

Santhera Pharmaceuticals Holding AG

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English translation of the German version, which is legally binding.

Invitation to the Annual Shareholders' Meeting

Monday, May 13, 2013, 10:30 a.m. CET, at Santhera Pharmaceuticals, Hammerstrasse 49, 4410 Liestal, Switzerland

Agenda

- 1. Approval of the Annual Report, the Annual Financial Statements, the Consolidated Financial Statements for 2012; Acknowledgment of the Reports of the Auditors**
- 2. Appropriation of the Results**
- 3. Set-off of Capital Contribution Reserves and of Free Reserves with Loss Carry Forwards**
- 4. Discharge of the Members of the Board of Directors and of the Executive Management from Liability for the Financial Year 2012**
- 5. Elections of Members of the Board of Directors**
- 6. Re-election of Auditors**
- 7. Continuation of Business Operations or Liquidation/Opting-out/Other Financial and Strategic Measures**
 - (a) Continuation of Business Operations**
 - (b) Liquidation of the Company**
 - (c) Opting-out of Mandatory Offer Rules**
 - (d) Other Financial and Strategic Measures**

Agenda, Motions and Explanations

1. Approval of the Annual Report, the Annual Financial Statements, the Consolidated Financial Statements for 2012; Acknowledgment of the Reports of the Auditors

Motion

The Board of Directors (**Board**) proposes to approve the Annual Report, the Annual Financial Statements and the Consolidated Financial Statements for 2012.

Explanation

As explained in the Annual Report, the Board thoroughly assessed the operating and financial situation of the Company and the question of continuation of the business operations (going concern). Although there are considerable uncertainties and the going concern cannot be assured based on the financial means available to the Company today, the Board has decided to present the Financial Statements 2012 on a going concern basis. This decision is based on indications, which made the Board, in consideration of all facts and circumstances believe that the going concern assumption may be sustained. Further information regarding the Financial Statements and the going concern assumption as well as the respective uncertainties is available in the Annual Report on pages 7 et seq.

2. Appropriation of the Results

Motion

The Board proposes to carry forward the entire loss for the year of CHF 164,299,838 to new account.

Explanation

For 2012, the Company's losses amounted to CHF 164,299,838. The Board proposes to carry forward these losses entirely to new account.

3. Set-off of Capital Contribution Reserves and of Free Reserves with Loss Carry Forwards

Motion

The Board proposes to set-off:

- an amount of CHF 151,045,795 from capital contribution reserves with an equivalent amount of the net loss carry forwards, and
- an amount of CHF 11,722,815 from free reserves with the remaining equivalent amount of the net loss carry forwards.

Explanation

The proposed set-offs are accounting measures and do not have any cash impact. As a result of such bookings, the capital contribution reserves would decrease to CHF 0 and the

free reserves would decrease to CHF 3,769,587, while the loss carry forwards would be eliminated entirely. If and to the extent the capital contribution reserves could be used for tax free distributions, setting off the loss carry forwards would eliminate this potential.

4. Discharge of the Members of the Board of Directors and of the Executive Management from Liability for the Financial Year 2012

Motion

The Board proposes to grant discharge to the members of the Board and of the Executive Management for the financial year 2012.

5. Election of Members of the Board of Directors

Motions

The Board proposes to re-elect Martin Gertsch for a period of one year until the Annual Shareholders' Meeting 2014.

Explanation

The terms of office of Mr. Klaus Schollmeier and Mr. Martin Gertsch expire on the occasion of this year's Annual Shareholders' Meeting. Mr. Klaus Schollmeier is not standing for re-election.

Mr. Martin Gertsch is standing for re-election. Martin Gertsch, up to now Vice-Chairman of the Board and Chairman of the Audit Committee, became a member of the Board in 2006. He has long-time experience as a financial expert and is currently CFO of Acino with vast experience in the Pharma industry. In case of his re-election, the Board intends to appoint Martin Gertsch as the new Chairman. Additional biographical information is available in the 2012 Corporate Governance Report and at www.santhera.com/board.

Mr. Timothy Rink submitted his resignation as per the date of the Annual Shareholders' Meeting.

Subject to the shareholders' approval, the Board would therefore consist of two members, Martin Gertsch and Jürg Ambühl.

6. Re-election of Auditors

Motion

The Board proposes to re-elect Ernst & Young AG, Basel, as auditors for an additional period of one year until the Annual Shareholders' Meeting 2014.

Explanation

Ernst & Young, Basel, assumed the existing auditing engagement for Santhera's predecessor company in 2002 and has ever since been Santhera's auditors without interruption.

7. Continuation of Business Operations or Liquidation/Opting-out/Other Financial and Strategic Measures

(a) Continuation of Business Operations

Motion

The Board proposes to approve the continuation of business operations in a consultative vote as follows:

The continuation of business operations, while implementing operational restructuring measures and continuously pursuing financial and strategic options, is approved.

(b) Liquidation of the Company

Motion

Should the Annual Shareholders' Meeting not approve the continuation of business operations in the consultative vote as per agenda item 7(a), the Board will propose the liquidation of the Company as follows:

- (1) The Company shall be dissolved and liquidated.
- (2) Copartner Revision AG with its registered office in Basel is elected as liquidator.
- (3) The Board of Directors shall apply for entry of the dissolution of the Company in the Commercial Register (art. 737 CO).

(c) Opting-out of Mandatory Offer Rules

Motion

The Board proposes to resolve an opting-out of the mandatory offer rules of articles 32 and 52 of the Swiss Stock Exchange and Securities Trading Act (**SESTA**) based on article 22 para. 3 SESTA by adopting a new Article 6a of the articles of incorporation of the Company as follows:

"Article 6a

Opting-out

The obligations to submit a public offer for the equity securities of the Company pursuant to articles 32 and 52 of the Swiss Stock Exchange and Securities Trading Act and any relating

obligation are hereby waived."

(d) Other Financial and Strategic Measures

Currently, the Board does not propose any other measures, but reserves to submit further proposals in view of possible future events, or proposals submitted by the shareholders.

Explanations to agenda items 7(a) through (d)

General Explanations

Further to Santhera's filing of a Marketing Authorization Application (MAA) with the European Medicines Agency's (EMA) Committee for Medicinal Products for Human Use (CHMP) for Raxone® as a potential therapy for Leber's Hereditary Optic Neuropathy (LHON) in 2011, the CHMP issued a negative opinion on the MAA in January 2013. In March 2013, Santhera dropped its intent to apply for re-examination for strategic reasons, thereby withdrawing its MAA.

Santhera plans to file a new MAA based on emerging clinical evidence further evaluating the efficacy of Raxone® in the treatment of LHON. Santhera currently anticipates that a new MAA can likely be filed in or around December 2013. If the new MAA can be filed as planned, Santhera believes that the opinion of the CHMP could be available in or around December 2014. Santhera expects that in case of a positive CHMP opinion, the marketing authorization would likely be granted within the two following months. Based on new clinical evidence and internal and external assessments, the Board came to the conclusion that the chances for a positive CHMP opinion are given despite the previous dismissal.

Explanations regarding Agenda Item 7(a) – Continuation of Business Operations

Prior to the date of this invitation, Santhera announced additional restructuring measures with the goal of further reducing its costs and liabilities.

Despite these measures, Santhera's actual funding at the time of this invitation is not sufficient to ensure the continuation of business operations for at least twelve months. Therefore, Santhera depends on additional funding in order to be able to implement the above-mentioned strategy, which is to file a new MAA and to continue operations up to and through the CHMP opinion on the new MAA, the market authorization and beyond. Currently, such additional funding is not secured.

In case of approval of agenda item 7(a) by the Annual Shareholders' Meeting to continue the Company's business operations, it can therefore, despite the going concern assumption underlying the Financial Statements 2012 (cf. explanation to agenda item 1), not be excluded that Santhera will have to initiate the liquidation procedure in the course of the year

2013, if Santhera will not be successful in securing the additionally required funding. In such event, based on current projections and taking into account the above-mentioned restructuring measures, the Company would presumably have to initiate its (solvent) liquidation in November 2013 at the latest, and, likely, no liquidation dividend could be paid to shareholders. In contrast, if the liquidation was initiated today, current assumptions suggest that a small liquidation dividend in an amount below the nominal value could be expected.

Having thoroughly considered all options, the Board came to the conclusion that it is in the interest of the Company to pursue the remaining opportunities for a continuation of the Company's business operations. However, in view of the risks of loss, the Board also decided to consult the shareholders by means of the consultative vote pursuant to agenda item 7(a).

In case of an approval of agenda item 7(a), the Board would continue to look into and pursue all financial and strategic options of the Company. These options are not limited to capital and financing measures supporting the continuation of business operations as described above, but do also include merger, sale, licensing, takeover and similar transactions and scenarios, which depending on the circumstances might exclude a continuation of business operations within the framework of today's Santhera. The Board has received expressions of interest and letters of intent regarding different types of transactions, which the Board would continue to pursue.

The Board would like to make the shareholders aware that Santhera will continue to monitor the financial situation and take measures if needed (including to convene another shareholders' meeting or to notify the court in case of over-indebtedness pursuant to article 725 CO), irrespective of the shareholders' meeting's vote on the continuation of business operations.

Explanations regarding Agenda Item 7(b) – Liquidation of the Company

In case the Annual Shareholders' Meeting does not approve the continuation of business operations as per agenda item 7(a), the Board will bring to a vote and propose the liquidation of the Company pursuant to agenda item 7(b).

Certain types of transactions mentioned above in the context of agenda item 7(a) could possibly also be implemented in case of a dissolution and liquidation of the Company pursuant to agenda item 7(b) or themselves result in a liquidation of the Company.

Explanations regarding Agenda Item 7(c)

The Board proposes the introduction of an opting-out clause pursuant to agenda item 7(c) in order to facilitate a possible further financing of the Company and in order not to deter shareholders or other investors from subscribing for shares or submitting a takeover offer

due to the applicability of the mandatory offer rules pursuant to the SESTA. Without such opting-out, investors would run a potential risk of having to submit an offer to all shareholders of the Company. In addition, without an opting-out, they may have to comply with minimum price rules that reflect historic stock prices which may not be realistic anymore.

The Board thus believes that in the current situation it is in the best interest of the Company to opt out of the mandatory offer rules and related obligations (in particular minimum price rules) pursuant to the SESTA.

The Board is currently not in possession of proposals of shareholders or investors requesting the introduction of an opting-out. However, shareholders should be aware that by approving an opting-out, they will forego the protections offered by the mandatory offer rules pursuant to the SESTA, including the obligation of bidders to extend an offer to all holders of listed equity securities of the Company and the obligation to offer a minimum price.

With an opting-out, the following minimum price rules would no longer apply with respect to the Company: Pursuant to the current article 32 para. 4 SESTA, the offer price (i) must not be lower than the stock exchange price or a valuation, and (ii) it must not be more than 25% below the highest price paid by the offeror for equity securities of the target company in the preceding twelve months (prior to announcing the offer). Pursuant to the revised article 32 para. 4 SESTA which has not yet come into effect, subtest (ii) will be amended and provide that the offer price must at least amount to the highest price paid by the offeror for equity securities of the target company in the preceding twelve months (prior to submitting the offer). According to the website of the Federal Finance Department, the revised SESTA will presumably enter into force on May 1, 2013.

Explanations regarding Agenda Item 7(d)

In view of the Annual Shareholders' Meeting, the Board also considered to propose specific capital measures, in particular a capital increase. For the time being, the Board has refrained from proposing such capital measures as the pursued options are not sufficiently advanced and the Board is currently also not in possession of a sufficiently materialized expression of interest for investment from existing shareholders or other investors which would justify the resulting costs.

Agenda item 7(d) would, however, allow the Board to make proposals addressing other measures that may come up prior to the shareholders' meeting and shareholders to come up with their own proposals with respect to a financing or otherwise with respect to the future of the Company.

Liestal, April 16, 2013

For the Board of Directors

Klaus Schollmeier

Chairman

Organizational Remarks

Documents

The Company's 2012 Annual Report is available for inspection at the Company's registered domicile at Hammerstrasse 49, 4410 Liestal, Switzerland. It can also be downloaded from Santhera's website (www.santhera.com/reports). Shareholders who wish to receive a printed copy of the Annual Report are kindly requested to tick the corresponding box on the reply form.

Admission cards/voting materials

Shareholders entered into the share register with the right to vote as of May 8, 2013, at 5:00 p.m. CET, will receive the invitation and – upon request – the admission card and the voting materials. A shareholder selling her/his shares before the Annual Shareholders' Meeting is not entitled to participate in the Annual Shareholders' Meeting.

Book closure period

The Company's share register will close on May 8, 2013, at 5:00 p.m. CET, and will reopen on May 14, 2013, at 7:00 a.m. CET.

Powers of attorney

Shareholders may give proxy to (i) the independent proxy (*unabhängiger Stimmrechtsvertreter*), Dr Caspar Zellweger, attorney-at-law, Elisabethenstrasse 2, 4010 Basel, Switzerland, (ii) the Company representative (*Organvertreter*), (iii) a depositary (*Depotvertreter*) or (iv) another shareholder entitled to vote. Proxy may be given by completing and returning the registration form or by filling out the respective form on the admission card.

Location

Santhera's offices (address on page 1) are in walking distance from the SBB railway station Frenkendorf-Füllinsdorf and about an hour from Basel airport (EuroAirport) by public transport.

Admission

At the day of the meeting, the admission desk will be open from 10:00 a.m. CET.

Language

The language of the meeting will be German.

Forward-looking statements

This document contains certain forward-looking statements concerning Santhera and its business. Such statements involve certain risks, uncertainties and other factors which could cause the actual results, financial condition, performance or achievements of Santhera to be materially different from those expressed or implied by such statements. Readers should therefore not place undue reliance on these statements, particularly not in connection with any contract or investment decision. Santhera disclaims any obligation to update these forward-looking statements.