

English convenience translation of the authoritative German version.

## **Invitation to the Annual General Meeting of Shareholders**

Thursday, April 12, 2018, 10:30 a.m.

Congress Center Basel, Hall Singapore (2nd floor), Messeplatz 21, 4058 Basel, Switzerland

### **Agenda (Overview)**

- 1. Approval of the Annual Financial Statements and the Consolidated Financial Statements 2017**
- 2. Appropriation of the Annual Results and Allocation of Reserves**
- 3. Consultative Vote on the 2017 Compensation Report**
- 4. Discharge of the Members of the Board of Directors and of the Executive Management from Liability for the Financial Year 2017**
- 5. Amendments to the Articles of Incorporation with respect to the Authorized Capital**
- 6. Amendment to the Articles of Incorporation regarding Relocation of Seat**
- 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 Auditors**
- 12. Re-election of the Independent Proxy**

## Agenda, Motions and Explanations

### 1. Approval of the Annual Financial Statements and the Consolidated Financial Statements 2017

#### Motion

The Board of Directors (**Board**) proposes to approve the Annual Financial Statements and the Consolidated Financial Statements for 2017.

### 2. Appropriation of the Annual Results and Allocation of Reserves

#### Motions

##### (a) Loss Carryforward

The Board proposes to carry forward the annual loss for 2017 of CHF 7,300,395 to the new account.

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

The Board proposes to allocate an amount of CHF 7,000,000 from the reserves from capital contributions to the free reserves.

#### Explanations

##### (a) Loss Carryforward

*The net annual loss for 2017 amounted to CHF 7,300,395. The loss carryforward of the previous financial years amounts to CHF 6,451,188. After appropriation of the annual loss, the total loss carryforward will amount to CHF 13,751,583.*

In CHF	2017	2016
Loss carryforward of previous financial years	-6,451,188	-5,556,524
Annual loss	-7,300,395	-894,664
<b>Total loss carryforward</b>	<b>-13,751,583</b>	<b>-6,451,188</b>

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

*The Board proposes to re-allocate an amount of CHF 7,000,000 from the reserves from capital contributions that amount to CHF 7,449,947 to the free reserves. To the extent that the reserves from capital contributions could be used for tax-free distributions, this possibility would be eliminated by the allocation to the free reserves. As a result of such allocation, the free reserves would be increased from CHF 50,494,714 to CHF 57'494'714 and the reserves from capital contributions would be reduced to CHF 449,947.*

### 3. Consultative Vote on the 2017 Compensation Report

#### Motion

The Board proposes to approve the 2017 Compensation Report in a non-binding consultative vote.

### Explanations

*The 2017 Compensation Report covers the fundamentals of the compensation for the members of the Board and of the Executive Management as well as the compensation paid to them during the financial year 2017. In line with Article 25 of the Articles of Incorporation and the recommendations of the Swiss Code of Best Practice for Corporate Governance, the Board submits the 2017 Compensation Report to the annual general meeting of shareholders (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 2017

### Motions

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

The Board proposes to grant discharge to the members of the Board for their activities during the financial year 2017.

#### (b) Discharge of the Members of the Executive Management

The Board proposes to grant discharge to the members of the Executive Management for their activities during the financial year 2017.

## 5. Amendments to the Articles of Incorporation with respect to the Authorized Capital

### Motion

The Board proposes to increase the authorized capital from CHF 1,261,076 to CHF 1,500,000 and to extend the period during which this authorization is valid until April 11, 2020, by an amendment to Article 3a of the Articles of Incorporation as per the separate enclosure.

### Explanations

*After parts of the authorized capital have been used successfully for a licensing transaction, the Board proposes to increase the former to the previous amount of CHF 1,500,000 (23.0% of the share capital) and to renew it for another two-year period, which is the maximum that the Swiss Code of Obligations allows. Without an extension of its term, the authorization to increase the Company's capital would expire as of May 10, 2018.*

## 6. Amendment to the Articles of Incorporation regarding Relocation of Seat

### Motion

The Board proposes to move the seat of the Company to Pratteln, Canton Basel-Landschaft by an amendment to Article 1 of the Articles of Incorporation as per the separate enclosure.

### Explanations

*According to the articles of incorporation, the seat of the Company is in Liestal, Canton Basel-Land. Due to an increase in employees, the Company moved to new offices in Pratteln in February 2018. This move makes it advisable to also relocate the seat of the Company to the place of the Company's headquarters in Pratteln.*

## 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 as Member of the Board

The Board proposes to re-elect Elmar Schnee as a member of the Board until the end of the 2019 AGM.

#### (b) Re-election of Martin Gertsch as Member of the Board

The Board proposes to re-elect Martin Gertsch as a member of the Board until the end of the 2019 AGM.

#### (c) Re-election of Philipp Gutzwiller as Member of the Board

The Board proposes to re-elect Philipp Gutzwiller as a member of the Board until the end of the 2019 AGM.

#### (d) Re-election of Thomas Meier as Member of the Board

The Board proposes to re-elect Thomas Meier as a member of the Board until the end of the 2019 AGM.

#### (e) Re-election of Patrick Vink as Member of the Board

The Board proposes to re-elect Patrick Vink as a member of the Board until the end of the 2019 AGM.

#### (f) Re-election of Elmar Schnee as Chairman of the Board

The Board proposes to re-elect Elmar Schnee as chairman of the Board until the end of the 2019 AGM.

### Explanations

*The members and the chairman of the Board have to be elected individually. The terms of office of all members of the Board expire at the end of this year's AGM. All members of the Board are standing for re-election. In addition, Elmar Schnee stands for re-election as chairman of the Board.*

***Elmar Schnee** has been a member and the chairman of the Board since 2017. He is also member of the Compensation Committee. Elmar Schnee is advisor to management of MindMaze, a neuro-technology company spun off from the Swiss Federal Institute of Technology in Lausanne (EPFL). Prior to that, he was Chairman, CEO and board member of Cardiorientis in Zug, Switzerland. Previously, he was a General Partner and member of the Executive Board of Merck KGaA, responsible for its worldwide pharmaceutical business. He also led the major restructuring of the business including the acquisition and integration of Serono. Prior to Merck, Elmar Schnee held senior roles as managing director and in marketing, licensing, strategy and business development with UCB Pharma, Sanofi-Synthelabo, Migliara Kaplan and Fisons. He currently serves on the board of directors of listed Jazz Pharmaceuticals and Stallergenes Greer as well as of several privately held life science companies.*

***Martin Gertsch** became a member of the Board in 2006 and has been its vice-chairman since 2017. He is also chairman of the Audit Committee. Up to January 2014, he served as Chief Financial Officer of Acino Holding. Prior to this he was Vice President Head of Finance EMEA at Synthes and held Chief Financial Officer and Chief Operating Officer positions at Delenex Therapeutics and ESBATech, two privately held biotech companies. From 2002 to the beginning of 2006, he was Chief Financial Officer of Straumann, which he had joined in 1997 as Head of Group Controlling and Reporting. Between*

1986 and 1997, Martin was an Audit Engagement Manager at PricewaterhouseCoopers, Basel, Switzerland. Martin is a Swiss-certified fiduciary and Swiss-certified public accountant. He has also completed several executive-level development programs at IMD (the International Institute for Management Development) in Lausanne, Switzerland. He serves as a Board Member of Evolva Holding, and the University Center of Dentistry, Basel (UZB). He is also Chairman of two diagnostic start-up companies.

**Philipp Gutzwiller** has been a member of the Board since 2017. He is also a member of the Audit Committee. Philipp Gutzwiller is Global Head Healthcare at Lloyds Banking Group plc in London. He has accumulated over 15 years of experience as a banker to the broader healthcare industry, advising corporate and private equity clients on the assessment, financing and execution of acquisitions and capital market transactions. He started his career at Roche as a finance controller and later worked as an executive in Roche's corporate mergers and acquisitions team.

**Thomas Meier**, PhD, has been a member of the Board since 2017. He was appointed Chief Executive Officer of Santhera in October 2011, having served for 7 years as Chief Scientific Officer for the Company. Thomas Meier is the founder and was CEO of MyoContract, a Basel-based research company focused on orphan neuromuscular diseases. In 2004, Thomas Meier played a key role in the merger with Graffinity Pharmaceuticals (Heidelberg, Germany) forming today's Santhera. Thomas Meier received his PhD in biology from the University of Basel in 1992 and subsequently joined the University of Colorado Health Sciences Center, Denver, CO, USA. He has a distinguished scientific track record and has written several important publications in the field of neuromuscular research.

**Patrick Vink**, MD, has been a member of the Board since 2017 and is chairman of the Compensation Committee. He has over 25 years of life science industry experience. In his latest assignment, he was employed as Chief Operating Officer at Cubist Pharmaceuticals, overseeing all worldwide commercial and technical operations as well as global alliance management. Previously, Patrick Vink held several senior management positions with Mylan Inc., Novartis Generics / Sandoz, Biogen and Sanofi-Synthelabo. He currently is chairman of privately held NMP Pharma and Acacia Pharma and a member of the board of directors of listed Concordia International Corp. and several privately held life science companies.

## 8. Re-election of the Members of the Compensation Committee

### Motions

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

The Board proposes to re-elect Elmar Schnee as a member of the Compensation Committee until the end of the 2019 AGM.

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

The Board proposes to re-elect Patrick Vink as a member of the Compensation Committee until the end of the 2019 AGM.

### Explanations

*The members of the Compensation Committee will have to be elected individually. Only members of the Board 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 proposes to approve a maximum total amount of fixed compensation of the Board of CHF 1,001,000 until the 2019 AGM.

### Explanations

*The proposed maximum total fixed compensation of the Board is unchanged compared to the previous year. It is expected to consist of:*

- *a fixed annual cash fee in the maximum amount of CHF 500,500, including employer's social security contributions; and*
- *a grant of Share Appreciation Rights (SAR) with a maximum market value of CHF 500,500, including employer's social security contributions. The number of SAR to be granted to the members of the Board will be calculated on the first trading day at grant on the basis of this amount, applying the Hull-White model.*

*Including social security contributions, the compensation of the chairman of the Board is expected to amount to CHF 286,000, the compensation of the vice chairman of the Board to CHF 242,000 and the compensation of the other members of the Board to CHF 198,000 each. The CEO of the Company, Thomas Meier, will not receive additional compensation for his Board membership. The chairman of the Audit Committee is expected to receive an additional amount of CHF 33,000; and the chairman of the Compensation Committee an additional CHF 22,000. The members of the Audit Committee and of the Compensation Committee are expected to receive an additional amount of CHF 11,000 each.*

*The table below sets out the approved maximum compensation for the Board for the period from the 2017 AGM to the 2018 AGM, the estimated actual compensation for the same period and the proposed maximum compensation for the period from the 2018 AGM to the 2019 AGM. The expected total compensation and grants for the period of the 2017 AGM to the 2018 AGM amounts to CHF 937,871. This amount is lower than the maximum amount approved by the 2017 AGM of CHF 1,001,000.*

	<b>Approved April 5, 2017 – April 12, 2018 (AGM)</b>	<b>Remunerated (estimation) April 5, 2017 – April 12, 2018 (AGM)</b>	<b>Proposal for April 13, 2018 – 2019 AGM</b>
Fixed compensation (CHF)	500,500	474,425	500,500
Share Appreciation Rights (CHF)	500,500	463,446	500,500
<b>Total (CHF)</b>	<b>1,001,000</b>	<b>937,871</b>	<b>1,001,000</b>

All numbers include employer's social security contributions

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

### Motions

#### **(a) Fixed Compensation of the Members of the Executive Management for 2019**

The Board proposes to approve a maximum total amount of fixed compensation of the members of the Executive Management for the period from January 1, 2019 to December 31, 2019 of CHF 3,200,000.

#### **(b) Variable Compensation of the Members of the Executive Management for 2017**

The Board proposes to approve a maximum total amount of variable compensation of the members of the Executive Management for the period from January 1, 2017 to December 31, 2017 of CHF 2,150,000.

#### **(c) Conditional special Compensation of the Members of the Executive Management**

The Board proposes to approve a maximum total amount of conditional special compensation of the members of the Executive Management of CHF 515,000.

### Explanations

*According to Art. 25 of the Articles of Incorporation, the maximum total amount of fixed compensation for the Executive Management is to be approved for the following year, i.e. for the period from January 1 to December 31, 2019. In addition, the Board has to submit the variable compensation of the Executive Management of the preceding year, i.e. the period from January 1 to December 31, 2017, for approval. According to Art. 25 of the Articles of Incorporation, the Board may make proposals that deviate from the aforementioned proposals or that are in addition thereto or conditional. The Board makes use of this possibility in agenda item 10(c).*

#### **(a) Fixed Compensation of the Members of the Executive Management for 2019**

*The fixed compensation for the Executive Management includes the base salary, the employer's social security contributions and contributions to the Company's pension fund. The estimated social security and pension fund contributions amount to approximately 28% of the base salary in aggregate.*

*At the 2016 AGM, the shareholders approved an amount of CHF 2,600,000 as fixed compensation for the Executive Management for 2017. The actual compensation of the members of the Executive Management who already worked for Santhera before January 1, 2017 amounted to CHF 2,555,344 in 2017. As of January 1, 2017, Kristina Sjöblom Nygren was appointed as a member of Executive Management. Her fixed compensation in 2017 amounted to CHF 883,282, comprising base salary, retention award, retention SAR, and social security payments. According to Art. 26 of the Articles of Incorporation ("Additional Amount"), the Company may pay compensation of up to 50% of the total compensation last approved by the AGM to joining or internally promoted members of the Executive Management in case the total amount already approved does not suffice. Therefore, for 2017 the additional amount was CHF 1,300,000. Out of this amount, CHF 838,626 were used for the fixed compensation of Kristina Sjöblom Nygren.*

*The maximum total amount of fixed compensation 2018 for the members of the Executive Management, amounting to CHF 3,200,000, was already approved by the 2017 AGM.*

*For the period between January 1, 2019 and December 31, 2019, the Board proposes an unchanged maximum total amount of fixed compensation for the Executive management of CHF 3,200,000.*

**(b) Variable Compensation of the Members of the Executive Management for 2017**

*The proposed maximum total amount of variable compensation of the members of the Executive Management for the period from January 1, 2017, to December 31, 2017, of CHF 2,150,000 (2016: CHF 2,400,000) corresponds to the variable compensation as disclosed in the compensation report for 2017 where additional details are available.*

*The total amount is based on an average corporate target achievement of 60% and individual goal achievements of the members of the Executive Management between 95% and 100%. The total amount mentioned consists of an amount of CHF 575,000 for the payment of a cash bonus and of an amount of CHF 1,575,000 for the allocation of 104,711 SAR.*

*The valuation of the allocation of 104,711 SAR to the members of the Executive Management which is subject to the approval of the shareholders, has already been made, applying the Hull-White model as per January 1, 2018. The SAR shall provide a positive incentive to ensure the long-term motivation and retention of Santhera's Executive Management.*

**(c) Conditional special Compensation of the Members of the Executive Management for 2018**

*The proposed maximum amount of CHF 515,000 shall be granted to members of Executive Management by way of an SAR grant. The vesting of SAR would only take place if Santhera obtains a positive opinion of the Committee for Medicinal Products for Human Use (CHMP) with respect to the marketing authorization of idebenone for the treatment of patients with Duchenne muscular dystrophy in the EU. The later the date of this opinion is, the more of these SAR lapse: 33⅓% on January 1, 2019, and each calendar quarter thereafter 8⅓%. This shall be an incentive to obtain a positive CHMP opinion as soon as possible.*

## **11. Re-election of Auditors**

**Motion**

The Board proposes to re-elect Ernst & Young AG, Basel, as auditors for another one-year term until the end of the 2019 AGM.

**Explanations**

*According to Article 22 para. 2 of the Articles of Incorporation, the general meeting of shareholders shall elect the auditors for a term until the next AGM.*

## **12. Re-election of the Independent Proxy**

**Motion**

The Board proposes to re-elect Dr. Balthasar Settelen, Basel, as independent proxy until the end of the 2019 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, March 20, 2018  
For the Board of Directors

Elmar Schnee  
Chairman

## Organizational Remarks

### Documents

The 2017 Annual Report (in English) can be downloaded from <http://www.santhera.com/investors-and-media/investor-toolbox/financial-reports> and is available for inspection at the Company's new headquarters at Hohenrainstrasse 24, 4133 Pratteln, Switzerland. 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 registered in the share register with the right to vote as of **March 19**, 2018, at 5:00 p.m. CET, will receive the invitation to the AGM and – upon request – the admission card and the voting materials. Shareholders selling their shares before the AGM will not be entitled to participate in the AGM.

### Book closure period

The Company's share register will close on **April 4**, 2018, at 5:00 p.m. CEST, and will reopen on **April 13**, 2017, at 7:00 a.m. CEST.

### 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 Board.

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

Shareholders may participate in the votes and elections by giving instructions to the independent proxy electronically via [www.netvote.ch/santhera](http://www.netvote.ch/santhera). 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 Saturday, **April 9**, 2018, 11:59 am (CEST).

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](http://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](http://www.netvote.ch/santhera) at any time.

#### Venue

The Congress Center Basel, Hall Singapore (2nd floor), Messeplatz 21, 4058 Basel, Switzerland, is located about 15 minutes from Basel SBB railway station and about a half an hour from Basel Airport (EuroAirport).

#### Admission

On the day of the meeting, the admission desk will be open from 10:00 a.m. CEST. The language of the meeting will be German.

#### Proposed Amendments to the Articles of Incorporation (convenience translation from the German original)

<b>Article 1 (current)</b>	<b>Article 1 (proposed change)</b>
<p><b>Corporate Name, Seat, Duration</b></p> <p>[...]</p> <p>The seat of the Company is in Liestal, Canton Basel-Land.</p> <p>[...]</p>	<p><b>Corporate Name, Seat, Duration</b></p> <p>[...]</p> <p>The seat of the Company is in Pratteln, Canton Basel-Landschaft.</p> <p>[...]</p>

<b>Article 3a (current)</b>	<b>Article 3a (proposed changes)</b>
<p><b>Authorized Share Capital</b></p> <p>The Board of Directors shall be authorized, at any time until May 18, 2018, to increase the share capital in a maximum amount of CHF 1,261,076.-- through the issuance of up to 1,261,076 fully paid registered shares with a nominal value of CHF 1 each.</p> <p>[...]</p>	<p><b>Authorized Share Capital</b></p> <p>The Board of Directors shall be authorized, at any time until April 11, 2020, to increase the share capital in a maximum amount of CHF 1,500,000.-- through the issuance of up to 1,500,000 fully paid registered shares with a nominal value of CHF 1 each.</p> <p>[...]</p>