

Santhera Pharmaceuticals Holding AG Hohenrainstrasse 24, 4133 Pratteln Schweiz

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English convenience translation of the authoritative German version.

Invitation to the Annual General Meeting of Shareholders (AGM)

Thursday, June 30, 2022, 10:30 a.m. Hohenrainstrasse 24 4133 Pratteln, Switzerland

Pursuant to Art. 27 para. 1 of the COVID-19 Ordinance 3, companies may provide that their shareholders exercise their rights solely by way of giving voting instructions to the independent proxy (*unabhängiger Stimmrechtsvertreter*).

On this basis, the Company hereby mandates that all shareholders exercise their rights at the ordinary General Meeting of shareholders solely via the independent proxy. There is no possibility to attend the AGM in person.

Information on how to issue power of attorney and instructions to the independent proxy, electronically or in writing, can be found on page 11 of this invitation.

The Board of Directors and the Executive Management of Santhera look forward to holding the 2023 Annual General Meeting again in the presence of shareholders.

Agenda (Overview)

- 1. Approval of the Annual Financial Statements and the Consolidated Financial Statements 2021
- 2. Appropriation of the Annual Result and Allocation of Reserves
- 3. Consultative Vote on the Compensation Report 2021
- 4. Discharge of the Members of the Board of Directors and of the Executive Management from Liability for the Financial Year 2021
- 5. Increase of Authorized Capital and Amendment to the Articles of Incorporation
- 6. Capital Reduction by Reduction of Nominal Value and Amendments to the Articles of Incorporation
- 7. Re-election of the Members of the Board of Directors and of the Chairman of the Board
- 8. Re-election of the Members of the Compensation Committee
- 9. Approval of the Compensation of the Members of the Board of Directors
- 10. Approval of the Compensation of the Members of the Executive Management
- 11. Re-election of the Statutory Auditors
- 12. Re-election of the Independent Proxy

Agenda Items, Motions and Explanations

1. Approval of the Annual Financial Statements and the Consolidated Financial Statements 2021

Motions

The Board of Directors proposes to approve the Annual Financial Statements and the Consolidated Financial Statements for the 2021 financial year.

2. Appropriation of the Annual Result and Allocation of Reserves

Motions

(a) Loss Carryforward

The Board of Directors proposes to carry forward the annual loss for 2021 of CHF 1,613,379 to the new account.

(b) Allocation of Reserves from Capital Contribution to Free Reserves

The Board of Directors proposes to allocate the amount of CHF 19,000,000 from the reserves from capital contributions to the free reserves.

Explanations

(a) Loss Carryforward

The net annual loss for 2021 amounted to CHF 1,613,379. The loss carryforward of the previous financial years amounts to CHF 39,801,095. After appropriation of the annual loss, the total loss carryforward amounts to CHF 41,414,474.

In CHF	2021	2020
Loss carryforward of previous financial years	-39,801,075	-32,377,063
Annual loss	-1,613,379	-7,424,032
Total loss carryforward	-41,414,474	-39,801,095

(b) Allocation of Reserves from Capital Contributions to Free Reserves

The Board of Directors proposes to re-allocate an amount of CHF 19,000,000 from the reserves from capital contributions (which amount to CHF 19,187,643) to the free reserves. To this extent, the reserves from capital contributions could no longer be used for tax-free distributions. As a result of such allocation, the free reserves would be increased from CHF 95,994,714 to CHF 114,994,714 and the reserves from capital contributions would be reduced from CHF 19,187,643 to CHF 187,643.

3. Consultative Vote on the Compensation Report 2021

Motion

The Board of Directors proposes to approve the Compensation Report 2021 in a non-binding consultative vote.

Explanations

The Compensation Report 2021 covers the fundamentals of the compensation for the members of the Board of Directors and of the Executive Management as well as the compensation paid to them during the financial year 2021. In compliance with Article 25 of the Articles of Incorporation and the recommendations of the Swiss Code of Best Practice for Corporate Governance, the Board of Directors submits the Compensation Report 2021 to the AGM for a non-binding consultative vote.

4. Discharge of the Members of the Board of Directors and of the Executive Management from Liability for the Financial Year 2021

Motions

(a) Discharge of the Members of the Board of Directors

The Board of Directors proposes to grant discharge from liability to the members of the Board of Directors for their activities during the financial year 2021.

(b) Discharge of the Members of the Executive Management

The Board of Directors proposes to grant discharge from liability to the members of the Executive Management for their activities during the financial year 2021.

5. Increase of Authorized Capital and Amendment to the Articles of Incorporation

Motion

The Board of Directors proposes to increase the authorized share capital by CHF 2,656,782 and an extension thereof until June 29, 2024, and to amend Article 3a of the Articles of Incorporation as set out in the Annex.

Explanations

Currently, the authorized share capital amounts to CHF 34,203,905. The Board of Directors proposes to increase the authorized share capital to the statutory maximum, being 50% of the share capital registered in the commercial register, and to extend the authorized share capital to the statutory maximum, i.e., two years. With such increase and extension, Santhera would have additional flexibility with respect to its foreseen financing in the near future.

6. Capital Reduction by Reduction of Nominal Value and Amendments to the Articles of Incorporation

Motion

- (1) The Board of Directors proposes to reduce the Company's share capital by CHF 72,984,160.26 from CHF 73,721,374.00 to CHF 737,213.74 through the reduction of the nominal value of the registered shares from CHF 1.00 to CHF 0.01 per share, and to allocate the aggregate reduction amount to the reserves from capital contribution. The amount of the reduction of the Company's share capital shall be increased by the reduced nominal value of the registered shares that may be issued from the Company's authorized and conditional share capital until the consummation of the capital reduction.
- (2) The Board of Directors proposes to reduce the nominal value of the shares issuable from the authorized share capital (art. 3a of the Articles of Incorporation) and conditional share capital (art. 3b and art. 3c of the Articles of Incorporation) from CHF 1.00 to CHF 0.01 per share, and to reduce the authorized share capital (art. 3a of the Articles of Incorporation) and the conditional share capital (art. 3b and art. 3c of the Articles of Incorporation) by a corresponding amount, and to amend the Articles of Incorporationaccordingly, all subject to and with effect as of the registration of the reduction of the Company's share capital as proposed in motion (1) with the commercial register of the Canton of Basel-Landschaft.
- (3) The Board of Directors proposes to determine the result of the audit report pursuant to art. 732 para. 2 of the Swiss Code of Obligations (CO) by Ernst & Young AG, Basel, which has been made available to the shareholders and is available at the General Meeting, according to which the claims of the creditors are fully covered despite the reduction of the share capital.

Explanations

The shares of Santhera are currently trading slightly above the nominal value of CHF 1 per share. Swiss law does not allow for the issue of shares below their nominal value. Therefore, the Board of Directors recommends reducing the nominal value in order to have more flexibility in light of the intended financings.

The proposed reduction of the nominal value is a purely technical measure. The total equity base remains unchanged, as does the number of shares currently issued. The amount by which the Company's share capital will be reduced shall be allocated to the reserves from capital contribution.

According to Swiss law, an audit report is required for the proposed capital reduction. The corresponding report by Ernst & Young AG, Basel, can be found on Santhera's website at https://www.santhera.com/investors-and-media/investor-toolbox/share-bondholder-meetings.

7. Re-election of the Members of the Board of Directors and of the Chairman of the Board

Motions

(a) Re-election of Elmar Schnee to the Board of Directors

The Board of Directors proposes the re-election of Elmar Schnee as a member of the Board of Directors until the end of the 2023 AGM.

(b) Re-election of Philipp Gutzwiller to the Board of Directors

The Board of Directors proposes the re-election of Philipp Gutzwiller as a member of the Board of Directors until the end of the 2023 AGM.

(c) Re-election of Thomas Meier to the Board of Directors

The Board of Directors proposes the re-election of Thomas Meier as a member of the Board of Directors until the end of the 2023 AGM.

(d) Re-election of Patrick Vink to the Board of Directors

The Board of Directors proposes the re-election of Patrick Vink as member of the Board of Directors until the end of the 2023 AGM.

(e) Re-election of Elmar Schnee as Chairman of the Board of Directors

The Board of Directors proposes the re-election of Elmar Schnee as Chairman of the Board of Directors until the end of the 2023 AGM.

Explanations

The members and the Chairman of the Board of Directors will have to be elected individually. The term of office of all members of the Board of Directors will end at this year's AGM. All members of the Board of Directors stand for re-election. Elmar Schnee is proposed for re-appointment as chairman of the Board of Directors. His experience provides crucial support to the management, in particular as the Company is moving towards a first launch of vamorolone in the USA.

8. Re-election of Members of the Compensation Committee

Motions

(a) Re-election of Elmar Schnee as member of the Compensation Committee

The Board of Directors proposes the re-election of Elmar Schnee as a member of the Compensation Committee until the end of the 2023 AGM.

(b) Re-election of Patrick Vink as member of the Compensation Committee

The Board of Directors proposes the re-election of Patrick Vink as a member of the Compensation Committee until the end of the 2023 AGM.

Explanations

The members of the Compensation Committee will have to be elected individually by the AGM. Only members of the Board of Directors are eligible. It is intended that Patrick Vink will be re-appointed as Chairman of the Compensation Committee.

9. Approval of the Compensation of the Members of the Board of Directors

Motion

The Board of Directors proposes the approval of a maximum total amount of fixed compensation of the Board of Directors of CHF 625,000 (excl. employer contributions to social security) until the 2023 AGM.

Explanations

The current members of the Board of Directors are nominated for re-election at the 2022 Annual General Meeting.

In case of approval of the proposed total compensation of the Board of Directors by the AGM, the Board of Directors plans to compensate the chairmanship and the membership of the Board of Directors, and its Committees as follows:

Function	Compensation (CHF)*	Number	Total (CHF)*
Chairman of the Board of Directors	180,000	1	180,000
Member of the Board of Directors	115,000	3	345,000
Chairman of the Audit Committee	30,000	1	30,000
Member of the Audit Committee	10,000	1	10,000
Chairman of the Compensation Committee	20,000	1	20,000
Member of the Compensation Committee	10,000	1	10,000
Chairman of the Scientific Committee	20,000	1	20,000
Member of the Scientific Committee	10,000	1	10,000
Total			625,000

^{*} excluding employer contributions to social security

The total compensation of CHF 625,000 (excluding legally required employer's contributions to social security) would be paid at a minimum of 50% in the form of restricted shares and the remainder in cash fees. The value of the restricted shares represents a market value that takes into account the following elements, among others: Share price on the day of the AGM, vesting on the day before the 2023 AGM and thereafter a restriction period until June 30, 2025, during which the restricted shares may generally not be sold. Members of the Board of Directors may elect to receive up to 100% of their compensation in the form of restricted shares.

The table below sets out the approved maximum compensation for the Board of Directors for the period from the 2021 AGM to the 2022 AGM, the estimated actual compensation for the same period and the proposed maximum compensation for the period from the 2022 AGM to the 2023 AGM.

	Proposed AGM 2022 - 2023	Approved AGM 2021 – AGM 2022	Paid/payable AGM 2021 – AGM 2022
Board fees (CHF) cash			223,125
Board fees (CHF)			
elected to be received			
as RSU	312,500		89,375
Board fees (CHF)			
mandatory RSU			
portion	312,500	312,500	312,500
Total ordinary			
compensation (CHF)	625,000	625,000	625,000
Extraordinary			
compensation (CHF) ¹	0	400,000	400,000
Total compensation			
(CHF)	625,000	1,025,000	1,025,000

¹⁾ Extraordinary compensation approved at the AGM on June 22, 2021, in the form of restricted shares.

10. Approval of the Compensation of the Members of the Executive Management

Motions

(a) Fixed Compensation of the Members of the Executive Management for 2023

The Board of Directors proposes the approval of a maximum total amount of fixed compensation of the members of the Executive Management for the period from January 1, 2023 to December 31, 2023 of CHF 2,950,000 (incl. employer contributions to social security and pension plans).

(b) Variable Cash Compensation of the Members of the Executive Management for 2021

The Board of Directors proposes a variable compensation for the financial year 2021 for the Executive Management in the maximum total amount of CHF 1,200,000 (incl. employer contributions to social security and pension plans).

(c) Variable Compensation of the Members of the Executive Management for 2022

The Board of Directors proposes a variable compensation for the financial year 2022 for the Executive Management in the maximum total amount of CHF 2,400,000 (incl. employer contributions to social security and pension plans).

Explanations

According to Article 25 of the Articles of Incorporation, the maximum fixed compensation of the Executive Management must be approved in each case for the following year, i.e. for the period from January 1 to December 31, 2023. In addition, the Board of Directors must submit for approval any variable compensation of the Executive Management for the previous year, i.e., for the period from January 1 to December 31, 2021.

(a) Fixed Compensation of the Members of the Executive Management for 2023

At the 2021 AGM, the maximum amount of CHF 4,100,000 was approved as fixed compensation for the Executive Management for 2022.

For the period from January 1, 2023 to December 31, 2023, the Board of Directors proposes a fixed compensation for the Executive Management of CHF 2,950,000 (incl. employer contributions to social security and pension plans). This amount would be used for the currently six members of the Executive Management. The amount also includes a reserve that would allow the Board of Directors to increase the fixed compensation of the currently six members of the Executive Management, if necessary, which has not changed since 2018, or since joining the Company, if later.

(b) Variable Compensation of the Members of the Executive Management for 2021 (Cash Gratification)

Santhera's variable compensation consists of the compensation elements of the short-term incentive plan (STI) in cash and the share-based long-term incentive plan (LTI).

The cash gratification is based on the achievement of corporate and individual objectives and the financial situation of the Company. Corporate objectives included the successful completion of a financing, achieving milestones to pursue the development of vamorolone as well as to prepare partnering of vamorolone in China.

Overall, Company targets were substantially achieved. The Company was successfully turned around in 2021 following the futility of the Phase 3 SIDEROS study with Puldysa (idebenone) in 2020 and the subsequent restructuring. The existing debt of CHF 60 million was successfully restructured. Following the full in-licensing of the rights for vamorolone, the development of vamorolone was pursued to enable the start of the filing process of vamorolone in Duchenne Muscular Dystrophy (DMD) in the US and the EU in 2022 and the subsequent commercialization in 2023.

The proposal to the shareholders at the AGM 2022 is for a maximum cash bonus payment of CHF 1,200,000 (incl. social security contributions) after two consecutive years of no cash bonus payment (for the year 2019 and for the year 2020).

(c) Variable Compensation of the Members of the Executive Management for 2022

The Company has discontinued its Share Appreciation Rights (SAR) program and replaced it with a time and performance dependent equity-based plan, a combination of options and Performance Share Units (PSU) from the year 2021 onwards.

The objective of the variable long-term remuneration is to align the executive management's long-term compensation with the strategy of Santhera. The Long-Term Incentive (LTI) program is designed to motivate eligible managers to ensure that their actions and decisions promote the achievement of the mediumand long-term value-based targets. Santhera seeks to align the interests of management and the Group with the interests of its shareholders beyond share price appreciation. In addition, the LTI program aims to strengthen the loyalty of its managers to Santhera, identification with the company and motivation among its key talents to stay with the Company.

Based on article 25 para. 2 of the Company's Articles of Incorporation, the Board proposes a variable compensation for the financial year 2022 for the Executive Management in the maximum total amount of CHF 2,400,000. The Company plans to issue stock options and PSUs to the currently six members of the Executive Management as an annual grant under the LTI program with an aggregate value of up CHF

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2,400,000. The Company plans to propose additional variable compensation for the financial year 2022 for the Executive Management to the 2023 AGM.

11. Re-election of the Statutory Auditors

Motion

The Board of Directors proposes the re-election of Ernst & Young AG, Basel, as auditors for another one-year term until the end of the 2023 AGM.

Explanations

According to Art. 22 para. 2 of the Articles of Incorporation, the AGM shall elect the auditors for a term until the next AGM.

12. Re-election of the Independent Proxy

Motion

The Board of Directors proposes the re-election of Dr. Balthasar Settelen, Basel, as independent proxy until the end of the 2023 AGM.

Explanations

According to Article 13a of the Articles of Incorporation, the independent proxy is elected annually for a term until the end of the next AGM.

Pratteln, June 9, 2022 For the Board of Directors

Elmar Schnee Chairman

Organizational Remarks

Documents

The 2021 Annual Report can be downloaded from www.santhera.com/financial-reports and is available for inspection until the day of the AGM at the Company's headquarters at Hohenrainstrasse 24, 4133 Pratteln. Shareholders who wish to receive a printed copy of the Annual Report (in English) are kindly requested to tick the corresponding box on the reply form.

Voting Right and Share Register Closure

Shareholders registered in the share register with the right to vote as of June 22, 2022, at 5:00 p.m. CEST, will receive the proxy form and will be able to vote by these means. The Company's share register will close on June 22, 2022, at 5:00 p.m. CEST, and will be reopened on July 1, 2022, at 7:00 a.m. CEST.

Granting of Power of Attorney to the Independent Proxy

All shareholders can exercise their rights at the AGM solely via the independent proxy, **Dr. Balthasar Settelen**, Advokat, Centralbahnstrasse 7, P.O. Box 206, 4010 Basel, Switzerland. There is no possibility to attend the AGM in person or to grant a power of attorney to another shareholder with voting rights.

Shareholders may grant powers of attorney and give instructions to the independent proxy as follows:

- by filling out and sending back the power of attorney on the proxy form; or
- electronically https://santhera.netvote.ch. The required login information will be sent to shareholders together with the written documents for the Annual General Meeting. Changes to the electronically transferred instructions can be made until June 28, 2022, 11:59 p.m. (CEST) at the latest.

Unless other instructions are given to the independent proxy, the independent proxy, in accordance with the proxy form or the electronic instruction form, is appointed to vote for the proposals made by the Board of Directors.

Invitation to the AGM via E-mail

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

Venue

The Annual General Meeting takes place at the headquarters of Santhera. There is no possibility to attend the AGM in person.

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Proposed Amendment to the Articles of Incorporation according to Agenda Item 5 (convenience translation)

Article 3a (current)	Article 3a (proposed)
Authorized Share Capital	Authorized Share Capital
The Board of Directors shall be authorized, including	The Board of Directors shall be authorized, including
in connection with an intended takeover, at any time	in connection with an intended takeover, at any time
until December 14, 2023, to increase the share	until June 29, 2024, to increase the share capital in
capital in an amount not to exceed	an amount not to exceed CHF 36,860,687.00
CHF 34,203,905.00 through the issuance of up to	through the issuance of up to 36,860,687 fully paid
34,203,905 fully paid registered shares with a	registered shares with a nominal value of CHF 1.00
nominal value of CHF 1.00 each.	each.
[]	[]