

# 2018

## **Interim Condensed Report January to September 2018**

## **Interim Financial Statements as of September 30, 2018 of Santhera Pharmaceuticals Holding AG**

### Note

The financial statements as of and for the nine months ended September 30, 2018 have been prepared and are being published exceptionally on the occasion of and in connection with the capital increase proposed to and approved by the EGM held on December 11, 2018.

Santhera publishes results in line with the disclosure requirements of the SIX Swiss Exchange. Santhera does not plan on publishing quarterly reports in the future.



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# Interim Condensed Consolidated Financial Statements

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## Interim Consolidated Balance Sheet

	in CHF thousands	Notes	Sep. 30, 2018 (unaudited)	Dec. 31, 2017 (audited)
<b>Assets</b>				
Tangible assets		10	2,344	2,157
Intangible assets		8, 10	27,443	23,560
Financial assets long-term			775	713
Deferred tax assets			1,245	1,242
Restricted cash long-term		6	1,500	4,500
<b>Noncurrent assets</b>			<b>33,307</b>	<b>32,172</b>
Prepaid expenses and accrued income			804	853
Inventories		5	9,319	10,147
Trade and other receivables			6,953	5,402
Financial assets short-term		9	5,735	13,011
Restricted cash short-term		6	3,000	3,000
Cash and cash equivalents		6	19,654	45,195
<b>Current assets</b>			<b>45,465</b>	<b>77,608</b>
<b>Total assets</b>			<b>78,772</b>	<b>109,780</b>
<b>Equity and liabilities</b>				
Share capital		7	6,528	6,289
Capital reserves and share premium			403,159	392,002
Retained earnings			-400,004	-360,081
Employee benefit reserve			-2,969	-4,905
Treasury shares			-1,145	-335
Other components of equity			-766	-714
<b>Total equity</b>			<b>4,803</b>	<b>32,256</b>
Senior unsecured convertible bonds		9	54,193	53,111
Derivative financial instruments		9	1,069	2,792
Pension liabilities			6,879	8,375
<b>Total noncurrent liabilities</b>			<b>62,141</b>	<b>64,278</b>
Trade and other payables			3,463	4,734
Accrued expenses			8,365	8,512
<b>Total current liabilities</b>			<b>11,828</b>	<b>13,246</b>
<b>Total liabilities</b>			<b>73,969</b>	<b>77,524</b>
<b>Total equity and liabilities</b>			<b>78,772</b>	<b>109,780</b>

## Interim Consolidated Income Statement (Unaudited)

For the three and nine months ended September 30, in CHF thousands	Notes	Three months, ended Sep. 30, 2018	Three months, ended Sep. 30, 2017	Nine months, ended Sep. 30, 2018	Nine months, ended Sep. 30, 2017
<b>Net sales</b>	<b>10</b>	<b>7,607</b>	<b>5,488</b>	<b>23,634</b>	<b>16,347</b>
Cost of goods sold		-1,165	-1,074	-3,606	-3,028
<i>Of which amortization intangible asset</i>		-760	-760	-2,279	-2,279
Other operating income		1	8	1	243
Development	11	-8,244	-6,465	-27,098	-18,168
Marketing and sales	11	-5,716	-7,287	-18,637	-19,909
General and administrative	11	-3,241	-3,460	-11,292	-9,573
Other operating expenses	11	-112	0	-169	-68
<b>Operating expenses</b>	<b>11</b>	<b>-17,313</b>	<b>-17,212</b>	<b>-57,196</b>	<b>-47,718</b>
<b>Operating result</b>		<b>-10,870</b>	<b>-12,790</b>	<b>-37,167</b>	<b>-34,156</b>
Financial income		344	2,687	2,856	3,533
Financial expenses		-1,759	-1,470	-5,232	-3,605
<b>Result before taxes</b>		<b>-12,285</b>	<b>-11,573</b>	<b>-39,543</b>	<b>-34,228</b>
Income taxes	12	-287	952	-380	895
<b>Net result</b>		<b>-12,572</b>	<b>-10,621</b>	<b>-39,923</b>	<b>-33,333</b>
Basic and diluted loss per share (in CHF)		-1.94	-1.69	-6.19	-5.32

## Interim Consolidated Statement of Comprehensive Income (Unaudited)

For the three and nine months ended September 30, in CHF thousands	Three months, ended Sep. 30, 2018	Three months, ended Sep. 30, 2017	Nine months, ended Sep. 30, 2018	Nine months, ended Sep. 30, 2017
<b>Net result</b>	<b>-12,572</b>	<b>-10,621</b>	<b>-39,923</b>	<b>-33,333</b>
<i>Items never to be reclassified subsequently to net income in subsequent periods:</i>				
Net actuarial gains/(losses) from defined benefit plans	781	4	1,936	137
<i>Items to be reclassified subsequently to net income in subsequent periods:</i>				
Currency translation differences	-41	57	-52	60
<b>Other comprehensive result</b>	<b>740</b>	<b>61</b>	<b>1,884</b>	<b>197</b>
<b>Total comprehensive result</b>	<b>-11,832</b>	<b>-10,560</b>	<b>-38,039</b>	<b>-33,136</b>

## Interim Consolidated Statement of Cash Flows (Unaudited)

for the nine months ended September 30, in CHF thousands	Notes	2018	2017
<b>Result before taxes</b>		<b>-39,543</b>	<b>-34,228</b>
Depreciation of tangible assets		455	175
Amortization of intangible assets		2,360	2,338
Expenses for equity rights plans		5,241	5,518
Change in fair value of derivatives	9	-1,723	-2,344
Change in fair value of financial assets short-term	9	249	-117
Other non-cash items (Polyphor clinical material)	8	290	0
Change in pension liabilities		440	584
Taxes paid		-383	-293
Change in net working capital		311	-2,046
Total financial result		2,376	71
Interest received		1	0
Interest paid		-3,033	-1,541
<b>Cash flow from operating activities</b>		<b>-32,959</b>	<b>-31,883</b>
Investments in tangible assets		-1,271	-439
Investments in intangible assets		-33	-104
Investments in other financial assets short-term		0	-12,915
Disposal of other financial assets short-term		7,027	0
Investments in other financial assets long-term		-70	-426
Change in restricted cash		3,000	-7,500
<b>Cash flow from investing activities</b>		<b>8,653</b>	<b>-21,384</b>
Proceeds from options exercised		0	21
Proceeds from sale of treasury shares		1,894	7,437
Purchase of treasury shares		-3,049	-7,626
Proceeds from convertible bonds		0	57,269
<b>Cash flow from financing activities</b>		<b>-1,155</b>	<b>57,101</b>
Effects of exchange rate changes on cash and cash equivalents		-80	94
<b>Net increase/(decrease) in cash and cash equivalents</b>		<b>-25,541</b>	<b>3,928</b>
Cash and cash equivalents at January 1		45,195	49,815
<b>Cash and cash equivalents at September 30</b>		<b>19,654</b>	<b>53,743</b>

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

## Interim Consolidated Statement of Changes in Equity (Unaudited)

	in CHF thousands	Notes	Share capital	Capital reserves and share premium	Retained earnings	Employee benefit reserve	Treasury shares	Translation differences	Total
<b>Balance at January 1, 2017</b>			<b>6,280</b>	<b>382,322</b>	<b>-308,549</b>	<b>-4,734</b>	<b>-172</b>	<b>-796</b>	<b>74,351</b>
Net result			0	0	-33,333	0	0	0	-33,333
Other comprehensive income			0	0	0	137	0	60	197
<b>Total comprehensive result for the period</b>			<b>0</b>	<b>0</b>	<b>-33,333</b>	<b>137</b>	<b>0</b>	<b>60</b>	<b>-33,136</b>
Share-based payment transactions		11	0	5,518	0	0	0	0	5,518
Capital increase from options exercise			5	16					21
Change in treasury shares			0	11	0	0	-200	0	-189
<b>Balance at September 30, 2017</b>			<b>6,285</b>	<b>387,867</b>	<b>-341,882</b>	<b>-4,597</b>	<b>-372</b>	<b>-736</b>	<b>46,565</b>
<b>Balance at January 1, 2018</b>			<b>6,289</b>	<b>392,002</b>	<b>-360,081</b>	<b>-4,905</b>	<b>-335</b>	<b>-714</b>	<b>32,256</b>
Net result			0	0	-39,923	0	0	0	-39,923
Other comprehensive income			0	0	0	1,936	0	-52	1,884
<b>Total comprehensive result for the period</b>			<b>0</b>	<b>0</b>	<b>-39,923</b>	<b>1,936</b>	<b>0</b>	<b>-52</b>	<b>-38,039</b>
Share-based payment transactions		11	0	5,241	0	0	0	0	5,241
Capital increase Polyphor		8	239	6,261	0	0	0	0	6,500
Change in treasury shares			0	-345	0	0	-810	0	-1,155
<b>Balance at September 30, 2018</b>			<b>6,528</b>	<b>403,159</b>	<b>-400,004</b>	<b>-2,969</b>	<b>-1,145</b>	<b>-766</b>	<b>4,803</b>

## Notes to the Interim Condensed Consolidated Financial Statements (Unaudited)

### 1 General Information

Santhera Pharmaceuticals Holding AG (the **Company**, together with its subsidiaries **Santhera** or **Group**) is a Swiss specialty pharmaceutical company focused on the development and commercialization of products for the treatment of neuro-ophthalmological, neuromuscular and pulmonary diseases, areas which include many orphan and rare indications with high unmet medical needs.

The Company, having the listing of its registered shares (**Shares**) on the SIX Swiss Exchange (**SIX**), is a Swiss stock corporation and the parent company of the Group. Its purpose is to acquire, dispose and manage investments. The Company has its registered offices at Hohenrainstrasse 24 in 4133 Pratteln, Switzerland.

The consolidated interim financial statements were approved for publication by the Board of Directors (**Board**) on November 29, 2018.

### 2 Summary of Significant Accounting Policies

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

#### ***Basis of preparation***

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

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

#### ***Material uncertainties and going concern***

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

Santhera continues to generate increasing income from product sales for the indication Leber's hereditary optic neuropathy (**LHON**). Moreover, the Company has collected additional data in patients with Duchenne muscular dystrophy (**DMD**) needed for submission of a Marketing Authorization Application for this indication in the European Union (**EU**) in early 2019. Santhera also prepares for submission of a New Drug Application for patients with DMD in the United States. Lastly, Santhera entered into an option for a sub-licensing agreement with Idorsia for the steroid vamorolone (see note 15 "*Subsequent Events*" for further discussion), which will require an upfront cash payment of USD 20 million within forty days after November 20, 2018.

Santhera's cash, cash equivalents and short-term financial assets amounted to CHF 25.4 million as of September 30, 2018. A material uncertainty remains as to whether Santhera's funding is sufficient to support its going concern for another twelve months.

In order to effect the upfront cash payment to Idorsia for the rights to vamorolone and to fund its ongoing activities, Santhera's Board of Directors has called an Extraordinary General Meeting (**EGM**) to be held on December 11, 2018, and is proposing to the EGM an ordinary capital increase of up to 3,500,000 registered Shares of the Company with a nominal value of CHF 1 each. Santhera plans to raise approximately CHF 50 million of gross proceeds (see note 15 "Subsequent Events" for further discussion). Shareholders should note that whilst the Board and Executive Management continue to apply best efforts to raise additional funds, there is no guarantee that such funds can be raised. The availability of sufficient funds is crucial for Santhera and its ability to continue and grow its operations, including the financing of the upfront cash payment to Idorsia. Based on the Board's and the Executive Management's plan as discussed above, the Board of Directors is confident to ensure business continuation and meet its obligations for a further twelve months. Hence, the interim consolidated financial statements have been prepared on a going concern basis.

### ***Changes in accounting policies***

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

The Group applies, for the first time, IFRS 15 Revenue from Contracts with Customers and IFRS 9 Financial Instruments. Several other amendments and interpretations apply for the first time in 2018, but do not have an impact on the interim consolidated financial statements of Santhera.

IFRS 15 supersedes IAS 11 Construction Contracts, IAS 18 Revenue and related interpretations and it applies to all revenue arising from contracts with customers, unless those contracts are in the scope of other standards. Under IFRS 15, revenue is recognized at an amount that reflects the consideration to which an entity expects to be entitled in exchange for transferring goods or services to a customer. The Group adopted IFRS 15 applying the modified retrospective approach. Revenue from sales of products is recognized at the point in time when the customer obtains control of the goods or services, which is generally upon delivery at the customer. The adoption of IFRS 15 had no material impact on the Group's revenue and profit or loss.

IFRS 9 Financial Instruments replaces IAS 39 Financial Instruments: Recognition and Measurement for annual periods beginning on or after January 1, 2018, bringing together all three aspects of the accounting for financial instruments: classification and measurement, impairment and hedge accounting. The application of the classification and measurement requirements of IFRS 9 had no material impact on the Group's equity and profit or loss. Furthermore, the Group does not apply hedge accounting. IFRS 9 requires Santhera to record expected credit losses (**ECL**) on all of its trade receivables, either on a 12-month or lifetime basis. The Group applied the simplified approach and records lifetime expected losses. Based on the nature of its receivables, the application of the impairment model under IFRS 9 had no material impact on the Group's balance sheet or equity. Consequently, no impact in equity was recorded.

## **3 Seasonality**

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

## 4 Exchange Rates of Principal Currencies

	Income statement in CHF		Balance sheet in CHF	
	average rates for nine months ended		as of period end	
	Sep. 30, 2018	Sep. 30, 2017	Sep. 30, 2018	Dec. 31, 2017
1 Euro (EUR)	1.1610	1.0943	1.1349	1.1691
1 US dollar (USD)	0.9720	0.9840	0.9773	0.9753
1 British pound (GBP)	1.3134	1.2542	1.2775	1.3173
1 Canadian dollar (CAD)	0.7549	0.7531	0.7509	0.7777

## 5 Inventories

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

## 6 Cash and Cash Equivalents and Restricted Cash

### 6.1 Cash and cash equivalents

	in CHF thousands	Sep. 30, 2018	Dec. 31, 2017
Cash at banks and on hand			
in CHF		8,902	34,730
in EUR		7,801	8,152
in USD		1,678	1,496
in GBP		1,033	697
in CAD		152	120
other currencies		88	0
<b>Total at period end</b>		<b>19,654</b>	<b>45,195</b>

### 6.2 Restricted cash

	in CHF thousands	Sep. 30, 2018	Dec. 31, 2017
Long-term		1,500	4,500
Short-term		3,000	3,000
<b>Total at period end</b>		<b>4,500</b>	<b>7,500</b>

Restricted cash is designated for interest payments due related to the convertible bonds during the first 3 years (starting 2017). These funds are kept in an escrow account with the bond agent.

## 7 Share Capital

### 7.1 Ordinary share capital

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

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

### 7.2 Authorized share capital

In February 2018, 238,924 Shares were issued out of the authorized share capital in connection with the agreement with Polyphor (see note 8 *“Transaction with Polyphor”*). On the occasion of the Annual General Meeting (AGM) on April 12, 2018, Santhera’s shareholders approved the increase of the authorized share capital of the Company.

The Board is authorized to increase the share capital at any time until April 11, 2020, through the issuance of up to 1,500,000 Shares with a nominal value of CHF 1 each (see note 15 *“Subsequent Events”*).

### 7.3 Conditional share capital

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

- (i) a maximum amount of CHF 691,302 (2017: CHF 700,000) through the issuance of up to 691,302 (2017: 700,000) Shares, under the exclusion of shareholders’ pre-emptive rights, for equity rights being exercised under the Company’s equity rights plans (see note 13 *“Equity Rights Plans”*).
- (ii) a maximum amount of CHF 930,000 (2017: CHF 930,000) by issuing up to 930,000 (2017: 930,000) Shares through the exercise of warrants/options and/or notes granted in connection with bonds or similar debt instruments linked with option and/or conversion rights granted by the Company.

## 8 Transaction with Polyphor

On February 15, 2018, Santhera announced that it had entered into a license agreement with Polyphor Ltd., Allschwil, Switzerland, for POL6014, a clinical stage selective inhibitor of human neutrophil elastase with the potential to treat cystic fibrosis (CF) and other neutrophilic pulmonary diseases. Under the terms of the agreement, Santhera may be required to make cash payments due to future development, regulatory and sales milestones of up to CHF 121 million (i.e. contingent payments). Consistent with existing licensing agreements, such contingent payments have not been capitalized.

### Significant non-cash transaction

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

## 9 Financial Assets and Liabilities

### 9.1 Financial assets short-term

Financial assets (units in a fund) are measured at fair value through profit or loss and are based on quoted prices (Level 1). A loss of TCHF 254 (financial expenses) resulted during the reporting period (2017: gain of TCHF 117).

### 9.2 Financial liabilities

On February 17, 2017, Santhera issued senior unsecured convertible bonds in the nominal amount of CHF 60 million. The bonds, listed on the SIX, are interest bearing (5%) with a maximum term of 5 years and are convertible into registered Shares of Santhera with a nominal value of CHF 1 each. The initial conversion price was fixed at CHF 86.4006 and has been reset in accordance with the terms of the bond in February 2018 to CHF 64.80. In addition, Santhera may call the convertible bonds at any time on or after the second anniversary of the issue date at par, plus accrued interest, if any, if the VWAP of the Shares is at least 160% of the conversion price. The convertible bonds are measured at amortized costs applying the effective interest method. The fair value of the bonds (Level 1) at September 30, 2018, amounts to CHF 46.7 million (December 31, 2017: CHF 51.6 million).

The embedded financial derivatives (conversion right, reset mechanism and early redemption option) are valued by an independent consultant initially and at period end at fair value, applying a simulation-based valuation approach. The valuation of the embedded derivatives is based on input parameters, classified as Level 3. The simulation is mainly based on the historical volatility of Santhera shares. The period of volatility data used is measured according to the remaining life of the convertible bonds. The volatility used as per September 30, 2018, was at 66.7% (September 30, 2017: 89.3%).

The embedded conversion right and the reset mechanism are directly related and have the same risk exposure. Therefore, these two derivatives are accounted for as a single instrument (i.e. a compound derivative). Due to the reset mechanism, the compound derivative is not settled for a fixed number of equity and hence classifies as a financial liability.

The value of the derivatives initially amounted to CHF 5.3 million (February 17, 2017). At December 31, 2017, the value was CHF 2.8 million and at period end CHF 1.1 million (September 30, 2018). The change in the fair value (CHF 1.7 million) was recognized in financial income.

Sensitivity analysis:

	September 30, 2018		September 30, 2017	
	Increase/decrease in volatility assumption	Effect on result before taxes in CHF thousands	Increase/decrease in volatility assumption	Effect on result before taxes in CHF thousands
Change in volatility	+5%	-111	+5%	163
	-5%	141	-5%	-112

	In CHF thousands	Convertible bonds	Derivative financial instruments
<b>January 1, 2017</b>		<b>0</b>	<b>0</b>
Proceeds from convertible bonds		60,000	0
Transaction costs relating to convertible bonds		-2,731	0
<b>Cash flows in 2017</b>		<b>57,269</b>	<b>0</b>
Non-cash changes			
Initial recognition derivative financial instruments		-5,332	5,332
Change in fair value of derivative financial instruments		0	-2,344
Effective interest/amortized cost calculation		827	0
<b>September 30, 2017</b>		<b>52,764</b>	<b>2,988</b>
Change in fair value of derivative financial instruments		0	-196
Effective interest/amortized cost calculation		347	0
<b>December 31, 2017</b>		<b>53,111</b>	<b>2,792</b>
Change in fair value of derivative financial instruments		0	-1,723
Effective interest/amortized cost calculation		1,082	0
<b>September 30, 2018</b>		<b>54,193</b>	<b>1,069</b>

## 10 Segment and Geographic Information

### 10.1 Segment information

Santhera operates in one business segment, namely development and commercialization of products for the treatment of neuro-ophthalmological, neuromuscular and pulmonary diseases. The Board, the Executive Management and senior managers, being the chief operating decision makers, assess the reporting data and allocate resources as one segment on an aggregated consolidated level according to operating expenses by function. Santhera generates revenue from sales of Raxone for the treatment of LHON. Geographic revenue information is based on location of the customer.

### 10.2 Geographic information

#### Net sales

	nine months ended September 30, in CHF thousands	2018	2017
Europe		23,547	16,282
Rest of the world		87	65
<b>Total</b>		<b>23,634</b>	<b>16,347</b>

In the reporting period 2018, net sales amounted to CHF 23.6 million. Raxone was sold in more than 20 European countries, with the majority of sales generated in France and Germany (in the reporting period 2017, sales went into 18 European countries, with a majority of sales in France and Germany).

**Noncurrent assets (excluding financial instruments and deferred tax assets)**

	in CHF thousands	<b>Sep. 30, 2018</b>	Dec. 31, 2017
Switzerland		29,577	25,451
Rest of Europe		136	171
North America		74	95
<b>Total</b>		<b>29,787</b>	<b>25,717</b>

**11 Operating Expenses by Nature**

	nine months ended September 30, in CHF thousands	<b>2018</b>	<b>2017</b>
External development expenses		-17,248	-10,678
Patent and license expenses		-329	-204
Marketing expenses		-7,538	-9,115
Employee expenses		-26,459	-23,834
<i>Of which non-cash-relevant expenses for share-based payments</i>		-5,241	-5,518
General and administrative expenses		-3,920	-3,074
Depreciation and amortization		-536	-234
Lease expenses		-997	-511
Other operating expenses		-169	-68
<b>Total operating expenses</b>		<b>-57,196</b>	<b>-47,718</b>

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

**12 Income Taxes**

	nine months ended September 30, in CHF thousands	<b>2018</b>	<b>2017</b>
Current income taxes		-382	-293
Deferred taxes		2	1,188
<b>Total</b>		<b>-380</b>	<b>895</b>

Movements on deferred taxes relate to temporary differences on inventory.

**13 Equity Rights Plans**

Santhera has established equity rights plans to align the long-term interests of the members of the Board, the Executive Management and employees. Rights granted under these plans are equity-settled. New grants are only possible under Share Appreciation Rights Plans (**SARP**).

The fair value of equity rights (share appreciation rights (**SAR**) or stock options) is determined at each grant date by using the Hull-White pricing model. For the calculation of the fair value of SAR granted during the reporting period in 2018 the same range of valuation parameters as disclosed in the financial statements as of December 31, 2017, was applied, except for the exercise prices (equal to the Share prices at grant) which were between CHF 16.20 and CHF 36.70. The non-cash-relevant expenses for all unvested SAR and stock options in the reporting period per September 30, 2018, amounted to CHF 5.2 million (2017: CHF 5.5 million).

### 13.1 Share Appreciation Rights Plans

Santhera has established a Board Share Appreciation Plans (**BSARP**), the BSARP 2016, the BSARP 2017, for the members of its Board and Employee Share Appreciation Rights Plans (**ESARP**), the ESARP 2016 and the ESARP 2017, for the Executive Management, employees and consultants. SAR grants are made mainly periodically at the full discretion of the Board or as contractually agreed with employees. SARP introduced in 2017 foresee vesting of 1/3 of the SAR on the first anniversary; the remaining 2/3 vest each following quarter end through the second and third year after the grant date (8 times 1/12 of the SAR granted). In January 2018, Santhera has introduced ESARP 2018 in order to provide a special grant for the Executive Management and employees. Besides the usual terms, this grant contains an additional vesting condition, which is based on Santhera obtaining a positive opinion of the Committee for Medicinal Products for Human Use (**CHMP**) with respect to the marketing authorization of idebenone for the treatment of patients with DMD in the European Union (**EU**).

In the reporting period ended September 30, 2018, a total of 622,282 SARs with exercise prices between CHF 16.20 and CHF 36.70 were granted. In the same period ending September 30, 2017, a total of 316,986 SAR with exercise prices between CHF 54.85 and CHF 77.80 were granted.

#### Number of SAR outstanding

nine months ended September 30, number of SAR	<b>2018</b>	<b>2017</b>
<b>Outstanding at January 1</b>	<b>360,110</b>	<b>56,581</b>
Granted <sup>1</sup>	622,282	316,986
Exercised	0	0
Forfeited	-213,873	-1,231
Expired	0	0
<b>Outstanding September 30</b>	<b>768,519</b>	<b>372,336</b>

<sup>1</sup> The weighted average fair value of the SAR granted during the reporting period in 2018 was CHF 12.11 (in the comparative reporting period 2017 the weighted average fair value of SAR granted was CHF 27.18).

### 13.2 Stock Option Plans

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

In the reporting period ended September 30, 2018, no stock options were granted. In the same period ending September 30, 2017, no stock options were granted.

**Number of stock options outstanding**

nine months ended September 30, number of stock options	2018	2017
<b>Outstanding at January 1</b>	<b>288,442</b>	<b>313,365</b>
Granted	0	0
Forfeited	-19,643	-10,533
Expired	0	-753
Exercised	0	-5,300
<b>Outstanding at September 30</b>	<b>268,799</b>	<b>296,779</b>

**14 Related Party Transactions**

During the reporting period 2018, a total of 62,659 SAR were granted to members of the Board and 139,194 SAR were granted to members of the Executive Management. In the same period in 2017, a total of 15,120 SAR were granted to members of the Board and 104,033 SAR to members of the Executive Management.

**15 Subsequent Events**

On November 20, 2018, Santhera announced the signing of an agreement with Idorsia Pharmaceuticals Ltd, Allschwil, Switzerland (Idorsia). Under the terms of the agreement, Idorsia will grant Santhera the option to an exclusive sub-license for vamorolone in all indications and all territories except Japan and South Korea. Idorsia will receive as consideration for entering into the agreement 1,000,000 (one million) new registered shares from Santhera's existing authorized share capital and an upfront cash component of USD 20 million. Santhera may exercise the option upon receipt of data from the Phase IIb VISION-DMD study (VBP15-004) and following a one-time consideration to Idorsia of USD 30 million.

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

In connection with the above announcement, Santhera informed that it intends to finance the cash payment through an ordinary capital increase.

In order to effect the upfront cash payment to Idorsia for the rights to vamorolone and to fund its ongoing activities, Santhera's Board of Directors has called an Extraordinary General Meeting (EGM) to be held on December 11, 2018, and is proposing to the EGM an ordinary capital increase of up to 3,500,000 registered Shares of the Company with a nominal value of CHF 1 each.

Santhera plans to raise approximately CHF 50 million of gross proceeds. The Company intends to use the net proceeds of the capital increase, together with available liquid funds, to finance the upfront cash payment to Idorsia, to further invest in the development of vamorolone and to fund ongoing business activities including the commercialization of Raxone® in Leber's hereditary optic neuropathy (LHON), market entry preparations for idebenone in DMD, and the development of POL6014 for the treatment of cystic fibrosis.

## About Santhera

Santhera Pharmaceuticals (SIX: SANN) is a Swiss specialty pharmaceutical company focused on the development and commercialization of innovative medicines for rare and other diseases with high unmet medical needs. The portfolio comprises clinical stage and marketed treatments for neuro-ophthalmologic, neuromuscular and pulmonary diseases. Santhera's Raxone® (idebenone) is authorized in the European Union, Norway, Iceland, Liechtenstein and Israel for the treatment of Leber's hereditary optic neuropathy (LHON) and currently commercialized in more than 20 countries. For further information, please visit [www.santhera.com](http://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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# 2018

**Interim Financial Statements  
as of September 30, 2018 of  
Santhera Pharmaceuticals Holding AG**

# Statutory Financial Statements of Santhera Pharmaceuticals Holding AG

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## Balance Sheet

	in CHF thousands	Notes	Sep. 30, 2018
<b>Assets</b>			
Cash and cash equivalents			6,472
Financial assets short-term			5,735
Other receivables from third parties			94
Prepaid expenses and accrued income			202
Restricted cash short-term			3,000
<b>Current assets</b>			<b>15,503</b>
Loans to shareholdings		3.1	91,068
Investments in shareholdings		3.2	216
Restricted cash long-term			1,500
<b>Noncurrent assets</b>			<b>92,784</b>
<b>Total assets</b>			<b>108,287</b>
<b>Liabilities and equity</b>			
Trade accounts payable to third parties			37
Other accounts payable to third parties			43
Accrued expenses			730
<b>Current liabilities</b>			<b>810</b>
Senior unsecured convertible bonds <sup>1</sup>		2	60,000
<b>Noncurrent liabilities</b>			<b>60,000</b>
<b>Total liabilities</b>			<b>60,810</b>
Share capital		3.3	6,528
<i>Reserves from capital contributions</i>			450
<i>Other capital reserves</i>			2,916
Statutory capital reserves			3,366
<i>Accumulated result</i>			-18,767
<i>Results carried forward</i>			-13,752
<i>Net result for the period</i>			-5,015
<i>Other voluntary reserves (free reserves)</i>			57,495
Voluntary accumulated result and other reserves			38,728
Treasury shares		3.4	-1,145
<b>Total equity</b>			<b>47,477</b>
<b>Total liabilities and equity</b>			<b>108,287</b>

<sup>1</sup> interest bearing

## Income Statement

for the nine month period ended September 30, in CHF thousands	<b>Notes</b>	<b>2018</b>
Income from shareholdings	3.5	890
Other operating income		0
<b>Total operating income</b>		<b>890</b>
General and administrative expenses	3.6	-2,179
Employee costs		-952
Other operating expenses		-18
<b>Total operating expenses</b>		<b>-3,149</b>
<b>Operating result</b>		<b>-2,259</b>
Financial income		272
Financial expenses		-3,046
<b>Financial result</b>		<b>-2,774</b>
Reversal on allowance of investment		18
<b>Result before taxes</b>		<b>-5,015</b>
Direct taxes		0
<b>Net result</b>		<b>-5,015</b>

## Notes to the Statutory Financial Statements

### 1 Introduction

Santhera Pharmaceuticals Holding AG (the Company or Santhera) is the parent company of Santhera Group. The Company has its business offices at Hohenrainstrasse 24 in 4133 Pratteln, Switzerland.

### 2 Principles

#### General

The statutory financial statements of the Company are prepared in accordance with the general accepted accounting principles as set out in Art. 957 to Art. 963b, of the Swiss Code of Obligations (**CO**). Since Santhera prepares consolidated financial statements in accordance with International Financial Reporting Standards (**IFRS**) of the International Accounting Standards Board (**IASB**), a recognized accounting standard, the Company has, in accordance with the CO, elected to forego presenting the statement of cash flows, the additional disclosures and the management report otherwise required by the CO.

#### Material uncertainties and going concern

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

Santhera Group continues to generate increasing income from product sales for the indication Leber's hereditary optic neuropathy (**LHON**). Moreover, the Company has collected additional data in patients with Duchenne muscular dystrophy (**DMD**) needed for submission of a Marketing Authorization Application for this indication in the European Union (**EU**) in early 2019. Santhera also prepares for submission of a New Drug Application for patients with DMD in the United States. Lastly, Santhera Pharmaceuticals (Switzerland) AG entered into an option for a sub-licensing agreement with Idorsia for the steroid vamorolone (see note 4.6 "*Events after the reporting date*" for further discussion), which will require an upfront cash payment of USD 20 million within forty days after November 20, 2018.

Santhera Group's cash, cash equivalents and short-term financial assets amounted to CHF 25.4 million as of September 30, 2018. A material uncertainty remains as to whether Santhera Group's funding is sufficient to support its going concern for another twelve months.

In order to effect the upfront cash payment to Idorsia for the rights to vamorolone and to fund its ongoing activities, Santhera's Board of Directors has called an Extraordinary General Meeting (**EGM**) to be held on December 11, 2018, and is proposing to the EGM an ordinary capital increase of up to 3,500,000 registered Shares of the Company with a nominal value of CHF 1 each. Santhera plans to raise approximately CHF 50 million of gross proceeds (see note 4.6 "Events after the reporting date" for further discussion). Shareholders should note that whilst the Board and Executive Management continue to apply best efforts to raise additional funds, there is no guarantee that such funds can be raised. The availability of sufficient funds is crucial for Santhera and its ability to continue and grow its operations, including the financing of the upfront cash payment to Idorsia. Based on the Board's and the Executive Management's plan as discussed above, the Board of Directors is confident to ensure business continuation and meet its obligations for a further twelve months. Hence, the financial statements have been prepared on a going concern basis.

### **Cash**

Santhera holds cash balances, denominated mainly in Swiss francs (**CHF**) which include cash deposited in demand bank accounts, money market investment accounts and other liquid investments and interest earned on such cash balances.

### **Financial assets short-term**

Financial assets (units in a fund) are held for trading and measured at fair value. In case of gains and losses from such assets are recognized through the income statement as financial income or financial expense.

### **Current assets and liabilities**

Current assets are recorded at historical cost less adjustments for impairment of value and current liabilities at historical cost.

### **Loans to shareholdings**

These are valued at their acquisition cost adjusted for impairment losses.

### **Investments in shareholdings**

Investments in shareholdings are recorded at acquisition cost less adjustments for impairment of value. Investments in subsidiaries are evaluated for impairment annually and an impairment loss is recorded when the carrying amount of such assets exceeds the fair value. Fair value estimates of investments are predominantly based on the income approach.

### **Convertible bonds**

On February 17, 2017, Santhera issued senior unsecured convertible bonds in the nominal amount of CHF 60 million. The bonds, listed on the SIX, are interest bearing (5%) with a maximum term of 5 years and are convertible into registered Shares of Santhera with a nominal value of CHF 1 each. The initial conversion price was fixed at CHF 86.4006 and has been reset in accordance with the terms of the bond in February 2018 to CHF 64.80. In addition, Santhera may call the convertible bonds at any time on or after the second anniversary of the issue date at par, plus accrued interest, if any, if the VWAP of the Shares is at least 160% of the conversion price.

### Treasury shares

Treasury shares are recognized at acquisition cost and deducted from shareholders' equity at the time of acquisition. Santhera holds treasury shares for market making which is maintained by an external bank. In case of a resale, the gain or loss is recognized through the income statement as financial income or financial expenses.

### Related parties

In the meaning of the Swiss Accounting Law, related parties are only considered to be shareholders, direct and indirect subsidiaries (shareholdings) and the Board of Directors.

## 3 Information on Balance Sheet and Income Statement Items

### 3.1 Loans to shareholdings

Loans are granted to shareholdings primarily to fund the development and marketing activities of the Santhera Group (September 30, 2018: CHF 263.4 million). Until the end of 2015 the balance consisted of fully impaired and subordinated loans to Santhera Pharmaceuticals (Schweiz) AG. To finance the activities in development and the commercialization of LHON, starting 2016 the loan granted to Santhera Pharmaceuticals (Schweiz) AG was increased (with the additional loans also being subordinated). As part of the reassessment as of September 30, 2018, Executive Management concluded that approximately 35% of the total loan balance is recoverable considering a more positive outlook, both in terms of market success of the developed and launched product (Raxone in LHON) and the development progress in other indications (e.g. idebenone in DMD).

### 3.2 Investments in shareholdings

In 2018, the following companies are direct subsidiaries of Santhera Pharmaceuticals Holding AG (100% ownership and 100% voting rights):

	Share capital	Sep. 30, 2018
Santhera Pharmaceuticals (Schweiz) AG Pratteln, Switzerland	CHF	125,000
Santhera Pharmaceuticals (Deutschland) GmbH Lörrach, Germany	EUR	25,000
Santhera Pharmaceuticals (USA), Inc. Burlington, US	USD	1,000
Santhera Pharmaceuticals (Canada), Inc. Montréal, Canada	CAD	1,000
Oy Santhera Pharmaceuticals (Finland) Ltd Helsinki, Finland	EUR	2,500

Santhera Pharmaceuticals (Schweiz) AG is the primary operational entity while Santhera Pharmaceuticals (Deutschland) GmbH holds the market authorization for the EU. Oy Santhera Pharmaceuticals (Finland) Ltd is not employing any personnel.

The following companies are 100% direct subsidiaries (100% voting rights) of Santhera Pharmaceuticals (Schweiz) AG:

	Share capital	Sep. 30, 2018
Santhera Pharmaceuticals (Liechtenstein) AG Ruggell, Fürstentum Liechtenstein	CHF	50,000
Santhera (Italy) S.r.l. Milano, Italy	EUR	50,000
Santhera (Germany) GmbH München, Germany	EUR	50,000
Santhera (Netherlands) B.V. Nieuwegein, The Netherlands	EUR	50,000
Santhera (UK) Limited London, United Kingdom	GBP	50,000
Santhera Pharmaceuticals (Spain), S.L.U. Bilbao, Spain	EUR	50,000

### 3.3 Share capital

During 2018, the share capital was increased by a total amount of CHF 238,924 to CHF 6,527,479 as of September 30, 2018 (December 31, 2017: CHF 6,288,555) through the issuing of shares from the authorized share capital.

### 3.4 Treasury shares

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

	No of Shares	TCHF
December 31, 2017	9,921	335
Purchase	128,080	3,049
Sale	-81,897	-2,239
September 30, 2018	56,104	1'145

### 3.5 Income from shareholdings

Income from shareholdings represents reimbursement for management services provided by the Company to its major shareholding Santhera Pharmaceuticals (Schweiz) AG.

### 3.6 General and administrative expenses

For the nine months, ended September 30, 2018, In CHF thousands	<b>1.-9.2018</b>
Administrative expenses	997
Consulting expenses	1,182
<b>Total</b>	<b>2,179</b>

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## 4 Other Information

### 4.1 Full-time equivalents

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

### 4.2 Significant shareholders (>2%)

Pursuant to information from the Company's share register and the disclosure of participations made to the Company in accordance with applicable stock exchange regulation, the following shareholders owned 2% or more of the Company's share capital as registered in the commercial register at September 30, 2018: 6,527,479 shares:

	<b>2018</b> <b>Shares<sup>1</sup></b>	<b>2018</b> <b>%</b>
Bertarelli Ernesto, Guichard-Bertarelli Donata and Bertarelli Maria-Iris, Switzerland <sup>3</sup>	536,278	8.2
Iglu Group, Switzerland <sup>3</sup>	533,350	8.2
Roderick Wong (RTW Master Fund, LTD, US)	325,638	5.0
JPMorgan Chase & Co., US	197,761	3.0
Polyphor AG, Switzerland	149,590	2.3

<sup>1</sup> Including disclosures until September 30, 2018

#### 4.3 Disclosure of shares and equity rights (share appreciation rights and stock options) held by members of the Board and Executive Management (and their respective related party)

As of September 30, 2018:

	Number of Shares	Number of vested equity rights	Number of un-vested equity rights	Total number of equity rights
<i>Board of Directors</i>				
Elmar Schnee, Chairman	2,000	21,207	1,870	23,077
Martin Gertsch, Vice-Chairman	38,109	22,026	5,623	27,649
Philipp Gutzwiller, Director	500	14,923	1,317	16,240
Thomas Meier, Director		--- See below ---		
Patrick Vink, Director	1,000	18,502	1,385	19,887
<i>Executive Management</i>				
Thomas Meier, CEO	75,562	39,953	22,833	62,786
Günther Metz, Head Business Development	0	24,905	22,252	47,157
Christoph Rentsch, Chief Financial Officer	0	35,096	21,803	56,899
Kristina Sjöblom Nygren, Chief Medical Officer & Head Development	0	36,937	0	36,937
Giovanni Stropoli, Chief Commercial Officer Europe & Rest of World until September 30, 2018 <sup>1</sup>	250	0	20,751	20,751
Oliver Strub, General Counsel and Secretary to the Board	0	25,472	14,498	39,970

<sup>1</sup> Number of Shares as of September 30, 2018

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

	1.-9.2018	2018
	Quantity	Value (in TCHF) <sup>1</sup>
Board of Directors	62,659	463
Executive Management	139,194	1,266
Employees of Santhera Group	420,429	5,809
<b>Total</b>	<b>622,282</b>	<b>7,538</b>

<sup>1</sup> Value of the equity rights calculated in accordance with the Hull-White model at the date of allocation in accordance with the terms of the award. The tax value of equity rights is 0 until they would be exercised. Such equity rights values are theoretical values and do not reflect income tax values and do also take into consideration certain vesting provisions.

#### 4.5 Contingencies and guarantees

##### *Guarantee towards Swiss VAT authorities*

The Company is part of the value-added tax group of the Swiss affiliated companies of Santhera Pharmaceuticals and is therefore jointly and severally liable to the Swiss federal tax administration for their value-added tax liabilities.

##### *Guarantee towards Santhera Pharmaceuticals (Schweiz) AG*

The Company guarantees to pay for the liabilities of its subsidiary Santhera Pharmaceuticals (Schweiz) AG until the Annual General Meeting in 2019.

##### *Declaration of liability towards Arval Deutschland GmbH*

The Company guarantees to pay for the liabilities of its subsidiary Santhera (Germany) GmbH for contractual duties and obligations.

#### 4.6 Events after the reporting date

On November 20, 2018, Santhera announced the signing of an agreement with Idorsia Pharmaceuticals Ltd, Allschwil, Switzerland (**Idorsia**). Under the terms of the agreement, Idorsia will grant Santhera Pharmaceuticals (Switzerland) AG the option to an exclusive sub-license for vamorolone in all indications and all territories except Japan and South Korea. Idorsia will receive as consideration for entering into the agreement 1,000,000 (one million) new registered shares from Santhera's existing authorized share capital and an upfront cash component of USD 20 million. Santhera Pharmaceuticals (Switzerland) AG may exercise the option upon receipt of data from the Phase IIb VISION-DMD study (VBP15-004) and following a one-time consideration to Idorsia of USD 30 million.

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

In connection with the above announcement, Santhera informed that it intends to finance the cash payment through an ordinary capital increase.

In order to effect the upfront cash payment to Idorsia for the rights to vamorolone and to fund its ongoing activities, Santhera's Board of Directors has called an Extraordinary General Meeting (EGM) to be held on December 11, 2018, and is proposing to the EGM an ordinary capital increase of up to 3,500,000 registered Shares of the Company with a nominal value of CHF 1 each.

Santhera plans to raise approximately CHF 50 million of gross proceeds. The Company intends to use the net proceeds of the capital increase, together with available liquid funds, to finance the upfront cash payment to Idorsia, to further invest in the development of vamorolone and to fund ongoing business activities including the commercialization of Raxone® in Leber's hereditary optic neuropathy (LHON), market entry preparations for idebenone in DMD, and the development of POL6014 for the treatment of cystic fibrosis.