**), a China-based company specializing in orphan diseases. Under the terms of the agreement, Santhera grants Sperogenix exclusive development and commercialization rights to vamorolone for the treatment of DMD and all other rare disease indications in Greater China (including mainland China, Hong Kong, Macau, and Taiwan). As consideration, Santhera;

- received a non-refundable initial payment of USD 12 million;
- is entitled to contingent regulatory-based milestone non-refundable payments of up to USD 22 million; and
- is entitled to contingent sales-based milestone non-refundable payments of up to USD 80 million, in addition double-digit royalties on net sales

Santhera assessed whether the performance obligation(s) promised in the agreement are distinct goods or services or represent a series of distinct goods or services to determine whether revenue is recognized at a point in time or when (or as) the performance obligation is satisfied. According to this assessment, Santhera identified one distinct performance obligation:

- Santhera grants a right to use license to Sperogenix for the development and commercialization of vamorolone in the agreed territory. This performance obligation is satisfied at the point in time when Sperogenix is granted the right of use license.

The regulatory-based milestone payments are contingent upon Santhera obtaining regulatory approval. Therefore, revenue is recognized when the regulatory milestones are achieved. For the sales-based milestone payments, as well as the further double-digit royalties on net sales, these considerations are contingent on Sperogenix achieving sales milestones. As such, revenue for the sales-based milestone payments is recognized if and when the sales threshold is met, with the same exception as for the royalties.

During the six months ended June 30, 2022, Santhera recognized the non-refundable initial payment of USD 12 million (CHF 11.2 million) as revenue from out-licensing transactions.

12 Operating Expenses by Nature

In CHF thousands	Six months ended	
	June 30, 2022	**June 30, 2021**
External development expenses	13,540	8,946
Patent and license expenses	202	263
Marketing and sales expenses	2,547	277
Employee expenses	7,220	7,739
Share-based compensation	1,451	1,152
General and administrative expenses	3,747	3,806
Depreciation and amortization	297	333
Facility related and lease expenses	150	126
Other	836	448
Total operating expenses	**29,990**	**21,938**

13 Financial Income/(Expense)

13.1 Financial income

In CHF thousands	Six months ended	
	June 30, 2022	**June 30, 2021**
Change in fair value of financial instruments, net	4,968	312
Realized and unrealized foreign exchange gains, net	323	206
Recognized gain on exchange of 2017/22 Bonds	-	13,439
Total financial income	**5,291**	**13,957**

13.2 Financial expense

In CHF thousands	Six months ended	
	June 30, 2022	**June 30, 2021**
Interest and make-whole expenses	(8,332)	(10,927)
Interest expense on lease liabilities	(22)	(32)
Transaction costs on financial instruments	(5)	(3,167)
Realized and unrealized foreign exchange losses, net	(528)	(220)
Total financial expense	**(8,887)**	**(14,346)**

14 Income Tax Expense

To determine income tax expense or benefit, various internal and external factors are considered, which may have favorable or unfavorable effects on the future effective tax rate. These factors include, but are not limited to, changes in tax laws, regulations and/or rates, changing interpretations of existing tax laws or regulations, results of tax audits, and changes in the overall level of pre-tax results. The table below summarizes the provision for income taxes for the six months ended June 30, 2022, and June 30, 2021. Movements in deferred taxes relate to temporary differences on inventory.

In CHF thousands	Six months ended	
	June 30, 2022	**June 30, 2021**
Current income taxes	514	85
Deferred taxes	78	568
Total income tax expense	**592**	**653**

15 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. The table below summarizes the equity rights plans' compensation transactions carried out during the six months ended June 30, 2022, and June 30, 2021:

In CHF thousands (except no. of grants)	Six months ended			
	June 30, 2022		**June 30, 2021**	
	Number	Fair value	Number	Fair value
Restricted stock units (A) granted	-	-	200,000	400
Restricted stock units (B) granted	-	-	156,250	313
Performance stock units granted	71,250	56	2,148,225	3,954
Stock options granted	71,250	66	385,225	686

The fair value of equity rights granted is measured on the grant date. For the six months ended June 30, 2022, the fair value measurement range of valuation parameters remained relatively similar with those disclosed in the Group's audited consolidated financial statements for the year ended December 31, 2021, except for the volatility, exercise price (equal to the Share prices at grant), which range between CHF 1.45 and CHF 1.48 (2021: CHF 2.73 and CHF 2.85).

The restricted stock units (A) comprise of three equal tranches with annual vesting on the anniversary of grant and remain restricted until June 21, 2024. The restricted stock units (B) vested on April 27, 2022.

The performance stock units granted during the six months ended June 30, 2022, vest between February 13, 2025 and March 31, 2025 (2021: between April 1, 2024 and June 23, 2024), with staggered vesting and subject to the achievement of pre-defined performance targets. One of such performance targets dependent on satisfying a market condition of a Share price exceeding CHF 9.00 by the vesting date.

The stock options granted during the six months ended June 30, 2022, vest annually over a three-year period and have an exercise price of CHF 2.73 (2021: CHF 2.73).

For the six months ended June 30, 2022, and June 30, 2021, non-cash share-based compensation expense recognized for all long-term equity incentive plans totaled CHF 1.5 million and CHF 1.1 million, respectively.

16 Earnings/(Loss) per Share

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

In CHF thousands (except per share data)	Six months ended	
	June 30, 2022	**June 30, 2021**
Net result attributable to shareholders	(29,724)	(20,519)
Weighted average number of shares issued and outstanding	56,720	22,244
Basic and diluted net result per share	**(0.52)**	**(0.92)**

17 Transactions with Related Parties

The Company's related parties include members of the Board and Executive Management. The table below summarizes the Board and Executive Management compensation expense for the six months ended June 30, 2022, and June 30, 2021:

In CHF thousands	Six months ended	
	June 30, 2022	**June 30, 2021**
Short-term employee benefits	1,426,021	966,351
Post-employment benefits	145,885	95,613
Share-based compensation	346,262	86,132
Total compensation expense	**1,918,168**	**1,148,096**

Detailed remuneration disclosures are provided in the Group's audited consolidated financial statements for the year ended December 31, 2021.

18 Subsequent Events

On September 28, 2022, Santhera and Highbridge agreed to amend the existing financing arrangement that has been previously announced on June 2, 2022, to provide for the immediate drawdown of a CHF 10 million tranche in senior secured Exchangeable Notes and amend certain provisions. Of this amount, approximately CHF 5 million was used to repurchase part of the outstanding convertible bonds issued to Highbridge in 2021 and due in 2024 at a 25 percent discount to its nominal value plus interest. The Exchangeable Notes can be exchanged by Highbridge for Shares at a discount to VWAP, subject to a reduced floor price. As part of this new money financing and further commitments, Santhera has agreed on a new conversion price of CHF 1.20 for the remaining outstanding private convertible bond and a new exercise price of CHF 0.80 per Share for the existing warrants held by Highbridge. A further tranche of CHF 10 million available for drawdown is conditional on management achieving certain milestones and other conditions.

On October 27, 2022, Santhera announced the completion of the rolling submission of a new drug application (NDA) to the U.S. Food and Drug Administration (FDA), seeking priority review for vamorolone for the treatment of Duchenne muscular dystrophy (DMD). Subject to FDA approval, vamorolone is set to become available to patients in the U.S. in H2-2023.

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. The Company has an exclusive license for all indications worldwide to vamorolone, a dissociative steroid with novel mode of action, which was investigated in a pivotal study in patients with Duchenne muscular dystrophy as an alternative to standard corticosteroids. Santhera has submitted a new drug application (NDA) to the U.S. FDA and a marketing authorization application (MAA) to the European Medicines Agency (EMA) for vamorolone for the treatment of DMD. The clinical stage pipeline also includes lonodelestat to treat cystic fibrosis (CF) and other neutrophilic pulmonary diseases. Santhera out-licensed rights to its first approved product, Raxone® (idebenone), outside North America and France for the treatment of Leber's hereditary optic neuropathy (LHON) to Chiesi Group. For further information, please visit www.santhera.com.

Trademarks

Raxone® is a trademark of Santhera Pharmaceuticals.

Forward-Looking Statements

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

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