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Ad hoc announcement pursuant to Art. 53 LR

Santhera Announces Preliminary 2022 Half-year Financial Results Ahead of Full Report Publication by End of October

Pratteln, Switzerland, September 29, 2022 – Santhera Pharmaceuticals (SIX: SANN) publishes preliminary financial results for the half-year ended June 30, 2022. Santhera also announces the amendment of its existing financing arrangement with certain funds managed by Highbridge Capital Management, LLC ("Highbridge"), providing for immediate disbursement of a CHF 10 million tranche and making a further CHF 10 million tranche available, subject to certain milestones and conditions, to fund Santhera's development and strategic initiatives. Further, as permitted by SIX Exchange Regulation, the Company will publish the full 2022 Half-year Report by the end of October at the latest.

"2022 has seen us work towards registration and approval of vamorolone and I am happy to report that we are approaching key regulatory milestones for our lead therapy candidate. Our EU filing of a marketing authorization application (MAA) for vamorolone in Duchenne muscular dystrophy (DMD) is planned in the coming weeks and in the U.S., we expect to complete the corresponding new drug application (NDA) submission in Q4-2022," said **Dario Eklund, CEO of Santhera**. "With equally high priority, we are pursuing additional near-term financing to allow us to fund market entry preparations for vamorolone and advance lonodelestat. We are evaluating various options which besides equity-based funding also includes a rights offering, debt financing, royalty financing, standby equity distribution agreement as well as the monetization of assets."

PRELIMINARY 2022 HALF-YEAR FINANCIAL RESULTS

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 the ongoing Raxone reimbursement negotiations in France. Until an agreement is reached 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 the U.S., 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 payment from the out-licensing of vamorolone for the Greater China Region to Sperogenix.

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 and an accrual of CHF 2.1 million in relation to the ongoing Raxone reimbursement negotiations in France. The operating result amounted to CHF -25.5 million (H1-2021: CHF -19.5 million).

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.

The net result for the half-year ended June 30, 2022, was a loss of CHF 29.7 million (H1-2021: net loss of CHF 20.5 million).

For half-year ended June 30, in CHF thousands	**H1-2022** (preliminary, unaudited)	H1-2021 (unaudited)
Net sales	**(5,873)**	2,853
Revenue from out-licensing transactions	**11,190**	-
Revenue from contracts with customers	**6,250**	4,492
Operating expenses	(29,990)	(21,938)
Operating result	**(25,536)**	(19,477)
Net financial result	**(3,596)**	(389)
Net result	**(29,724)**	(20,519)

In CHF thousands	**June 30, 2022** (preliminary, unaudited)	Dec 31, 2021 (audited)
Cash and cash equivalents	**12,697**	21,208
Other current assets	**1,866**	3,433
Noncurrent assets	**65,219**	66,476
Total assets	**79,782**	91,117
Equity	**(13,845)**	1,328
Noncurrent liabilities	**55,051**	57,007
Current liabilities	**38,576**	32,782
Total equity and liabilities	**79,782**	91,117

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

AMENDMENT OF FINANCING BY HIGHBRIDGE TO MEET IMMEDIATE LIQUIDITY REQUIREMENTS

Santhera and Highbridge have amended the existing financing arrangement with Santhera that the Company previously announced on June 2, 2022. Highbridge has agreed to the immediate disbursement of a CHF 10.0 million tranche in senior secured exchangeable notes. Of this amount, around CHF 5 million will be 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 disbursement of further tranches of CHF 10.0 million in aggregate is conditional on certain milestones and other conditions. The exchangeable notes can be exchanged by Highbridge for shares at a discount to VWAP (volume-weighted average price), 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. Santhera has also agreed to establish a new management incentive plan with the goal to maximize value to all stakeholders and will soon announce further steps in this regard.

DECISION OF SIX EXCHANGE REGULATION

SIX Exchange Regulation has permitted Santhera to publish its 2022 Half-year Report on October 31, 2022, at the latest. Santhera is in the process of completing the 2022 Half-year Report and the postponement enables Santhera to reflect its new arrangements with Highbridge. As required by SIX Exchange Regulation, Santhera hereby reprints the following extract of the decision of SIX Exchange Regulation (translation from the German original):

> *The exemption from the obligations for maintaining listing and thus the deferral of the publication of the interim report for the first half of 2022 as well as the filing of this report with SIX Exchange Regulation Ltd by Monday, October 31, 2022 at the latest is hereby approved subject to the following provision (lit. a) and conditions (lit. b):*
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> *a. SIX Exchange Regulation Ltd reserves the right to potentially suspend trading in the securities of Santhera Pharmaceuticals Holding Ltd for a certain period of time if it does not publish its interim report for the first half of 2022 in accordance with the provisions on ad hoc publicity (art. 53 Listing Rules in conjunction with the Directive on Ad hoc Publicity) and submit it to SIX Exchange Regulation Ltd by 23:59 on Monday, October 31, 2022, at the latest.*
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> *b. Santhera has to publish a media release regarding the present decision in accordance with the provisions on ad hoc publicity (art. 53 Listing Rules in conjunction with the Directive on Ad hoc Publicity) by 07:30 a.m. on Friday, September 30, 2022 at the latest. Such media release*
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> - *has to include the full text of clause I of the present decision in a prominent place;*
> - *must mention the reasons for postponing the publication and filing of the interim report for the first half of 2022;*
> - *must mention the unaudited key figures such as net sales, EBITDA, EBIT, net profit/loss, total assets, equity etc. with regard to the business results of the first half of 2022.*

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 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 plans to complete the rolling submission of its filing for approval for vamorolone with the U.S. FDA in Q4-2022. 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.

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Disclaimer / Forward-looking statements

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