Santhera Pharmaceuticals Holding AG
Hohenrainstrasse 24, 4133 Pratteln, Switzerland

Phone: +41 61 906 89 50 | Fax: +41 61 906 89 51
www.santhera.com

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Ad hoc announcement pursuant to Art. 53 LR

Santhera Announces Preliminary 2021 Financial Results and Increases Issued Share Capital to Create Treasury Shares for Future Financing

- ***Initiation of rolling US NDA submission for vamorolone in Duchenne muscular dystrophy (DMD) on track for end of March 2022***
- ***Cash and cash equivalents at February 28, 2022 of CHF 18 million***
- ***Increases in issued share capital to provide future financing flexibility***
  - ***Issuance of 3,100,000 treasury shares from authorized capital on March 10, 2022***
  - ***Planned additional issuance of 15,500,000 treasury shares in ordinary capital increase on March 14, 2022***

Pratteln, Switzerland, March 11, 2022 – Santhera Pharmaceuticals (SIX: SANN) announces preliminary 2021 financial results and operational progress made. The Company is on track to start a rolling NDA submission for vamorolone in DMD at the end of March. With a cash balance of CHF 18 million (February 28, 2022), the announced increase of share issuances to treasury stock will provide Santhera with financing flexibility, enabling it to raise additional funds.

"Santhera started the year 2021 with the strong foundation of a refreshed clinical and operational strategy. Today, I am pleased to confirm that we continued to make strong progress on all fronts, advancing our initiatives as planned. Most importantly, the filing of a new drug application (NDA) for vamorolone in DMD is on track and we expect to start the rolling submission to the FDA this month. In parallel, we are stepping up market readiness preparations in the U.S., our first launch country for the product," said **Dario Eklund, Chief Executive Officer of Santhera**. "With regard to business development, we closed two agreements, one of which already resulted in cash receipts in January 2022. On the financing side, we extended Santhera's cash reach beyond key milestones and until mid-2022, reduced the short-term debt overhang and strengthened the balance sheet through a bond restructuring. The share capital increase, approved by Santhera's shareholders at the last EGM in December 2021, provides Santhera with additional financing flexibility to be able to raise funds."

2021 OPERATIONS HIGHLIGHTS AND UPCOMING MILESTONES

Throughout 2021 and into 2022, Santhera made progress on all fronts. Santhera has further advanced its core pipeline products, started expanding its U.S. operations, concluded agreements potentially giving rise to non-dilutive cash inflows, and implemented various measures to secure funding and strengthen its capital structure. Ahead of the 2021 Annual Report publication in April 2022, the highlights are summarized below:

Vamorolone. Evidence of clinical efficacy and safety of vamorolone in patients with DMD is based on the positive outcome of the pivotal Phase 2b VISION-DMD study and additional Phase 2a data covering a vamorolone treatment period of up to 30 months. The VISION-DMD study was designed to demonstrate efficacy and safety of vamorolone compared to placebo in addition to comparisons to prednisone as an active control in period 1(initial 24 weeks) followed by the treatment of all patients with vamorolone in period 2 (latter 24 weeks). In this study, vamorolone met the primary endpoint at week 24 and showed sustained efficacy across multiple endpoints over 48 weeks. Treatment was very well tolerated with 54/60 (90%) of subjects continuously on vamorolone completing the 48-week study, of whom only 1 subject (1.6%) discontinued due to an adverse event. In study participants starting on prednisone 0.75 mg/kg/day and switching to vamorolone 6 mg/kg/day after 24 weeks, efficacy was maintained across all functional endpoints. Fewer adverse events commonly associated with corticosteroids were reported with vamorolone compared to prednisone such as growth stunting, negative effect on biomarkers of bone health and behavioral problems, which are some of the most frequent reasons cited for treatment discontinuation with current corticosteroids. Importantly, stunting of growth and negative effect on biomarkers of bone health observed with prednisone over the initial 24 weeks on prednisone were quickly reversed when subjects were switched to vamorolone over the latter 24 weeks of the study.

In November 2021, Santhera announced that the U.S. FDA considered the proposed efficacy and safety data sufficient in support of the filing of a new drug application (NDA). Acceptance of the NDA will be subject to the FDA's review of the complete filing. Santhera and ReveraGen expect to commence the rolling NDA submission at the end of March 2022.

Lonodelestat. Lonodelestat has the potential to treat cystic fibrosis (CF) and other lung diseases associated with increased neutrophil elastase activity. In March 2021, Santhera announced that a Phase 1b multiple ascending dose (MAD) trial of orally inhaled multiple doses of lonodelestat in patients with CF had identified doses with good tolerability of lonodelestat. No serious patient-reported adverse events occurred in the trial. On this basis, Santhera is currently completing the design of the further clinical development program to advance lonodelestat for the treatment of CF and potentially for other inflammatory pulmonary conditions, whether chronic or acute. A 12-week Phase 2a clinical trial of lonodelestat in CF together with a second Phase 2a study in an acute pulmonary indication are under preparation to commence during the second half of 2022.

U.S. operations. Santhera plans to launch vamorolone in the U.S., the first country, shortly after FDA approval with its own organization. Under the leadership of Stephanie Brown, who joined Santhera in December 2021 as President North America and Member of the Executive Management Team, a launch readiness program is being advanced which includes organizational readiness, hiring and building the team and intensifying the pre-commercialization activities.

Business development. Santhera is focusing on its lead development products vamorolone and lonodelestat and is proactively pursuing collaborations with partners to assess and exploit the potential of both clinical stage compounds in other disease areas, beyond DMD and CF, as well as for regions outside the US and main European markets.

In January 2022, Santhera entered into an exclusive license agreement with Sperogenix Therapeutics for vamorolone in the Greater China region. Under this agreement, Sperogenix has in-licensed vamorolone for rare disease indications for a total consideration of up to USD 124 million, including a double-digit upfront cash compensation received at deal closing as well as DMD-related US-regulatory milestone payments and further double-digit royalties on net sales.

In order to focus on its core clinical assets vamorolone and lonodelestat, Santhera discontinued research agreements with University of Basel as well as Rutgers in the area of gene therapy and, in February 2022, entered into a new agreement with SEAL Therapeutics who will further develop a gene therapy approach intended for the treatment of LAMA2-deficient congenital muscular dystrophy. Santhera will be eligible for payments based on future proceeds of SEAL Therapeutics.

Financing. Santhera repaid the remaining CHF 15 million Convertible Bonds (2017/22 CB). Convertible Bonds in the nominal amount of CHF 32.3 million will mature in August 2024. Santhera continues to evaluate various non-dilutive and dilutive financing alternatives, using existing share capital authorizations and treasury shares, which may include existing shareholders' subscription rights, depending on market conditions. This, in combination with cash balances as at February 28, 2022 of CHF 18 million and existing facilities, provides flexibility to secure approximately CHF 100 million (including the USD 50 million milestone payments due upon FDA approval of vamorolone in DMD) which is required to reach breakeven anticipated during H2 2024.

PRELIMINARY UNAUDITED 2021 FINANCIAL RESULTS

Total revenue for the twelve months ended December 31, 2021, amounted to CHF 6.6 million (2020: CHF 15.0 million). The decrease in revenues is mainly attributable to an agreement with the regulatory authorities in France to supply Raxone free of charge from August 2021 while reimbursement discussions are ongoing. Operating expenses of CHF 46.1 million (2020: CHF 58.4 million) were 21% lower due to reduced expenses for development, marketing and sales, and general administrative purposes following the termination of the Puldysa program in 2020 and the subsequent restructuring. The net financial expense of CHF 8.5 million (2020: CHF 14.4 million) reflects the costs associated for financing, partially offset by the effects of the exchange of the 17/22 convertible bond. For 2021, the Company recorded a net loss of CHF 52.6 million (2020: CHF 67.6 million).

As of December 31, 2021, the Company had cash and cash equivalents of CHF 21.2 million compared to CHF 12.4 million as of December 31, 2020.

Total consolidated equity as of December 31, 2021 amounted to CHF 9.9 million compared to a net equity deficit of CHF 6.4 million as of December 31, 2020.

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In CHF thousands	**2021** (preliminary unaudited figures)	**2020** (audited)
Revenue from contracts with customers	**6,644**	**15,008**
Operating expenses	-46,090	-58,347
Operating result	-43,290	-53,076
Net financial result	-8,462	-14,380
Net result	**-58,805**	**-67,659**

In CHF thousands	**Dec 31, 2021** (preliminary unaudited figures)	**Dec 31, 2020** (audited)
Cash and cash equivalents	21,208	12,411
Other current assets	3,263	5,312
Noncurrent assets	66,529	70,964
Total assets	**91,000**	**88,687**
Total equity	**9,909**	**-6,354**
Convertible bonds	49,444	57,875
Other noncurrent liabilities	6,137	8,097
Current exchangeable notes	1,569	10,595
Other current liabilities	23,941	18,474
Total liabilities	**81,091**	**95,041**
Total equity and liabilities	**91,000**	**88,687**

The preliminary key financial figures presented in this press release are subject to change. The Company plans to publish its 2021 Annual Report, with an operational progress update, during April 2022.

INCREASE OF ISSUED SHARE CAPITAL TO BE HELD AS TREASURY STOCK

In order to provide Santhera with additional financing flexibility, enabling it to raise, without delay, new funds, Santhera has issued and plans to issue additional treasury shares. On March 10, 2022, Santhera issued 3,100,000 treasury shares with a nominal value of CHF 1 each out of its existing authorized capital. In addition, Santhera updated its articles of association to reflect past share issuances out of the conditional capital for employee participation and out of the conditional capital for financings. As a result, Santhera's issued share capital currently amounts to CHF 58,225,702. All of these shares are listed on the SIX Swiss Exchange. Santhera will hold the newly issued shares as treasury shares until market conditions permit for a favorable financing transaction.

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On March 14, 2022, Santhera plans to complete the ordinary capital increase resolved by its shareholders on December 15, 2021, and issue 15,500,000 additional treasury shares. The SIX Swiss Exchange has approved a postponement of the listing of these 15,500,000 treasury shares, subject to certain conditions, until July 29, 2022 at the latest. After the completion of the ordinary capital increase, Santhera's issued share capital is expected to amount to CHF 73,725,702 and Santhera expects to hold 21,718,515 shares in treasury.

Concurrently with the ordinary capital increase and as decided by the EGM on December 15, 2021, Santhera's authorized capital will increase from CHF 24,203,905 to CHF 34,203,905 and its conditional capital for financing will increase from CHF 21,374,664 to CHF 31,374,664.

SWISS TAKEOVER BOARD DECISION

Upon completion of the ordinary capital increase, Santhera will exceed for a few hours the mandatory offer threshold of 33 1/3% of the voting rights. Santhera is obliged to publish the decision of the Swiss Takeover Board (the **TOB**) of March 8, 2022:

*"**The Takeover Board decides**:*

1. *Santhera Pharmaceuticals Holding AG and Santhera Pharmaceuticals (Schweiz) AG and any persons acting in concert with them will be exempt from the obligation to make a public takeover offer for Santhera Pharmaceuticals Holding AG as a result of the acquisition of shares in Santhera Pharmaceuticals Holding AG in connection with the ordinary capital increase of Santhera Pharmaceuticals Holding AG resolved at the extraordinary general meeting of December 15, 2021. This exemption from the mandatory offer obligation is granted under the condition that the registration of the newly created shares of Santhera Pharmaceuticals Holding AG in the commercial register takes place on March 14, 2022.*
2. *Santhera Pharmaceuticals Holding AG is required to inform the Takeover Board about the registration in the commercial register of the ordinary capital increase resolved at the extraordinary general meeting of Santhera Pharmaceuticals Holding AG on December 15, 2021.*
3. *Santhera Pharmaceuticals Holding AG is required to publish the provisions of the present decision as well as the reference to the right of objection of qualified shareholders pursuant to Articles 6 and 7 TOO.*
4. *This decision will be published on the website of the Swiss Takeover Board following the publication by Santhera Pharmaceuticals Holding AG in accordance with clause 3 above.*
5. *The fees payable by Santhera Pharmaceuticals Holding AG and Santhera Pharmaceuticals (Switzerland) AG with joint and several liability amounts to CHF 15,000."*

Santhera's Board of Directors has decided not to publish a separate report within the meaning of Article 61 para. 3 lit. a of the Takeover Ordinance.

Shareholders of the Company who have been holding at least 3% of the voting rights of Santhera, whether exercisable or not (a “qualified participation”), since the date of publication of the decision, may file an objection against the decision of the TOB. The objection must be filed with the TOB (Stockerstrasse 54, 8002 Zurich; fax: +41 44 283 17 40) within five (5) trading days from the date of publication of the

decision. The first trading day after the publication of the decision on the TOB's website will be the first day of the filing period. The objection must contain a motion, summary reasons and proof of the qualified participation as from the date of the publication of the decision.

About Santhera

Santhera Pharmaceuticals (SIX: SANN) is a Swiss specialty pharmaceutical company focused on the development and commercialization of innovative medicines for rare neuromuscular and pulmonary diseases with high unmet medical need. Santhera has an exclusive license for all indications worldwide to vamorolone, a first-in-class dissociative steroid with novel mode of action, which was investigated in a pivotal study in patients with DMD as an alternative to standard corticosteroids. The Company is planning for filing for approval with the U.S. FDA starting at the end of March 2022. 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.

Raxone® is a trademark of Santhera Pharmaceuticals.

For further information please contact:

public-relations@santhera.com or
Eva Kalias, Head External Communications
Phone: +41 79 875 27 80
eva.kalias@santhera.com

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