

English convenience translation of the authoritative German version
Invitation to the Extraordinary General Meeting of Shareholders

December 11, 2018, 10.30 am
Hohenrainstrasse 24
4133 Pratteln, Switzerland

Agenda (Overview)

1. Ordinary Capital Increase

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Agenda Items, Motions and Explanations

1. Ordinary Share Capital Increase

Motion

The Board of Directors (BoD) proposes to increase the Company's ordinary share capital from CHF 7,527,479 by a maximum of CHF 3,500,000 to a maximum of CHF 11,027,479 as per the following provisions:

1(a)	Total nominal value by which the share capital shall be increased	Up to CHF 3,500,000.
1(b)	Amount of contribution to be paid	CHF 1 per registered share.
2(a)	Number, nominal value and type of shares	A maximum of 3,500,000 registered shares with a nominal value of CHF 1 each.
2(b)	Preferential rights of individual classes of shares	None.
3(a)	Issue Price and Subscription Price	The BoD is authorized to determine the issue price and (if the shares are placed indirectly) subscription price. The total proceeds of the capital increase after deduction of cost, commissions and taxes shall be no less than USD 20,000,000.
3(b)	Beginning of dividend entitlement	January 1, 2019.
4	Type of Contributions	In cash.
5	Takeover of assets (also planned takeovers of assets)	None.
6	Special benefits	None.
7	Restriction on transferability of Shares	The new shares are subject to the restrictions on transferability in accordance with article 5 of the Company's articles of incorporation.
8	Restriction or exclusion of subscription rights and allocation of subscription rights that have not been exercised or that have been withdrawn	The subscription rights have been excluded to allow for a placement of the shares on short notice in the form of a private placement and/or an accelerated book building. The shares will be allocated to investors who are willing to subscribe for a tranche of the newly issued shares at short notice.
9	Prerequisites for the exercise of contractually acquired subscription rights	Do not exist.

Explanations

Background

On November 20, 2018, Santhera Pharmaceuticals Holding Ltd (Santhera) announced the signing of an agreement with Idorsia Pharmaceuticals Ltd, Allschwil (Idorsia). Under the agreement, Santhera is granted an option to obtain from Idorsia an exclusive sub-license to the active ingredient vamorolone for all indications and in all territories, except Japan and South Korea. The granting of the option under said agreement only occurs if and when Santhera has issued 1,000,000 registered shares in Santhera to Idorsia and Santhera has made a payment of USD 20,000,000 to Idorsia, each as set out in further detail below.

Content of the agreement with Idorsia

ReveraGen, the developer of the active ingredient vamorolone, has granted Idorsia a world-wide, unlimited and exclusive option on a license agreement for the commercialization of vamorolone. Under the agreement, Idorsia has committed to grant Santhera an option to a sub-license for vamorolone as soon as Santhera has issued to Idorsia 1,000,000 registered shares in Santhera and the payment of USD 20,000,000 has been made. As a result, Santhera obtains the option to acquire an exclusive sub-license for the commercialization of vamorolone in all territories except Japan and South Korea. The exercise of this option will have to be made no later than the data of pivotal Phase IIb study in patients with Duchenne muscular dystrophy (DMD) are available which is currently expected to be the case in the second half of 2020.¹

Consideration

In accordance with the agreement, the consideration due by Santhera for the granting of the option by Idorsia to acquire a sub-license consists of an equity and a cash component to be issued and paid, respectively, to Idorsia – as described in more detail below. Such consideration does not include payments to be made in the future to Idorsia, at the time of the exercise of the option, when milestones are achieved, and when royalty payments are due. See also the press releases of November 20, 2018.

Equity component

Santhera further agreed to issue to Idorsia 1,000,000 registered Santhera shares with a nominal value of CHF 1 each. These shares will be issued out of Santhera's existing authorized share capital and will be transferred to Idorsia in the coming days under the exclusion of the subscription rights of the existing shareholders. As a result, Idorsia will become the largest shareholder of Santhera with a participation of approximately 13.3% of Santhera's share capital, which after the issue to Idorsia will amount to CHF 7,527,479, divided into 7,527,479 registered shares with a nominal value of CHF 1 each.

Cash component

Additionally, Santhera agreed to make a cash payment to Idorsia in the amount of USD 20,000,000. This cash payment shall *inter alia* reimburse Idorsia for its payment of USD 15,000,000 already made to ReveraGen to finance the ongoing pivotal Phase IIb study for patients with DMD. The granting of

¹ <https://clinicaltrials.gov/ct2/show/NCT03439670>

the option for the sub-license from Idorsia will become effective only if and when the 1,000,000 Santhera shares have been issued and the USD 20,000,000 have been paid to Idorsia.

Financing and extraordinary general meeting

Santhera intends to raise the funds for the payment of the cash component and additionally, cash to continue its business activities by way of the proposed ordinary capital increase. It is currently planned to place the new shares after the extraordinary general meeting (**EGM**) in an accelerated book building (**ABB**).

During the last couple of weeks, banks mandated by Santhera and Santhera Management have engaged in discussions with potential investors who have indicated their interest to participate in this financing and who might potentially be ready to subscribe for a tranche of the newly issued registered shares at short notice.

Should Santhera be unsuccessful in placing shares for at least USD 20,000,000 by the end of the year, the ordinary capital increase would not be effected and the option to acquire an exclusive sublicense under the agreement would not be granted by Idorsia to Santhera; instead, the rights for the distribution of vamorolone would remain with Idorsia under its agreement with ReveraGen. Notwithstanding this outcome, however, in such case, Idorsia would keep the 1,000,000 Santhera shares of the equity component issued to it under the agreement.

In order to raise the required capital at short notice, to be able to address certain investor constituencies, and to allocate to such investors the required participation quota, the subscription rights of the existing shareholders shall be excluded in favor of those investors participating in the financing.

In accordance with the motions proposed to the EGM, the determination of the issue price is delegated to the BoD. The BoD intends to set such price within the offers received from investors, which can differ from the share price at the time of the placement.

While with a minimum placement amount of USD 20,000,000 the amount required as consideration for the option agreement is secured, the Company requires additional funds for its business activities. This is why the BoD intends to raise approximately an additional CHF 30,000,000 beyond the amount of USD 20,000,000 by way of the ordinary capital increase. Even if the Company is successful to raise such additional funds, it cannot be excluded that at a later point in time, the Company will require additional funds.

The BoD is convinced that the agreement with Idorsia is the foundation for the strategic and financial progress of Santhera and that, therefore, the required transactions are in the best interests of the Company and its shareholders.

Pratteln, November 20, 2018
For the Board of Directors

Elmar Schnee
Chairman

Organizational Matters

Admission Cards/Voting

Shareholders registered in the share register with the right to vote on November 20, 2018 at 5 p.m. CET, will receive the invitation to the EGM and – upon request – the admission card and the voting material. Shareholders selling their shares before the EGM will not be entitled to participate in the EGM.

Book closure period

The Company's share register will close on December 3, 2018 at 5:00 p.m. CET and will reopen on December 12, 2018 at 7:00 a.m. CET.

Powers of attorney

Shareholders may give proxy to the independent proxy (*unabhängiger Stimmrechtsvertreter*), Dr. Balthasar Settelen, attorney-at-law, Centralbahnstrasse 7, P.O. Box 206, 4010 Basel, Switzerland or to another shareholder entitled to vote. Proxy may be given by completing and returning the registration form for the admission card and voting material or by filling in the respective proxy form on the admission card. In the absence of specific instructions given to the independent proxy, the proxy form will provide that the independent proxy will be deemed to be instructed to vote in favor of the proposals of the BoD.

Electronic remote votes by proxy and voting instructions to the independent proxy (Netvote)

Shareholders may participate in the vote by giving instructions to the independent proxy electronically via www.netvote.ch/santhera. The required login information will be sent to the shareholders together with the written documents to the EGM. Changes to the electronically transferred instructions can be made until Friday, December 7, 2018, 11.59 a.m. CET.

If you would like to receive the invitation to the general meeting of shareholders by e-mail in the future, you can select the option "select dispatch method" on www.netvote.ch/santhera. The login details are printed on the enclosed reply form. You may change the dispatch method on www.netvote.ch/santhera at any time.

Venue

The meeting place is located at the Hohenrainstrasse 24 in Pratteln, in the offices of the Company on the ground floor. The location can be reached by foot in about five minutes from Pratteln railway station.

Admission

On the day of the EGM, the admission desk will be open from 10.00 a.m. CET. The language of the EGM will be German.