IMPORTANT NOTICE: You must read the following before continuing.

THE PROSPECTUS ATTACHED TO THIS DISCLAIMER IS AVAILABLE ONLY TO INVESTORS WHO ARE EITHER (A) "QUALIFIED INVESTORS" (AS DEFINED IN THE PROSPECTUS REGULATION, (REGULATION (EU) 2017/1129) ("**EEA QUALIFIED INVESTORS**") IN THE EUROPEAN ECONOMIC AREA (THE "**EEA**"), OR (B) "QUALIFIED INVESTORS" (AS DEFINED IN REGULATION (EU) 2017/1129 AS IT FORMS PART OF DOMESTIC LAW BY VIRTUE OF THE EUROPEAN UNION (WITHDRAWAL) ACT 2018 ("**UK QUALIFIED INVESTORS**"), OR (C) OUTSIDE THE EEA AS PERMITTED UNDER APPLICABLE SECURITIES LAWS.

IMPORTANT: You must read the following before continuing. The following disclaimer applies to the attached prospectus (the "**Prospectus**"), which has been accessed via internet or otherwise received and you are therefore advised to read this disclaimer carefully before reading, accessing or making any other use of the Offering Document. In accessing the Offering Document, you agree to be bound by the following terms and conditions, including any modifications to them from time to time, any time you receive any information, as the case may be, as a result of such access. Capitalized terms used but not defined in this notice have the meanings ascribed to such terms in the Offering Document.

NOTHING IN THIS ELECTRONIC TRANSMISSION CONSTITUTES AN OFFER OF SECURITIES FOR SALE IN ANY JURISDICTION WHERE IT IS UNLAWFUL TO DO SO. NO ACTION HAS BEEN OR WILL BE TAKEN THAT WOULD, OR IS INTENDED TO, PERMIT A PUBLIC OFFERING OF THE SECURITIES DESCRIBED IN THE PROSPECTUS IN ANY JURISDICTION OTHER THAN SWITZERLAND.

The Prospectus is being provided to you on a confidential basis for informational use solely in connection with your consideration of the purchase of the securities referred to therein. Its use for any other purpose is not authorized, and you may not, nor are you authorized to, copy or reproduce the Prospectus in whole or in part in any manner whatsoever or deliver, distribute or forward the Prospectus or disclose any of its contents to any other person. Failure to comply with this directive may result in a violation of the applicable laws of your or other jurisdictions. If you are not the intended recipient of the Offering Document, you are hereby notified that any dissemination, distribution or copying of the Prospectus is strictly prohibited.

Confirmation of your representation: In order to be eligible to view the Prospectus or make an investment decision with respect to the securities described therein, potential investors must be either (a) EEA Qualified Investors or (b) UK Qualified Investors or (c) outside the EEA. You have been sent this disclaimer with the attached Prospectus on the basis that you have confirmed to the relevant parties that you and any customers that you represent are in compliance with this disclaimer and the applicable laws.

You are reminded that the Prospectus has been delivered to you on the basis that you are a person into whose possession the Prospectus may be lawfully delivered in accordance with the laws of the jurisdiction in which you are located and you may not, nor are you authorized to, deliver the Prospectus to any other person.

This disclaimer does not constitute or contain any offer to sell or invitation to subscribe or make commitments for or in respect of any security in any jurisdiction where such an offer or invitation would be unlawful. There are restrictions on the distribution of this disclaimer and the Prospectus and/or the offer or sale of the securities described in the Prospectus in certain jurisdictions. If a jurisdiction requires that the offering be made by a licensed broker or dealer and a bank involved in the offering or any affiliate of such bank is a licensed broker or dealer in that jurisdiction, the offering shall be deemed to be made by such bank or any affiliate of such bank on behalf of the Issuer in such jurisdiction.

The Prospectus has been sent to you in electronic form. You are reminded that documents transmitted via this medium may be altered or changed during the process of electronic transmission and, consequently, none of the involved parties in the offering, or their respective affiliates, directors, officers, employees or agents accepts any liability or responsibility whatsoever in respect of any difference between the Prospectus distributed to you in electronic format and any hard copy version that may have been delivered to you by third parties.

You are responsible for protecting against viruses and other destructive items. Your receipt of the Prospectus via electronic transmission is at your own risk and it is your responsibility to take precautions to ensure that it is free from viruses and other items of a destructive nature.

Santhera Pharmaceuticals Holding AG
(incorporated in Pratteln, Switzerland, with limited liability)

CHF 30,270,375 Senior Unsecured Convertible Bonds due 2024

This prospectus (the "**Prospectus**") relates to the of 7.50 % bonds in the aggregate principal amount of Swiss francs ("**CHF**") 30,270,375 due 2024 (each and collectively the "**Bonds**") of Santhera Pharmaceuticals Holding AG (the "**Issuer**" or the "**Company**"), convertible into registered shares with a nominal value of CHF 1.00 each of the Issuer (the "**Shares**", and each a "**Share**"), and the listing of the Bonds on the SIX Swiss Exchange.

Exchange Offer	On March 25, 2021, the Company announced an exchange offer (the "**Exchange Offer**") in respect of the outstanding CHF 60 million Convertible Bonds due 2022 (the "**2017/22 Bonds**"). The press release and the "notice of a repurchase offer" containing the details of the Exchange Offer are incorporated in this Prospectus by reference. The holders of the 2017/22 Bonds who have accepted the Exchange Offer will, for each of their 2017/22 Bond, receive one Bond and 26 Shares on the date the Exchange Offer is settled (the "**Exchange Date**"). The entire issuance of the Bonds is used for the settlement of the Exchange Offer.
Principal Amount/ Denomination	CHF 3,375 per Bond.
Issue Price	100% of the Principal Amount.
Interest Rate	7.50% per annum, payable semi-annually in arrears on February 17 and August 17, for the first time on August 17, 2021. The first interest payment on August 17, 2021 shall be CHF 126.5625 per Bond, as of the Bonds had been issued on February 17, 2021.
Issue Date	May 4, 2021.
Maturity Date	August 17, 2024.
Redemption Price	100% of the Principal Amount.
Conversion Price	CHF 3.0029.
Conversion Ratio	1,123.91243 Shares per Bond.
Status	Senior, unsecured.
Form of the Bonds	The Bonds will be issued as uncertificated securities (*Wertrechte*) in accordance with article 973c of the Swiss Code of Obligations (the "**CO**") and registered as intermediated securities (*Bucheffekten*) in the main register (*Hauptregister*) with SIX SIS Ltd. Neither the Bondholders (as defined in the Terms of the Bonds) nor any other parties will have the right to request printing and physical delivery of individually certificated Bonds.
Early Redemption at the option of the Issuer	The Issuer may, by giving not less than 30 and not more than 60 calendar days' prior notice, redeem all but not only some of the outstanding Bonds at their Principal Amount, together with unpaid accrued interest, if any, at any time after the second anniversary of the Payment Date, if the volume weighted average price of a Share on each of at least 20 out of 30 consecutive Trading Days ending not earlier than 5 Trading Days prior to the giving of notice of redemption is at least 150% of the prevailing Conversion Price.
Clean-up Call	The Issuer may, by giving not less than 30 and not more than 60 calendar days' prior notice, redeem all but not only some of the outstanding Bonds at their Principal Amount, together with unpaid accrued interest, if any, at any time after the Payment Date and prior to the Maturity Date if less than 15% of the aggregate Principal Amount of the Bonds originally issued is outstanding.
Assurances	Negative pledge clause (with restrictions), *pari passu* clause, cross default clause (subject to CHF 3 million threshold), events of default clause, anti-dilution provision, all as provided in the Terms of the Bonds.
Trading	The Bonds have been provisionally admitted to trading on the SIX Swiss Exchange as of May 7, 2021. The last trading day is expected to be August 15, 2024.
Listing	Listing of the Bonds will be applied for at the SIX Swiss Exchange. The Shares are listed on the SIX Swiss Exchange in accordance with the International Reporting Standard.
Selling Restrictions	**United States of America, U.S. persons, European Economic Area, United Kingdom**, in particular. See Selling Restrictions of this Prospectus.
Governing Law and Jurisdiction	Swiss law; place of jurisdiction is the city of Zurich, Switzerland.

	Bonds	**Shares**
Swiss Security Number:	56334874	2714864
ISIN:	CH0563348744	CH0027148649
Ticker symbol:	SAN21	SANN

This Prospectus has been approved by SIX Exchange Regulation Ltd in its capacity as review body pursuant to article 52 of the Swiss Financial Services Act on June 23, 2021.

I. IMPORTANT INFORMATION

This Prospectus will not be updated for any developments that occur after its date. In particular, this Prospectus is not required to be updated as of the date of the approval by SIX Exchange Regulation Ltd in its capacity as Swiss Review Body. Consequently, neither the delivery of this Prospectus nor the offering, sale or delivery of any Bonds shall in any circumstances imply that the information contained herein concerning the Issuer is correct at any time subsequent to the date hereof or that any other information supplied in connection with the issue of the Bonds is correct as of any time subsequent the date indicated in the document containing the same.

This Prospectus has been prepared by the Issuer solely for use in connection with the offering of the Bonds and for the admission to trading and listing of the Bonds on the SIX Swiss Exchange. The Issuer has not authorized the use of this Prospectus for any other purpose.

This Prospectus is to be read in conjunction with all documents incorporated by reference herein. This Prospectus shall be read and construed on the basis that such documents are incorporated into and form part of this Prospectus. See "*ABOUT THIS PROSPECTUS—Documents incorporated by reference*" on page 6 of this Prospectus.

An investment in the Bonds will involve certain risks, including the risk that Bondholders will lose their entire investment in the Bonds. For a discussion of certain risks that potential investors should carefully consider before deciding to invest in any Bonds, see "*RISK FACTORS*" beginning on page 11 of this Prospectus.

II. SELLING RESTRICTIONS

A. United States of America / U.S. persons

(a) The offering of the Bonds is being made in the United States in reliance on, and compliance with, Section 14(e) of the US Securities Exchange Act of 1934 and Regulation 14E thereunder.

(b) The Company, certain affiliated companies and the nominees or brokers (acting as agents) may make certain purchases of, or arrangements to purchase outside the Exchange Offer during the period in which the Exchange Offer remains open for acceptance the 2017/22 Bonds. If such purchases or arrangements to purchase are made they will be made outside the United States and will comply with applicable law, including the Exchange Act.

(c) The Company as the offeror is a Swiss company. Information distributed in connection with the offering of the Bonds is subject to Swiss disclosure requirements that are different from those of the United States. Financial statements and financial information included herein are prepared in accordance with Swiss accounting standards that may not be comparable to the financial statements or financial information of United States companies.

(d) It may be difficult for you to enforce your rights and any claim you may have arising under the U.S. federal securities laws in respect of the offering of the Bonds, since the Company is located in Switzerland and all of its officers and directors are residents of Switzerland or elsewhere outside of the United States. You may not be able to sue the Company or its officers or directors in a Swiss court or another court outside the United States for violations of the U.S. securities laws. Finally, it may be difficult to compel the Company and its affiliates to subject themselves to a U.S. court's judgment.

B. Prohibition of sales to EEA Retail Investors

The Bonds are not intended to be offered, sold or otherwise made available and should not be offered, sold or otherwise made available, and will not offer, sell or otherwise make available to any retail investor in the European Economic Area. For the purposes of this provision the expression "**retail investor**" means a person who is one (or more) of the following:

(a) a retail client as defined in point (11) of Article 4(1) of Directive 2014/65/EU (as amended, "**MiFID II**");

(b) a customer within the meaning of Directive (EU) 2016/97 (the Insurance Distribution Directive or "**IDD**"), where that customer would not qualify as a professional client as defined in point (10) of Article 4(1) of MiFID II; or

(c) not a qualified investor as defined in Regulation (EU) 2017/1129 (the "**Prospectus Regulation**").

C. Prohibition of Sales to UK Retail Investors

The Bonds are not intended to be offered, sold or otherwise made available and should not be offered, sold or otherwise made available to any retail investor in the United Kingdom (the "**UK**"). For these purposes, a retail investor means a person who is one (or more) of: (i) a retail client as defined in point (8) of Article 2 of Regulation (EU) No 2017/565 as it forms part of domestic law by virtue of the European Union (Withdrawal) Act 2018 (the "**EUWA**"), (ii) a customer within the meaning of the provisions of the United Kingdom Financial Services and Markets Act 2000, as amended (the "**FSMA**"), and any rules or regulations made under the FSMA to implement the IDD, where that customer would not qualify as a professional client as defined in point (8) of Article 2(1) of Regulation (EU) No 600/2014 as it forms part of domestic law by virtue of the EUWA, or (iii) not a qualified investor as defined in the Prospectus Regulation as it forms part of domestic law by virtue of the EUWA.

D. General

The Issuer does not represent that Bonds may at any time lawfully be sold in compliance with any applicable registration or other requirements in any jurisdiction, or pursuant to any exemption available thereunder, or assumes any responsibility for facilitating such sale. The distribution of this Prospectus and the offering of the Bonds in certain jurisdictions may be restricted by law. Persons into whose possession this Prospectus comes are required by the Issuer to inform themselves about and to observe any such restrictions. This Prospectus does not constitute, and may not be used for or in connection with, an offer or solicitation by anyone in any jurisdiction in which such offer or solicitation is not authorized or to any person to whom it is unlawful to make such offer or solicitation and no action is being taken in any jurisdiction that would permit a public offering of the Bonds or the distribution of this Prospectus in any jurisdiction where action for that purpose is required.

III. ABOUT THIS PROSPECTUS

A. Documents incorporated by reference

The following documents shall be deemed to be incorporated in, and form part of this Prospectus:

- Santhera Annual Report 2020
- Santhera Press Release of February 16, 2021 titled “Santhera Announces Corporate Update and Proposal to Strengthen Capital Structure”
- Notice of a Repurchase Offer (Exchange of CHF 60,000,000 Senior Unsecured Convertible Bonds due 2022), dated March 25, 2021
- Santhera Press Release of March 25, 2021 titled "Santhera launches Exchange Offer for its CHF 60 Million Convertible Bonds"
- Santhera Press Release of April 27, 2021 titled “Santhera Reports End Results on Convertible Bond Exchange Offer”
- Santhera Press Release of May 3, 2021 titled “Santhera Announces Settlement of Convertible Bond Exchange Offer on May 4, 2021”
- Santhera Press Release of May 4, 2021 titled “Santhera Announces Settlement of Exchange Offer and Issuance of New Senior Unsecured Convertible Bonds due 2024”
- The Issuer's articles of association, dated April 30, 2021

B. Availability of documents

Copies of this Prospectus as well as the documents incorporated by reference are available free of charge at Santhera Pharmaceuticals Holding AG, Hohenrainstrasse 24, 4133 Pratteln, Switzerland (telephone number: +41 61 906 89 50; email: daniela.glatz@santhera.com, during regular business hours.

The documents incorporated by reference are also available free of charge at the Issuer’s website at http://www.santhera.com/investors-and-media.

IV. FORWARD-LOOKING STATEMENTS

This Prospectus contains certain forward-looking statements. A forward-looking statement is a statement that does not relate to historical facts and events. Forward-looking statements are based on analyses or forecasts of future results and estimates of amounts not yet determinable or foreseeable. These forward-looking statements are identified by the use of terms and phrases such as “anticipate”, “believe”, “could”, “estimate”, “expect”, “intend”, “may”, “plan”, “predict”, “project”, “will” and similar terms and phrases, including references and assumptions. This applies, in particular, to statements in this Prospectus containing information on future earnings capacity, plans and expectations regarding the Issuer’s business and management, its growth and profitability and general economic and regulatory conditions and other factors that affect the Issuer.

Forward-looking statements in this Prospectus are based on current estimates and assumptions that the Issuer makes to the best of its present knowledge. These forward-looking statements are subject to risks, uncertainties and other factors that could cause actual results, including the Issuer’s financial condition and results of operations, to differ materially from and be worse than results that have expressly or implicitly been assumed or described in these forward-looking statements. The Issuer’s business is also subject to a number of risks and uncertainties that could cause a forward-looking statement in this Prospectus to become inaccurate. Accordingly, investors are strongly advised to read the following sections of this Prospectus: “*RISK FACTORS*” and “*THE COMPANY AND ITS BUSINESS*”. These sections include more detailed descriptions of factors that might have an impact on the Issuer's business and the markets in which it operates.

In addition, the Issuer does not assume any obligation to update any forward-looking statement or to conform any forward-looking statement to actual events or developments.

V. TABLE OF CONTENTS

VI. SUMMARY

This summary should be read as an introduction to this Prospectus. Any decision to invest in the Bonds should be based on a consideration of this Prospectus as a whole, including any documents incorporated by reference into this Prospectus. Potential investors in the Bonds should be aware that liability under article 69 FinSA for any false or misleading information contained in this summary is limited to any such information that is false or misleading when read together with, or that is inconsistent with, the other parts of this Prospectus.

A. Information on the Issuer

Issuer:	The Issuer, Santhera Pharmaceutical Holding AG, is a stock corporation (*Aktiengesellschaft*), incorporated under the laws of Switzerland and registered since July 16, 2002, in the commercial register of the Canton of Basel-Landschaft (register number CHE-105.388.338). The domicile of the Issuer is Pratteln and its registered office is at Hohenrainstrasse 24, 4133 Pratteln, Switzerland.
Independent Auditors:	The auditors are Ernst & Young AG, Aeschengraben 27, 4051 Basel, Switzerland, who have acted as the Issuer's auditors for more than a decade. Ernst & Young AG is supervised and regulated by the Federal Audit Oversight Authority.

B. Information on the Terms of the Bonds

Issue Date:	May 4, 2021
Maturity Date:	August 17, 2024
Redemption Price:	100% of the Principal Amount.
Interest Rate:	7.50% per annum, payable semi-annually in arrears on February 17 and August 17, for the first time on August 17, 2021. The first interest payment on August 17, 2021 shall be CHF 126.5625 per Bond, as of the Bonds had been issued on February 17, 2021.
Denomination:	CHF 3,375
Status:	Senior, unsecured.
Form of the Bonds:	The Bonds will be issued as uncertificated securities (*Wertrechte*) in accordance with article 973c CO and registered as intermediated securities (*Bucheffekten*) in the main register (*Hauptregister*) with SIX SIS Ltd. Neither the Bondholders (as defined in the Terms of the Bonds) nor any other parties will have the right to request printing and physical delivery of individually certificated Bonds.
Principal Paying Agent:	Basler Kantonalbank
Governing Law and Jurisdiction:	Swiss law; place of jurisdiction is the city of Zurich, Switzerland.

C. Information on the Offering

Offering:	On March 25, 2021, the Company announced its Exchange Offer in respect of the 2017/22 Bonds. The press release and the "notice of a repurchase offer" containing the details of the Exchange Offer are incorporated in this Prospectus by reference. The holders of the 2017/22 Bonds who have accepted the Exchange Offer will, for each of their 2017/22 Bond, receive one Bond and 26 Shares on the Exchange Date. The entire issuance of the Bonds is used for the settlement of the Exchange Offer.
Issue Price:	100% of the Principal Amount.
Clearing and Settlement:	SIX SIS Ltd.

D. Information on the Admission to Trading and Listing

Swiss Trading Venue:	SIX Swiss Exchange
Trading:	The Bonds have been provisionally admitted to trading on SIX Swiss Exchange with effect from May 7, 2021. Application will be made for the Bonds to be listed on SIX Swiss Exchange. The last day of trading is expected to be August 15, 2024.
Swiss Security Number:	56334874
ISIN:	CH0563348744
Ticker symbol:	SAN21

E. Information on the Prospectus Approval

Swiss Review Body:	SIX Exchange Regulation Ltd, Hardturmstrasse 201, 8005 Zurich, Switzerland.
Prospectus Date and Approval:	This Prospectus is dated May 4, 2021, and has been approved by the Swiss Review Body on the date appearing on the cover page of this Prospectus. This Prospectus will not be updated for any developments that occur after its date. In particular, this Prospectus is not required to be updated as of the date of the approval by the Swiss Review Body.

VII. RISK FACTORS

Any investment in our securities involves a high degree of risk. Prospective investors should carefully consider the risks and uncertainties described below, together with all other information contained in this Prospectus, prior to making an investment decision.

The risks and uncertainties described below represent those we consider to be material as of the date of this Prospectus. However, these risks and uncertainties are not the only ones we are facing. Additional risks and uncertainties not presently known to us, or that we currently consider not to be significant, could also materially and adversely affect our business, results of operations, financial condition or prospects. If any or a combination of these risks actually occurs, our business, results of operations, financial condition and/or prospects could be materially and adversely affected. In such case or cases, prospective investors may lose all or part of their investment. This Prospectus contains forward-looking statements that involve risks and uncertainties. The actual results could differ materially from those anticipated in such forward-looking statements as a result of certain factors, including the risks we face that are described below or elsewhere in this Prospectus. The selected sequence of the risk factors mentioned below represents neither a statement about the probability of the risks' realization nor an assessment of the extent of the economic effects or the importance of the risks.

Investment decisions should not be made solely on the basis of the risk warnings set out in the Prospectus since such information cannot serve as a substitute for individual advice and information that is tailored to the requirements, objectives, experience, knowledge and circumstances of each prospective investor individually. Therefore, before entering into any transaction, each prospective investor should consult with its own legal, regulatory, tax, financial and accounting advisors to the extent it considers necessary in order to determine whether an investment in our securities is a fit, proper and suitable investment for it with a view to its financial situation, its constitutional documents, its internal policies and guidelines, the laws and regulations applicable to it and the impact an investment in our securities will have on its overall investment portfolio. Only prospective investors who are fully aware of the risks associated with an investment in our securities and who are financially able to bear any losses that may arise in connection therewith should consider engaging in any transactions in our securities.

Capitalized terms used but not defined herein have the meanings ascribed to them elsewhere in this Prospectus.

A. Risks related to our business and financial situation

1. Risks related to our financial position and capital needs

We will require additional capital to continue to fund our operations and to finance the further advancement of our product candidates. Without further funding, we currently expect to run out of cash in the third quarter of 2021. Our ability to meet our financial obligations, including under the Bonds, depends on our ability to obtain additional funding by way of equity and/or debt financings in the short term. If we default under any existing financing arrangement, we may have to file for bankruptcy or insolvency. There is a material uncertainty as to whether we will be able to continue as a going concern for another twelve months. This uncertainty is further aggravated by material uncertainties in connection with the development of our product candidates and other risks set out elsewhere in these risk factors.

As a research, development and commercialization company, our operations have consumed substantial amounts of cash since inception. We currently have one late-stage product candidate, vamorolone for the treatment of Duchenne muscular dystrophy ("**DMD**"), in development, the outcome of which is highly uncertain. Our operations have used substantial amounts of cash since our inception and we continue to require significant amounts of cash for operating our business and to satisfy our obligations. As at April 27, 2021, we had cash and cash equivalents of CHF 11.7 million (unaudited). Our current net cash outflow is approximately CHF 2.2 to 3.2 million per month. We currently expect to run out of cash in the third

quarter of 2021. We have based these estimates on assumptions that may change or prove to be inaccurate, and we could run out of cash sooner than we currently expect.

Our projected cash runway assumes that we will be able to draw further tranches in the aggregate principal amount of at least CHF 6 million under our financing arrangement with Highbridge Tactical Credit Master Fund L.P., Grand Cayman, Cayman Islands ("**Highbridge**") providing for the issuance of senior secured notes exchangeable at the option of Highbridge (the "**Highbridge Facility**", see "*THE COMPANY AND ITS BUSINESS—Additional information on our business—Material agreements—Financing Arrangements—Equity-linked financing arrangement with Highbridge*" beginning on page 81 for further information). If we are unable to draw down a sufficient amount of such tranches, we will run out of cash sooner than we currently expect (see also risk factor "*We may be unable to receive additional funds under the Highbridge Facility. If we do not receive such additional funds, we will face imminent illiquidity within weeks or a few months and we may have to further restructure, cease operations or file for bankruptcy or insolvency.*").

In addition to our monthly cash outflow, we anticipate significant additional cash outflows in the coming twelve months:

- When we acquired our license from ReveraGen BioPharma, Inc.'s ("**ReveraGen**") to vamarolone in September 2020, we issued as part of the consideration an interest-free exchangeable note in the principal amount of CHF 10 million (the "**Idorsia Exchangeable Note**") to the previous license holder, Idorsia Pharmaceuticals Ltd ("**Idorsia**"). The Idorsia Exchangeable Note will become due on September 1, 2021, or earlier if and when the U.S. Food and Drug Administration (the "**FDA**") supports a new drug application ("**NDA**") for vamorolone in DMD in the U.S. Upon the Idorsia Exchangeable Note becoming due, we will be required to pay Idorsia a cash amount of at least CHF 3.5 million, and up to CHF 10.0 million if and to the extent we are unable or choose not to make such payment in discounted Shares up to the agreed maximum of 65% of the principal value of the note.
- We will have to pay interest under the Bonds and under the remaining portion of our CHF 60 million Senior Unsecured Convertible Bonds due 2022 (ISIN CH0353955195; the "**2017/22 Bonds**", see "*THE COMPANY AND ITS BUSINESS—Additional information on our business—Material agreements—CHF 60 million Senior Unsecured Convertible Bonds due* 2022" beginning on page 81 for more information) of approximately CHF 1.5 million in August 2021 and again in February 2022. Under the Terms of the Bonds set out in Section VIII (but not under the terms and conditions of the 2017/22 Bonds), we will be able to pay interest in Shares, but we may not be able to do so if we do not have a sufficient amount of Shares available for such payment.
- The portion of the 2017/22 Bonds that will remain outstanding upon completion of the Exchange Offer, with an aggregate principal value of CHF 15.2 million, will become due for redemption on February 17, 2022.

Further, in connection with the offering of the Bonds and previous financings, certain financial advisors may have contractual rights to fees in the aggregate amount of several million Swiss francs.

We currently expect that we will need to be able to raise additional funds by way of equity and/or debt financing—in addition to the Highbridge Facility—in the short term in order to meet the cash requirements for operating our business and to meet our financial obligations, including under the Bonds, and to continue our operations as planned. If we are unable to meet our financial obligations and if we default under the Idorsia Exchangeable Note, under the 2017/22 Bonds, under the Bonds, under the Highbridge Facility or under any other contract, our obligations under these instruments and potentially under other contracts may become immediately due and payable. In this event, we would immediately become insolvent if no agreement with the respective creditor can be reached, and the Bond investment may be lost. For additional risks in connection with our ability to raise additional funds see the following risk factors.

Furthermore, our ability to raise additional funds highly depends on a successful development of our product candidates. For example, if the six-month top line data of our licensor ReveraGen's ongoing pivotal Phase IIb clinical trial of vamorolone in DMD (the "**VISION-DMD Trial**"), which we currently

expect for the second quarter of 2021, show that such trial does not meet its endpoints after six months of treatment, our ability to obtain further financing and to meet our obligations in connection with the Bonds would be materially impaired (see next risk factor).

In any case, there is a material uncertainty as to whether we will be able to continue as a going concern for another twelve months. Pursuant to Note 2 to our 2020 consolidated financial statements, a material uncertainty exists as to whether our current funding is sufficient to support its going concern for another twelve months. The inclusion of a going concern qualification or emphasis of matter paragraph in the audit opinion with respect to our 2020 consolidated financial statements, as well as any audit opinion related to our financial statements in the future, may materially adversely affect our trading and our ability to raise new capital that we need to fund our operations.

We will not receive any cash proceeds from the issuance of the Bonds.

The Bonds were offered as part of the consideration for the exchange of the 2017/22 Bonds in the Exchange Offer. Therefore, we will not receive any cash proceeds from the issuance of the Bonds. If we cannot secure and maintain cash through other sources to ensure our going concern and maintain a positive equity, we may have to cease our operations and be forced into bankruptcy or insolvency.

The six-month top line data of the VISION-DMD Trial, which we currently expect for the second quarter of 2021, might show that such trial does not meet its endpoints after six months of treatment. Should that be the case, and if we are unable to raise additional funds and meet our financial obligations, including under the Bonds, we will need to enter a further restructuring leading to a possible ceasing of operations and insolvency.

The six-month top line data of the VISION-DMD Trial, which we currently expect for the second quarter of 2021, might show that such trial does not meet its endpoints after six months of treatment. Given that the VISION-DMD Trial is a pivotal trial, we believe that it is important in view of obtaining marketing authorization in the U.S. and the EU that the trial (also) meets its secondary endpoints.

If the six-month top line data of the VISION-DMD Trial show that such trial does not meet its endpoints after six months of treatment, and if we are unable to raise additional funds to and meet the cash requirements for operating our business as well as our financial obligations, including without limitation under the Bonds, under the Idorsia Exchangeable Note and/or under the 2017/22 Bonds, we will need to enter a further restructuring leading to a possible ceasing of operations and bankruptcy or insolvency. Holders of the Bonds and of the 2017/22 Bonds may lose some or all of their investment.

If the six-month top line data of the VISION-DMD Trial show that such trial meets its endpoints after six months of treatment, we will require additional capital to continue to fund our operations and to finance the further advancement of our product candidates. We may not be able to raise additional funds on acceptable terms, at all or in time. If we fail to raise additional necessary funds, we may be unable to meet our financial obligations, including under the Bonds, and may be required to delay, limit or terminate development efforts, further restructure, cease operations or file for bankruptcy or insolvency. Additional funds may only be available on terms that significantly dilute the Company's shareholders (and make the Conversion Price unattractive) and/or restrict our flexibility to operate.

If the six-month top line data of the VISION-DMD Trial show that such trial meets its endpoints after six months of treatment (a "**Positive Interim Readout**"), we expect our operational expenses and operational cash outflow to increase significantly both in the short- and long term in connection with our ongoing development activities as well as the commencement of our pre-commercialization activities relating to vamorolone in DMD. Moreover, we will need to raise substantial additional funds in the coming twelve months to become able to redeem the 2017/22 Bonds and to meet our other financial obligations.

We may not be able to raise additional funds at all or within the short period of time in which we will need them, or on terms acceptable to us. Also, the 2017/22 Bonds and the Bonds prohibit us from issuing any secured marketable debt instruments or incurring any secured financial debt (including bank debt)

with an aggregate outstanding amount exceeding CHF 10 million at any point in time (subject to exceptions) unless the 2017/22 Bonds and the Bonds, respectively, are secured equally and rateably, or the Paying and Conversion Agent under the 2017/22 Bonds and the Bonds consents, which could adversely impact our ability to raise additional debt financing. Further, according to the terms of the Highbridge Facility, the Issuer and its direct and indirect subsidiaries (collectively, the "**Group**") are not permitted—other than under the Highbridge Facility and subject to certain exceptions—to create security over their assets or incur additional financial indebtedness.

If we fail to raise sufficient additional funds in a timely manner, we may need to (i) significantly descale, scale back or discontinue development activities, (ii) seek strategic alliances for research and development programs at an earlier stage than we would otherwise desire or on less favorable terms than might otherwise be available, and (iii) relinquish, or license on unfavorable terms, our rights to technologies or any future product candidates that we otherwise would seek to develop or commercialize ourselves. If none of these efforts succeed, we may be required to further restructure, cease operations, or file for bankruptcy or insolvency.

Even if we are able to raise additional funds in time, we currently expect that we will continue to require additional capital to fund our operations and to finance the further advancement of our product candidates. Fundraising efforts may divert our management from our day-to-day activities, which may adversely affect our ability to develop and commercialize our product candidates, or we may default our existing financial obligations. In addition, we cannot guarantee that future financing will be available in sufficient amounts or on terms acceptable to us, if at all.

If we raise additional equity or issue equity-linked instruments, the Company's shareholders could be significantly diluted. If we incur additional debt, the terms of such debt may subject us to restrictive covenants or security obligations that limit our flexibility in conducting future business activities, such as incurring additional debt or acquiring or licensing intellectual property rights. Additional debt may limit our potential to redeem the Bonds upon maturity even more.

We may be unable to receive additional funds under the Highbridge Facility. If we do not receive such additional funds, we will face imminent illiquidity within weeks or a few months and we may have to further restructure, cease operations or file for bankruptcy or insolvency.

The drawing of additional tranches under the Highbridge Facility is subject to certain conditions, many of which are not in our control. If not all of these conditions are met, we may be unable to receive additional funds under the Highbridge Facility, on which we rely for short-term liquidity. If we do not receive additional funds under the Highbridge Facility for any reason, we will face imminent illiquidity within weeks or months and we may have to further restructure, cease operations or file for bankruptcy or insolvency.

We may be unable to redeem the Bonds.

Upon maturity of the Bonds, in the event of a delisting of the Shares or in other situations, the Bondholders may require the Company to redeem all of the outstanding Bonds (see Conditions 5, 7 and 11 of the Terms of the Bonds in Section VIII). If such an event were to occur, or at maturity of the Bonds, no assurance can be given that the Company will have sufficient funds or would be able to arrange financing to pay the redemption amount for all Bonds that are to be redeemed. The Company's ability to redeem the Bonds in such event may be limited by law or the terms of other debt instruments. Also, the Company may be required to refinance its debt in order to make such payments. If we are unable to redeem the Bonds or otherwise suffer financial distress, the Shares into which Bondholders may convert the Bonds may not have a significant value, either.

In addition, the portion of the 2017/22 Bonds that will remain outstanding after completion of the Exchange Offer will become due for redemption on February 17, 2022, *i.e.*, 30 months prior to the maturity date of the Bonds. It is uncertain whether the Company will be able to satisfy its payment obligations under the 2017/22 Bonds. Even if the Company is able to do that, the Company may not be able to redeem the Bonds when they become due.

If we are unable to maintain an asset value covering our liabilities, including the Bonds, we will be required to further restructure, cease operations or file for bankruptcy or insolvency, even long before maturity of the Bonds.

If we are unable to maintain an asset value covering our liabilities, including those under the Bonds and under the 2017/22 Bonds still outstanding after completion of the Exchange Offer, our amount of equity will decrease. As a result, we may have to restructure our balance sheet or raise further equity, or otherwise file for bankruptcy or insolvency. This may occur even long before the maturity of the Bonds.

We have not received marketing authorization for any of our current product candidates for any country. We have incurred significant losses since our inception and expect to incur substantial losses and negative operating cash flows for the foreseeable future and may never achieve or maintain profitability.

Other than Raxone®, which we have outlicensed to Chiesi Group, we currently have no products approved for commercial sale. Our remaining lead product candidate is vamorolone in DMD, which we have in-licensed from Reveragen for DMD and other indications. We have not received marketing authorization for our current product candidates for any country (whereby references herein to "our" product candidates include vamorolone in DMD as well as in-licensed product candidates such as lonodelestat, unless otherwise stated or the context requires otherwise).

We have incurred consistent cash-outflow and significant losses since our inception, including a net loss of CHF 54.2 million in 2018, of CHF 19.0 million in 2019 (which was reduced due to the one-off effect of the out-licensing of Raxone®) and of CHF 67.7 million in 2020. As of December 31, 2020, our total equity on a consolidated basis was negative CHF 6.4 million. We expect to continue to incur significant operating losses for the foreseeable future, as we continue our development and commercialization efforts and make investments. We expect our expenses to increase substantially over the coming years, primarily due to higher operating expenses in connection with our ongoing development activities as well as the ramping up of our commercialization activities relating to vamorolone if the results of the VISION-DMD Trial show a pathway to obtaining marketing authorization. To become and remain profitable, we must successfully complete the development of our product candidates, obtain marketing authorizations and pricing and reimbursement approvals for them, expand our development pipeline, maintain and manage our manufacturing arrangements with third parties, maintain and build up an effective internal sales and marketing organization, establish and maintain sales and marketing arrangements with third parties and raise sufficient funds to finance our activities. We may never succeed in these activities, and even if we do, we may never generate sales that are significant enough to achieve profitability.

We only have one late-stage product candidate, vamorolone in DMD. Even if we should receive a Positive Interim Readout, our future profitability, if any, will depend on us being able to obtain marketing authorization and, thereafter, pricing and reimbursement approvals for vamorolone in DMD and our other product candidates. Any setbacks impacting the development and potential commercialization of vamorolone in DMD or our other product candidates may have a material adverse effect on us.

We had to discontinue the development of our late-stage product candidate, Puldysa®, in fall 2020 as a result of unfavorable results of an interim analysis of our phase III clinical trial of Puldysa® in certain DMD patients. As a result, we only have one remaining late-stage product candidate, vamorolone in DMD. Even if we should receive a Positive Interim Readout, our future success and profitability (if any) will depend on our ability to obtain marketing authorization and, thereafter, pricing and reimbursement approvals for vamorolone in DMD in the United States of America (the "**U.S.**") and in the European Union (the "**EU**"), as well as on other factors. We may never receive a marketing authorization for vamorolone in DMD. Even if we eventually obtain such marketing authorization for the U.S. or the EU, we may not receive it on terms acceptable to us, we may not receive pricing and reimbursement for our product at all or on satisfactory terms, or our product may not be commercially viable. In addition, any setbacks impacting the further development and potential commercialization of vamorolone in DMD or any difficulty in the manufacture, or problem with the supply, of vamorolone, or any measures taken by regulators in relation to vamorolone, may have a material adverse effect on our business, results of operations, financial condition, or prospects.

If we are unsuccessful or significantly delayed in obtaining marketing authorization for vamorolone in DMD or its subsequent commercialization, we would have to rely on our early stage pipeline comprising lonodelestat in cystic fibrosis ("**CF**") (with respect to which we recently completed a phase Ib multiple ascending dose trial) and our pre-clinical collaborations regarding a potential gene therapy for laminin-alpha 2 (LAMA2)-deficient congenital muscular dystrophy (LAMA2-MD). Given the uncertainties around the development and commercialization of pharmaceuticals, we may not be able to develop and commercialize any such product candidates in a timely manner or at all.

News on our development and commercialization efforts that we expect to receive in 2021 and in the longer term may have a significant and potentially material adverse effect on the value of the Group and, as a consequence, the market price of our securities.

The value of the Group strongly depends on the results of our clinical trials and on the decisions by regulatory authorities. We expect to receive material new information on such matters in 2021 and in the longer term. In particular, we currently expect six-month top line data of the VISION-DMD Trial in the second quarter of 2021. Any negative outcomes or delays of our development activities may have a material adverse effect on the value of the Group and may adversely affect its business and prospects. As a consequence, the market price of our securities is expected to be very volatile. Should any such news be unfavorable, the market price of our securities may significantly decline and, potentially, not recover.

2. Risks related to the development of our product candidates

Our product candidates must prove their efficacy and safety in rigorous clinical testing. Drug development involves a lengthy and expensive process, even more so in the case of pediatric medications like vamorolone in DMD, and in any case with an uncertain outcome. Failure may occur at any stage of clinical development.

Before we may seek marketing authorization for any product candidate, we must conduct extensive clinical trials to demonstrate its safety and efficacy in humans. Clinical testing is expensive, difficult to design and implement, can take many years to complete and is inherently uncertain as to its outcome. A failure of one or more clinical trials can occur at any stage of testing. Promising results in preclinical studies of a product candidate may not be predictive of similar results in humans during clinical trials, and successful results from early clinical trials may not be replicated in later and larger clinical trials or in clinical trials for different indications or different patient populations.

As our lead product candidate, vamorolone in DMD, is a pediatric medication, a marketing authorization application ("**MAA**") in the EU would have to include the results of studies as described in an agreed pediatric investigation plan ("**PIP**") aimed at ensuring that the necessary data are obtained through studies in children, unless the medicine is exempt because of a deferral or waiver. Similarly, the FDA would require us to conduct studies under a PIP as a post-authorization measure, if it approves a future NDA with regard to vamarolone in DMD. Preparing a PIP and conducting studies under a PIP is time consuming and expensive, and we may fail to complete such studies or such studies could have negative outcomes.

The conduct of clinical trials may be prevented, delayed, or even futile, and delays in the commencement, enrollment or completion of clinical trials for any of our product candidates could result in increased costs or prevent us from commercializing our product candidates on a timely basis, or at all.

Before a clinical trial may begin, we or our partners must obtain approval from the competent regulatory authority and/or the competent ethics committee. We or our partners may not obtain authorization for further testing of our product candidates. Clinical trials of our product candidates may not be conducted as planned, and commencement, enrollment or completion may not occur on our planned schedule, if at all, for many reasons, which could result in increased costs and could negatively affect our or our partners' ability to complete the clinical trial. We have experienced delays in clinical trials, also due to the COVID 19 pandemic, and cost overruns in the past and may do so again in the future. If we or our partners are not able to successfully design, operate, complete and correctly evaluate the results of the clinical

trials for our product candidates, we will not be able to seek marketing authorization or commercialize them.

If we or our partners experience delays or difficulties in the enrollment of patients in clinical trials, the conduct and completion of clinical trials may be delayed or prevented.

Initiation and successful and timely completion of clinical trials requires us to enroll a sufficient number of eligible patients in these trials. Given our focus on orphan drugs, our clinical trials look to enroll patients with characteristics that are found in a small number of patients and are likely to compete with other clinical trials for product candidates targeting treatment of patients with the same characteristics. As the number of qualified clinical investigators is limited, we expect to conduct some of our clinical trials at the same clinical trial sites that some of our competitors use, which reduces the number of patients available for our clinical trials at these clinical trial sites.

Delays in patient enrollment may result in increased costs or may affect the timing or outcome of our clinical trials, which could prevent their completion.

We may not be successful in our efforts to build up our development pipeline or to spend our limited resources on the most promising product candidates.

We may not be able to develop our existing product candidates or to identify and develop further product candidates that are safe and effective despite spending substantial technical, financial, and personnel resources thereon. Because we have limited resources, we may forgo or delay pursuit of opportunities with certain product candidates or indications that later prove to have a greater potential than the product candidates or indications on which we have chosen to focus. Even if we are successful in continuing to build our development pipeline, the product candidates that we identify may not be suitable for clinical development, including as a result of being shown to have harmful side effects or other characteristics that indicate that they are unlikely to receive marketing authorization and achieve market acceptance.

We rely and will in the future rely on third parties to conduct clinical trials for our product candidates, and if these third parties do not properly and successfully perform their obligations, we may not be able to successfully complete the respective development of our product candidates.

We rely on Clinical Research Organizations ("**CROs**") and other third parties to assist in managing, monitoring and otherwise carrying out clinical trials for our product candidates. Together with the salaries paid to our employees in the product development department, the fees and expenses of these CROs account for a significant part of our development expenses. We compete with many other companies for the resources of these third parties. These third parties generally may terminate their engagements with us at any time.

If the quality or accuracy of the data that these third parties obtain is compromised due to the failure on the part of these parties to adhere to clinical trial protocols or to regulatory requirements, or if these third parties otherwise fail to comply with clinical trial protocols or meet expected deadlines, the clinical trials of our product candidates may not meet regulatory requirements. If clinical trials do not meet regulatory requirements or if these third parties need to be replaced for any reason, the development of our product candidates may be delayed or suspended, may be more expensive than planned or may ultimately fail.

Although we rely extensively on third parties to conduct our product development work, we remain responsible for ensuring that each of our clinical trials is conducted in accordance with its general investigational plan, protocol, legal and regulatory requirements and scientific standards. We may incur financial liabilities or suffer negative regulatory consequences as a result of any shortcoming in meeting such responsibilities irrespective of whether we have delegated such responsibility to a CRO or other third party.

We may not realize the benefits of our in-licensing of vamorolone from ReveraGen, of lonodelestat from Polyphor, of any other product candidates or compounds that we may in-license or acquire, of

any strategic alliances that we may form, joint ventures that we may create, or strategic transactions that we may enter into in the future.

We have in-licensed all of our current product candidates, typically against payment of upfront consideration and milestone and royalty payments.

In September, 2020 we acquired a worldwide exclusive, royalty-bearing and sub-licensable license from ReveraGen (in a tripartite agreement including the previous license holder, Idorsia) to develop, commercialize and manufacture vamorolone for all indications. In February 2018, we in-licensed the compound lonodelestat from Polyphor. In each case, we made significant upfront payments and we agreed to make substantial additional payments, some of which are contingent and some are not, to our respective counterparties.

We may not be able to realize the benefit of our past or future acquisitions or in-licensing transactions, or they may turn out to have been made at too high a price. Among other things, ReveraGen is financially dependent on regular agreed payments from us and may be require to file for bankruptcy if we defaulted on such payments or if its cash needs increased significantly. In that event, we would likely lose the benefit of our license from ReveraGen.

Any strategic alliances, joint ventures or strategic transactions that we may enter into in the future may fail to achieve the expected results and may divert capital resources and management time. It is unclear whether and when any product candidates may generate revenues for the Company.

We may not be successful in maintaining existing or establishing and maintaining additional collaborations, and we may not fulfil our obligations vis-à-vis our collaboration partners.

We have entered into a number of collaborations and licensing arrangements, for example with other pharmaceutical companies, clinical research centers and other research institutions. For summaries of our material license and collaboration agreements see "*THE COMPANY AND ITS BUSINESS—Additional information on our business—Material agreements—License and collaboration agreements*" beginning on page 77. These existing collaborations are, and any future collaborations or partnerships may be, important to our business. Generally, such collaborations allow us to share the development costs with our collaboration partners, thereby significantly reducing our own costs, and to utilize the expertise and know-how of our development partners.

In collaboration agreements, we often undertake to take certain specified efforts to develop a product candidate. If we are unable to fulfil, or otherwise do not fulfil, these obligations, we may be in breach of our obligations vis-à-vis the respective collaboration partner, which may result in a termination of the collaboration or in our liability vis-à-vis the collaboration partner.

Generally, we may not be able to maintain our current or any future collaborations or partnerships, including for reasons beyond our control. In the event of termination of a collaboration, we may be unable to progress the relevant product candidate on our own or may be unable to successfully find a new partner with which to do so on terms favorable to us or at all. Also, any termination of a collaboration by our partner could make it difficult for us to attract new strategic partners or adversely affect how we are perceived in scientific and financial communities.

We will face significant competition in seeking partners for future product development collaborations. In order for us to successfully partner our product candidates, potential partners must view us and the respective product candidate as attractive, also in light of the terms that we are seeking. Even if we successfully establish new collaborations, their terms may not be favorable to us.

If we fail to establish or maintain a collaboration related to a particular product candidate, we will bear all of the related development cost and risk and may be unable to develop that product candidate on our own for lack of resources or other reasons.

If serious adverse events or undesirable or unacceptable side effects are identified during the development of any of our product candidates or after commercialization of any product or any future products, we may need to discontinue the development of the product candidates or withdraw the product from the market.

If any of our product candidates cause undesirable or unacceptable side effects in clinical trials or have characteristics that are unexpected, we may decide or be required to interrupt, delay or discontinue the relevant product candidate's development or may choose to limit its development to more narrow uses or patient subpopulations in which such side effects or other characteristics are less prevalent, less severe or more acceptable from a risk-benefit perspective. Serious procedure- or treatment-related side effects may occur at any stage of product development and even after commercialization. Any such side effects may cause us to discontinue or limit the development of the relevant product candidate or we may decide or be required to withdraw the relevant product from the market, which may result in a sudden and sharp drop of our net sales and/or significant impairment charges.

3. Risks related to marketing approval of our product candidates and legal compliance matters

Following clinical development, our product candidates will require marketing authorization. If we are unable to obtain marketing authorization for a particular product candidate in a timely manner, on terms acceptable to us or at all, we will not be able to commercialize it, and our ability to generate sales will be materially impaired.

Our product candidates require marketing authorizations from the FDA in the U.S., from the European Commission in the EU and from comparable regulatory authorities in other relevant jurisdictions (such as Swissmedic in Switzerland), prior to commercialization. In most jurisdictions, the process of obtaining marketing authorization for a product candidate is expensive and may take many years, if approval is obtained at all, and can vary substantially based upon a variety of factors, including the type, complexity and novelty of the product candidates involved. Failure to obtain marketing authorization for a product candidate will prevent us from commercializing the product candidate.

Our experience in filing and supporting the applications necessary to gain marketing authorization for a product candidate is limited and some of our MAAs were not successful in the past. We have never filed an NDA with, or obtained marketing authorization from, the FDA in the U.S., which is a significant pharmaceutical market. We have started (but may fail) to build up our in-house capacity for purposes of obtaining marketing authorization for vamorolone in DMD in the U.S., and we continue to rely on external advisors to assist us with the marketing authorization process in the U.S.

Regulatory authorities have substantial discretion in the timing and substance of the approval process and may refuse to accept any application or may decide that our data are insufficient for approval and require additional preclinical, clinical or other studies. Even if we receive a Positive Interim Readout and the FDA support an NDA based thereon, for which there is no assurance, it is possible that the FDA may still not consider the VISION-DMD Trial, or any additional studies for vamorolone in DMD performed and completed that it may request, sufficient to approve any NDA for vamorolone in DMD that we may submit. If our clinical data are found insufficient, we may be forced to abandon seeking marketing authorization in the U.S. or the EU for vamorolone in DMD.

Regulatory authorities may also narrow the uses or patient subpopulations for which the product is approved or require extensive warnings on the label. For example, the FDA, the EMA or any other regulatory authority could, if at all, approve vamorolone in DMD for a narrower age group of patients than requested. Any of these restrictions could materially limit the potential market for or interest in the respective product.

If we experience delays in obtaining or fail to obtain marketing authorizations for any of our product candidates in any key jurisdiction, especially in the U.S. and the EU, their commercial prospects may be harmed or they may no longer be commercially viable. As a result, our ability to generate sales will be materially impaired.

Fast track, breakthrough therapy and similar designations for some of our product candidates may not lead to a faster development or regulatory review or approval process, will not increase the likelihood of receiving marketing authorization and may be revoked.

ReveraGen has received orphan drug designation for vamorolone in DMD from both the FDA and the EMA, as well as fast track designation from the FDA (see "*THE COMPANY AND ITS BUSINESS—Our lead product candidate: vamorolone for the treatment* of DMD—*Vamorolone in DMD—Market exclusivity, regulatory status*" beginning on page 71, respectively, for more information). We may seek, but may not necessarily receive fast track or similar designations for lonodelestat in CF and/or any future product candidates, or designations comparable to breakthrough therapy designations designations in certain and for certain products or product candidates.

Regulatory authorities typically have broad discretion in granting fast track, break through therapy and similar designations and may rescind or revoke such designations. Even if such designation is granted, such designation is not predictive of future clinical trial results, does not necessarily (and in the case of certain designations will not) result in a faster development process, review or marketing approval compared to conventional approval procedures and does not increase the likelihood that a product candidate will receive marketing authorization. Many drugs that have received such designations have failed to obtain marketing authorization. If we fail to obtain any such designation for a product candidate that we think meets the criteria or any existing designations is revoked, further development of that product candidate and, ultimately, its commercialization could be materially adversely affected.

Raxone® is, and any product candidate for which we may obtain marketing authorization will be, subject to extensive post-marketing regulatory requirements and could be subject to post-marketing restrictions, post-marketing studies or withdrawal from the market, and we may be subject to penalties if we or the third parties with which we collaborate fail to comply with regulatory requirements or experience unanticipated problems with that product.

The commercialization activities by Chiesi Group with respect to Raxone® under our outlicensing agreement are, and our own commercialization activities with respect to any product candidates for which we may receive marketing authorization will be, subject to comprehensive regulation by regulatory authorities in each jurisdiction in which it is authorized. This regulation includes requirements regarding the testing, manufacture, recordkeeping, labeling, storage, approval, advertising, promotion, sale and distribution of the relevant product. For example, we need to with respect to Raxone®, and we will need to for any product for which we may receive marketing authorization in the future, submit safety and other post-marketing information and reports, ensure that our contract manufacturers observe current Good Manufacturing Practice ("**cGMP**") requirements and comply with requirements regarding safety monitoring and pharmacovigilance.

Regulatory authorities may also impose requirements for expensive post-marketing studies or clinical trials and surveillance to monitor the safety or efficacy of the product. For example, under the European Commission's marketing authorization that was granted for Raxone® "under exceptional circumstances" because the European Commission found that comprehensive efficacy and safety data cannot be obtained, we are required to conduct several post-authorization measures (see "*THE COMPANY AND ITS BUSINESS—Our product outlicensed to Chiesi Group: Raxone®—Clinical development, efficacy, and post-authorization measures*" beginning on page 72 for more information). We have a contractual obligation to Chiesi Group to conduct the post-authorization measures with regard to Raxone® that are required under our marketing authorization for the EU at our own cost.

Any such requirements for any other products for which we may receive marketing authorization in the future, including vamorolone in DMD, may adversely affect our profit and cash flow generated from the relevant products, and such additional clinical trials involve the risks associated with any clinical trials. For example, if our phase IV clinical trial of Raxone® does not establish the product's long-term efficacy, this may adversely impact its commercial success and thus the likelihood that our rights to milestone payments are triggered, as well as the timing thereof, under our agreement with Chiesi Group. Also, later discovery of previously unknown adverse effects or other problems with our products, manufacturers or

manufacturing processes, or non-compliance with regulatory requirements may have serious consequences for us, including legal or regulatory actions such as warning letters, suspension of manufacturing, seizure of product, injunctions, withdrawal of the relevant product from the market and sanctions.

Our relationships with customers and third-party payers and our general business operations are and will be subject to applicable anti-kickback, fraud and abuse and other healthcare laws and regulations, which could expose us to criminal sanctions, civil penalties, contractual damages, reputational harm or diminished earnings, among other penalties.

Healthcare providers and third-party payers play a primary role in the recommendation and prescription of Raxone® and any product candidates for which we may obtain marketing authorizations. The arrangements with healthcare professionals, third-party payers and customers that we or our distributors have entered or will enter into may expose us to broadly applicable fraud and abuse and other healthcare laws and regulations that may constrain the business or financial arrangements and relationships through which we or our distributors market, sell and distribute our products (for which we receive marketing authorization). Efforts to ensure that our business arrangements with third parties will comply with applicable healthcare laws and regulations will involve substantial costs. If authorities conclude that our or our distributors' business practices do not comply with applicable laws and regulations, we or our employees or distributors may be subject to significant civil, criminal and administrative penalties, damages, fines, imprisonment, exclusion of products from government-funded healthcare programs such as Medicare and Medicaid, disgorgement, contractual damages, reputational harm, diminished profits and future earnings, or the curtailment or restructuring of our operations.

If we or our third-party contractors or employees fail to comply with environmental, health and safety laws, we could become subject to civil or criminal penalties, other remedial measures or incur costs that could harm our business.

We are subject to a variety of environmental, health and safety laws and regulations, including those governing laboratory procedures and the handling, use, storage, treatment and disposal of biological materials and hazardous materials and wastes. The operations of our third-party manufacturers and suppliers involve the use of hazardous and flammable materials, including chemicals and biological materials, and also produce hazardous waste products. We cannot eliminate the risk of contamination or injury from these materials or wastes. In the event of such contamination or injury, we could be held liable for any resulting damages, and any liability could exceed our insurance coverage and our own resources. Non-compliance by us or our third-party contractors or employees with environmental, health and safety laws and regulations may result in substantial fines, civil or criminal penalties or other sanctions. In addition, we may incur substantial costs in order to comply with such laws and regulations.

4. Risks related to the commercialization of our product candidates and marketing and sale of our products

We have outlicensed our only commercial product, Raxone®, to Chiesi Group in August 2019 and we no longer generate significant revenue from the sale of Raxone® by ourselves. We may not receive any of the milestone payments of up to EUR 49 million agreed with Chiesi Group, which would have a negative impact on our financial situation and timeline towards profitability.

In August 2019, we outlicensed our rights in our only product, Raxone®, which is approved for commercial sale for the treatment of Leber's hereditary optic neuropathy ("**LHON**") in the EU, the United Kingdom (the "**UK**") and certain other jurisdictions, to Chiesi Group. Under the agreement, we have the right to receive up to EUR 49 million in staggered milestone payments if and when Chiesi Group meets certain sales thresholds. It is uncertain whether and when Chiesi Group will meet any of these sales thresholds. Consequently, we may not receive any of these milestone payments or may receive only partial milestone payments.

Following the outlicensing of Raxone® to Chiesi Group, we continue to sell Raxone® in France under a transitory regime agreed with Chiesi Group. However, as the French Ministry for Solidarity and Health

has finally refused to register Raxone® on the list of reimbursed products in France, we do not expect sales of Raxone® in France to continue from August 2021 onwards.

Our product, Raxone®, which is outlicensed to Chiesi Group, and any of our product candidates (to the extent we receive marketing authorization for them) may fail to achieve the degree of market acceptance by physicians, patients, third-party payers and others in the medical community necessary for commercial success despite having received marketing authorization.

Raxone®, our product outlicensed to Chiesi Group, and any product candidates for which we may receive marketing authorization in the future, may fail to gain sufficient market acceptance by physicians, patients, third-party payers and others in the medical community despite having received marketing authorization. For example, other novel products may be preferred to our product. If any such products do not achieve an adequate level of acceptance, we may not generate significant sales of our own products (if we receive marketing authorization for them) and we may not become profitable.

We may be required to refund to the French Social Security part of our revenue generated from the sale of Raxone® in France since January 1, 2016. If we are required to make such a refund in cash, our financial situation, results of operations and prospects may be materially adversely affected.

In France, Raxone® has been reimbursed by the French Social Security under the so-called post-*autorisation temporaire d'utilisation*, or post-ATU, financing scheme (*dispositif pérenne*) since the product was launched. As a result of the refusal of the French Ministry for Solidarity and Health to register Raxone® on the lists of reimbursed products in France for patients and hospitals, applicable rules require that we as the holder of the ATU refund to the French Social Security the difference between the price at which we sold Raxone® under the ATU and a reference price to be set by the *Comité économique des produits de santé* (CEPS). Therefore, the CEPS could require us to refund part of our revenue generated from the sale of Raxone® in LHON in France since January 1, 2016. In the assessment of our French legal counsels, it is impossible as of the date of this Prospectus to assess the amount of a potential refund because the reference price to be set by the CEPS is unknown. Also, the process of establishing a potential refund and its modalities could be protracted. As of the date of this Prospectus, our discussions with French authorities about the reimbursement status of Raxone® in France are ongoing. If we are unable to reach an agreement with French authorities on a potential refund of part of our revenue generated from the sale of Raxone® in LHON in France since January 1, 2016, the authorities may require such refund from us. Should we be required to make a significant cash payment, our financial situation, results of operations and prospects may be materially adversely affected. Should the CEPS accept an alternative way of compensation by way of discounts on future sales to be determined with the CEPS, which management believes to be likelier scenario, we currently estimate the cost of goods and supply for the products to be provided under such refund scheme to be less than CHF 0.6 million *per annum*.

Off-label and unlicensed uses of currently available forms of idebenone may adversely affect the sales of Raxone®.

Physicians may prescribe available products containing idebenone (the active ingredient in Raxone®) for uses for which they are not approved, such as the treatment of LHON, if they view such products as a less expensive treatment or a better alternative to Raxone®. A considerable number of physicians in Europe, and to a lesser degree in the U.S. and other countries, have been prescribing or recommending products containing idebenone to their patients on an off-label basis. The off-label product is either acquired from internet sources or in countries where it is approved and marketed for a different indication. By way of example, and without any claim to completeness:

- Takeda's Mnesis®, 45mg tablets containing idebenone, is registered in Italy for the treatment of "cognitive-behavioral deficits resulting from cerebral pathologies whether from vascular or degenerative origin" and is used off-label and prescribed as an unlicensed medicine for the treatment of other (non-approved) indications in Italy and in certain other countries.

- Sweden's Medical Products Agency ("**MPA**") has granted several licenses to individual patients for the prescription and reimbursement of Mnesis® for the treatment of LHON and so far we have not been able to successfully challenge these decisions.
- Pharmacies have been compounding idebenone. See risk factor "*Pharmacies have been compounding idebenone. Future compounding may adversely affect sales by Chiesi Group* of Raxone®."

Any off-label or unlicensed use of idebenone, especially from inexpensive sources, and any reimbursement for such use granted by third-party payers may reduce the likelihood that our rights to milestone payments from Chiesi Group in connection with Chiesi Group's sale of Raxone® are triggered.

We have limited experience in marketing products and do not expect to have significant marketing synergies between our current product candidates, if and when approved. If we are unable to establish and expand marketing and sales capabilities or enter into distribution agreements with third parties, we may not be able to generate product sales.

We have limited experience in marketing products in Europe and have no experience in marketing products in the U.S. and elsewhere. In the U.S., our team currently manages our patient advocacy interactions, prepares for market entry in the U.S. and is the source of our U.S. regulatory and medical affairs expertise, whereas commercialization will only be possible if we file an NDA with, and receive marketing authorization from, the FDA regarding vamorolone in DMD. In fall 2020, as a result of the discontinuation of the development of our late-stage product candidate, Puldysa® and the resulting need for cost savings, we had to cease our commercial capabilities in the EU. A commercial launch of vamorolone in DMD (if and when approved) or of any of our early-stage product candidates will require us to develop in-house marketing, sales and distribution capabilities. All of these activities are associated with an increase in the headcount of our marketing and sales personnel and of the related overhead and higher overall fixed costs, and will also require significant management resources and time. At the same time, we may engage additional third-party distributors to perform marketing, sale and/or distribution services. Any income that we may receive from third-party distributors will depend upon the efforts of such distributors, over which we may have little or no control. We may not be able to develop in-house marketing, sales and distribution capabilities or establish or maintain relationships with third-party collaborators to successfully commercialize vamorolone in DMD (if and when approved) in key markets and for particular indications, which would adversely impact our ability to generate product sales.

Our product candidates (if approved) will have different prescriber bases: primarily neurologists in the case of vamorolone in DMD, and primarily pulmonologists in the case of lonodelestat in CF. As a result, we expect to have somewhat limited marketing synergies between our products and may have to build separate sales channels for each of our products, which is expensive and may result in our products suffering from low profit margins or a lack of profitability.

We face substantial competition, which may result in others discovering, developing or commercializing competing products before or more successfully than we do, as well as reducing the price at which we are able to sell our products.

The development and commercialization of new products is highly competitive. For an overview of the competitive landscape of our product and product candidates see "*THE COMPANY AND ITS BUSINESS—Additional information on our business—Competition*" beginning on page 83 and the references cited therein. Notably, vamorolone is being developed to compete with steroid-based treatments of DMD patients and, therefore, will by design face competition from steroid-based treatments, some of which are performed with inexpensive generic drugs. Moreover, we are aware of five approved treatments for DMD developed by third parties that are not based on steroids, and of several dozen clinical and preclinical programs to develop DMD treatments, some of which are at a late stage of clinical development and some of which we consider to be direct competitors with vamorolone. Also, several companies are currently developing new products for the treatment of LHON, which may compete with Raxone®. If these competitors are granted marketing authorization for their products, sales by Chiesi Group of Raxone® could be reduced and the likelihood and timing of milestone payments by Chiesi Group to us could be adversely

impacted. Also, the fact that neither idebenone (the active ingredient in Raxone®) nor vamorolone enjoys composition of matter patent protection lowers entry barriers for competitors.

Our commercial opportunity could be reduced or eliminated if our competitors develop and commercialize products that are safer, more effective, have fewer or less severe side effects, are more convenient or are less expensive or are better marketed than Raxone® or any other product candidates for which we receive marketing authorization. Our competitors may obtain marketing authorizations for their products more rapidly than we do, which could result in them establishing a strong market position before we are able to enter the market. In addition, our ability to compete may be affected by third-party payers seeking to encourage the use of products that are less expensive than ours.

Should we or our distributors be found to have improperly promoted off-label uses, we may become subject to significant liability.

Our products, if and when approved, may be suitable for certain off-label uses. In particular, we believe that vamorolone might be used as a substitute for glucocorticoids, which are widely a used across a broad range of indications. If we cannot successfully manage the marketing of our products by restricting off-label promotion or if we or our current or future distributors promote our products beyond their approved indications, we could become subject to enforcement action for off-label promotion and significant liability.

The insurance coverage and reimbursement status of newly-approved products is uncertain. Failure to obtain or maintain coverage and adequate reimbursement for our marketed product (which is out-licensed to Chiesi Group) or any product for which we receive marketing authorization in the future and price controls could limit our ability to market those products and decrease our ability to generate sales.

The availability and extent of coverage and reimbursement by governmental and private third-party payers is essential for most patients to be able to afford expensive treatments. Sales of Raxone® (with respect to which we have a right to milestone payments under our agreement with Chiesi Group if and when Chiesi Group meets certain sales thresholds) and potential sales of vamorolone in DMD (if and when approved) and any other products for which we receive marketing authorization in the future will depend substantially on the extent to which the costs will be paid by third-party payers. Also, we may rely on the efforts of third-party distributors to obtain pricing and reimbursement approvals in certain countries. We may have little or no control over the efforts of such third parties.

Seeking third-party reimbursement is a time-consuming and expensive process, which typically requires us to provide scientific and clinical support and pharmaco-economic arguments for the use of the relevant product to each third-party payer separately. We may need to conduct expensive pharmaco-economic studies in order to demonstrate the medical necessity and cost-effectiveness of our products, and third-party payers may nonetheless refuse reimbursement. Also, one third-party payer's decision to provide coverage for a product does not assure that other payers will also provide coverage, and pricing negotiations may continue after reimbursement has been obtained. Third-party payers may reject our requests for pricing and reimbursement of any future product and we may have to initiate legal proceedings in relation to such decisions. Even where reimbursement may be approved in the future, we may have to grant a significant discount on the list price and may have to reduce the price further in the future. Irrespective of the level of initial pricing, we expect the prices of our current and any future products to erode substantially during any market exclusivity period. We expect such price erosion to be accelerated after we have lost any such market exclusivity.

If reimbursement is not available or only to limited levels, we may not succeed in commercializing a product even if marketing authorization has been obtained. Even if coverage is provided, the approved reimbursement amount may not allow us to realize a sufficient return on our investment. All of this also applies to sales of Raxone®, *mutatis mutandis*, with respect to which we have a right to milestone payments under our agreement with Chiesi Group if and when Chiesi Group meets certain sales thresholds.

Recently enacted and future healthcare reform legislation involves a high degree of uncertainty and may adversely affect our business.

We operate in a highly regulated industry. New laws, regulations or judicial decisions, or new interpretations of existing laws, regulations or decisions, related to healthcare availability, the method of delivery or payment for healthcare products and services could adversely affect pricing, reimbursement, marketing or sales of our marketed product (which is outlicensed to Chiesi Group) or any product candidates for which we may receive marketing authorization in the future. In the U.S. and other jurisdictions, there have been a number of legislative and regulatory changes, proposed changes and statements by the government regarding the pharmaceutical industry and the healthcare system that could prevent or delay marketing authorization and pricing and reimbursement approvals of our product candidates or make them more expensive, or their terms less attractive, or restrict or regulate post-approval activities. All of these enacted or future measures may prevent us from generating sales, attain profitability, commercialize or market our products.

Pharmacies have been compounding idebenone. Future compounding may adversely affect sales by Chiesi Group of Raxone®.

Compounding (also called pharmacy or magistral preparation) is a practice in which a licensed pharmacist prepares medicines in a pharmacy by combining, mixing, or altering pharmaceutical ingredients. Under certain conditions, the sale of compounded idebenone (the active ingredient of Raxone®) is legal. In the EU, such compounding exemption is based on Article 3 of the EU Directive 2001/83. We are aware of pharmacies in Germany and the Netherlands that advertised compounded idebenone on the internet for the treatment of LHON, DMD and other indications at considerably lower prices than we previously charged, and Chiesi Group currently charges, for Raxone®, sometimes making reference to the clinical trials that we have conducted.

In the past, compounding of idebenone has also resulted in litigation: a pharmacist in Germany filled capsules with generic idebenone purchased from a third party and advertised their sale on his website. We settled a lawsuit against the pharmacist, who has undertaken to refrain from advertising and selling idebenone capsules.

Compounding of idebenone still continues and, consequently, reduces sales by Chiesi Group of Raxone® and thereby the likelihood that our rights to milestone payments are triggered, as well as the timing thereof, under our agreement with Chiesi Group.

Product liability lawsuits against us could cause us to incur substantial liabilities and to limit commercialization of our marketed product (which is outlicensed to Chiesi Group) or any product candidates for which we receive marketing authorization in the future.

We face an inherent risk of product liability exposure related to the commercialization of Raxone® by Chiesi Group and any product candidate for which we receive marketing authorization in the future, as well as to the use of our product candidates in humans in clinical trials. If we cannot successfully defend ourselves against claims that our products or product candidates caused injuries, we may incur substantial liabilities. In addition, if our product liability risk in relation to Raxone® materializes, we may also be contractually liable to Chiesi Group.

We currently have product liability insurance for Raxone® and insurance for human clinical trials covering all clinical trials conducted by us. However, our current product liability coverage may not be adequate in scope to protect us in the event of a successful product liability claim, nor does it cover any contractual liability to Chiesi Group we may incur in connection with product liability matters. Further, we may not be able to maintain our current insurance or obtain product liability insurance for any products for which we may receive marketing authorization in the future on reasonable terms and at acceptable cost, and our insurance may in any event provide insufficient coverage against potential liabilities. As a result, we may have to bear substantial uninsured losses.

Our future profitability may be adversely affected if our estimates regarding the size of the market for our product candidates are inaccurate.

To evaluate the commercial potential of our product candidates, we have to estimate the size of the market for these product candidates. We base these estimates on the frequency of the respective condition, on our evaluation of market conditions and on other factors, using publicly available information. In formulating these estimates, we have to make certain assumptions, which have not been verified by third parties. If our assumptions are incorrect, there is a risk that our estimates could be wrong and our future profitability may be adversely affected.

5. Risks related to market exclusivity rights and intellectual property

Our business model relies on orphan drug exclusivity for our current or future product candidates. Orphan drug designation can be difficult to obtain and maintain, and it provides only limited protection from competition.

It is our strategy to develop and commercialize product candidates in indications qualifying for orphan drug designation in order to obtain marketing exclusivity. For more information on orphan drug designation and its potential benefits see "*THE COMPANY AND ITS BUSINESS—Additional information on our business—Market exclusivity and intellectual property*" beginning on page 83. The market exclusivity period of an orphan drug designation is generally shorter than a patent protection period. In the U.S., for instance, such period is seven years, and in the EU, it is ten years (reduced to six years if the relevant drug no longer meets the criteria or is sufficiently profitable) after receipt of marketing authorization. Also, orphan drug exclusivity may be lost if the applicable regulatory authority determines that the request for designation was materially defective, if the manufacturer is unable to assure sufficient quantity of the drug to meet patient needs, or for other reasons.

To date, we have obtained orphan drug designations for our marketed product, Raxone® (which is outlicensed to Chiesi Group), in the EU (maximum duration until fall 2025), the U.S. and South Korea, and for lonodelestat in alpha-1 antitrypsin deficiency, primary ciliary dyskinesia and CF in the EU. ReveraGen has received orphan drug designation for vamorolone in DMD in the U.S. and in the EU. We have not filed for orphan drug designation in all national and regional jurisdictions where such protection may be available; instead, we have sought such protections only with respect to jurisdictions that we currently anticipate being key to our business.

Obtaining an orphan drug designation can be difficult, and we may not be successful in obtaining or maintaining orphan drug designations for our marketed product (which is outlicensed to Chiesi Group) or any of our product candidates. The procedure for obtaining orphan drug designation is an independent procedure in each jurisdiction, and applications might be denied in some jurisdictions, but granted in others. Further, orphan drug designation may be obtained for the same product in the same indication by several parties, and only the first such party to obtain marketing approval will receive marketing exclusivity for the relevant product in the relevant indication. Consequently, despite us having obtained an orphan drug designation for a product candidate in a particular indication, if a third party were to obtain orphan drug designation and marketing authorization and the correspondence market exclusivity for the same product in the same indication, we would be excluded from marketing such product in such indication during the applicable exclusivity period.

If we lose orphan drug designation or fail to maintain that designation for the duration of the applicable exclusivity period in relation to our marketed product (with respect to which we have a right to milestone payments under our outlicensing arrangement with Chiesi Group if and when Chiesi Group meets certain sales thresholds) or, after receipt of marketing authorization (if any), any of our product candidates, we may be unable to generate sufficient sales to become profitable.

Our marketed product, Raxone® (which is outlicensed to Chiesi Group), is not patent protected and there are only method of use patents for the use of vamorolone in DMD and certain other indications.

Even granted patents will eventually expire, may not be enforceable, and we may be subject to ownership disputes over patents or other intellectual property.

Raxone® (which is outlicensed to Chiesi Group and with respect to which we have a right to milestone payments if and when Chiesi Group meets certain sales thresholds), is not patent protected. There are to our knowledge no composition of matter patents or patent applications with respect to vamorolone, which we have in-licensed from ReveraGen. ReveraGen holds method of use patents for the use of vamorolone in DMD and other indications in the U.S., the EU, China and other jurisdictions. The main patents relating thereto are due to expire in 2029. The composition of matter patents with respect to lonodelestat held by Polyphor and certain other parties and exclusively licensed or sublicensed, as applicable, to us, are due to expire in 2025, subject to potential extended market protection. Further, we may not be able to rely on patent protection for any of our future product candidates.

Typically, the protection derived from method of use patents is not as strong as the protection derived from composition of matter patents. Method of use patents do not prevent a third party from using, applying or manufacturing the same compound for other indications and may not prevent a third party from finding a way to circumvent the patent. For these reasons, a third party may be able to use vamorolone in different or comparable formulas, applications or indications. Further, method of use patents are, in general, more susceptible to invalidity attacks by third parties than composition of matter patents.

The patent applications that we own or in-license may fail to result in issued patents with claims that cover our product candidates in certain countries. There is no assurance that all potentially relevant prior art relating to such patents and patent applications has been identified. We may be unaware of prior art that could be used to invalidate an issued patent or prevent pending patent applications from issuing as patents. Even if patents do successfully issue and even if such patents cover our product candidates, third parties may challenge their validity, enforceability or scope, which may result in such patents being narrowed or invalidated. Without extensive patent protection, we will only be able to rely upon the limited market exclusivity, if any, resulting from any orphan drug designation, which may be revoked, will only apply for a limited time period and will be subject to other conditions and limitations (see risk factor "*Our business model relies on orphan drug exclusivity for our product candidate, Puldysa® in DMD, and most of our current or future clinical product candidates. Orphan drug designation can be difficult to obtain and maintain, and it provides only limited protection from competition*."). If we are unable to obtain, or if we or our licensors or sublicensors lose, patent protection with respect to any of our products or product candidates, we may be unable to prevent competitors from entering the market with a product that is similar to or the same as our product or product candidate. Further, we may be subject to ownership disputes over patents or other intellectual property with licensors, sublicensors, former employees, collaborators or other third parties.

We have in-licensed all of our product candidates and other intellectual property from third parties. We could lose our rights to use the licensed intellectual property in the event of termination of or dispute relating to the relevant agreement or if such intellectual property is unenforceable for any reason. In addition, enforcement of in-licensed intellectual property and defending against third-party claims in relation thereto are more complex than in the case of our owned intellectual property.

We have in-licensed all of our product candidates, including vamorolone from ReveraGen and lonodelestat from Polyphor (see "*THE COMPANY AND ITS BUSINESS—Additional information on our business—Material agreements—License and collaboration agreements*" beginning on page 77 for more information).

The same risks that apply to the intellectual property rights we own generally apply with respect to protection of intellectual property that we license. If we or our licensors fail to prosecute, maintain and enforce such intellectual property or if such intellectual property is unenforceable or if a licensor would enter bankruptcy or similar distressed status, we could lose our rights to use such intellectual property or our exclusivity with respect to those rights. The same may be the case if the agreements by which we have in-licensed or under which we have the option to in-license intellectual property are terminated or if a dispute arises between us and our licensing partners in relation to our rights or obligations under the license or option agreements, including any such conflict, dispute or disagreement arising from our failure

to satisfy payment obligations under any such agreements. In addition, the enforcement of in-licensed intellectual property in case of violations or misappropriation by third parties and defending against third-party claims in relation to in-licensed intellectual property are more complex than in the case of owned intellectual property. Such proceedings may require coordination with the licensor, and licensors typically have rights to intervene or veto rights. As a result of these factors, our ability to develop and commercialize the affected product candidates may be adversely affected and we may not be able to prevent competitors from making, using, and selling competing products.

Third-party claims of intellectual property infringement or misappropriation may prevent or delay our product development and commercialization efforts.

There is a substantial amount of litigation involving patent and other intellectual property rights in the pharmaceutical and biotechnology industries, including patent infringement lawsuits, interferences, oppositions and *inter partes* reexamination proceedings before the government patent offices. Numerous patents and pending patent applications owned by third parties exist in the fields in which we are active. Third parties may assert that we infringe their intellectual property, and patent applications covering our product candidates could have been filed by others without our knowledge. We may also face a claim of misappropriation if a third party believes that we inappropriately obtained and used trade secrets of such third party. Parties making claims against us may obtain injunctive or other equitable relief that could effectively prevent us from further developing or commercializing our product candidates or marketing our product or any future products. We have not conducted a freedom-to-operate search or analysis for our own or in-licensed products (including vamorolone). Thus, we may not be aware of third parties' intellectual property that our products, or our sale or commercialization thereof, may infringe or that, if issued, would block us from selling or otherwise commercializing our products. If any third-party patents were held by a court of competent jurisdiction to cover aspects of our materials, formulations, methods of manufacture or methods of treatment, the holder of any such patents would be able to block our ability to develop and commercialize the applicable product candidate until such patent expired or unless we or our partners obtain a license. These licenses may not be available on acceptable terms, if at all. Even if we or our partners were able to obtain a license, the rights may be non-exclusive, which could result in our competitors gaining access to the same intellectual property. Ultimately, we could be prevented from commercializing a product candidate, or be forced to cease some aspect of our business operations, if as a result of actual or threatened patent infringement claims, we or our partners are unable to enter into licenses on acceptable terms. Defending against claims of patent infringement or misappropriation of trade secrets could be costly and time-consuming, regardless of the outcome, and result in significant demands on the time and attention of the management teams. In the event of a successful claim of infringement, we may be required to pay substantial damages, royalties or other financial remedies and incur other significant costs, redesign our infringing products or obtain one or more licenses from third parties, which may be impossible, or require substantial time and monetary expenditure, or incur other significant costs and lose the patent protection to which we thought we were entitled.

During the course of any patent or other intellectual property litigation, there could be public announcements of the results of hearings, rulings on motions, and other interim proceedings in the litigation. If potential and current partners or collaborators or securities analysts or investors regard these announcements as negative, the perceived value of our technology, product candidates and products, development programs or intellectual property could be diminished.

Furthermore, as set out in "*THE COMPANY AND ITS BUSINESS—Additional information on our business—Market exclusivity and intellectual property*" beginning on page 83, the U.S. government has reserved certain rights to vamorolone. As a consequence, ReveraGen is required to comply with certain formalities, including in particular the filing of certain information with governmental databases. Whether or not ReveraGen complies with this requirement is beyond our control. Should ReveraGen be found to be, or have at any point been, in breach of such filing or other obligations in connection with vamorolone, this could result in the retransfer of intellectual property rights in connection with vamorolone to the U.S. Army Medical Research and Materiel Command (USAMRMC) or any successor or other governmental entity or authority and/or in ReveraGen and/or ourselves being involved in a litigation relating to intellectual property rights in connection with vamorolone, each of which could have a materially adverse

effect on our business, results of operations, financial position and cash flows and potentially damage our reputation.

We enjoy only limited geographical protection with respect to patents and may face difficulties in certain jurisdictions, which may diminish the value of intellectual property rights in those jurisdictions.

We and our licensors have not filed patent applications in all national and regional jurisdictions where such protection may be available; instead, we have sought such protection only with respect to jurisdictions that we currently anticipate being key to our business, in particular the U.S. and the EU. In addition, we may decide to abandon national and regional patent applications before grant. Finally, the grant proceeding of each patent in each jurisdiction is an independent proceeding, and applications might in some jurisdictions be refused, while granted in others, which may ultimately limit our ability to rely on jurisdictional exclusivity, if any, for our marketed product (which is outlicensed to Chiesi Group) or our product candidates in certain jurisdictions. Depending on the jurisdiction, the scope of patent protection may vary for the same product candidate or technology.

The laws of some jurisdictions do not protect intellectual property rights to the same extent as the laws in the U.S. and Europe, and many companies have encountered significant difficulties in protecting and defending such rights in such jurisdictions. If we or our licensors encounter difficulties in protecting, or are otherwise precluded from effectively protecting, the intellectual property rights important for our business in such jurisdictions, the value of these rights may be diminished, and we may face additional competition from others in those jurisdictions.

Many countries have compulsory licensing laws under which a patent owner may be compelled to grant licenses to third parties. In addition, many countries limit the enforceability of patents against government agencies or government contractors. In these countries, the patent owner may have limited remedies, which could materially diminish the value of such patent. If we are or any of our licensors is forced to grant a license to third parties with respect to any patents relevant to our business, our competitive position may be impaired and our business and results of operation may be adversely affected.

If we are unable to protect the confidentiality of our trade secrets, our business and competitive position would be harmed.

We rely on trade secrets and confidentiality agreements to protect proprietary know-how that is not patentable or that we elect not to patent, processes for which patents are difficult to enforce and any other elements of our technology and development processes that involve proprietary know-how, information or technology that is not covered by patents. In addition, we rely on our employees, advisors, third party contractors such as CROs, consultants and collaboration partners to develop and manufacture our product and product candidates, which is why we must, at times, share our intellectual property and trade secrets with them.

Trade secrets can be difficult to protect. We seek to protect our proprietary and in-licensed technology and processes, in part, by entering into confidentiality agreements with our employees, consultants, and outside scientific advisors, contractors and collaborators, but our employees, consultants, outside scientific advisors, contractors and collaborators may intentionally or inadvertently disclose our trade secret information to competitors. In addition, our competitors may gain access to our trade secrets through legal or illegal means or independently develop substantially equivalent information and techniques. We may not be able to protect trade secrets effectively and we may not have adequate remedies against misappropriation of trade secrets. Enforcing a claim that a third party illegally obtained and is using any of our trade secrets is expensive and time consuming and the outcome is unpredictable. In addition, courts outside the U.S. sometimes are less willing than U.S. courts to protect trade secrets. Misappropriation, unauthorized disclosure or a competitor's discovery of our trade secrets could materially impair our competitive position or our business.

Many of our employees were previously employed at universities or other pharmaceutical companies, including our competitors or potential competitors. We may be subject to claims that we or our employees have inadvertently or otherwise used or disclosed trade secrets or other proprietary information of our

employees' former employers or other third parties. We may also be subject to ownership disputes in respect of intellectual property created by these employees during the course of their employment with us. Any of such claims could result in our competitive position being impaired and our business and results of operation may be adversely affected.

We may become involved in lawsuits to protect or enforce our patents and other exclusivity rights, which could be expensive, time-consuming, and unsuccessful.

Competitors may infringe our intellectual property, the intellectual property of our licensors, or the market exclusivity resulting from orphan drug designations that we have achieved. To counter or defend against such claims can be expensive and time-consuming. In an infringement proceeding, a court may decide that a patent owned or in-licensed by us is invalid or unenforceable and/or may refuse to stop the other party from using the technology at issue. An adverse result in any litigation over exclusivity rights could put one or more of our or our licensors' patents at risk of being invalidated or interpreted narrowly or an orphan drug designation of being revoked. Furthermore, because of the substantial amount of discovery required in connection with intellectual property litigation in many jurisdictions, there is a risk that some of our confidential information could be compromised by disclosure during this type of litigation.

We may also be subject to claims that former employees, collaborators or other third parties have an ownership interest in the patents or other intellectual property we own or license-in. We may be subject to ownership disputes in the future arising from, for example, conflicting obligations of consultants or others who are involved in developing our product candidates. Litigation may be necessary to defend against these and other claims challenging inventorship or ownership. If we or our licensors fail in defending any such claims, in addition to paying monetary damages, we may lose valuable intellectual property rights, such as exclusive ownership of, or right to use, valuable intellectual property. Such an outcome could have a material adverse effect on our business. Even if we are successful in defending against such claims, litigation could result in substantial costs and be a distraction to management and employees.

6. Risks related to manufacturing, employment matters, operations, managing growth, corporate structure and financial reporting

We have no manufacturing capabilities or capacity of our own and rely on third parties for production of our compounds and finished drug products.

We have no manufacturing capabilities or capacity of our own and have outsourced the entire manufacture, formulation, packaging, storage and distribution of Raxone® and other drug products to third parties. Under our outlicensing arrangement with Chiesi Group, we have undertaken to supply Raxone® to Chiesi Group in the necessary quantities.

For the production of Raxone® we rely on a manufacturer of active pharmaceutical ingredient ("**API**") (with two manufacturing sites), with whom we have agreed on a seven-year exclusivity period (subject to exceptions) starting with the first launch of our product in the EU and the U.S., respectively. In the EU, such exclusivity period will lapse in October 2022. We recently switched the finished drug product manufacturer of Raxone®. If the respective agreements are terminated or not renewed, we may not be able to timely negotiate a new agreement with that or another third-party provider in time. Furthermore, switching a supplier of the API or the finished drug product is an expensive and time-consuming process.

For the production of vamorolone, we rely on third party manufacturers and we have only started to manage such production by third party manufacturers in September 2020. We have to perform additional technical development and formulation work in view of the assembly of the technical section (the so-called common technical document, CTD) of a potential NDA for vamorolone in DMD, since bioequivalence between ReveraGen's formulation used in clinical trials and the formulation that we intend to use for upscaling production in the run-up to commercial production has not yet been demonstrated. Therefore, we decided to submit a potential NDA for vamorolone in DMD, and to subsequently launch the product, if and when we receive marketing authorization, using the process for producing the API as well

as the formulation used in the clinical trials. We are required to validate the process for producing the API and the formulation used in the clinical trials before we can use them for a marketed product, but we may fail to do so in time or at all, the latter of which could require us to delay a potential launch of vamorolone in DMD. After a potential launch of vamorolone in DMD, we currently plan to introduce a new process for producing the API and establish a formulation that is more suitable for marketed products. We will be required to show equivalence (by either demonstrating bioequivalence or by other technical or clinical data) of the drug product so produced with the drug product used in the clinical trials but we may fail to do so in time or at all.

The API and the finished drug product of lonodelestat are manufactured by a third party, and we rely on PARI Pharma GmbH, Gräfelfing, Germany, as the manufacturer of the nebulizer called eFlow® with which lonodelestat is administered. We currently have no plans to build up or acquire manufacturing capacity and the related know-how of our own in relation to lonodelestat.

The facilities used by our suppliers to manufacture our compounds and the finished products containing such compounds are subject to approval and inspections by regulatory authorities. Even as we conduct regular cGMP audits with our manufacturers, we do not have full control over their manufacturing facilities, quality control or compliance with laws, regulations or cGMP standards, and any non-compliance could result in sanctions being imposed also on us, including fines, injunctions, civil penalties, delays, suspension, withdrawal or non-grant of market of approvals, seizures or recalls of products, operating restrictions and criminal prosecutions.

The compounds we use are complex and difficult to manufacture. Only a few specialized manufacturers are able to manufacture these compounds, and our manufacturers may experience production problems.

The manufacturing of our compounds necessitates compliance with regulatory requirements, such as cGMP, and is complex, time-consuming and expensive. In particular, only a few specialized manufacturers are able to manufacture idebenone, vamorolone or lonodelestat in compliance with all regulatory requirements. Manufacturing idebenone involves heavy metal catalysts, the incomplete removal of which in the manufacturing process would result in toxic amounts of these impurities remaining in the drug substance. Manufacturing vamorolone requires specific know-how, experience and capabilities in steroid chemistry and capabilities to handle toxic raw materials. Manufacturing lonodelestat requires very specific know-how, experience and capabilities in peptide chemistry at large scale as well as in specialized purification processes and analytical methods.

Problems with the manufacturing process of the compounds we use, even minor deviations from the normal process, could result in contamination, product defects or manufacturing failures that could result in harm to patients, lot failures, product recalls, product liability claims, or insufficient inventory. Regulatory authorities may require us to submit samples of any lot or may require that we do not distribute a lot until the agency authorizes its release. Our contract manufacturers may be unable to achieve adequate quantities and quality of clinical-grade materials, and their supply chain could be interrupted from time to time. Any such problems could materially harm our business, financial condition, results of operations, and prospects.

If we lose the services of any member of our top management or other key members of our management, scientific or commercial staff, or if we fail to attract and retain key scientific or other personnel, we may be unable to successfully develop and commercialize our product candidates or any future products for which we obtain marketing authorization.

We are highly dependent on the performance and expertise of members of our top management, especially our CEO, CFO and other key members of our management, scientific and commercial staff. We are a small company with many key functions being carried out by one person only. The loss of the services of any of our key personnel for any reason or our inability to attract new highly qualified and experienced employees could harm our business. Furthermore, we do not currently maintain “key person” insurance for any of our executives or other employees.

A limited number of people have experience and know-how in neuromuscular and pulmonary diseases and the product and product candidates developed by us. To foster retention, we have established employee participation plans, but there is intense competition for skilled personnel. If our product candidates are granted marketing authorizations or if we expand our development activities, we would need to hire additional personnel, which may be difficult to recruit and retain on acceptable terms given such competition.

Our cash-preserving measures have resulted in a loss of know-how and capabilities and legal action against us and thus adversely impacted our operations and prospects.

Starting in fall 2020, as a result of the discontinuation of the development of our late-stage product candidate, Puldysa®, and the resulting need for cost savings, we had to significantly reduce our operations and had to take many other cash-preserving and cost-saving measures. For instance, we ceased our commercial capabilities in the EU, which has led to a reduction of our headcount of approximately 50%.

These measures may have had a significant adverse impact on our operations and prospects, and we expect this effect to adversely impact our ability to develop our product candidates the future. For instance, the loss of know-how and capabilities resulting from the redundancies is difficult to replace in a timely manner. Also, some former employees have taken legal action against us. We will incur substantial defense costs and may be required to make payments to such employees under a potential settlement or if the competent court decides against us. Moreover, certain key employees whom we had planned to retain have resigned or left the company. Our current financial situation limits our ability to provide competitive employment terms for skilled personnel. For example, as a consequence of a recent decrease in the market price of the Shares, most equity-linked rights that we had granted to our directors, officers and employees currently provide only limited incentives to remain in our service. We may continue to lose further key employees in the near future.

If we receive a Positive Interim Readout, we will need to grow the size and capabilities of our organization, and we may experience difficulties in managing this growth.

If we receive a Positive Interim Readout, we expect our headcount to increase significantly in connection with the preparations for a potential launch of vamorolone in DMD (if and when approved) in the U.S. and later in the EU, and also as a result of our planned clinical development of lonodelestat. Our future financial performance will depend, in part, on our ability to effectively manage any future growth. We will need to expand and effectively manage our organization, personnel, operations and facilities in order to successfully develop and commercialize our product candidates. We will only be able to organize operations efficiently and avoid a misallocation of resources if we continue to improve our operational, financial and management controls, reporting systems and procedures. Our management may have to divert a disproportionate amount of its attention away from day-to-day activities in order to manage these growth activities. If we are unable to effectively expand our organization, we may not achieve our development and commercialization goals and our operational efficiency may be materially adversely affected.

Our and our partners' computer systems may fail or suffer security breaches, which could result in a material disruption of our product development programs and our business operations.

Despite the implementation of security measures, our computer systems and those of our current and any future suppliers, CROs and other contractors, consultants and collaborators are vulnerable to damage from computer viruses, ransomware, unauthorized access, natural disasters, terrorism, war and telecommunication and electrical failures. If such an event were to occur and cause interruptions in our operations, it could result in a material disruption of our product development programs and our business operations, whether due to a loss of our trade secrets or other disruptions. For example, the loss of clinical trial data from completed or future clinical trials could result in delays in our regulatory approval efforts and significantly increase our costs due to efforts to recover or reproduce the data. To the extent that any disruption or security breach were to result in data loss or inappropriate disclosure of confidential or proprietary information or personal data of patients or other persons, we may be exposed to liability and suffer reputational damage.

7. Risks related to general economic and financial market conditions

Our business could be adversely affected by the effects of health epidemics, including the COVID-19 pandemic, in regions where we or third parties on which we rely have significant manufacturing facilities, concentrations of clinical trial sites or other business operations.

Our business could be adversely affected by health epidemics. For example, the coronavirus disease "**COVID-19**" pandemic caused by the novel strain of coronavirus that was discovered in 2019 has continued to spread, impacting the economies of most countries around the world. Our operations have been impacted by the COVID-19 pandemic. The outbreak has resulted in governments implementing numerous measures to contain the COVID-19 pandemic, which are subject to change and the respective government authorities may tighten the restrictions at any time. The outbreak has caused us and our business partners to modify business practices including restricting employee travel, developing social distancing plans for employees and canceling physical participation in meetings, events and conferences. We and our business partners may take further actions in this regard. Such modifications may negatively impact productivity, divert resources away from product development, disrupt our and our business partners' business operations and delay and disrupt our development activities.

In addition, the pandemic and the resulting government actions have adversely impacted our clinical trials and may continue to do so in the future. Clinical site initiation, including difficulties in recruiting clinical site investigators and clinical site staff, and patient enrollment may be delayed due to prioritization of hospital resources toward the COVID-19 pandemic. Some patients may not be willing and/or able to comply with clinical trial protocols due to the COVID-19 pandemic, particularly if quarantines impede patient movement or interrupt healthcare services. Similarly, our ability to recruit and retain patients and principal investigators and site staff who, as healthcare providers, may have heightened exposure to COVID-19 may be impeded, which would adversely impact our clinical trial operations. All of these factors may significantly disrupt our development activities. As a result, the expected timeline for data readouts of our clinical trials and certain regulatory filings have been and may be negatively impacted. For example, the completion of our phase Ib multiple ascending dose (MAD) trial of lonodelestat in patients with CF, whose results we announced on March 1, 2021, was delayed by roughly nine months. The VISION-DMD Trial was delayed by several months, and some patient visits were missed or had to be made remotely due to travel restrictions. We expect such possible delays to continue for as long as the COVID-19 pandemic exists. Furthermore, we could face the interruption of key clinical activities such as trial site data monitoring, which may impact the integrity of clinical data. As a result of disruptions caused by the COVID-19 pandemic, we may require additional capital to continue our research activities, which we may be unable to secure on favorable terms, if at all.

The response to the COVID-19 pandemic may redirect resources with respect to regulatory matters in a way that would adversely impact our ability to progress regulatory approvals. In addition, we may face impediments to regulatory meetings and approvals due to measures intended to limit in-person interactions. For example, certain regulatory authorities have postponed inspections of manufacturing facilities and products and are conducting only teleconference meetings. If global health concerns continue to prevent regulatory authorities from conducting their regular inspections, reviews or other regulatory activities, it could significantly impact the ability of regulatory authorities to timely review and process our regulatory submissions, including a potential NDA submission for vamorolone in DMD, which could have a material adverse effect on our business.

As the COVID-19 pandemic continues to evolve, we believe that the extent of its impact to our business, results of operations, financial condition and prospects cannot be reasonably predicted. As a result, all anticipated milestones set forth in this Prospectus are subject to further future adjustment based on the impact of the COVID-19 pandemic. In any case, the COVID-19 pandemic could heighten many of our known risks described elsewhere in this "Risk Factors" section.

Changes in the macro-economic environment and political developments in Europe, the United States and elsewhere may have a material adverse effect on the Group and may reduce the value of our securities.

Over the past years, there has been a series of political, economic and societal events such as the COVID-19 pandemic, increased geopolitical instability as well as increased financial and political instability in the U.S. and serval EU countries, the UK leaving the EU (commonly known as "Brexit"), increased trade disputes and warfare in the Middle East and elsewhere. These events have impacted the global economy at large, the economies and financial situation of governments in many of our current and potential future markets, as well as exchange rates (in particular the euro/Swiss franc rate) and have been associated with, among other things, instability of financial institutions, high market volatility, liquidity problems, limited availability of financing and legal uncertainty. Recession or rising inflation and other effects may also be a consequence of these events. This uncertain macro-economic environment may have a material adverse effect on our business, results of operations, financial condition, prospects, or the market price of our securities.

We are exposed to currency fluctuation risks and other financial risks.

While we incur costs mainly in Swiss francs, a significant proportion of our costs are required to be paid in euros and in the future in United States dollars ("**USD**"). To the limited extent we generate sales, we receive payments primarily in euros. If and as our business grows, we expect that a significant part of our sales and a significant part of our expenses will be denominated USD and euros. Our reporting currency is the Swiss franc and, as a result, financial line items are converted into Swiss francs at applicable foreign exchange rates. Further, we are subject to interest rate risks. Unfavorable developments in the value of the Swiss franc as compared to the euro, in interest rates and in the capital markets could have a material adverse effect our financial condition and results. For a discussion of our foreign exchange rate, interest rate, credit and liquidity risks please also refer to Note 28 to the 2020 consolidated financial statements.

B. Risks related to the offering of the Bonds and to our securities

Unless otherwise indicated, references to "our securities" in this section relate to the Bonds, the 2017/22 Bonds, the Shares and any other securities or derivatives thereof that we have issued or may issue in the future.

The Bonds are unsecured and structurally subordinated to other indebtedness.

The Bonds are unsecured and rank equally in right of payment with all of the Issuer's existing and future unsecured, non-privileged indebtedness. The Issuer is the holding company of the Group. In the event of bankruptcy of one or more of the Issuer's subsidiaries, the respective subsidiaries' creditors have preference over the Issuer (so-called "structural subordination"). Moreover, to the extent that the Issuer pledges or provides security over any of its assets, the respective pledgees or security holders have priority over the other creditors, including the Bondholders, with respect to the distribution of enforcement proceeds.

In particular, the Bonds are structurally subordinated to our indebtedness under the Highbridge Facility, which has been and will be issued by our largest operative subsidiary, Santhera Pharmaceuticals (Schweiz) AG ("**SPCH**"). Also, our indebtedness under the Highbridge Facility is guaranteed by the Issuer and certain subsidiaries of the Issuer and secured by a comprehensive security package, including security over all shares in SPCH and other subsidiaries of the Issuer as well as over the Group's material intellectual property and other assets of the Group. In addition, the Bonds rank *pari passu* with the 2017/22 Bonds still outstanding after completion of the Exchange Offer, as well as with the Idorsia Exchangeable Note in the principal amount of CHF 10 million. The Bondholders will have to share any enforcement proceeds pro rata with Idorsia and the holders of the 2017/22 Bonds.

In the event of our bankruptcy or insolvency, we would expect to lose the benefit of our rights to vamorolone and Bondholders would recover, if at all, only a fraction of the Bonds' principal value.

If we have to file for bankruptcy or insolvency, the net enforcement proceeds would in first order be used to satisfy those creditors with more senior claims than the Bondholders. Moreover, the opening of bankruptcy or insolvency proceedings and other insolvency-related events would constitute events of default under certain contracts, as a result of which, among other things, the Group would expect to lose the benefit of what we believe to be the Group's most important assets, namely the existing license from ReveraGen regarding vamorolone and our marketing authorization in the EU for Raxone®. As a result, Bondholders would, if at all, only recover a fraction of the principal amount of the Bonds in the event of our insolvency.

The Issuer may redeem the Bonds early under certain circumstances and Bondholders may be exposed to reinvestment risk.

The Issuer may redeem all, but not only some of the outstanding Bonds at their Principal Amount, subject to the Interest Make-Whole, at any time if the average VWAP of a Share over a certain period of time has been at least 150% of the prevailing Conversion Price, as set out in more detail in Condition 5(b) of the Terms of the Bonds in Section VIII. During any period when the Issuer may elect to redeem the Bonds, the market value of the relevant Bonds generally is not expected to rise substantially above the price at which they can be redeemed. This may also be the case prior to such period. In the event of an early redemption of the Bonds, Bondholders may be exposed to risks connected to the reinvestment of the proceeds received upon early redemption and may incur additional transaction costs as a consequence of reinvesting such proceeds.

The Issuer and/or other member of the Group can incur additional debt.

The Terms of the Bonds (Section VIII) do not limit the amount of additional indebtedness that the Issuer and/or its subsidiaries can create, incur, assume or guarantee. We may create, incur, assume or guarantee additional indebtedness, and, to the extent Terms of the Bonds in Section VIII permit, such debt may be privileged over the Bonds. We have done so in relation to the 2017/22 Bonds several times.

Bondholders' anti-dilution protection is limited.

The Conversion Price at which the Bonds may be converted into Shares will be adjusted only in the situations and to the extent provided in the Terms of the Bonds (see, *e.g.*, Condition 6 of the Terms of the Bonds in Section VIII). There is no requirement that there must be an adjustment for every corporate or other event that may affect the value of the Conversion Rights. Events in respect of which no adjustment must be made may adversely affect the value of the Conversion Rights and the Bonds.

Bondholders have no protection against a falling Share price.

The Conversion Price at which the Bonds may be converted into Shares will not be subject to any downward adjustment if the market price of the Shares falls below the market price by reference to which the Conversion Price will be set. The market price of the Shares may never be higher than the Conversion Price during the term of the Bonds, and Bondholders may never be able to convert he Bonds into Shares at a profit.

Upon conversion of the Bonds, Bondholders may be subject to additional expenses or taxes.

Upon conversion of a Bond, expenses, taxes, stamp, issue, registration, documentary, transfer and other duties may be due by the Bondholders.

Bondholders have no shareholder rights prior to exercising their Conversion Rights.

An investor in the Bonds is not a shareholder of the Issuer. No Bondholder (in his capacity as such) has any right to participate in shareholders' meetings, any voting rights, rights to receive dividends or other

distributions or any other rights with respect to the Shares unless such Bondholder has exercised his or her Conversion Right and has been recorded in the share register of the Issuer as a shareholder with voting rights in relation to the Shares received upon conversion.

The Company has the right to make interest payments in the form of Shares.

The Company at its option can choose to make interest payments in the form of Shares. The number of Shares would be determined by dividing the interest amount by 90% of the share price at such time. The Bondholders do not have a choice as the form in which they receive the interest payments. Further, in case the Company pays interest in the form of Shares, concurrent sales of Shares by Bondholders could result in a decline of the Share price.

We expect that no active and liquid trading market for the Bonds will develop.

The Bonds are a new issue of securities for which there is no established trading market. Application will be made for listing of the Bonds on the SIX Swiss Exchange. We expect that no active and liquid trading market for the Bonds will develop. Even if such trading market should develop, it may not provide enough liquidity to allow a Bondholder to trade or sell the Bonds easily, or the Bonds may trade at unfavorable prices. Such trading market may also fail to continue throughout the term of the Bonds. The Company is not under an obligation to provide a bid or offer price for the Bonds. Therefore, Bondholders may not be able to sell the Bonds easily at prices reasonably acceptable to them, or at all, and potential investors should only invest in the Bonds if they can hold them until their Maturity Date.

We expect the market price of the Bonds to be volatile.

The market price of the Bonds will depend on many factors, including our ability to meet our obligations under the Bonds, fluctuations in the market price of the Shares, on which the market price of the Bonds is expected to partly depend, and price and volume fluctuations of bond markets in general, which have from time to time been substantial. The market price of the Bonds may drop significantly at any time during their term. The price at which the Bonds will trade will depend upon many factors within and outside of our control. Bondholders may have to sell their Bonds at a substantial discount from the original purchase price and may lose some or all of their initial investment.

Neither the Issuer nor the Bonds have a credit rating. Even if a credit rating agency were to rate the Bonds, such rating may not reflect all risks.

The Issuer does not have a credit rating and the Bonds are not expected to be rated. Even if a credit rating agency were to assign a credit rating to the Bonds, such rating may not reflect all risk factors that may affect the value of the Bonds. Further, a credit rating is not a recommendation to buy, sell or hold securities and may be revised, suspended or withdrawn at any time. Such actual or anticipated revision, suspension or withdrawal may adversely affect our financing costs or the market price and trading of the Bonds.

Investors in our securities may be exposed to exchange rate risks.

The settlement currency of the Bonds is the Swiss franc. Shares are and will be quoted in Swiss francs only, and future dividends, if any, will be denominated in Swiss francs. If the Swiss franc depreciates against a foreign currency that is the home currency of an investor in our securities, the value of such securities, expressed in such foreign currency, will decrease accordingly. Exchange rates between currencies are highly volatile. Exchange rate fluctuations between an investor's home currency and the Swiss franc may adversely affect investors who intend to convert the proceeds from the sale of securities or future dividends, if any, into their home currency. As a result, such investors may lose some or all of their initial investment.

The Company may not be able to issue Shares out of its conditional or authorized share capital and may be unable to deliver Shares to Bondholders upon conversion at all or on time.

As of the date of this prospectus, the Company's conditional share capital is insufficient to cover a conversion in full of all Bonds at the option of the Bondholders. Therefore, the Company needs to rely on its authorized share capital for such purpose and will need to issue a sufficient number of Shares from its authorized share capital to a subsidiary or to itself. Swiss law does not permit an authorized share capital to subsist, without being extended by the company's shareholders, for more than two years after it was approved or extended by the shareholders. If our current authorized share capital expires for any reason, the Company may not be able to source a sufficient number of Shares to cover a conversion in full of all Bonds at the option of the Bondholders. Even if the number of available Shares is sufficient, the Company may fail to deliver Shares upon conversion within two Business Days after delivery of the respective Conversion Notice, as would be required under the Terms of the Bonds set out in Section VIII.

In any event, the issuance of new Shares out of the authorized share capital is subject to a number of legal and regulatory requirements, including successful registration in the commercial register. The issuance of Shares by the Company out of its authorized capital may be blocked by the registrar or any shareholder, which may prevent or delay such Share issuance and may prevent the Company from fulfilling its obligations under the Bonds in a timely manner, or at all.

A majority or supermajority of Bondholders could modify the Terms of the Bonds (Section VIII) on behalf of all Bondholders.

Articles 1157 et seq. CO contain provisions for calling meetings of Bondholders to consider matters affecting their interests generally, including, without limitation, modifications of the Terms of the Bonds set out in Section VIII. These provisions permit defined majorities to bind all Bondholders including Bondholders who did not attend and vote at the relevant meeting and Bondholders who voted in a manner contrary to the majority. For example, if the meeting of the holders of the 2017/22 Bonds held on March 8, 2021, had approved the Issuer's proposal to restructure such bonds with the requisite majority of two-thirds of 2017/22 Bonds in circulation, the 2017-22 Bonds would have been mandatorily restructured with respect to all holders of such bonds.

Investors in our securities may suffer dilution as a result of further issuance of equity, conversions of our convertible instruments or further issuances of other securities convertible into equity.

We have issued various equity-linked instruments. In aggregate, as of April 30, 2021 and applying the reporting rules set forth in art. 120 et seq. FMIA, as applied by us, we had sale positions corresponding to 88.1% of our share capital registered in the commercial register. These include, without limitation, the following instruments:

- Under the Highbridge Facility (see "THE COMPANY AND ITS BUSINESS—Additional information on our business—Material agreements—Financing Arrangements—Equity-linked financing arrangement with Highbridge" for more information), based on the closing price of the Shares on the SIX Swiss Exchange on April 30, 2021, we would be required to issue, at a discount, an aggregate of up to 3,889,606 Shares. In addition, we have agreed to issue warrants to Highbridge, which will be exercisable for an aggregate of 984,769 Shares.
- We have the right to repay up to 65% of the CHF 10 million principal amount of the Idorsia Exchangeable Note in Shares at a discount. Based on the closing price of the Shares on the SIX Swiss Exchange on April 30, 2021, this would correspond to up to 3,071,108 Shares.
- We have issued various equity-linked rights under our employee participation plans and have made conditional share grants, and we may issue new such rights in the future. The number of Shares we would be required to issue or deliver under such plans depends on the market price of our Shares.
- Holders of the 2017/22 Convertible Bonds that will remain outstanding after completion of the Exchange Offer have the right to convert such bonds into Shares at a conversion price of CHF 64.80 per Share.

We may issue other rights to acquire Shares at prices significantly below the Offer Price in the future. In particular, we will need to raise additional equity or equity-linked financing in the immediate short-term in order to continue our operations as planned. The Company has authorized and conditional share capital, which we plan to use to cover our equity-linked financings, including our obligations to deliver Shares under the Bonds. We will likely propose to our shareholders to approve additional capital increases or authorizations therefor in the near future.

If and whenever a capital increase is implemented, existing holders of our securities will incur substantial dilution. Additional dilution may occur to holders of our securities if and to the extent our convertible instruments will be converted and such rights to acquire Shares will ultimately be exercised and settled in Shares. Moreover, to the extent that we issue additional shares or equity-linked instruments (*e.g.*, for financing purposes or for employee participations), investors' ownership interest will be further diluted, and the terms of such issued shares may include liquidation or other preferences that adversely affect investors' rights as a shareholder.

The Share price has been and is expected to be volatile, and investors may not be able to resell their Shares at or above the price they have paid for them.

The market price of the Shares has historically been subject to substantial fluctuations. We expect the market price of the Shares to continue to be highly volatile after the completion of the offering of the Bonds. Such volatility may depend upon many factors within and beyond our control, including the risk factors listed in this Prospectus, our or our competitors' financial and business performance, general market conditions and the volatility in financial and other markets (*i.e.*, the degree to which prices fluctuate over a particular period in a particular market, regardless of market levels) in general. In some cases, the markets have produced downward pressure on share prices for certain issuers seemingly without regard to those issuers' underlying financial strength. As a result, investors may not be able to resell their Shares at or above the price they have paid for them on the market or in connection with the conversion of the Bonds or other financial instruments.

The trading market for the Shares may be illiquid and shareholders may not trade or sell their Shares easily or at all. There currently is no market making regarding the Shares.

The volume of the trading market for the Shares on the SIX Swiss Exchange has historically been low and may be low in the future. Therefore, the trading market may not provide enough liquidity to allow shareholders to trade or sell their Shares easily or at all. The Company is not obliged to provide a bid or offer price for the Shares. Further, the Company's market making arrangement with Kepler Cheuvreux SA is currently inactive, and the Company does not plan to arrange for a market making in the future. There is no assurance that shareholders will be able to trade or sell their Shares easily or at all.

Future sales of a substantial number of Shares or derivative instruments by us or our investors could adversely affect the market price of our securities.

Sales, or the possibility or perceived possibility of sales, of a substantial number of Shares in the market may have had, and could in the future have a material adverse effect on the market price of the Shares and other securities. Our counterparty to the Highbridge Facility (see "*THE COMPANY AND ITS BUSINESS—Additional information on our business—Material agreements—Financing Arrangements—Equity-linked financing arrangement with Highbridge*" for more information) is managed by a hedge fund that, to our knowledge, sells a corresponding number of Shares in the market before or after it exchanges the exchangeable notes that we issue to to Highbridge from time to time. Holders of Bonds—including Highbridge, which held 32% of the principal value of the 2017/22 Bonds and exchanged such bonds for Bonds in the Exchange Offer—will have the right to convert the Bonds into Shares at any time before the maturity of the Bonds in 2024 and will be able to sell some or all of the Shares issued by the Company upon such conversion in the market or otherwise at any time. The current lock-up undertaking of Idorsia vis-à-vis us with respect to 1,000,000 Shares will terminate with the Company's consent or when we obtain FDA approval for sale of vamorolone in DMD in the U.S.

The Company may issue additional Shares out of its existing authorized share capital or may propose to its shareholders to approve additional capital increases, in each case excluding shareholders' preemptive rights. As a result of the respective issuances or sales of Shares, or if such issuances or sales are anticipated by investors, the market price of our securities could fall substantially.

We do not expect to pay dividends in the foreseeable future.

Since its inception, the Company has never paid any dividends and it does not anticipate paying dividends in the foreseeable future. Further, according to the terms of the Highbridge Facility, the members of the Group are not permitted to pay dividends. Investors cannot rely on dividend income from the Shares, and any returns on an investment in the Shares will likely depend entirely upon any future appreciation in the price of the Shares and the ability of investors to sell Shares in the market.

Shareholders outside Switzerland may not be able to exercise preemptive rights in future issuances of equity or other securities that are convertible into equity.

Under Swiss law, shareholders may have certain preemptive rights to subscribe on a pro rata basis for issuances of newly issued equity or other securities that are convertible into equity. Due to laws and regulations in their respective jurisdictions, non-Swiss shareholders may not be able to exercise such rights unless we take action to register or otherwise qualify the particular rights offering under the laws of that jurisdiction. There can be no assurance that we would take any such action, and we will have the full discretion to decide not to take such action in one or more jurisdictions, including the EU, the UK and the U.S. If shareholders in such jurisdictions are unable to exercise their subscription rights, their ownership interest in the Company would be diluted.

If securities or industry analysts do not publish research at all or publish inaccurate or unfavorable research about the Group's business, the market price and/or the trading volume of our securities could remain on a low level and even decline further.

The trading market for our listed securities depends in part on the research and reports that securities or industry analysts publish about the Group or its business. If no or few securities or industry analysts cover the Company, the market price for our securities could be adversely affected. If one or more of the analysts who cover the Group downgrades a recommendation with regard to the Shares, publishes inaccurate or unfavorable research about the Group's business, ceases to cover the Group or fails to publish reports on it regularly, the market price and/or the trading volume of the Shares and our other securities could decline.

Our largest investors, including our largest shareholders and our equity-linked financing provider and largest Bondholder, Highbridge, are able to exert influence over the Company, and their interests may not necessarily be the same as those of other investors.

As of the date of this Prospectus, our two largest shareholders, Idorsia and a group of shareholders around Mr. Ernesto Bertarelli, held an aggregate of 2,459,371 Shares according to their latest disclosure notices pursuant to art. 120 et seq. FMIA, corresponding to an aggregate of 9.4% of the voting rights in the Company registered in the commercial register. Any of our investors may start acting in concert, and any existing or new investors may acquire significant ownership interests in the Company in the future. Such shareholders or groups of shareholders may be able to exert influence over, and potentially block, certain matters that must be decided by the Company's general meeting of shareholders, in particular those matters that require the consent of two-thirds of voting rights represented. The influence of significant shareholders or groups of shareholders is accentuated by the low historic rates of participation at the Company's past three annual general meetings of shareholders, which were 32.0% in 2020, 36.8% in 2019 and 33.3% in 2018.

In addition, Highbridge, which will hold almost 43% of the principal value of the Bonds upon their issuance, will hold a blocking stake, and perhaps even a majority of votes, in some or all meetings of Bondholders that we may call in the future in order to modify the Terms of the Bonds. In addition, Highbridge will be able to request the calling of a meeting of Bondholders to declare an Event of Default under the

Terms of the Bonds set out in Section VIII. Highbridge is also our counterparty to the Highbridge Facility, on which we currently rely to obtain short-term funding.

The interests of influential investors, including of Highbridge, may not be the same as the interests of other investors in our securities, and such investors might be able to bring about or prevent corporate decisions that materially adversely affect the interests of other investors in our securities.

U.S. shareholders may not be able to obtain judgments or enforce civil liabilities against the Company or its directors or executive officers.

The Company is organized under the laws of Switzerland and has its seat in the Canton of Basel-Landschaft, Switzerland. Most of its assets are located outside the U.S. In addition, none of its directors and executive officers are residents of the U.S. and all of or a substantial portion of their assets are located outside the U.S. As a result, it may not be possible to enforce against the Company or such persons judgments of U.S. courts, including judgments in actions predicated upon the civil liability provisions of the federal securities laws of the U.S. We have been advised by our Swiss counsel that there is doubt as to the enforceability in Switzerland of original actions, or in actions for enforcement of judgments of U.S. courts, of civil liabilities to the extent predicated upon the federal and state securities laws of the U.S. Original actions against persons in Switzerland based solely upon the U.S. federal or state securities laws are governed, among other things, by the principles set forth in the Swiss Federal Act on International Private Law of December 18, 1987, as amended ("**PILA**"). This statute provides that the application of provisions of non-Swiss law by the courts in Switzerland shall be precluded if the result is incompatible with Swiss public policy. Also, mandatory provisions of Swiss law may be applicable regardless of any other law that would otherwise apply.

Switzerland and the U.S. do not have a treaty providing for reciprocal recognition and enforcement of judgments in civil and commercial matters. The recognition and enforcement of a judgment of the courts of the United States in Switzerland is governed by the principles set forth in the PILA. This statute provides in principle that a judgment rendered by a non-Swiss court may be enforced in Switzerland only if (i) the non-Swiss court had jurisdiction pursuant to the PILA; (ii) the judgment of such non-Swiss court has become final and non-appealable; (iii) the judgment does not contravene Swiss public policy; (iv) the court procedures and the service of documents leading to the judgment were in accordance with the due process of law; and (v) no proceeding involving the same position and the same subject matter was first brought in Switzerland, or adjudicated in Switzerland, or was earlier adjudicated in a third state and this decision is recognizable in Switzerland.

If the Company is classified as a passive foreign investment company for U.S. federal income tax purposes, U.S. investors that hold the Company's securities could be subject to potentially significant adverse tax consequences.

If the Company is classified as a passive foreign investment company ("**PFIC**") in any taxable year in which a U.S. Holder, holds Shares, such U.S. Holder may be subject to significant adverse tax consequences. For the purposes of this risk factor, a "**U.S. Holder**" is a beneficial owner of Shares that is for U.S. federal income tax purposes (i) an individual who is a citizen or resident of the United States, (ii) a corporation created in, or organized under the laws of, the United States or any state thereof, including the District of Columbia, (iii) an estate the income of which is includible in gross income for U.S. federal income tax purposes regardless of its source or (iv) a trust that is subject to U.S. tax on its worldwide income regardless of its source.

The Company has not conducted an analysis to determine whether it is or may be a PFIC for the current taxable year or any other taxable year. Therefore, there can be no assurance that it is not a PFIC or that it will not become a PFIC in the future. All U.S. Holders are therefore urged to consult their tax advisors regarding the consequences of owning Shares.

VIII. TERMS OF THE BONDS

The terms and conditions (each a "**Condition**", and together the "**Terms of the Bonds**") of the 7.50 per cent senior unsecured convertible bonds due 2024 (the "**Bonds**" and each a "**Bond**"), conferring a conversion right with reference to registered shares of Santhera Pharmaceuticals Holding AG, Hohenrainstrasse 24, 4133 Pratteln, Switzerland (the "**Issuer**") in the aggregate principal amount of Swiss Francs ("**CHF**") 30,270,375 are as follows.

The Terms of the Bonds govern the rights and obligations of the Issuer and of each holder of Bonds (a "**Bondholder**", collectively the "**Bondholders**") in relation to the Bonds (defined terms used herein have the meaning ascribed to them in Condition 18):

1. Denomination, Form and Delivery of the Bonds

(a) The aggregate principal amount of the Bonds of CHF 30,270,375 is divided into 8,969 Bonds with denominations of CHF 3,375 (three thousand three hundred and seventy five) each (the "**Principal Amount**").

(b) The Bonds and all rights in connection therewith are issued in uncertificated form in accordance with article 973c of the Swiss Code of Obligations of March 30, 1911, as amended (the "**CO**") as uncertificated securities (*Wertrechte*) that will be created by the Issuer by means of a registration in its register of uncertificated securities (*Wertrechtebuch*). Such uncertificated securities (*Wertrechte*) will then be entered into the main register (*Hauptregister*) of SIX SIS Ltd ("**SIS**") or any other intermediary in Switzerland recognized for such purposes by the Relevant Exchange (SIS or any such other intermediary, the "**Intermediary**"). Once the uncertificated securities (*Wertrechte*) are registered in the main register (*Hauptregister*) of the Intermediary and entered into the accounts of one or more participants of the Intermediary, the Bonds will constitute intermediated securities (*Bucheffekten*) (the "**Intermediated Securities**") in accordance with the provisions of the Swiss Intermediated Securities Act of 3 October 2008, as amended (*Bucheffektengesetz*) (the "**FISA**").

(c) So long as the Bonds are in the form of Intermediated Securities, the Bonds may only be transferred or otherwise disposed of in accordance with the provisions of the FISA.

(d) The records of the Intermediary will determine the number of Bonds held through each participant of the Intermediary. In respect of Bonds held in the form of Intermediated Securities, the Bondholders will be the persons holding the Bonds in a securities account (*Effektenkonto*) which is in their own name or, in the case of intermediaries (*Verwahrungsstellen*), the intermediaries holding the Bonds for their own account in a securities account (*Effektenkonto*) which is in their name.

(e) The conversion of the uncertificated securities (*Wertrechte*) into a permanent global certificate (*Globalurkunde auf Dauer*) or individually certificated bonds (*Wertpapiere*) is excluded. Neither the Issuer nor the Bondholders nor BKB as paying and conversion agent (the "**Paying and Conversion Agent**") nor any third party shall at any time have the right to effect or demand the conversion of the uncertificated securities (*Wertrechte*) into, or the delivery of a permanent global certificate (*Globalurkunde auf Dauer*) or individually certificated securities (*Wertpapiere*).

2. Interest

The Bonds bear interest at the rate of 7.50 per cent per annum of their Principal Amount. The interest is payable semi-annually in arrears on each Interest Amount Payment Date (the CHF amount in respect of each Bond so calculated being the "**Interest Amount**"). Interest on the Bonds is computed on a 30E/360 basis, i.e., on the basis of a year consisting of twelve (12) months of thirty (30) days each. The first interest payment on August 17, 2021 shall be CHF 126.5625 per Bond, as if the Bonds had been issued on February 17, 2021.

The Issuer may elect to pay all or part of any Interest Amount in the form of Shares. Notice of such election (the "**Election Notice**") shall be given (i) to the Bondholders in accordance with Condition 10 at least 5 Business Days before the date on which the respective Interest Amounts shall be paid, if interest payments to all Bondholders are concerned or (ii) directly to the Bondholder(s) concerned at least on the date on which the respective Interest Amounts shall be paid, if interest payments are concerned that are not made to all Bondholders (such as upon a conversion). In case of such election, each Bondholder is deemed to have exercised the right to convert the respective Interest Amounts into Shares in accordance with this Condition 2.

The Shares in respect to such an interest payment shall be delivered in ten (10) installments. On each of the ten (10) Trading Days following (but excluding) the date of the Election Notice, one tenth of the Interest Amount shall be divided by 90% of the VWAP of one Share on that day to determine the number of Shares deliverable in respect to that installment, with the resulting number of Shares being rounded down to the nearest whole number of Shares. Each of the ten (10) installments of Shares shall be delivered to the relevant Bondholders within two (2) Business Days after the date on which the number of Shares to be delivered in respect of the relevant installment was determined. The Cash Payment for Fractions, if any, shall be paid in respect to all ten (10) installments at the same time as the delivery of the last installment of Shares for the respective Interest Amount.

Each Bond will cease to bear interest from the due date for redemption or repayment of such Bond, provided that if, upon due presentation, delivery of the Shares or payment of any amount due is improperly withheld or refused, such Bond shall continue to bear interest as provided in these Terms of the Bonds. In such case, interest will accumulate until the day on which all Shares and/or all sums due in respect of such Bonds are received by the Paying and Conversion Agent on behalf of the relevant Bondholder.

3. Conversion

(a) Conversion Right, Conversion Ratio and Conversion Price

(i) Each Bond in the Principal Amount of CHF 3,375 (three thousand three hundred and seventy five Swiss Francs) will be convertible on any Business Day during the Conversion Period into Shares at the Conversion Ratio.

(ii) The Conversion Ratio will be determined by dividing CHF 3,375 (three thousand three hundred and seventy five Swiss Francs), the Principal Amount, by the Conversion Price prevailing on the Conversion Date. The number of Shares to be delivered upon conversion of one Bond shall be calculated to five decimal places, provided that if more than one Bond is converted at any one time by the same Holder, the number of Shares to be delivered upon conversion will be determined by dividing the aggregate Principal Amount of the Bonds converted by the same Bondholder at any one time by the Conversion Price prevailing at the Conversion Date, such number of Shares to be calculated to five decimal places.

(iii) Fractions of Shares will not be issued and delivered on conversion. Instead, cash payments in CHF based on the VWAP of a Share on the Relevant Exchange on the Trading Day immediately preceding the relevant Conversion Date will be made in respect thereof (the "**Cash Payment for Fractions**"), except where any individual entitlement would be less than CHF 10.00 (ten), in which case, no such payment shall be required to be made. If the resulting amount of CHF is not an integral multiple of CHF 0.01 (one hundredth of a Swiss Franc), it shall be rounded to the nearest whole or multiple of CHF 0.01 (one hundredth of a Swiss Franc) with 0.005 being rounded upwards.

(iv) A Conversion Right may not be exercised following the giving of a default notice pursuant to Condition 9 nor in respect of a Bond which has been redeemed pursuant to Conditions 5, 7 (d) or 9.

(v) Where a Conversion Right is exercised during a Change of Control Period, the provisions in Condition 7(c) shall apply.

(vi) In the case of a conversion by a Bondholder in accordance with this Condition 3 (a), the Issuer shall at the same time as and in addition to the delivery of the Shares, pay the converting Bondholder (i) the accrued interest on the relevant Bond up to the Conversion Date as well as (ii) an amount equal to the interest amount that would be payable on the relevant Bond for a period of the shorter of (a) three years from the relevant Conversion Date and (b) the time from the relevant Conversion Date to the Maturity Date (the "**Make-whole Amount**"). The right of the Issuer to elect to pay Interest Amounts in Shares in accordance with Condition 2 also applies to Make-whole Amounts, provided that the conversion price calculated under Condition 2 paragraph 3, i.e. by 90% of the VWAP of one Share on relevant day, shall in any event not be less than CHF 2.50.

(b) Conversion Procedures

(i) Conversion Notices

To exercise the right to convert all or any of its Bonds pursuant to this Condition 3, a Bondholder must deposit with the Paying and Conversion Agent at its own expense during the Conversion Period a duly completed notice of conversion (the "**Conversion Notice**") in a form satisfactory to the Paying and Conversion Agent together with clearing instructions in a form satisfactory to the Paying and Conversion Agent allowing for the transfer of the relevant Bond(s) through the Intermediary to the Paying and Conversion Agent at the Specified Office.

By depositing the Conversion Notice, a Bondholder is deemed to represent and warrant that (x) it understands that the Shares to be transferred upon conversion of the Bonds have not been and will not be registered under the U.S. Securities Act of 1933 (the "**Securities Act**") and (y) it is not a U.S. person (as defined in Regulation S under the Securities Act ("**Regulation S**")) and is located outside the United States within the meaning of Regulation S, is acquiring the Shares to be transferred upon conversion of the Bonds in an offshore transaction (as defined in Regulation S) in accordance with Rule 903 or 904 of Regulation S and understands that the Shares may not be delivered within the United States upon conversion of the Bonds and may not be resold in the United States except pursuant to an exemption from the registration requirements of the Securities Act.

A Conversion Notice, once duly completed and deposited as aforesaid, shall be irrevocable. Bonds duly presented and/or transferred for conversion shall be cancelled in their entirety by the Paying and Conversion Agent and, upon delivery of the relevant Shares and the payment of the Cash Payment for Fractions, if any, shall be considered redeemed.

The Conversion Right shall be exercised only in respect of the whole of the Principal Amount of a Bond.

A Conversion Notice shall be deemed to be presented on a Business Day if received in a form satisfactory to the Paying and Conversion Agent before 4.00 p.m. CET on that Business Day at the Specified Office. Any Conversion Notice presented after 4.00 p.m. CET will be deemed to have been received on the following Business Day.

The conversion date in respect of a Bond (the "**Conversion Date**") shall be the date on which a Conversion Notice has been received or deemed to have been received in accordance with this Condition 3(b)(i).

(ii) Delivery of Shares and Cash Payments for Fractions

The Shares to be delivered upon conversion of Bonds in accordance with this Condition 3, if any, will be Shares issued in an ordinary capital increase, from the conditional capital or authorised capital or existing Shares of the Issuer, in each case with the same entitlements as the other outstanding Shares, except that the Shares so delivered will not be entitled to any dividend or other distribution declared, paid or made by reference to a Record Date prior to the relevant Conversion Date and except that the voting rights may not be exercised unless the person designated in the Conversion Notice as recipient of the Shares is registered as the holder of the Shares with voting rights in the Issuer's share register.

The Issuer will (x) effect delivery of the Shares and (y) make Cash Payments for Fractions, if any, within two (2) Business Days after the Conversion Date through the Intermediary in accordance with directions given by the relevant Bondholder in the relevant Conversion Notice. At the time of such delivery of the Shares, the then valid share registration rules of the Issuer will apply; the Issuer does not offer any assurance or guarantee that the exercising Bondholder will be accepted as a shareholder with voting rights in its share register.

(iii) Taxes and other Costs

Any Swiss Federal Stamp Duty, if due, as well as the fee of the Relevant Exchange, if any, payable upon the delivery in Switzerland of Shares to the Bondholder upon the conversion of Bonds will be paid or reimbursed by the Issuer. The Issuer will, however, not pay (a) any tax payable in connection with any subsequent sale or transfer of Shares by the respective Bondholder, or (b) any tax or other cost payable in connection with the sale, transfer or delivery of Share(s) in or to a country other than Switzerland.

4. Payments

The amounts required for the payment of the Interest Amounts (after deduction of the then applicable Swiss withholding tax) and the Principal Amount and any other payments in cash to be made under these Terms of the Bonds will be made available in good time in freely disposable CHF, which will be placed at the free disposal of the Paying and Conversion Agent in Switzerland. If the due date for any payment by the Issuer does not fall on a Business Day, the Issuer undertakes to effect payment for value the Business

Day immediately following such due date and Bondholders will not be entitled to any additional sum in relation thereto.

Upon receipt of the funds in Switzerland, the Paying and Conversion Agent will arrange for payment to the Bondholders.

The Issuer undertakes that payments shall be made in freely disposable CHF without collection cost to the Bondholders, and, unless otherwise provided for by applicable law, without any restrictions and whatever the circumstances may be, irrespective of nationality, residence or domicile of the Bondholders and without requiring any affidavit or the fulfilment of any other formality.

The receipt by the Paying and Conversion Agent of funds in CHF in Switzerland from the Issuer shall release the Issuer from its obligations under the Bonds to the extent of the amounts received by the Paying and Conversion Agent.

5. Redemption and Purchase

(a) Repayment at Maturity Date

Unless previously converted, redeemed, or purchased and cancelled as provided below, the Issuer undertakes to repay the Bonds on the Maturity Date, without further notice, at the Principal Amount together with unpaid accrued interest to such date (such repayment of any Bond on the Maturity Date, as well as any early redemption in accordance with this Condition 5, with Condition 7 or with Condition 9, in these Terms of the Bonds being referred to as the "**Redemption**").

(b) Early Redemption at the Option of the Issuer

Subject to not less than thirty (30) nor more than sixty (60) calendar days' prior notice, the Issuer may redeem all but not only some of the Bonds outstanding at the Principal Amount (together with unpaid accrued interest, if any):

(i) at any time after the Issue Date and prior to the Maturity Date, if less than fifteen (15) per cent of the aggregate Principal Amount of the Bonds issued pursuant to the Terms of the Bonds are outstanding at the time of the notice; or

(ii) at any time, if the VWAP of a Share on the Relevant Exchange on each of at least twenty (20) out of thirty (30) consecutive Trading Days ending not earlier than five (5) Trading Days prior to the date the relevant notice of Redemption is given has been at least 150 per cent of the Conversion Price in effect on each such Trading Day, respectively.

A redemption notice according to this Condition 5 (b) does not prevent the Bondholders from converting Bonds in accordance with Condition 3.

(c) Early Redemption at the Option of the Bondholders in Case of Delisting of Shares

If the Shares are delisted from the Relevant Exchange without being listed on another Relevant Exchange, each Bondholder may, acting in accordance with this Condition 5(c), require the Issuer to redeem all or any of the Bonds held by such Bondholder at their Principal Amount (together with unpaid accrued interest) at the Relevant Put Date.

At the latest on the date the Shares are delisted from the Relevant Exchange, the Issuer shall give notice of that fact (the "**Notice of Delisting**").

To exercise its right pursuant to this paragraph c), the Bondholder must deposit at its own expense a duly completed and signed notice (a "**Put Notice**") in a form satisfactory to the Paying and Conversion Agent during the period starting on the date of the Notice of Delisting and ending sixty (60) calendar days thereafter. A Put Notice shall be irrevocable.

(d) Purchases

The Issuer and any of its Subsidiaries may at any time purchase Bonds at any price in the open market or otherwise. Any purchase shall be made in accordance with applicable laws or regulations, including applicable stock exchange regulations. Such Bonds may be held, resold or, at the option of the Issuer, surrendered to the Paying and Conversion Agent for cancellation in accordance with Condition 5(e) below.

Any Bonds while held by or on behalf of the Issuer or any of its Subsidiaries, shall not entitle their Bondholder to vote at any meetings of the Bondholders and shall not be deemed to be outstanding for the purposes of calculating quorums at meetings of the Bondholders.

(e) Cancellation

All Bonds which are converted, redeemed, or surrendered, shall forthwith be cancelled. All Bonds which are to be cancelled cannot be reissued or resold.

6. Adjustments to the Conversion Price

(a) Events leading to adjustments to the Conversion Price

(i) Increase of capital by means of capitalization of reserves, profits or premiums by distribution of Shares, or division or consolidation of Shares:

In the event of a change in the Issuer's share capital as a result of capitalization of reserves, profits or premiums, by means of the distribution of Shares, save for a distribution of Shares as a Dividend as set out in Condition 6(a)(iv) below, and in the event of division or consolidation of Shares, the Conversion Price shall be adjusted by multiplying the Conversion Price in force immediately prior to such change by the result of the following formula:

NOld/NNew

where:

NOld is the number of Shares existing before the change in share capital; and

NNew is the number of Shares existing after the change in share capital.

Such adjustment shall become effective on the date on which such Shares are distributed or, in the event of division or consolidation of Shares, on the first day the Shares are traded on the new basis on the Relevant Exchange.

(ii) Issues of Shares or Other Securities by way of conferring subscription or purchase rights:

If (a) the Issuer grants to holders of Shares any rights or options, warrants or other rights to subscribe for or acquire Shares, Other Securities or securities convertible or exchangeable into Shares or Other Securities, or (b) any third party with the agreement of the Issuer issues to holders of Shares any rights, options or warrants to purchase any Shares, Other Securities or securities convertible or exchangeable into Shares or Other Securities (the rights referred to in (a) and (b) collectively and individually being the "**Purchase Rights**"), the Conversion Price shall be adjusted by multiplying the Conversion Price in force immediately prior to such issue or grant by the result of the following formula:

(Pcum – R)/Pcum

where:

Pcum is the Current Market Price by reference to whichever is the later of (x) the date on which the Shares are first traded ex-Purchase Rights on the Relevant Exchange and (y) the Trading Day when the subscription or purchase price for Shares or Other Securities under the Purchase Right is announced, or, if the day the subscription or purchase price is announced is not a Trading Day, the next following Trading Day; and

R is the value of the Purchase Right(s) relating to one Share or Other Security, such value to be calculated as follows:

(A) in the event the Purchase Rights relate to Shares:

R = Pcum - TERP

where:

TERP = (Nold x Pcum + Nnew x (Prights + Div)) / (Nold + Nnew)
and:

TERP is the theoretical ex-Purchase Rights price; and

Nold is the number of Shares existing before the change in share capital; and

Nnew is the number of offered Shares contemplated to be newly issued; and

Prights is the price at which one new Share can be subscribed, exercised or purchased; and

Div is the amount (in CHF) by which the entitlement to Dividends per existing Share exceeds the entitlement to Dividends per new Share, (x) if Dividends have already been proposed to the general meeting of shareholders but not yet paid, based on the proposed amount of the Dividends, or (y) if Dividends have not yet been proposed, based on the Dividends paid in the immediately preceding financial year;

provided, however, that no such adjustment shall be made if the subscription or purchase price at which one new Share can be subscribed or purchased is at least ninety-five (95) per cent of the Current Market Price on whichever is the later of (x) the date on which the Shares are first traded ex-Purchase Rights on the Relevant Exchange or (y) the Trading Day when the subscription or purchase price for the Purchase Right is announced, or, if the day the subscription or purchase price is announced is not a Trading Day, the next following Trading Day;

(B) in the event the Purchase Rights relate to Other Securities or to securities convertible or exchangeable into Shares or Other Securities and where such Purchase Rights are traded on a regulated stock exchange in Switzerland, the European Union, the United Kingdom, the United States of America, Canada or Japan:

R = Nrights x Prights

where:

Nrights is the number of Purchase Rights granted per Share; and

Prights is the VWAP of the Purchase Rights on the Relevant Exchange (or, if no dealing is recorded, the arithmetic mean of the bid and offered prices) during the time Purchase Rights are traded, but not longer than the first ten (10) Trading Days.

(C) in all other cases where neither of the previous paragraphs (A) or (B) is applicable, R will be determined by a Common Expert.

Such adjustment shall become effective

i. in the case of Condition 6(a)(ii)(A), on the first day on which the Shares are traded ex-Purchase Rights on the Relevant Exchange;

ii. in the case of Condition 6(a)(ii)(B), five (5) Trading Days after (x) the end of the period during which the Purchase Rights are traded or (y) the tenth (10^{th}) Trading Day of the Purchase Rights, whichever is sooner; and

iii. in the case of Condition 6(a)(ii)(C), on the date determined by the Common Expert.

(iii) Spin-offs and capital distributions other than Dividends:

If, in respect of a spin-off or a capital distribution other than Dividends as referred to in Condition 6(a)(iv) below, the Issuer shall issue or distribute to holders of its Shares any assets, evidence of indebtedness of the Issuer, shares or other rights (other than as referred to in Condition 6(a)(ii) above) (the "**Distribution**"), the Conversion Price shall be adjusted as follows:

(A) In case the Distribution (x) consists of securities that will be traded on a regulated stock exchange in Switzerland, the European Union, the United Kingdom, the United States of America, Canada or Japan, (y) consists of securities that are traded on a regulated stock exchange in Switzerland, the European Union, the United Kingdom, the United States of America, Canada or Japan or (z) has otherwise a value which is determinable by reference to a stock exchange quotation or otherwise, by multiplying the Conversion Price in force immediately prior to such issue or distribution by the result of the following formula:

(Pcum – D)/Pcum

where:

Pcum is the Current Market Price by reference to the date on which the Shares are first traded ex-Distribution on the Relevant Exchange following the relevant Distribution; and

D is equal to (i) in case of iii)(A)(x), the current market price of the Distribution (in CHF) on the Relevant Exchange, calculated on a per Share basis, as determined by the Paying and Conversion Agent, or (ii) in case of iii)(A)(y), the current market price of the Distribution (in CHF) on the Relevant Exchange on the date by reference to which Pcum has been determined, calculated on a per Share basis, as determined by the Paying and Conversion Agent or (iii) case of iii)(A)(z), as determined by a Common Expert,

where for purposes of this provision, the current market price (to determine D) in case of iii)(A)(x) shall be deemed to be the average of the VWAPs on the five (5) consecutive Trading Days commencing on the date on which the Shares are first traded ex-Distribution on the Relevant Exchange, and in case of iii)(A)(y) shall be deemed to be the average of the VWAPs on the five (5) consecutive Trading Days ending on and including the Trading Day preceding the day on which the Shares are first traded ex-Distribution.

(B) In all other cases and where there is one (but not more than one) Distribution on a given Trading Day, by multiplying the Conversion Price in force immediately prior to such issue or distribution by the result of the following formula:

Pafter/Pbefore

where:

Pafter is the current market price per Share after the date of such Distribution (the "**Distribution Date**"); and

Pbefore is the current market price per Share before the Distribution Date;

whereby for purposes of this provision the current market price per Share shall be deemed to be the average of the VWAPs, (x) in the case of Pbefore, on the five (5) consecutive Trading Days before the Distribution Date, and (y) in the case of Pafter, on the five (5) consecutive Trading Days after the Distribution Date, as determined by the Paying and Conversion Agent. When calculating the average of the VWAPs the gross dividend amount (or any other entitlement), if any, of any dividend paid (or any other entitlement) during either of the above mentioned periods of five (5) consecutive Trading Days, shall be added back to the VWAPs on each of the Trading Days on which the Shares are traded ex-dividend (or any other entitlement).

(C) If the Issuer issues or distributes to its shareholders tradable put options as a Dividend with respect to any financial year, the Conversion Price shall be adjusted according to the formula set out in Condition 6(a)(iv).

(D) In all cases where there is more than one Distribution on a given Trading Day, the Common Expert will determine the necessary adjustment.

Such adjustment shall become effective, in the case of (A)(y), on the date on which the Distribution is made and, in the case of (A)(x) and (B), on the sixth (6^{th}) Trading Day after the Distribution Date and, in the case of (A)(z) and (D) as determined by a Common Expert.

(iv) Dividends:

If the Issuer pays a Dividend, the Conversion Price shall be adjusted by multiplying the Conversion Price in force immediately prior to such payment by the following fraction:

(Pcum – D)/Pcum

where:

Pcum is the Current Market Price with respect to the Effective Date; and

D is the Dividend attributable to one Share as set out below

Any reference to D in the above formula shall be a reference to

(A) the cash amount in case of a cash dividend or a repayment of paid-in capital;

(B) an amount as calculated by the following formula in case of a stock dividend in lieu of a cash dividend:

current market price - (current market price x (NOld/NNew))

where:

current market price is the average of the daily VWAP of one Share on each of the five (5) consecutive Trading Days ending on and including the Trading Day immediately prior to the Ex-Date;

NOld is the number of Shares existing before the change in share capital; and

NNew is the number of Shares existing after the change in share capital;

(C) an amount as calculated by the following formula in case of tradable put options in lieu of a cash dividend (the "**Put Options**"):

current market price x (P/N)

where:

current market price is the average of the daily VWAP of the Put Option on each of the five (5) consecutive Trading Days commencing on the Ex-Date;

P is the number of Put Options to be issued; and

N is the number of Shares existing prior to the Ex-Date.

Such adjustment shall become effective on the Ex-Date or, in case of Put Options according to (C) above, on the sixth (6^{th}) Trading Day following the Ex-Date.

(b) Calculation of Adjustments

(i) Each adjustment to be made pursuant to Condition 6(a) shall be calculated by the Paying and Conversion Agent and shall (in the absence of manifest error) be binding on all parties concerned. The Paying and Conversion Agent shall for the purpose of the foregoing provisions only be liable for making, or not making, adjustments or taking, or not taking, any other measures in connection with these Bonds, if and to the extent that it fails to act with due care according to established market practice. The Paying and Conversion Agent may engage the advice or services of any Common Expert whose advice or services it may consider necessary and rely upon any advice so obtained, and the Paying and Conversion Agent shall incur no liability as against the Issuer or the Bondholders in respect of any action taken, or not taken, or suffered to be taken, or not taken, in accordance with such advice and in exercising due care according to established market practice.

(ii) If in case of any adjustment the resulting Conversion Price is not an integral multiple of CHF 0.01 (one hundredth of a Swiss Franc), it shall be rounded to the

nearest whole or multiple of CHF 0.01 (one hundredth of a Swiss Franc) with 0.005 being rounded upwards.

(iii) The Issuer will procure that a notice is published in the manner described in Condition 10 as soon as practicable after either the date on which any adjustment to the Conversion Price becomes effective or, if no adjustment is required, the date on which it is possible to determine that such is the case.

(c) Retroactive Adjustments

If the Conversion Date in relation to any Bond is (i) before the relevant Record Date for any issue, sale, grant or offer leading to an adjustment pursuant to Condition 6(a)(ii) before publication of the event leading to such Record Date, and (iii) before the relevant adjustment to the Conversion Price becomes effective under Condition 6(a), and (iv) provided that the Shares will be delivered to the converting Bondholder after the Record Date, the Issuer shall (conditional upon the relevant adjustment becoming effective) procure that there shall be issued or paid to the converting Bondholder such an additional cash amount or number of Shares, if applicable (the "**Additional Consideration**") as, together with the cash amounts to be transferred and the Shares delivered or to be delivered, if any, on conversion of the relevant Bond is equal to the consideration (in form of cash amounts or Shares as set out in Condition 3(a)(ii) and (iii)) which would have been required to be delivered on conversion of such Bond if the relevant adjustment to the Conversion Price had in fact been made and become effective prior to the Conversion Date (the "**Retroactive Adjustment**").

Without prejudice to the provisions of Condition 3, upon a Retroactive Adjustment becoming effective in accordance with this Condition 6(c), the delivery of the relevant Additional Consideration shall be made within ten (10) Business Days after the first date it is possible to calculate such adjustment but not earlier than the Record Date. Without prejudice to the foregoing and to mandatory provisions of applicable law, in the event that an issue, sale, grant or offer leading to an adjustment pursuant to Condition 6(a) is effected between the above Conversion Date and the date of delivery of the relevant Additional Consideration, the Issuer shall request a Common Expert to determine the amount of the further consideration to be made to the converting Bondholder, whether in kind or in cash, so that the Bondholder may be substantially treated as if such Bondholder actually held the Additional Consideration on the Conversion Date.

(d) Events not giving rise to Adjustments

No adjustment to the Conversion Price will be made:

(i) as a result of any issue or distribution of Shares or Other Securities if the pre-emptive right (*Bezugsrecht*) in respect thereof under the CO has been validly excluded by resolution of the general meeting of shareholders or by the board of directors of the Issuer unless a preemptive right in respect thereof is granted indirectly to the shareholders by a third party with the agreement of the Issuer; or

(ii) as a result of any public issue of bonds convertible or exchangeable into Shares or bonds with options to subscribe for Shares, such issue being in connection with a conditional increase of the share capital of the Issuer, irrespective of whether in respect of such issue the advance subscription rights to acquire such bonds (*Vorwegzeichnungsrecht*) have been excluded or not, unless advance subscription rights have been granted to the shareholders of the Issuer and are traded on the Relevant Exchange; or

(iii) if Shares or Other Securities (including pre-emptive rights, options, warrants or stock appreciation rights in relation to Shares or Other Securities) are issued, offered or granted to, or for the benefit of, members of the board of directors, officers, employees or advisors of the Issuer or of any of its Subsidiaries or of any associated company or to trustees to be held for the benefit of any such person, in

any such case pursuant to any employee share or participation scheme of the Issuer or of any of its Subsidiaries; or

(iv) if an increase in the Conversion Price would result from such adjustment, except in case of an exchange of the Shares for Other Securities or a consolidation of Shares; or

(v) if the Conversion Price would fall below the nominal value of a Share. In this case, the Conversion Price will be adjusted to the nominal value of a Share and any remaining reduction of the Conversion Price resulting from such adjustment or from any further adjustment will be carried forward and only be applied if and to the extent the nominal value of a Share will be reduced.

(e) Other Events

If the Issuer determines, after consultation with the Paying and Conversion Agent, or the Paying and Conversion Agent determines after consultation with the Issuer, that notwithstanding Condition 6(a) and Condition 6(d) an adjustment should be made to the Conversion Price as a result of one or more events or circumstances not referred to in Condition 6(a) or circumstances including circumstances listed in Condition 6(d) have arisen which have an adverse effect on the right to convert Bonds and no adjustment to the Conversion Price under Condition 6(a) would otherwise arise or is excluded according to Condition 6(d), the Paying and Conversion Agent shall engage the advice or services of a Common Expert to determine as soon as practicable what adjustment, if any, to the Conversion Price or amendment, if any, to the terms of this Condition 6 is fair and reasonable to take account thereof and the date on which such adjustment should take effect. If several events occur which become effective on the same Trading Day and which would lead to an adjustment of the Conversion Price pursuant to Condition 6(a), the decision as to the manner of calculating the adjustment of the Conversion Price shall be taken by the Common Expert. The decision of the Common Expert shall be binding as set forth in Condition 18.16. The Paying and Conversion Agent shall have no responsibility to make any inquiries as to whether or not any event has occurred which might require an adjustment to the Conversion Price or amendment, if any, to the terms of this Condition 6.

(f) Correction of Adjustments

If an adjustment has been made in accordance with Condition 6(a) based on events or circumstances that subsequently are not implemented or are implemented in a manner materially different than anticipated when calculating the adjustment, then the Issuer and the Paying and Conversion Agent shall determine whether and to what extent the adjustment previously made shall be corrected. The Paying and Conversion Agent may engage the services of a Common Expert to determine whether and to what extent a correction shall be made. The decision of the Common Expert shall be binding. The Paying and Conversion Agent shall have no responsibility to make any inquiries as to whether or not any event has occurred which might require correction of an adjustment to the Conversion Price previously made.

7. Change of Control

(a) A "**Change of Control**" occurs when:

(i) an offer to acquire Shares, whether expressed as a public takeover offer, a merger or similar scheme with regard to such acquisition, or in any other way, is made in circumstances where (A) such offer is available to (aa) all holders of Shares, (bb) all holders of Shares other than the offeror and any persons acting in concert with such offeror, or (cc) all holders of Shares other than persons who are excluded from the offer by reason of being connected with one or more specific jurisdictions (or a combination of the exceptions pursuant to (bb) and (cc)), and (B) such offer having become or been declared unconditional with respect to acceptances,

the Issuer becomes aware that the right to cast more than fifty (50) per cent of all the voting rights (whether exercisable or not) of the Issuer has become or will become vested in the offeror and any persons acting in concert with the offeror; or

(ii) the Issuer consolidates with or merges into any other company, save where, following such consolidation or merger, shareholders of the Issuer immediately prior to such consolidation or merger have the right to cast at least fifty (50) per cent of the voting rights (whether exercisable or not) of such other company; or

(iii) the Issuer becomes aware that the right to cast more than fifty (50) per cent of all voting rights (whether exercisable or not) of the Issuer has become unconditionally vested directly or indirectly in any person (or in persons acting in concert with each other in respect of the exercise of such voting rights); or

(iv) the legal or beneficial ownership of all or substantially all of the assets owned directly or indirectly by the Issuer is acquired by one or more other persons (other than Subsidiaries).

(b) Upon a Change of Control, the Issuer shall give notice of the fact that a Change of Control occurred (the "**Change of Control Notice**") to the Bondholders no later than two (2) Trading Days after the occurrence of a Change of Control in the form set out in Condition 10. The Change of Control Notice shall:

(i) inform the Bondholders of their right to either require redemption of the Bonds pursuant to Condition 7(d) or, if applicable, exercise their Conversion Rights for a period of forty (40) Trading Days (the "**Change of Control Period**") starting on the Trading Day immediately following the date the Change of Control Notice is given, at the adjusted Conversion Price, as further described in Condition 7(c);

(ii) specify the date (the "**Change of Control Redemption Date**"), being not more than sixty (60) and not less than fifty-one (51) Trading Days after the date the Change of Control Notice is given, on which the Bonds may be redeemed at the option of the Bondholders pursuant to Condition 7(d);

(iii) if Condition 7(c) applies, specify the Conversion Price in effect immediately prior to the Change of Control and the adjusted Conversion Price applicable as a consequence of the Change of Control; and

(iv) provide details concerning the Change of Control.

(c) Adjustment of Conversion Price upon Change of Control

If a Change of Control occurs, the Conversion Price for Bonds converted on a Conversion Date falling within the Change of Control Period shall be adjusted as follows:

$$CPa = RP \times (1 + (CP \times (1 - c/t)))$$

where:

CPa Adjusted Conversion Price

RP Conversion Price prevailing five (5) calendar days before the Change of Control Notice is published, divided by (1 + CP);

CP Conversion premium of 15.0 per cent (expressed as a fraction);

cthe number of calendar days from and including the date on which the adjusted Conversion Price is applicable to but excluding the seventh (7^{th}) Trading Day prior to the Maturity Date; and

t the number of calendar days from and including the Issue Date to but excluding the seventh (7^{th}) Trading Day prior to the Maturity Date.

(d) Early Redemption at the Option of Bondholders upon Change of Control

Upon the occurrence of a Change of Control, the Issuer will, at the option of a Bondholder, redeem such Bondholders' Bond(s) on the Change of Control Redemption Date at their Principal Amount (together with unpaid accrued interest to such date). To exercise such option, a Bondholder must present, by not later than ten (10) Trading Days prior to the Change of Control Redemption Date, at the Specified Office a duly completed redemption notice in a form satisfactory to the Paying and Conversion Agent (a "**Change of Control Redemption Notice**"), together with clearing instructions in a form satisfactory to the Paying and Conversion Agent allowing for the transfer of the relevant Bond(s) through the Intermediary to the Paying and Conversion Agent. No Change of Control Redemption Notice so deposited may be withdrawn without the consent of the Issuer.

(e) Conversion after the Change of Control Redemption Date

With respect to the Bonds that remain outstanding after the Change of Control Redemption Date, in the case of a Change of Control as defined in Condition 7(a)(ii) and if the Issuer is not the surviving company, the Issuer shall use its commercially reasonable efforts to ensure that each Bond shall be convertible into such shares or other equity securities, including depositary receipts issued for the same and any other consideration (including cash) which such Bondholder would have received in the Change of Control transaction if such Bondholder had exercised its Conversion Rights immediately prior to the date of the Change of Control Notice (and then participated in the Change of Control transaction).

8. Status and Negative Pledge

(a) The Bonds constitute direct, unconditional, and (subject to Condition 8(b)) unsecured obligations of the Issuer and (subject to Condition 8(b)) rank and will rank *pari passu* among themselves and with all other unsecured and unsubordinated obligations of the Issuer, except for such preferences as are provided for by any mandatorily applicable provision of law.

(b) So long as any Bonds remain outstanding, the Issuer will not, and will procure that no Material Subsidiary will, create or have outstanding any mortgage, charge, pledge, lien or other form of encumbrance or security interest upon the whole or any part of its business, property, assets or revenues, present or future, to secure any Relevant Debt or to secure any guarantee or indemnity in respect of any Relevant Debt unless, at the same time or prior thereto, the Issuer's obligations under the Bonds (i) are secured equally and rateably therewith by such encumbrance or security interest or benefit from a guarantee or indemnity in substantially identical terms thereto, as the case may be or, (ii) have the benefit of such other security, guarantee, indemnity or other arrangement as shall be approved by the Bondholder Representative in its discretion.

For the purposes of this Condition, "**Relevant Debt**" means (i) any present or future indebtedness of the Issuer and its Subsidiaries represented or evidenced by notes, bonds, debentures or other securities which are for the time being, or are capable of being, quoted, listed or ordinarily dealt with on any stock exchange, over-the-counter-market or other securities market and (ii) any financial debt (including, for the avoidance of doubt, bank debt) of the Issuer and its Subsidiaries at any time outstanding exceeding CHF 10 million in the aggregate, other than (x) financial debt secured through or by receivables, and (y) any debt with a security interest over or affecting any asset or property of any company which becomes a Subsidiary after the Issue Date, where the security interest is created prior to the date on which that company becomes a Subsidiary (and with such security interest being limited to encumbering the assets or property acquired).

9. Event of Default

Each of (a) the Paying and Conversion Agent (in its capacity as initial bondholder representative) or any other Swiss bank appointed by the general meeting of Bondholders as bondholder representative in its place (the "**Bondholder Representative**") or (b) the general meeting of Bondholders by majority decision on the basis of the votes represented at the meeting (the "**Majority Bondholders**") has/have the right but not the obligation, on behalf of the Bondholders, to declare all Bonds to be immediately due and repayable at the Principal Amount (together with unpaid accrued interest to such date and any other amounts payable under the Bonds), by serving a written notice of default (the "**Default Notice**") upon the Issuer which shall have that effect, but only in case of the occurrence of any of the following events (each an "**Event of Default**"):

(a) there is a failure by the Issuer (i) to pay the Interest Amount or the Principal Amount or any other amount payable under the Bonds when due, or (ii) to deliver Shares and/or to make Cash Payments for Fractions, if and when due, upon conversion of a Bond; and such failure continues for a period of ten (10) Business Days in case of any payment of cash or ten (10) Trading Days in case of delivery of Shares; or

(b) a default is made by the Issuer in the performance or observance of any material covenant, condition or provision contained in the Terms of the Bonds which is to be performed or observed on its part, the Bondholder Representative or the Majority Bondholders consider(s) such default to be materially prejudicial to the interests of the Bondholders, and such default continues for a period of thirty (30) calendar days following the service by the Bondholder Representative or the Majority Bondholders on the Issuer of a notice requiring such default to be remedied; or

(c) any other present or future indebtedness of the Issuer, of Santhera Pharmaceuticals (Schweiz) AG or of any Material Subsidiary for or in respect of Moneys Borrowed from third parties (i.e., excluding any transactions between the Issuer and one or several of its Subsidiaries or among Subsidiaries) is not paid when due or, as the case may be, within any applicable grace period, or becomes due and payable prior to its stated maturity as a result of an event of default (howsoever described), or any security in respect of any such indebtedness becomes enforceable or any guarantee of, or indemnity in respect of, any such indebtedness given by the Issuer or any Material Subsidiary is not honoured when due and called upon or, as the case may be, within any applicable grace period, provided that no such event shall be taken into account for the purposes of this paragraph c) unless such indebtedness, either alone or when aggregated with other indebtedness subject to an event set out in this paragraph c) which has occurred and is continuing is equal to or exceeds an amount of CHF 3,000,000 (or its equivalent in another currency); or

(d) the Issuer or any Material Subsidiary is (or is deemed by law or a court to be) insolvent or bankrupt or unable to pay its debts, stops or suspends payment of all or a material part of its debts, or proposes or applies for a stay of execution; or

(e) a postponement of payments (*Stillhaltevereinbarung*), a general assignment or an arrangement or composition with or for the benefit of the relevant creditors in respect of any such debts or a moratorium or postponement of payments is agreed or declared in respect of or affecting all or any part of (or of a particular type of) the debts of the Issuer or any Material Subsidiary; or

(f) the Issuer or one or more Material Subsidiaries alters its or their legal or commercial structure through bankruptcy, liquidation, or disposal of its or their assets, the Issuer changes the objects of the company or its commercial activities or merges with a third party (other than the Issuer or any of its Subsidiaries) and such merger does not constitute a Change of Control, in so far as the relevant action in each of the above scenarios has or may have a material adverse effect on the capacity of the Issuer to meet its obligations in connection with the Bonds then or in the future, unless in the sole opinion of the Bondholder Representative the situation of the Bondholders as a consequence of the security

provided or other steps taken by the Issuer provide adequate protection to the Bondholders; or

(g) a dissolution or merger involving the Issuer as a result of which the Issuer is not the surviving company, unless the successor company assumes all the Issuer's liabilities.

The Issuer shall inform the Bondholder Representative without delay that any event mentioned under paragraphs a) through g) has occurred and provide the Bondholder Representative with all necessary documents. The Issuer accepts responsibility for the information contained in those documents.

Upon the occurrence or the potential occurrence of an Event of Default, the Bondholder Representative may (and if requested by Bondholders holding at least 25% of the aggregate Principal Amount of all outstanding Bonds, the Bondholder Representative shall without delay) invite the Bondholders in accordance with art. 1157 et seq. CO to a Bondholders' meeting for the taking of a resolution on the serving of a Default Notice, provided the Bondholder Representative has not served such Default Notice itself. The legally valid resolution of the Bondholders' meeting to serve a Default Notice, shall replace the right reserved by the Bondholder Representative according to these Terms of the Bonds to serve a Default Notice on behalf of the Bondholders. If the Bondholders' meeting votes against the serving of a Default Notice, the right to serve such Default Notice shall revert to the Bondholder Representative whereby the Bondholder Representative shall not be bound by the resolution of the Bondholders' meeting if and to the extent that new circumstances arise or become known which require a revised assessment of the facts.

For the purpose of this Condition 9, "**Moneys Borrowed**" means indebtedness, either alone or in aggregate, evidenced by bonds, notes, debentures or other securities, tradable on exchanges or over the counter, or bank loans or other interest bearing indebtedness of any type whatsoever.

10. Notices

All notices to Bondholders regarding the Bonds (the "**Notices**") shall be published by the Paying and Conversion Agent on behalf of, and in accordance with directions by and at the expense of the Issuer in due time in a daily newspaper nationally circulated in Switzerland, expected to be the Neue Zürcher Zeitung.

Upon the first Trading Day of the Bonds on the Relevant Exchange and for as long as the Bonds are admitted to trading or listed on the Relevant Exchange, all Notices shall be validly published according to the then applicable rules of the Relevant Exchange, in case of SIX Swiss Exchange currently electronically on its internet website (currently: www.six-group.com/en/products-services/the-swiss-stock-exchange/market-data/news-tools/official-notices.html .html), save as otherwise required by law, replacing the publication in a daily newspaper.

11. Listing

The Issuer will use its reasonable efforts to have the Bonds listed on the SIX Swiss Exchange and to maintain such listing until the Maturity Date or in case of an early redemption of the Bonds to the date of the early redemption. The Issuer will use all reasonable efforts to have the Shares listed and to maintain a listing for all the issued Shares on the SIX Swiss Exchange or any other Relevant Exchange.

12. Statute of Limitations

Claims for payment of the Principal Amount and for Cash Payments for Fractions, respectively, cease to be enforceable by legal action in accordance with the applicable Swiss statute of limitations (presently after ten (10) years from their relevant due dates for payment). Claims for pay-

ments of Interest Amounts cease to be enforceable by legal action in accordance with the applicable Swiss statute of limitations (presently after five (5) years from their relevant due dates for payment).

13. Governing Law and Jurisdiction

The Bonds and these Terms of the Bonds shall in every respect (including without limitation questions of form, content and interpretation) be subject to and governed by substantive Swiss law.

Any dispute arising out of or in connection with the Bonds or these Terms of the Bonds shall be submitted to the exclusive jurisdiction of the courts of the City of Zurich (Zurich 1), Switzerland.

The Issuer shall be discharged by and to the extent of any payment or delivery of Shares made in respect of any Bonds to a person recognized as a creditor by an enforceable judgement of a Swiss court.

14. Amendment to these Terms

The Terms of the Bonds may be amended from time to time by agreement between the Issuer and the Bondholder Representative, acting on behalf of and with effect for all present and future Bondholders, provided that in the sole opinion of the Bondholder Representative such amendment is of a formal, minor or technical nature, is made to correct a manifest error or is not materially prejudicial to the interests of the Bondholders.

Notice of any such amendment shall be published in accordance with Condition 10 above.

Any such amendment shall be binding on the Issuer and the Bondholders in accordance with its terms.

15. Role of BKB

BKB acts as Listing Agent, will act as Paying and Conversion Agent of this Bond issue and will or may also act as Bondholder Representative, but only in the cases stated explicitly in these Terms of the Bonds. In any other cases, BKB is not obliged to take or to consider any actions on behalf or for the benefit of the Bondholders.

16. Severability

If at any time any one or more of the provisions of the Terms of the Bonds is or becomes unlawful, invalid, illegal or unenforceable in any respect under any law, the validity, legality and enforceability of the remaining provisions shall not be in any way affected or impaired thereby.

17. Further Issues

The Issuer reserves the right to issue from time to time further bonds with identical terms as set out in these Terms of the Bonds without the consent of the Bondholders. In such a case such further issue shall form a single series with the then outstanding Bonds and the term "Bonds" shall comprise such additionally issued Bonds.

18. Definitions

1 "**Additional Consideration**" has the meaning given to it in Condition 6(c);

2 "**BKB**" means Basler Kantonalbank;

3 "**Bond(s)**" has the meaning given to it in the preamble;

4 "**Bondholder(s)**" has the meaning given to it in the preamble;

5 "**Bondholder Representative**" has the meaning given to it in Condition 9;

6 "**Business Day**" means any day (other than Saturday or Sunday) on which banks in Zurich are open for the whole day for business;

7 "**Cash Payment for Fractions**" has the meaning given to it in Condition 3(a)(iii);

8 "**Change of Control**" has the meaning given to it in Condition 7(a);

9 "**Change of Control Notice**" has the meaning given to it in Condition 7(b);

10 "**Change of Control Period**" has the meaning given to it in Condition 7(b)(i);

11 "**Change of Control Redemption Date**" has the meaning given to it in Condition 7(b)(ii);

12 "**Change of Control Redemption Notice**" has the meaning given to it in Condition 7(d);

13 "**CHF**" has the meaning given to it in the preamble;

14 "**CO**" has the meaning given to it in Condition 1(b);

15 "**Common Expert**" means an independent investment bank of international repute or an independent law firm or accounting firm of international repute or an independent financial advisor with relevant expertise of international repute (an "**Expert**") selected and instructed by the Issuer and the Paying and Conversion Agent by mutual agreement. If the Issuer and the Paying and Conversion Agent do not mutually agree on an Expert within seven (7) days from the beginning of the appointment process, each of the Issuer and the Paying and Conversion Agent shall select an Expert, whereby the so elected Experts shall select together a third Expert. In case the two selected Experts do not mutually agree on a third Expert within seven (7) days after being appointed, each of them shall select another Expert, whereby a Swiss Notary Public appointed by the Paying and Conversion Agent will pick one of these two Experts as third Expert by drawing lots. In the case of the appointment of three Experts references in the Terms of the Bonds to a Common Expert shall be deemed to refer to these three Experts, deciding by majority decision. Decisions of the Common Expert shall be final and binding on the Issuer, the Bondholders and the Paying and Conversion Agent. The Paying and Conversion Agent shall incur no liability against the Issuer or the Bondholders in respect of any action taken, or suffered to be taken, in accordance with such decision and in good faith. The fees and costs of the Common Expert shall be borne by the Issuer;

16 "**Condition**" has the meaning given to it in the preamble;

17 "**Conversion Date**" has the meaning given to it in Condition 3(b)(i);

18 "**Conversion Notice**" has the meaning given to it in Condition 3(b)(i);

19 "**Conversion Period**" means the period during which a Bondholder may exercise the Conversion Rights at his option, such period commencing on May 4, 2021 up to and including the earlier of (i) seven (7) Trading Days before the Maturity Date or (ii) in case of early redemption of the Bonds pursuant to Condition 5(b) ten (10) Trading Days prior to the date fixed for early redemption;

20 "**Conversion Price**" means CHF 3.0029, subject to adjustments in accordance with Condition 6 or 7(c);

21 "**Conversion Ratio**" has the meaning given to it in Condition 3;

22 "**Conversion Right**" means the right of a Bondholder to request conversion of any Bond in accordance with the provisions of these Terms of the Bonds;

23 "**Current Market Price**" means the average (mean) of the daily VWAP of one Share on each of the five (5) consecutive Trading Days ending on (and including) the Trading Day immediately preceding the date by reference to which such average is calculated, provided that when calculating the average (mean) of the VWAPs the gross dividend amount (or any other entitlement), if any, of any dividend (or any other entitlement) paid during either of the above mentioned period of five (5) consecutive Trading Days, shall be added back to the VWAPs on each of the Trading Days on which the Shares are traded ex-dividend (or any other entitlement);

24 "**Distribution**" has the meaning given to it in Condition 6(a)(iii);

25 "**Distribution Date**" has the meaning given to it in Condition 6(a)(iii)(B);

26 "**Dividend**" means a distribution per Share made by the Issuer to holders of the Shares at any time as (i) a cash dividend, (ii) a repayment of paid-in capital, (iii) a stock dividend in lieu of a cash dividend, or (iv) tradable put options in lieu of a cash dividend;

27 "**Effective Date**" means the last date on which the Shares are traded cum–dividend on the Relevant Exchange or, in the case of a purchase, redemption or buy back of Shares or any depositary or other receipts or certificates representing Shares, the date on which such purchase, redemption or buy back is made or in the case of a spin-off, the last date on which the Shares are traded cum–the relevant spin-off on the Relevant Exchange;

28 "**Event of Default**" has the meaning given to it in Condition 9;

29 "**Ex-Date**" means the first day on which the Shares are traded on the Relevant Exchange without entitlement (ex);

30 "**Expert**" shall have the meaning given to it in the definition of Common Expert;

31 "**FISA**" has the meaning given to it in Condition 1(b);

32 "**Interest Amount**" has the meaning given to it in Condition 2;

33 "**Interest Amount Payment Date**" means February 17 and August 17 in each year, for the first time on August 17, 2021;

34 "**Intermediary**" has the meaning given to it in Condition 1(b);

35 "**Intermediated Securities**" has the meaning given to it in Condition 1(b);

36 "**Issue Date**" means May 4, 2021

37 "**Issuer**" has the meaning given to it in the preamble;

38 "**Listing Agent**" means BKB, appointed as recognized representative pursuant to article 58a of the listing rules of SIX Swiss Exchange to file the listing application (including the application for provisional admission to trading) for the Bonds with SIX Swiss Exchange;

39 "**Material Subsidiary**" means any operating Subsidiary of the Issuer whose assets or net sales at any time represent ten (10) per cent or more of the consolidated assets, the consolidated net sales, the consolidated operating result or net result (profit after tax or net loss), as the case may be, of the Issuer and its consolidated Subsidiaries at any time, and for this purpose:

(a) the assets, net sales, the operating result and the net result (profit after tax or net loss) of any such Subsidiary shall be ascertained by reference to:

(i) the financial statement information of such Subsidiary, provided to the Issuer and adjusted for consolidation purposes (and thereby in particular excluding mere inter-company transactions) under generally accepted accounting principles applied by the Issuer, at the date to which the last audited consolidated annual financial statements of the Issuer and its consolidated Subsidiaries have been prepared;

(ii) if such corporate body becomes a Subsidiary of the Issuer after that date, the latest financial statements of such Subsidiary adjusted to take into account subsequent acquisitions and disposals or other changes in circumstances and adjustments required for consolidation purposes under generally accepted accounting principles applied by the Issuer;

(b) the consolidated assets, consolidated net sales, consolidated operating result and net result (profit after tax or net loss) of the Issuer shall be ascertained by reference to the last audited consolidated annual financial statements of the Issuer and its consolidated Subsidiaries; and

(c) once a subsidiary has become a Material Subsidiary, it shall be considered one until it has been demonstrated to the reasonable satisfaction of the Paying and Conversion Agent that it has ceased to be a Material Subsidiary, a written report from the Issuer's auditors to this effect being sufficient for this purpose;

40 "**Maturity Date**" means 17 August 2024;

41 "**Notices**" has the meaning given to it in Condition 10;

42 "**Notice of Delisting**" has the meaning given to it in Condition 5(c);

43 "**Other Securities**" means equity securities of the Issuer other than Shares;

44 "**Paying and Conversion Agent**" has the meaning given to it in Condition 1(e);

45 "**Principal Amount**" has the meaning given to it in Condition 1(a);

46 "**Purchase Rights**" has the meaning given to it in Condition 6(a)(ii);

47 "**Put Notice**" has the meaning given to it in Condition 5(c);

48 "**Put Options**" has the meaning given to it in Condition 6(a)(iv)(C);

49 "**Record Date**" means the last Business Day prior to the Ex-Date;

50 "**Redemption**" has the meaning given to it in Condition 5(a);

51 "**Regulation S**" has the meaning given to it in Condition 3(b)(i);

52 "**Relevant Debt**" has the meaning given to it in Condition 8(b);

53 "**Relevant Exchange**" means (i) in the case of Shares, the SIX Swiss Exchange or any successor thereof or, if the Shares are no longer admitted to trading on the SIX Swiss Exchange, the principal stock exchange or securities market on which the Shares are traded, and (ii) in the case of other securities, the principal stock exchange or securities market on which such other securities are traded;

54 "**Relevant Put Date**" means the fourteenth (14^{th}) day after the expiry of the period of sixty (60) days referred to in Condition 5(c). If such a due date does not fall on a Business Day, the Relevant Put Date shall be on the Business Day immediately following such due date;

55 "**Retroactive Adjustment**" has the meaning given to it in Condition 6(c);

60

56 "**Securities Act**" has the meaning given to it in Condition 3(b)(i);

57 "**Shares**" means issued and fully paid registered shares of currently CHF 1.00 (one Swiss Franc) par value each of the Issuer, or any other shares or stock resulting from any subdivision, consolidation or reclassification of such shares, which as between themselves have no preference in respect of dividends or of amounts payable in the event of any voluntary or involuntary liquidation or dissolution of the Issuer;

58 "**SIS**" has the meaning given to it in Condition 1(b);

59 "**SIX Swiss Exchange**" means SIX Swiss Exchange Ltd (or any successor to SIX Swiss Exchange Ltd), or the Swiss stock exchange operated by that company, as the context requires;

60 "**Specified Office**" means Basler Kantonalbank, Aeschenvorstadt 41, 4002 Basel, Switzerland

61 "**Subsidiary**" of the Issuer means a company the financial statements of which are, in accordance with applicable law or generally accepted accounting principles, fully consolidated with those of the Issuer;

62 "**Swiss Federal Stamp Duty**" means (a) the transfer stamp duty that may become due on the transfer of securities if a transfer is made by or through a Swiss securities dealer (*Effektenhändler*) within the meaning of the Swiss Stamp Duty Act of 27 June 1973, as amended (*Bundesgesetz über die Stempelabgaben*) and (b) the capital issuance stamp duty becoming due upon the issuance of any new Shares by the Issuer;

63 "**Terms of the Bonds**" has the meaning given to it in the preamble;

64 "**Trading Day(s)**" means any day (other than a Saturday or Sunday) on which (i) the Relevant Exchange is open for business and Shares may be dealt in or (ii) (if the Shares are not listed or admitted to trading on the Relevant Exchange) closing bid and offered prices are furnished for the Shares; and

65 "**VWAP**" means with respect to any Trading Day the volume-weighted average price of one Share (or other relevant security) published by Bloomberg Page HP (setting Weighted Average Line) or, if there is none, by such other source as shall be determined to be appropriate by the Common Expert on such Trading Day, provided that on any such Trading Day on which such price is not available or cannot otherwise be determined as provided above, the volume weighted average price of a Share (or other relevant security) in respect of such Trading Day shall be the VWAP, determined as provided above, on the immediately preceding Trading Day on which the same can be so determined.

IX. THE COMPANY AND ITS BUSINESS

References herein to "our" product candidates include product candidates that we have in-licensed or for which we have an option to in-license, unless otherwise stated or the context requires otherwise.

A. Business overview

We are a specialty pharmaceutical company, listed on the SIX Swiss Exchange, focused on the development and commercialization of innovative medicines for rare neuromuscular and pulmonary diseases with high unmet medical need. Our portfolio comprises the following:

Vamorolone. We acquired in 2020 a worldwide exclusive sub-licensable license to develop, commercialize and manufacture vamorolone, a novel anti-inflammatory drug candidate developed by ReveraGen BioPharma, Inc. ("**ReveraGen**"), for all indications. ReveraGen is currently conducting a pivotal Phase IIb clinical trial of vamorolone (the "**VISION-DMD Trial**") for the treatment of Duchenne muscular dystrophy ("**DMD**"), which is one of the most common types of inherited degenerative muscle weakness. With symptoms starting at young age, patients commonly are unable to walk by their teenage years and require mechanical ventilation to prolong survival beyond their late teenage years. Vamorolone initially targets early stage DMD patients who can still walk and aims at preserving upper and lower limb muscle strength and at delaying the time when they will need a wheelchair. With vamorolone, we have the ambition to replace steroids as standard of care in DMD patients due to a more favorable safety profile and comparable efficacy. Further, we believe that vamorolone has the potential to treat certain other inflammatory diseases with high unmet medical need beyond neuromuscular diseases.

We expect top-line data from the VISION-DMD Trial after six months of treatment to be available in the second quarter of 2021. If the U.S. Food and Drug Administration (the "**FDA**") supports the filing of a new drug application ("**NDA**") for vamorolone in DMD based on six-month or twelve-month top-line data of the VISION-DMD Trial and if all our other preparatory works for an NDA filing are completed according to plan, we currently expect to file such NDA in the first quarter of 2022, following which a decision of the FDA could be expected in the fourth quarter of 2022, with a potential launch of vamorolone in the U.S.shortly thereafter. In the EU, we may be able to file a marketing authorization application ("**MAA**") with respect to vamorolone in DMD in the second quarter of 2022 if the VISION-DMD Trial is completed successfully within the currently anticipated timeframe.

Lonodelestat. Our clinical stage pipeline also includes lonodelestat (formerly known as POL6014) to treat cystic fibrosis ("**CF**"). CF is a rare, life-threatening, progressive genetic disease that is typically diagnosed in young children and affects primarily the lungs but also the digestive system. In February 2018, we in-licensed lonodelestat from Polyphor Ltd, Allschwil, Switzerland ("**Polyphor**"). We believe that lonodelestat has the potential to treat CF and other lung diseases associated with increased neutrophil elastase activity. Based on prior development work by Polyphor, we recently completed a phase Ib multiple ascending dose (MAD) trial (the "**MAD Trial**") of orally inhaled multiple doses of lonodelestat in patients with CF, which showed a good tolerability of lonodelestat and no serious patient-reported adverse events across all doses and over all treatment durations.

Gene therapy approach to treat LAMA2-MD. We have established preclinical collaborations to explore the opportunities of a potential novel gene therapy approach for the treatment of so-called laminin-alpha 2 (LAMA2)-deficient congenital muscular dystrophy ("**LAMA2-MD**", also known as "MDC1A"), a form of inherited and progressive muscle weakness with symptoms starting in newborns or infants.

Raxone®. We also have an approved product, Raxone® for the treatment of Leber's hereditary optic neuropathy ("**LHON**") a rare, severe hereditary eye disease affecting primarily men in their 20s and 30s and leading to central vision loss in both eyes. We outlicensed our rights in Raxone®, including relating to development, commercialization and distribution, to Chiesi Group in August 2019.

We are managed by a capable, experienced and professional team. Our Chief Executive Officer, Dario Eklund, has a strong commercial focus and is in our view ideally suited to lead us into the future. All members of our Executive Management have comprehensive pharmaceutical industry experience.

Santhera was founded in September 2004. The Shares have been listed on the SIX Swiss Exchange since November 2006 and are included in the SXI Life Sciences® and SXI Bio+Medtech® indices of the SIX Swiss Exchange. Our headquarters are located in Pratteln, Switzerland, with subsidiaries in several European countries and the U.S. As of March 1, 2021, our headcount was 55 employees (52.4 full-time equivalent).

B. Pipeline and strategy

We are focused on the development and commercialization of innovative medicines for rare neuromuscular and pulmonary diseases with high unmet medical need. Our goal is to become a market leader in the development and commercialization of products for treatment of rare neuromuscular and pulmonary diseases.

We believe that our key strengths are:

— our dedication to rare neuromuscular and pulmonary diseases;

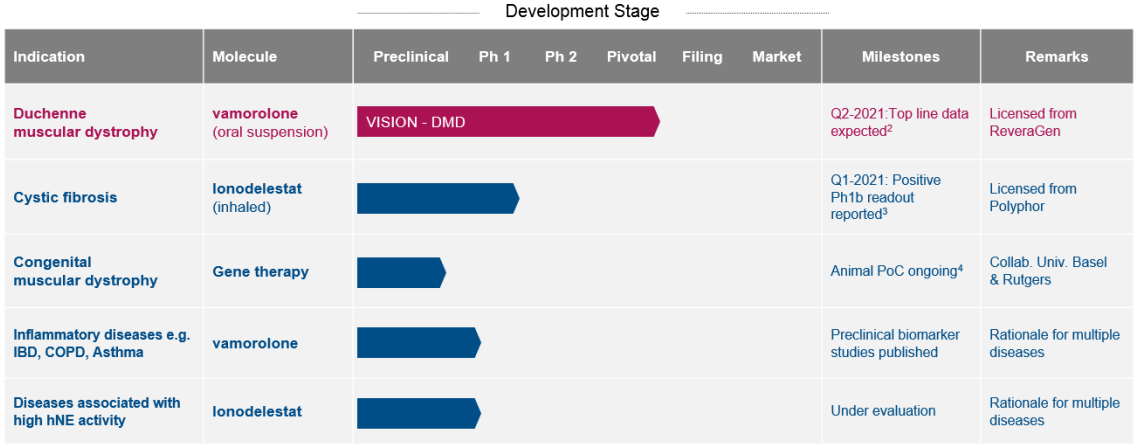
— our compound vamorolone (in-licensed from ReveraGen), which, being a dissociative steroid, is a new class of molecule that aims at having the structural properties required for its desired clinical efficacy, but only limited structural properties which may cause side effects or safety concerns observed with glucocorticoids;

— the advanced clinical development program (conducted by ReveraGen and supported by us) with regard to vamorolone for the treatment of DMD, which we believe has the potential to provide an alternative to treatment of DMD patients with standard glucocorticoids, thereby potentially addressing what we believe to be an attractive market for us; and

— our early stage product candidate lonodelestat, which we currently develop for the treatment of CF an in our view has the potential to treat other diseases characterized by high human neutrophil elastase (hNE) activity, as well as preclinical collaborations to develop a gene therapy approach to treat congenital muscular dystrophies ("**CMD**").

The following graph summarizes the status of our pipeline, which we regularly refocus and reprioritize:

Figure 1: Our current development pipeline[(1)]



IBD: Inflammatory Bowel Disease; COPD: Chronic Obstructive Pulmonary Disease
hNE: Human Neutrophil Elastase; Lonodelestat was formerly known as POL6014
PoC: Proof of Concept

(1) This chart sets out our current pipeline in summary form. For further details and the associated risks, please refer to the information set out elsewhere in this Prospectus, including in this section "*The Company and its Business*" and in "*Risk Factors*". The anticipated milestones set forth in this chart and in this prospectus are subject to further future adjustment based on, among other factors, the impact of the COVID-19 pandemic.

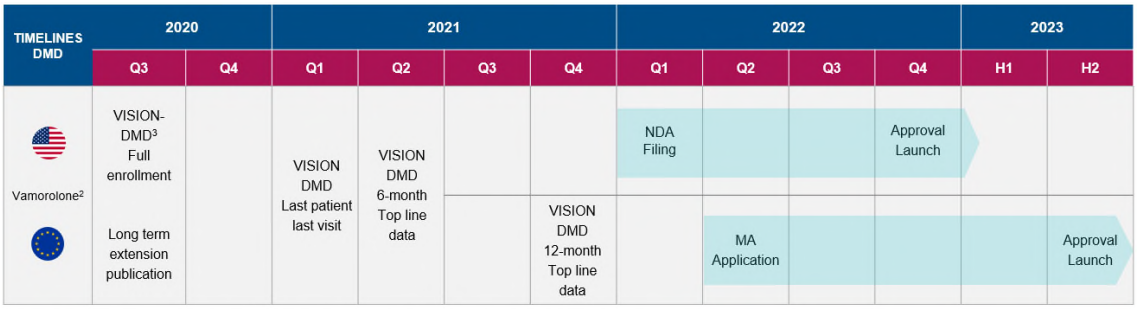
(2) We currently expect six-month top-line data from the VISION-DMD Trial to be available in the second quarter of 2021 and twelve-month top-line data in the fourth quarter of 2021.

(3) We recently completed a phase Ib multiple ascending dose (MAD) trial of lonodelestat in cystic fibrosis.

(4) We have established preclinicals collaborations with the University of Basel's Center for Molecular Life Sciences (*Biozentrum*) and with Rutgers, The State University of New Jersey, to explore the opportunities of a potential novel gene therapy approach for the treatment of LAMA2-MD.

The following graph summarizes our current view of the timeline in relation to the development and marketing authorization process with regard to vamorolone in DMD:

Figure 2: Our current timeline for vamorolone in DMD(1)



NDA: New Drug Application with the FDA; MA: Marketing Authorization

(1) This graph sets out our current timeline in summary form and only with respect to the product candidate indicated. For further detail and the associated risks, please refer to the information set out elsewhere in this Prospectus, including in this section "THE COMPANY AND ITS BUSINESS" and in "*RISK FACTORS*".

(2) We in-licensed vamorolone from Reveragen in September 2020.

(3) ReveraGen is currently conducting a pivotal Phase IIb clinical trial of vamorolone in DMD (VISION-DMD), whose six-month top-line data we currently expect to be available in the second quarter of 2021 and whose twelve-month top-line data we currently expect to be available in the fourth quarter of 2021. If the FDA supports the filing of an NDA based on six-month or twelve-month top-line data of the VISION-DMD Trial and if all our other preparatory works for an NDA filing are completed according to plan, we currently expect to file such NDA in the first quarter of 2022, following which a decision of the FDA could be expected in the fourth quarter of 2022, with a potential launch of vamorolone in the U.S. shortly thereafter. In the EU, we may be able to file an MAA after completion of twelve months of treatment within the VISION-DMD Trial if the trial's endpoints are met. If the VISION-DMD Trial is completed successfully within the currently anticipated timeframe, we could file such MAA in the second quarter of 2022, following which a decision on the MAA could be expected in the second half of 2023.

Our strategy includes the following key elements:

Offer novel treatment options for DMD patients independent of the specific genetic background of the disease

With vamorolone, which we have in-licensed from ReveraGen, we have the ambition to replace steroids as standard of care in DMD patients, typically young boys, who can still walk, aiming to preserve upper and lower limb muscle strength and to delay the time when they will need a wheelchair. If the FDA supports the filing of an NDA based on six-month or twelve-month top-line data of the VISION-DMD Trial and if all our other preparatory works for an NDA filing are completed according to plan, we currently expect to file such NDA in the first quarter of 2022, following which a decision of the FDA could be expected in the fourth quarter of 2022, with a potential launch of vamorolone in the U.S.shortly thereafter. In the EU, we may be able to file an MAA with respect to vamorolone in DMD in the second quarter of 2022 if the VISION-DMD trial is completed successfully within the currently anticipated timeframe.

Commercialize vamorolone, if and when marketing authorization is granted

We intend to build the commercial infrastructure in the U.S., and potentially major EU Markets or other territories where we decide that is necessary to support the commercialization of vamorolone if and when we believe a marketing authorization in a particular geographic market appears imminent. We are operating in the rare disease area in which patients typically form local, national and global patient groups that are well informed concerning the latest treatment possibilities. Because patients are typically treated at a limited number of expert reference centers, physicians can frequently be located and targeted efficiently by a small and highly skilled in-house team or by distribution partners.

Leverage vamorolone into other indications

We believe that vamorolone has the potential to treat certain other inflammatory diseases with high unmet medical need beyond neuromuscular diseases. Third parties have obtained preclinical data with vamorolone in vitro and in vivo models for asthma, multiple sclerosis, inflammatory bowel disease, rheumatoid arthritis, dysferlin muscular dystrophy, critical illness muscle disease, and brain tumor. In some of these diseases, the prescription of standard glucocorticoids is limited due to detrimental side-effects.

We plan to explore the use of vamorolone in select indications beyond DMD. For instance, we are planning to develop additional formulations of vamorolone that would allow for differential dosing and pricing in indications other than DMD as well as the potential for new intellectual property to be developed.

Advance our early-stage pipeline

We intend to advance our early-stage pipeline. Based on prior development work by Polyphor, we recently completed the phase Ib MAD Trial of lonodelestat in patients with CF, which showed a good tolerability of lonodelestat and no serious patient-reported adverse events across all doses and over all treatment durations. Further, we plan to advance our preclinical collaborations to explore the opportunities of a potential novel gene therapy approach for the treatment of LAMA2-MD.

Actively manage our portfolio as a potential additional source of future income

We intend to actively manage our portfolio of product candidates (and, if and when approved, products) as an additional source of future non-dilutive income streams. For instance, our worldwide exclusive sublicensable license to develop, commercialize and manufacture vamorolone for all indications in our view lends itself to increased partnering opportunities for further expansion outside the U.S. and the EU as well as with respect to indications other than DMD, for the benefit of patients worldwide. Similarly, we are currently seeking opportunities for outlicensing agreements for lonodelestat in pulmonary indications other than CF.

C. Our lead product candidate: vamorolone for the treatment of DMD

1. Duchenne muscular dystrophy (DMD)

The most advanced product candidate in our pipeline, vamorolone, which we have in-licensed from ReveraGen, targets DMD. DMD is one of the most common and devastating types of muscular degeneration. It is an inherited condition and primarily affects boys starting at an age between three and five years on average. DMD is characterized by a loss of a protein called dystrophin, which links the muscle cytoskeleton and extracellular matrix to maintain muscle integrity. This results in progressive muscle weakness, loss of muscle tissue and early illness and death due to cardio-respiratory failure. Patients are commonly unable to walk by their teenage years. Progressive respiratory muscle weakness leads to a need for mechanical ventilation to prolong the life of the patient beyond the late teenage years.

2. Market opportunity

Based on third-party studies, we estimate the prevalence (number of cases during a particular period or at a particular point in time) of DMD to be close to four per 100,000 individuals worldwide. Based on population figures, we estimate the number of DMD patients in the U.S. and the EU and the UK combined to be approximately 30,000-35,000. Third party studies have projected the worldwide overall market for all DMD drugs to be in excess of USD 4 billion by 2023.

Vamorolone initially targets a subgroup of DMD patients, namely young children with DMD who can still walk, and aims at preserving upper and lower limb muscle strength and at delaying the time when they will need a wheelchair. We see an opportunity that some DMD patients who would otherwise use glucocorticoids (a form of steroids) would use vamorolone rather than glucocorticoids, and also that some patients who would not use glucocorticoids may start using vamorolone for a certain period of time. Although it is generally difficult to estimate the size of the market for pharmaceutical products, we believe that the peak sales potential of vamorolone to treat DMD in the U.S., the EU and the UK combined could exceed USD 500 million if around one third of all DMD patients used vamorolone at any given point in time.

Generally, we believe that the market for DMD treatments has characteristics which may be favorable for us. There is a well-defined standard of care using glucocorticoids as the most common chronic treatment. Patients are routinely diagnosed at an early age and are thus accessible. There is a limited number of specialized centers for DMD treatment of around 300-400 in the U.S., the four largest EU members states and the UK combined. Also, we believe that the patient advocacy groups in the DMD field are well organized and influential. We believe there is a need for DMD treatments which are independent of the genetic background of the disease and for all disease stages. In particular, exon-skipping drugs address only limited patiens subpopulations and may potentially be used in combination with vamorolone in DMD patients. Last but not least, the only steroid that is, to our knowledge, approved for the treatment of DMD in the US, namely EMFLAZA™, achieves margins that we view as attractive.

3. Treatment options and competing therapeutic approaches

Steroid-based approaches

Currently, glucocorticoids (a form of steroids) are the standard of care in the U.S. and Europe for the treatment of DMD patients who are still able to walk. Glucocorticoids are prescribed in order to slow the decline in muscle strength and function caused by DMD regardless of the genetic mutation underlying DMD. However, the long-term use of steroids is hindered by their well-known side effects profile, which limits the long-term use of steroids. A recent third-party study showed that up to 42% of DMD patients aged 10 years or older had either never been treated with steroids or have discontinued their use. The use of steroids is often discontinued at some stage of disease progression. Loss of respiratory function with increasing age continues to be a major cause of illness and death in patients with DMD whether or not they are treated with steroids.

The current mainstays of medical therapy are prednisone and deflazacort. Prednisone is widely prescribed off-label to DMD patients and is recommended for use in standard of care guidelines for DMD. EMFLAZA™, whose active ingredient is deflazacort, is, to our knowledge, the first and only steroid that is explicitly approved for the treatment of DMD, but only in the U.S. EMFLAZA™ is currently marketed in the U.S. by PTC Therapeutics, Inc., South Plainfield, New Jersey, USA ("**PTC Therapeutics**").

Deflazacort is an oxazoline derivative of prednisolone (into which prednisone is transformed in the human body). Preliminary data from third-party studies indicate that deflazacort may show a lower risk of osteoporosis and growth rate retardation compared to prednisone. In Europe, physicians still prescribe prednisone and deflazacort on an off-label basis to treat DMD patients.

Both prednisone and deflazacort are associated with significant side effects, including obesity, delayed puberty, hypertension, cataracts, glaucoma, and emotional lability.

Non-steroid based approaches

To our knowledge, the most active company developing non-steroid based DMD therapies is Sarepta Therapeutics, Cambridge, Massachusetts, USA ("**Sarepta**"). To our knowledge, five such treatments for DMD, three developed by Sarepta, one by NS Pharma, Inc., Paramus, New Jersey, USA ("**NS Pharma**"), a subsidiary of Nippon Shinyaku, and one by PTC Therapeutics, have been approved to date. The most advanced approaches to treat DMD are directed against the genetic cause of the disease and typically address a subset of patients with certain genetic characteristics and who are still able to walk. In total, we are aware of several dozen clinical and preclinical programs to develop non-steroid based DMD treatments, some of which are at a late stage of clinical development and some of which we consider to be direct competitors with vamorolone.

In most of the DMD cases, the disease is caused by deletions in the gene that encodes dystrophin in the cell nucleus. EXONDYS 51™ (eteplirsen, SRP-4051), an oligonucleotide drug (i.e., a short chain of nucleotides) developed by Sarepta Therapeutics, Cambridge, Massachusetts, USA ("**Sarepta**"), targets a subgroup of these patients who have a mutation that is amenable to so-called exon 51 skipping. Clinical trials conducted by Sarepta have shown that eteplirsen may allow the expression of a truncated version of dystrophin. Studies by third parties estimate that eteplirsen has the potential to benefit 13% of patients with DMD. Sarepta has been marketing EXONDYS 51™ in the U.S. since 2016. Sarepta has also filed an MAA for EXONDYS 51™ with the EMA for the EU, on which it received a negative opinion from the CHMP in May 2018 and a negative decision on the re-examination procedure in September 2018.

In December 2019, the FDA granted Sarepta marketing authorization for a second oligonucleotide drug, VYONDYS 53™ (golodirsen, SRP-4053), which targets a subgroup of DMD patients who have a mutation that is amenable to so-called exon 53 skipping. In August 2020, the FDA granted NS Pharma, marketing authorization for Viltespo™, another exon skipping oligonucleotide drug (viltolarsen) that addresses the same patient population. NS Pharma has also received conditional marketing authorization for Viltespo™ in Japan.

In June 2020, Sarepta filed an NDA with regard to AMONDYS 45™ (casimersen, SRP-4045), another exon-skipping drug which targets a subgroup of DMD patients who have a mutation that is amenable to so-called exon 45 skipping. The FDA has granted marketing authorization for the product on a priority review basis in February 2021.

Sarepta is also developing experimental RNA-targeted treatments for DMD patients with the same gene mutation. Its most advanced program apears to be SRP-5051, targeting DMD patients who have a mutation that is amendable to exon 51 skipping. Sarepta is currently conducting a phase II clinical trial of SRP-5051 and has announced positive interim results in December 2020.

In December 2019, Roche announced that it had entered into an agreement with Sarepta providing Roche exclusive commercial rights to SRP-9001, Sarepta's investigational micro-dystrophin gene therapy candidate for DMD, outside the U.S. for an upfront consideration of USD 1.15 billion plus certain milestone and royalty payments. As part of the agreement, Roche also obtained an option to acquire rights outside the U.S. to certain future DMD-specific programs from Sarepta. Sarepta is currently conducting phase II clinical trials of SRP-9001. In January 2021, Sarepta announced that one of such trials showed a treatment effect with a subgroup of patients but did not reach its primary endpoint.

So-called nonsense point mutations account for an additional 13% of DMD cases. These mutations in the gene that encodes dystrophin lead to a premature stop signal that prevents the complete translation of the affected gene and thus the expression of dystrophin. This group of mutations is targeted by Translarna™, a small molecule drug developed by PTC Therapeutics that it believes to increase readthrough at the premature stop signals to enable the production of the full-length dystrophin. PTC Therapeutics obtained a conditional marketing authorization for Translarna™ (ataluren) for the treatment of DMD in certain patients with such nonsense point mutation in the EU in 2014, which has subsequently been renewed, and PTC Therapeutics has been selling Translarna™ in the EU. Translarna™ is also approved in certain other jurisdictions outside the U.S. and the EU. We believe that PTC Therapeutics may file an NDA with respect to Translarna™ in the U.S. soon.

Several therapeutic approaches seek to enhance muscle regeneration by inhibiting fibrosis in DMD patients. Italfarmaco S.p.a., Cinisello Balsamo, Italy, is currently conducting a phase III clinical trial of givinostat, which targets a novel pathway to inhibit fibrosis in DMD patients using corticosteroids. The estimated study completion date is March 2022. FibroGen, Inc., San Francisco, USA, is currently recruiting patients for two phase III clinical trials of pamrevlumab, an antibody that inhibits the activity of a growth factor contributing to fibrosis, in DMD patients using corticosteroids who are still able to walk and who have lost their ability to walk, respectively. The estimated completion date of the phase III clinical trials of pamrevlumab in DMD patients September 2022 and March 2023, respectively. We believe givinostat and pamrevlumab could be add-ons to treatment with vamorolone.

The University Hospital of Basel, Switzerland, is currently conducting a phase III clinical trial of tamoxifen, a compound widely used to treat breast cancer, in DMD patients using corticosteroids who are still able to walk and in DMD patients not using corticosteroids who have lost their ability to walk. The estimated study completion date is September 2022. We believe tamoxifen to be a potential direct competitor to vamorolone in DMD.

Antisense Therapeutics Limited, Toorak, Victoria, Australia, announced in May 2020 that it had completed an open-label phase IIa clinical trial of ATL1102 in DMD patients who have lost their ability to walk and that it was planning to conduct a phase IIb clinical trial of ATL1102 in DMD patients. ATL1102 has been shown to block the expression of a certain antigen on immune cells in the blood that are associated with inflammation that exacerbates the damage of muscle fibres in DMD patients and may also have a disease modifying impact on muscle dystrophin loss. We believe ATL-1102 to be a potential direct competitor to vamorolone in DMD.

Pfizer Inc., New York City, USA ("**Pfizer**"), is currently conducting a phase III clinical trial of PF-06939926, an investigational gene therapy to carry a shortened version of the human dystrophin gene into patients' muscle tissue, in DMD patients aged 4 to 7 years who use corticosteroids. Pfizer has received fast track, orphan drug and a rare pediatric disease designation for PF-06939926 from the FDA.

4. Vamorolone in DMD

In September 2020, we acquired a worldwide exclusive license from ReveraGen to commercialize vamorolone for all indications. For a summary of our license arrangement see "*—Additional information on our business—Material agreements—License and collaboration agreements—License Agreement with* " beginning on page 80.

a. Potential clinical benefits of vamorolone

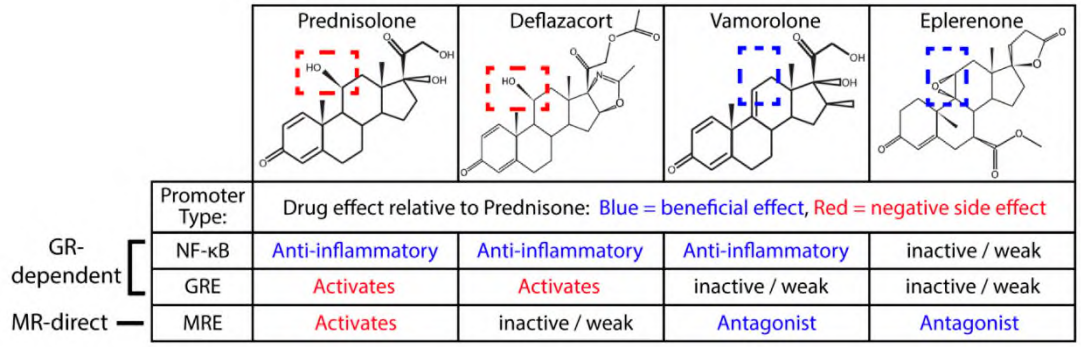
Glucocorticoids, the form of steroids currently used to treat certain DMD patients, while binding to their corresponding receptors, regulate many physiological processes, including metabolism, skeletal growth, cardiovascular activity, and immune response. They function as highly effective anti-inflammatory agents, which also have immune-suppressive activity. However, their long-term use as medication is associated with a series of clinically relevant side effects (e.g., stunted growth, hormonal imbalance, immunosuppression) that often limit their use and reduce patients' quality of life considerably. On the molecular level, glucocorticoids have many different mechanisms of action, comprising among others transcriptional repression or activation, physicochemical effects on cell membranes, cross-talk with other steroidal hormone receptors, tissue remodeling and synchronization of cell division. But each activity responsible for a desired treatment response is often associated with side effects, which sometimes counterbalance or even exceed its benefit.

Vamorolone (VBP15) has been developed by ReveraGen and is characterized as a dissociative steroid, i.e., it aims at having the structural properties required for its desired clinical action, but only limited structural properties which may cause side effects or safety concerns. We believe that vamorolone is a first-in-class multi-functional anti-inflammatory drug candidate that has shown in pre-clinical studies conducted by third parties potent inhibition (so-called trans-repression) of pro-inflammatory signaling pathways via the transcription factor NF-κB (so-called NF-κB pathway) through high-affinity binding to the glucocorticoid receptor. Other described activities such as muscle membrane stabilization properties,

and high affinity antagonism for the mineralocorticoid receptor may add to the favorable pharmacological profile of vamorolone.

On the structural level, vamorolone is similar to but distinct from prednisone, and they both show comparable powerful anti-inflammatory properties. Third-party pre-clinical studies have shown that like other glucocorticoids vamorolone induces trans-repression, thus retaining anti-inflammatory properties, but on the other hand through subtle modification to the steroidal backbone minimizes trans-activation, which is primarily responsible for undesirable side effects of glucocorticoid drugs. Figure 3 provides an overview of the differentiating pharmacological profiles for vamorolone compared to the standard glucocorticoids prednisolone (into which prednisone is transformed in the human body) and deflazacort as well as to one mineralocorticoid receptor antagonist, eplerenone.

Figure 3: Molecular structure of vamorolone as compared to prednisone



GR: glucocorticoid receptor; GRE: GR response element, MR: mineralocorticoid receptor, MRE: MR response element

Source: https://doi.org/10.26508/lsa.201800186

Developed with these properties, we believe that vamorolone has the potential for being a replacement for long-term glucocorticoid treatment of DMD patients and patients with other chronic inflammatory conditions. While retaining the anti-inflammatory efficacy via trans-repression of the NF-κB pathway and improving disease-related symptoms, ReveraGen and its collaborators have shown in multiple pre-clinical mouse models that vamorolone has the potential of an improved safety profile without key steroid side effects (e.g. stunted growth, hormonal imbalance, immunosuppression).

In mouse models of DMD (and of other conditions) conducted by ReveraGen and its collaborators, vamorolone has shown efficacy with respect to muscle strength and membrane stabilization (two key efficacy aspects in DMD therapy) similar or superior to prednisone. These in vivo pre-clinical studies also showed improvement of bone metabolism of vamorolone when benchmarked against prednisone tested in parallel, including loss of stunting of growth and osteopenia.

The first-in-human safety data from ReveraGen's phase I program VBP15-001 (see next subsection) indicate that vamorolone was safe to the highest doses tested (20.0 mg/kg/day; or about 10-15 times higher than typical glucocorticoid doses in DMD) and therefore suggest an improved safety profile relative to existing glucocorticoid drugs. Using multiple serum biomarkers bridged to later clinical safety concerns, such program has shown that vamorolone has lost key safety concerns regarding bone turnover, insulin resistance, and immunosuppression.

b. Clinical development

The clinical development program of vamorolone has been and is being conducted by ReveraGen and has been subject to, and is in agreement with, scientific advice by U.S. and EU regulators.

The phase I program (VBP15-001) consisted of a randomized, double-blind, placebo controlled single ascending dose (SAD) and multiple ascending dose (MAD) study to evaluate the safety, tolerability, and

pharmacokinetics of vamorolone in healthy adults. A total of 86 healthy adult males were enrolled, with single ascending doses (0.1–20.0 mg/kg), and multiple ascending doses (1.0–20 mg/kg/day; 14 days treatment) of vamorolone. The program showed that vamorolone was well-tolerated at all dose levels and that it showed pharmacokinetic and metabolism profiles similar to prednisone. Biomarker studies showed loss of side effects of traditional glucocorticoid drugs (bone fragility, metabolic disturbance, immune suppression) and that suppression of the adrenal axis was 10-fold less than prednisone. The researchers concluded that vamorolone retains high affinity binding and nuclear translocation of both glucocorticoid (agonist) and mineralocorticoid (antagonist) receptors, but does not show pharmacodynamic safety concerns of existing glucocorticoid drugs at up to 20 mg/kg/day.

VBP15-002 was a 2-week, open label phase IIa multiple ascending dose trial (0.25, 0.75, 2.0, and 6.0 mg/kg/day) with 48 boys with DMD aged four to seven years enrolled, with outcomes including clinical safety, pharmacokinetics and pharmacodynamic biomarkers. Twelve patients were enrolled in each of the vamorolone dose groups. The study design included pharmacodynamic biomarkers in three contexts of use: (1) secondary outcomes for pharmacodynamic safety (insulin resistance, adrenal suppression, bone turnover); (2) exploratory outcomes for drug mechanism of action; and (3) exploratory outcomes for expanded pharmacodynamic safety. The trial showed that vamorolone was safe and well-tolerated through the highest dose tested (6.0mg/kg/day) and pharmacokinetics of vamorolone were similar to prednisolone. Using pharmacodynamic biomarkers, the trial demonstrated improved safety of vamorolone versus glucocorticoids as shown by reduction of insulin resistance, beneficial changes in bone turnover, and a reduction in adrenal suppression. Exploratory biomarkers of pharmacodynamic efficacy showed an anti-inflammatory mechanism of action and a beneficial effect on plasma membrane stability, as demonstrated by a dose-responsive decrease in serum creatine kinase activity. All study participants completed the VBP15-002 trial and were invited to enroll in a 24-week phase IIa extension trial (VBP15-003) at the same dose level.

VBP15-003, a Phase IIa-extension to VBP15-002, was a 24-week open-label, multiple-dose extension study assessing the efficacy and safety of vamorolone in 48 boys with DMD aged four to seven years, 46 of whom completed the trial. The trial showed that oral administration of vamorolone at all doses tested was safe and well tolerated over the 24-week treatment period. The 2.0 mg/kg/day dose group met the primary efficacy outcome of improved muscle function (time to stand; 24 weeks of vamorolone treatment vs. natural history controls), without evidence of most adverse effects of glucocorticoids. A biomarker of bone formation, osteocalcin, increased in vamorolone-treated boys, which suggests that vamorolone does not have adverse effects on bone turnover to the same extent as glucocorticoids. Biomarker outcomes for adrenal suppression and insulin resistance were also lower in vamorolone-treated patients with DMD relative to published studies of glucocorticoid therapy.

All 46 patients who completed VBP15-003 opted to receive continued treatment with 2.0 to 6.0 mg/kg/day vamorolone under a 2-year long-term extension protocol (VBP15-LTE). The large majority of the 41 patients who completed their end-of-study visit have transitioned to expanded access programs or compassionate use programs to receive continued vamorolone treatment. In April 2021, ReveraGen and we announced data from these 41 patients treated with vamorolone for 30 months in the VBP15-LTE study, which showed that patients treated with vamorolone had improved standardized motor function outcomes (time to stand from supine, to run/walk 10 meters and to climb four stairs) from baseline to month 30. We believe that the data from the VBP15-LTE study support the conclusion that vamorolone treatment has the potential to improve motor function in DMD patients with an efficacy comparable to glucocorticoids, but with a more favorable safety profile.

The VISION-DMD Trial (VBP15-004) is an ongoing phase IIb randomized, double-blind, parallel group, placebo and active-controlled study to evaluate the efficacy, safety, pharmacodynamics, and population pharmacokinetics of vamorolone administered orally at daily doses of 2.0 mg/kg and 6.0 mg/kg versus prednisone 0.75 mg/kg/day and placebo over a treatment period of 24 weeks, and to evaluate persistence of treatment effect over a treatment period of 48 weeks in boys with DMD aged four to seven years who are able to walk. Study efficacy endpoints include timed function tests and measures of muscle strength and endurance. Safety endpoints include monitoring of weight gain, bone metabolism, cataracts, and biomarkers of metabolic disturbances. The VISION-DMD Trial, which is being conducted by ReveraGen as a pivotal clinical trial at over 30 sites across North America, Europe, Israel and Australia, is fully

enrolled with 121 patients. We currently expect top-line data after six months of treatment to be available in the second quarter of 2021 and top-line data after twelve months of treatment in the fourth quarter of 2021.

c. Market exclusivity, regulatory status and commercial formulation

ReveraGen has received orphan drug designation for vamorolone by both the FDA and the EMA, and has received fast track designation by the FDA. Upon granting of a marketing authorization in the U.S. or the EU, vamorolone would enjoy market exclusivity based on the orphan drug status by 7 years in the U.S. and 10 years in Europe from the date of the marketing authorization.

The use of vamorolone as treatment of DMD is also covered by method of use patents held by ReveraGen, which are set to expire in 2029 (see "*—Additional information on our business—Market exclusivity and intellectual property*" beginning on page 83).

In the U.S., ReveraGen has obtained rare pediatric disease designation for vamorolone in the U.S. with eligibility for a priority review voucher. Further, ReveraGen has received a USD 3.3 million grant from the National Institute of Neurological Disease and Strokes (the "**NINDS**"), an institute within the National Institutes of Health (NIH), to support the preparations for an NDA filing during a two-year period. In the UK, the MHRA has designated vamorolone in DMD as a Promising Innovative Medicine, a status similar to a breakthrough therapy designation by the FDA.

For the production of vamorolone, we rely on third party manufacturers and we have only started to manage such production by third party manufacturers in September 2020. We have to perform additional technical development and formulation work in view of the assembly of the technical section (the so-called common technical document, or "CTD") of a potential NDA for vamorolone in DMD, since bioequivalence between ReveraGen's formulation used in clinical trials and the formulation that we intend to use for upscaling production in the run-up to commercial production has not yet been demonstrated. Therefore, we decided to submit a potential NDA for vamorolone in DMD, and to subsequently launch the product, if and when we receive marketing authorization, using the process for producing the active pharmaceutical ingredient ("**API**") as well as the formulation used in the clinical trials. As we are required to validate the process for producing the API and the formulation used in the clinical trials before we can use them for a marketed product, we would only be able to submit an NDA in the first quarter of 2022 at the earliest, if the six-month top line data of the VISION-DMD Trial show that such trial meets its endpoints after six months of treatment (a "**Positive Interim Readout**"); in this case, we would submit such NDA based on the twelve-month results of the VISION-DMD Trial. After a potential launch of vamorolone in DMD, we currently plan to introduce a new process for producing the API and establish a formulation that is more suitable for marketed products.

In the EU, based on our discussions with the EMA, we are confident that the EMA would accept the filing of a MAA after completion of twelve months of treatment within the VISION-DMD Trial if the trial's endpoints are met. If the VISION-DMD Trial is completed successfully within the currently anticipated timeframe, we could file such MAA in the second quarter of 2022, following which a decision on the MAA could be expected in the second half of 2023.

D. Our product outlicensed to Chiesi Group: Raxone®

As described in more detail in "*—Additional information on our business—Material agreements—License and collaboration agreements—License agreement with Chiesi Group*" beginning on page 79, we have outlicensed our rights in Raxone®, including relating to development, commercialization and distribution, to Chiesi Group effective August 2019.

a. Leber's hereditary optic neuropathy (LHON)

LHON is a rare, severe hereditary eye disease that usually affects young, otherwise healthy individuals and is more common in men than women. Loss of central vision occurs in the majority of patients within

one year of their symptoms becoming apparent. The loss of vision in the first eye is typically sudden, abrupt, painless and profound. This is typically followed by loss of vision in the other eye 1–3 months later. The rapid loss of vision caused by LHON, which in most cases leads to profound loss or severe deterioration of vision, has a dramatic impact on the quality of life for patients, and is associated with considerable economic and social costs due to productivity loss, lower employment rates, income loss and costs of care.

LHON is caused by mutations in genes in the mitochondria. When such mutations are present, nerve cells in the retina at the back of the eye (so-called retinal ganglion cells) do not have enough energy to work properly, leading to loss of vision.

b. Treatment of LHON with Raxone®

Raxone® is to our knowledge the first and only approved treatment for LHON, and is approved in the EU, the UK, Israel, Serbia and South Korea for the treatment of visual impairment in adolescents and adults with LHON. Based on the results of our clinical trials we believe that Raxone® helps bypass the deficient mechanism caused by the mutated genes that prevents the mitochondria from working properly. As a result, Raxone® may reactivate nerve cells in the retina (retinal ganglion cells) that are still viable but no longer active. In addition, studies conducted by us as well as by third parties have shown that Raxone® also works as a cell-protecting antioxidant, preventing damage induced by so-called reactive oxygen species, commonly known as free radicals, which are by-products of the cell metabolism that may damage cells.

The results of our RHODOS trial (see "*—Clinical development*" beginning on page 72 for more information) suggest that Raxone® can prevent further vision loss associated with LHON and promote clinically relevant recovery of visual acuity in LHON patients with disease duration of up to five years. An early diagnosis of LHON offers patients the best chance for the treatment of their condition with Raxone® in the long term. There is a window of opportunity for the optimal treatment of LHON with Raxone® when the retinal ganglion cells are still viable: clinical data of our completed expanded access program (see "*—Clinical development*" beginning on page 72 for more information) suggest that up to 50% of patients with disease duration of less than one year experience a clinically relevant recovery of visual acuity following Raxone® treatment.

Raxone® is available in the form of film-coated tablets containing 150 mg idebenone.

c. Clinical development, efficacy, and post-authorization measures

We have demonstrated the efficacy and safety profile of Raxone® in what is, to our knowledge, the first clinical development program ever undertaken with regard to a drug for the treatment of LHON. Our clinical development program included a double-blind, randomized, placebo-controlled trial ("**RHODOS**"), an observational long-term follow-up study ("**RHODOS-OFU**"), an Expanded Access Program ("**EAP**"), and a natural history case record surveys (the "**CRS**"). The European Commission has approved Raxone® for the treatment of LHON in 2015. The EMA's European Public Assessment Report (EPAR) discloses relevant data and the basis for the EMA's recommendation. Under our marketing authorization in the EU we are required to conduct post-authorization measures, namely a phase IV clinical trial on the long-term effects and safety of Raxone® ("**LEROS**"), the continuation of the EAP, an additional CRS and a post-authorization safety study ("**PASS**"). The following is a short summary of the key characteristics of the above-mentioned studies:

— RHODOS was, to our knowledge, the first randomized, placebo-controlled clinical trial to be completed with regard to a drug for the treatment of LHON. RHODOS was conducted in 85 patients aged 14 to 66 years having all three major genetic mutations causing LHON and with an onset of vision loss within the last five years prior to enrollment.

— RHODOS-OFU was conducted as a single-visit observational follow-up study in 58 patients who had completed the RHODOS trial and who were assessed after a median of 30 months without treatment.

— The EAP aimed at providing insight into Raxone®'s therapeutic potential in a real-world setting and was conducted under the Named Patient Program in the EU and the Investigational New Drugs regulations in the U.S. Enrollment was limited to patients presenting for treatment within one year of onset of symptoms of LHON. Data from 69 patients at 36 centers worldwide were analyzed and reported at the time of filing the MAA. We continued the EAP as part of the post-authorization measures and completed the EAP in 2018 with a total of 111 patients receiving Raxone®.

— The first CRS, which was completed in 2014, was a collaboration between us and the European Vision Institute Clinical Research Network (EVICR.net) in which historically documented visual acuity data from LHON patients not treated with idebenone were collected from participating centers. We completed an additional CRS with data from 592 patients in the first quarter of 2019. The data from both CRSs form the basis for a comparator group for the ongoing LEROS phase IV clinical trial.

— LEROS is an additional phase IV clinical trial on the long-term effects and safety of Raxone®, designed as an open label interventional study. LEROS builds on the results from the RHODOS clinical trial. The primary objective of the LEROS clinical trial is to assess the efficacy of Raxone® to improve or stabilize visual acuity in patients starting treatment up to one year after the onset of vision loss, compared to an external natural history control group. Patient enrollment was completed in March 2019 with 197 participating patients. The participating patients are being treated with Raxone® (daily dose of 900 mg) for 24 months. LEROS is being conducted on 31 study sites across nine European countries and the U.S. We currently expect top-line results of the study by the end of 2021.

— In addition, we are currently conducting a multicenter, prospective, non-interventional PASS for patients with LHON treated with Raxone® called PAROS, top-line results of which we currently expect by the end of 2021.

d. Market exclusivity, regulatory status and outlicensing revenue

We have been granted orphan drug designation for Raxone® in the EU (until September 2025), the U.S., and South Korea. The product is not patent protected.

We received marketing authorization for Raxone® for the treatment of adolescents and adults with LHON in all EU countries (then including the UK), Norway, Iceland and Liechtenstein in September 2015, and the respective exclusive distributor has received marketing authorization in Israel in August 2017, in Serbia in January 2019 and in South Korea in September 2019.

Raxone® is, to our knowledge, the first therapy approved for treatment of LHON. The marketing authorization for the EU was granted under "exceptional circumstances". Such authorization may be given when comprehensive efficacy and safety data cannot be obtained, but it is still appropriate to grant the authorization. In the case of Raxone®, authorization under "exceptional circumstances" was given because the CHMP was of the view that it was not feasible to generate a comprehensive data set, mainly due to the rarity of LHON. Furthermore, at the time of the CHMP's determination, idebenone was already used by physicians on an off-label basis to treat LHON patients, which is why it was believed that neither physicians nor patients would be prepared to participate in a placebo-controlled trial. Under the EU marketing authorization granted to us for Raxone®, we are required to conduct the phase IV LEROS clinical trial as well as other post-authorization measures. For more information on the studies we have recently completed or are currently conducting, see "*—Clinical development*" beginning on page 72.

We launched Raxone® in Germany in October 2015.

Effective August 2019, we outlicensed our rights in Raxone®, including relating to development, commercialization and distribution, to Chiesi Group. See "*—Additional information on our business—Material agreements—License and collaboration agreements—License agreement with Chiesi Group*" beginning on page 79. The total consideration under our outlicensing arrangement with Chiesi Group with respect to Raxone® amounts to up to EUR 93 million, comprising an upfront cash payment of EUR 44

million, which was received at closing in August 2019, and near- to mid-term sales milestone payments of up to EUR 49 million. In our transaction with Chiesi Group, we retained the rights in Raxone® for the U.S. and Canada, for which jurisdictions we, however, do not have and currently do not plan to obtain marketing authorizations for Raxone®. We also agreed with Chiesi Group on a transitory regime pursuant to which we continue to sell Raxone® in France. Chiesi Group has an option to fully acquire our entire Raxone® business at any point in time.

In France, the French Ministry for Solidarity and Health has finally refused to register Raxone® on the lists of reimbursed products in France. As of the date of this Prospectus, our discussions with French authorities about the reimbursement status of Raxone® in France are ongoing. By invitation of the competent authority in France, we resubmitted an application to register Raxone® on the list of conditionally reimbursed products in April 2021. We have been asked by French authorities to provide Raxone® for free as from August 2021. In any case, we do not expect sales of Raxone® in France to continue from August 2021 onwards.

e. Competing therapeutic approaches

Gensight Biologics, Paris, France ("**Gensight**"), has a program for treatment of LHON based on gene therapy (LUMEVOQ™, GS010) that addresses patients within one year of symptom onset and carrying one of the major genetic mutations causing LHON. Gensight filed an MAA with regard to LUMEVOQ™ with the EMA in September 2020 and has announced that it expects a decision in the fourth quarter of 2021.

Wuhan Neurophth Biological Technology Limited Company, Wuhan Hubei, People's Republic of China ("**Neurophth Therapeutics**"), announced in February 2021 that it would initiate clinical trials for a gene therapy approach (NR082) to treat LHON in the first half of 2021. Neurophth Therapeutics has received an orphan drug designation for NR082 from the FDA in 2020.

In addition, Stealth BioTherapeutics, Inc., Newton, Massachusetts, USA, has completed a phase II clinical trial of elamipretide (MTP-131), a mitochondria targeting peptide, in LHON patients. An abstract of the results was published at a conference in 2019.

E. Our early stage pipeline

1. Lonodelestat as phase Ib product candidate in cystic fibrosis

In February 2018, we in-licensed the compound lonodelestat from Polyphor. lonodelestat is a selective inhibitor of an enzyme called human neutrophil elastase ("**hNE**") that we believe has the potential to treat CF and other lung diseases associated with increased neutrophil elastase activity such as non-cystic fibrosis bronchiectasis, alpha-1 antitrypsin deficiency, and primary ciliary dyskinesia. Based on prior development work by Polyphor, we recently completed a phase Ib multiple ascending dose (MAD) trial (the "**MAD Trial**") of orally inhaled multiple doses of lonodelestat in patients with CF, which showed a good tolerability of lonodelestat and no serious patient-reported serious or severe (grade 3 or higher) adverse events across all doses and over all treatment durations.

a. Cystic fibrosis (CF) and mode of action of lonodelestat

CF is a rare, life-threatening, progressive genetic disease affecting primarily the lung but also the digestive system. The symptoms in the lung are characterized by build up of mucus obstructing the airways leading to persistent infection and chronic inflammation, thereby limiting the ability to breathe over time. CF is typically diagnosed in young children mostly within the first year of age.

Third-party research suggests that activated neutrophils (a type of white blood cell) liberate high levels of hNE in the lung, which in turn causes damage to structural, cellular and soluble components of the microenvironment in the lung. These high levels of hNE play a central role in the deterioration of lung function associated with CF and correlate with the severity of CF as determined by measures of lung

function. We believe that inhibition of hNE may stop or slow damage to lung tissue, may help preserve lung function and may help improve the overall quality of life for individuals with CF.

Pre-clinical research by Polyphor suggests that lonodelestat, a cyclic peptide designed by Polyphor, is a highly potent and selective inhibitor of hNE. In the phase I clinical trials conducted by Polyphor, lonodelestat was administered by inhalation via an optimized eFlow® nebulizer (the "**Inhaler**") developed by PARI Pharma GmbH, Gräfelfing, Germany ("**PARI**").

Third-party research suggests that chronic inflammation is also present in lung diseases other than CF which are associated with increased neutrophil elastase activity, such as non-cystic fibrosis bronchiectasis, alpha-1 antitrypsin deficiency, and primary ciliary dyskinesia.

b. Market opportunity, treatment options and competing therapeutic approaches

We estimate that approximately 80,000 patients worldwide have CF at any point in time. We believe that lonodelestat has the potential to be applied in all CF patients, as it is not mutation-specific.

CF treatment typically consists of concomitant use of several approved medications, including CFTR (cystic fibrosis transmembrane conductance regulator) modulators (ivacaftor or ivacaftor combined with lumacaftor), mucolytics (Pulmozyme®, a drug marketed by Roche), inhaled antibiotics (tobramycin, aztreonam and azithromycin), and pancreatic enzyme products (Creon®, a drug marketed by BGP Products). Several dual and other triple combination CFTR modulators are advancing in their clinical development, as are some novel gene therapy agents. These medications, however, do not significantly reduce lung inflammation, which is why we believe that there is a clinical need for drugs that target inflammation. Hence, we believe that lonodelestat could also potentially be used as add-on therapy.

We are aware that a number of other product candidates to treat lung inflammation in CF patients are currently being developed.

A drug candidate which is, like lonodelestat, an hNE inhibitor, is Chiesi Group's CHF6333, which is also administered via a dry-powder inhaler. Chiesi Group completed a phase I clinical trial in healthy volunteers in July 2017. A phase Ib clinical trial in patients with CF and in patients with non cystic fibrosis (NCFB) bronchiectasis is currently ongoing.

Fenretinide (LAU-7b), which is being developed by Laurent Pharmaceuticals Inc., Montréal, Canada ("**Laurent**"), triggers the body's own pro-resolving mechanisms to terminate inflammation. Laurent is currently conducting a phase II clinical trial of fenretinide for the treatment of CF and has indicated a completion date in the third quarter of 2021.

Ensifentrine (RPL554), which is being developed by Verona Pharma PLC, London, UK ("**Verona Pharma**"), is an inhaled, dual inhibitor of the enzymes phosphodiesterase 3 and 4 for the treatment of respiratory diseases. Verona Pharma has completed a phase IIa clinical trial of ensifentrine for the treatment of CF, whose results were made public in 2019.

c. Clinical development, regulatory status and market exclusivity

Polyphor has completed two clinical trials of lonodelestat for the treatment of CF. A first-in-human single ascending dose phase I safety and tolerability trial in 48 healthy volunteers and a single ascending dose phase I safety and tolerability trial in 24 CF patients have shown that lonodelestat was well tolerated and safe. The trial in CF patients also showed that levels of concentration of lonodelestat in the sputum were around 1,000 fold higher than in the plasma and that lonodelestat strongly inhibited hNE in the sputum tested 3 hours after administration, while the inhibitory effect was lower to absent and more variable after 24 hours.

In the license agreement with Polyphor, we have undertaken to assume the global development, regulatory filings and commercialization of lonodelestat. In a first step, we conducted a MAD Trial that was previ-

ously planned by Polyphor. In the MAD Trial, 32 patients were randomized in four cohorts of eight patients each and received lonodelestat starting with 80 mg once a day, 80 mg twice a day, and 160 mg once a day, each administered for 15 days, followed by the fourth cohort with 40 mg once a day administered for 28 days, which was chosen after observing a reduction of forced expiratory volume in one second (FEV1) in some patients treated with the highest doses (80 mg twice a day and 160 mg once a day). In all four cohorts and over all treatment durations, lonodelestat demonstrated a good tolerability and no serious or severe (grade 3 or higher) adverse events were reported by the patients. Results showed a linear dose-exposure relationship over the dose range from 40 mg to 160 mg, with no accumulation in plasma or sputum. In all cohorts, a transient, near complete inhibition of hNE activity was observed after inhalation. In addition, in some patients in the 40 mg per day cohort, a constant level of near complete inhibition gradually developed over the 28 days of drug inhalation.

In parallel, we plan to collect additional preclinical safety data that, together with the results of the MAD Trial, may allow us to start a phase II efficacy trial of lonodelestat in CF in 2022.

Polyphor received financial support for its development program from Cystic Fibrosis Foundation Therapeutics Inc., Bethesda, Maryland, USA ("**CFFT**"), and we are entitled to financial support from CFFT in the form of milestone payments relating to the MAD Trial, in a currently remaining amount of up to USD 100,000 in aggregate.

We have a world-wide exclusive license or sublicense, as applicable, from Polyphor for the use of lonodelestat in any indication. In addition, we have an exclusive world-wide sublicense from Polyphor to use the PARI Inhaler to administer lonodelestat.

2. Activities regarding congenital muscle dystrophy

We explore the opportunities of a potential novel gene therapy approach for the treatment of so-called laminin-alpha 2 (LAMA2)-deficient congenital muscular dystrophy ("**LAMA2-MD**", also known as "MDC1A") in preclinical collaborations with the University of Basel's Center for Molecular Life Sciences (*Biozentrum*) (the "**Biozentrum**") and with Rutgers, The State University of New Jersey ("**Rutgers University**"). We previously conducted and have completed a phase I clinical trial of omigapil, a so-called deprenyl analogue that third-party research has shown to prevent cell death pathways (apoptosis), in CMD (called CALLISTO), which met its primary objective. However, we decided in 2020 to down-prioritize the further clinical development of omigapil in CMD because additional non-clinical work would be needed before a pivotal clinical trial of omigapil in CMD should be conducted, and also because of the emerging gene therapy approaches to treat some forms of CMD.

a. Congenital muscular dystrophies (CMD)

CMD is a group of inherited neuromuscular conditions that causes progressive and potentially life-threatening loss of muscle tissue, affecting frequently newborns and children. CMD conditions are caused by mutations in proteins required for the muscles, and sometimes the eyes and brain, to work properly. Children born with CMD often have muscle weakness or "floppiness" and can also have stiffness of the joints, hip dislocation and a type of curvature of the spine (known as kyphoscoliosis). Affected patients have difficulties in walking, maintaining stable body posture and lifting objects. These symptoms can be present at birth or develop during childhood or later in life.

Many types of CMD are stable or progress only slowly, allowing children to acquire new skills and to live a normal lifespan. However, some severe forms of CMD cause respiratory problems that can be life-threatening as they worsen over time.

b. Market opportunity, treatment options and competing therapeutic approaches

The British Muscular Dystrophy Campaign estimates that two to five in 100,000 children in the UK are born with CMD.

Currently, treatment options for CMD are confined to treatment of symptoms and include physiotherapy, speech therapy, occupational therapy, respiratory support and orthopedic spine surgery. Another important aspect of disease management is supplementary nutrition to avoid malnutrition. To our knowledge, no approved pharmaceutical treatments are available or in clinical development for CMD. As of the date of this Prospectus we are not aware of any ongoing or completed clinical trials in CMD other than the completed CALLISTO trial. However, we are aware of a few preclinical programs for pharmaceutical treatments of CMD.

c. Potential gene therapy for LAMA2-MD

In May 2019, we entered into a preclinical collaboration with Prof. Markus Rüegg of the Biozentrum, who pioneered a novel gene therapy approach for the treatment of Lama2-MD. Third-party research has shown that this condition is caused by a defect in the protein LAMA2, which sits on the membrane of muscle cells.

The preclinical research of Prof. Rüegg and his team suggests that the simultaneous transgenic expression of specifically designed small protein domains (structures which form part of a protein), so-called linker proteins, may help overcome the structural and functional loss of muscle fibers. Prof. Rüegg and his team have developed a novel gene therapy strategy in which they expect to use two linker proteins that are composed of domains derived from extracellular matrix proteins agrin, laminin and nidogen. The researchers expect that these linker proteins, if expressed simultaneously, could ameliorate muscle membrane stability by compensating the deficient functionality of LAMA2. In a mouse model for LAMA2-MD, simultaneous expression of these linker proteins has led to restoration of muscle fiber basement membranes, recovery of muscle force and size, increased overall body weight and markedly prolonged survival.

In our preclinical research collaboration with the Biozentrum, our collaborators and we plan to explore the feasibility of the above-described therapeutic approach by delivery of the genes encoding the two linker proteins via standard viral vectors into muscle tissue.

We, together with Prof. Rüegg, have successfully applied for a grant from Innosuisse, the government-backed Swiss Innovation Agency, for the above-described pre-clinical proof of concept studies by Prof. Rüegg and his team. Under the terms of this grant, each of Innosuisse and us has committed to invest CHF 0.65 million into this preclinical research collaboration.

In April 2020, we entered into two agreement with Rutgers University, under which we acquired an exclusive non-transferable sublicensable worldwide royalty-bearing license to two patent aplications regarding a gene construct. Rutgers University has agreed to further study such gene construct until the end of 2021. We fund such research with a fixed amount. We believe that this collaboration complements our collaboration with Prof. Rüegg.

F. Additional information on our business

1. Material agreements

For a description of certain key risks associated with our contractual relationships, see the risk factors included elsewhere in this Prospectus.

a. License and collaboration agreements

License Agreement with ReveraGen

In November 2018, we entered into an agreement (the "**Option Agreement**") with Idorsia, under which we acquired an option to obtain from Idorsia an exclusive sub-license to develop and commercialize (but not manufacture) vamorolone, which is being developed by ReveraGen. At the time, Idorsia had a worldwide, exclusive, sub-licensable license to develop and commercialize (but not manufacture) vamorolone

from ReveraGen under a "License, Collaborative Development and Commercialization Agreement" that had been entered into in April 2016 between ReveraGen and Actelion Pharmaceuticals, Inc. and was subsequently assigned by Actelion to Idorsia in 2017 and further amended in 2018 (the "**ReveraGen Agreement**"). As initial consideration for the option for such sub-license, we issued 1,000,000 Shares to Idorsia (the "**Idorsia Shares**") and paid USD 20 million in cash to Idorsia. Both the issuance of the Idorsia Shares and the payment of the USD 20 million in cash were non-refundable. The cash payment amount was intended to compensate Idorsia for having already paid USD 15 million to ReveraGen to fund ReveraGen's VISION-DMD Trial.

In September 2020, we, Idorsia and ReveraGen agreed to terminate the Option Agreement, to assign the ReveraGen Agreement to us, and to amend the terms of the ReveraGen Agreement. As a result, we acquired a worldwide exclusive, royalty-bearing and sub-licensable license to develop, commercialize and manufacture vamorolone for all indications. Idorsia agreed to transfer know-how, including any rights to inventions, relevant for the development and commercialization of vamorolone as well as material and know-how for the drug production process to us. As upfront consideration, we paid 366,667 Shares to Idorsia and we issued an interest-free exchangeable note in the principal amount of CHF 10 million to Idorsia (the "**Idorsia Exchangeable Note**"). The Idorsia Exchangeable Note will become due on September 1, 2021, or earlier if and when the FDA supports an NDA for vamorolone in DMD in the U.S. Upon the Idorsia Exchangeable Note becoming due, we will be required to pay Idorsia a cash amount of at least CHF 3.5 million and we have the right to repay the remaining CHF 6.5 million on cash or in Shares, valued at 85% of the then prevailing volume weighted average price.

We have undertaken to use commercially reasonable efforts to further develop and, when appropriate, seek regulatory approval for the marketing and sale of vamorolone in DMD and other additional indications. We agreed to fund ReveraGen's ongoing clinical development of vamorolone, including the VISION-DMD Trial, in monthly and, later on, quarterly instalments of up to USD 0.5 million and USD 7 million in total. We also have to bear the costs associated with certain other studies conducted by ReveraGen, to the extent such costs exceed a defined threshold. After a potential filing of an NDA with respect to vamorolone in DMD, we would have to bear certain additional development costs and expenses of ReveraGen.

We agreed to make a USD 5 million milestone payment to ReveraGen if and when the FDA supports an NDA for vamorolone in DMD in the U.S. based on a Positive Interim Readout. If the FDA does not support such NDA filing based on a Positive Interim Readout and if we decide to file an NDA at a later stage, we will have to make a milestone payment in a lower amount. Upon receiving marketing authorization for vamorolone in DMD in the U.S., we will have to make milestone payments to ReveraGen and Idorsia in the aggregate amount of USD 50 million. Further milestone payments in the aggregate amount of USD 40 million will be triggered upon certain regulatory and commercialization milestones outside the U.S. If we achieve certain milestones with respect to the development and commercialization of vamorolone in indications other than DMD, we will be required to make payments to Idorsia in the aggregate amount of up to USD 45 million. If we reach certain sales milestones with vamorolone products irrespective of the indication, we will be required to make milestone payments to Idorsia in the aggregate amount of up to USD 15 million and to ReveraGen in the aggregate amount of up to USD 120 million. Additionally, we agreed to tiered royalty payments on a progressive scale ranging from single digit % to low teens % on net sales of vamorolone products irrespective of the indication.

ReveraGen, having obtained rare pediatric disease designation for vamorolone in the U.S., is eligible for a rare pediatric disease priority review voucher. If we obtain such priority review voucher and subsequently sell such voucher (which would require the prior written approval by ReveraGen), we would have to pass along up to 90% of the sale proceeds to Idorsia and ReveraGen. If we use such voucher ourselves, we would have to pass along a similar percentage of the fair market value of such voucher to Idorsia and ReveraGen.

Idorsia has agreed not to increase its equity stake in the Company to a level above 20% of the voting rights, other than as a result of the Company delivering Shares to Idorsia in a partial redemption of the Idorsia Exchangeable Note, unless it publically announces its intention to submit a takeover offer for all Shares.

License agreement with Chiesi Group

In May 2019, we entered into an exclusive license and supply agreement with Chiesi Farmaceutici S.p.A., Parma, Italy (together with its consolidated subsidiaries, "**Chiesi Group**"), pursuant to which we granted to Chiesi Group an exclusive license to the rights to Raxone®, including relating to development, commercialization and distribution, for the treatment of LHON and all other potential ophthalmological indications worldwide (except for the U.S. and Canada, for which territories we retain the rights), for a total consideration of up to EUR 93 million, comprising an upfront cash payment of EUR 44 million payable at closing and staggered near- to mid-term sales milestone payments of up to EUR 49 million. Closing of the transaction occurred in August 2019.

During a transition phase, we agreed to continue providing medical, technical, logistical and scientific support with regard to ongoing market authorization activities and certain market access undertakings. In particular, we agreed with Chiesi Group on a transitory regime in France, under which we continue to sell Raxone® in France. However, we do not expect sales of Raxone® in France to continue from August 2021 onwards. Under our agreement with Chiesi Group, we remain responsible, at our own cost, for fulfilling our ongoing pharmacovigilance obligations and for completing the post-authorization measures that we are required to conduct under our marketing authorization from the EMA. Further, we have undertaken to supply Raxone® to Chiesi Group in the necessary quantities and to hold safety stock of Raxone®. We plan to explore a US FDA submission later in 2021 after we have seen the results of the LEROS and PAROS studies. In our agreement with Chiesi Group, we have granted Chiesi Group a right of first negotiation with respect to any license, sale or other disposition of the right to develop, manufacture and commercialize any idebenone-based product, including Raxone®, in non-ophthalmological indications in any territory except the U.S. and Canada.

License agreement with Polyphor

In February 2018, we entered into a license agreement with Polyphor, under which we in-licensed lonodelestat in any indication on an exclusive world-wide basis in any indication (except in relation to the sub-licensed technology produced by PARI, which we sub-license in relation to the pulmonary delivery of certain liquid formulations of lonodelestat via the Inhaler). Such license and sublicense (as applicable) extends to, among other things, the patents and patent applications with regard to lonodelestat held by Polyphor and a licensor of Polyphor, respectively, as well as the patents licensed or sublicensed to Polyphor by PARI with regard to the use of the Inhaler to administer lonodelestat. We made an initial payment of CHF 6.5 million, payable and paid in 238,924 Shares, as upfront payment and for the purchase of Polyphor's inventory of API and some finished drug product containing lonodelestat. In addition, we agreed to cash payments of up to CHF 121 million contingent on future development, regulatory and sales milestones in relation to the milestones achieved for the first indication. We have agreed to make additional milestone payments if milestones for additional indications are reached. In addition, Polyphor is entitled to tiered royalty payments at a medium to high single-digit percentage of future net sales of lonodelestat-based products.

We have undertaken to use commercially reasonable efforts to develop and commercialize at least one lonodelestat-based product throughout the U.S., Germany, France, the UK, Italy and Spain. The agreement will terminate upon the later of the expiry of the last to expire patent that is owned and licensed by Polyphor, the twelfth anniversary of the first commercial sale of the last licensed product, and the expiration of regulatory exclusivity of the last licensed product. Either party has the right to terminate the license agreement for material breach. Polyphor also has the right to terminate the license agreement if it is not feasible, or if we fail, to initiate the first clinical trial that is intended to result in an MAA or an NDA for lonodelestat by June 30, 2024 or if certain other events occur. We have the right to terminate the license agreement with or without cause at any time with 180 days' prior written notice, in which case we are required to comply with certain termination obligations unless the agreement is terminated due to a material breach of the agreement by Polyphor or an insolvency event experienced by Polyphor.

Polyphor has also transferred and assigned to us all of its current and future rights and obligations (with the exception of an obligation to make certain payments, which remains the obligation of Polyphor) under

an award between the Cystic Fibrosis Foundation Therapeutics, Inc. and Polyphor regarding a clinical trial for lonodelestat.

License agreement with Novartis

In June 2007, we entered into a license agreement with Novartis Pharma AG, Basel, Switzerland ("**Novartis**"), under which we in-licensed omigapil for treatment of CMD on an exclusive world-wide basis. We undertook to use commercially reasonable efforts to develop omigapil in CMD world-wide according to a defined development plan, to seek marketing authorization therefor and to commercialize and market the respective product, if and when approved. We are required to make certain development, regulatory and commercial milestone payments to Novartis (up to a single-digit million USD figure), as well as to pay royalties to Novartis on a jurisdiction-by-jurisdiction basis. Novartis has a one-time call back option to terminate the license granted to us and to obtain an exclusive world-wide license with regard to intellectual property in relation to omigapil in CMD generated by us. Such call back option becomes exercisable after completion of a clinical trial that is intended to result in an MAA or an NDA. The agreement with Novartis is due to terminate upon expiration of all royalty obligations that it provides. Novartis has a right to terminate the agreement for material breach or if we breach our development and commercialization obligations, and, under certain conditions, we also have the right to terminate the agreement early. In these cases, the license granted to us would terminate and we would be required to grant Novartis a worldwide, royalty-free, perpetual, non-exclusive license to use any intellectual property in relation to omigapil in CMD generated by us. In addition, Novartis may terminate the agreement if we are subject to a change of control where the acquirer owns or has in-licensed a product that is substitutable with omigapil in CMD; upon such termination, all licenses under the agreement would terminate.

Preclinical collaborations

For information on our preclinical collaborations to explore the opportunities of a potential novel gene therapy approach for the treatment of LAMA2-MD see "*— Our early stage pipeline—Potential gene therapy for LAMA2-MD*" beginning on page 77.

b. Distribution agreements

In November 2016, we entered into an exclusive distribution arrangement with Ewopharma AG, Schaffhausen, Switzerland ("**Ewopharma**"), to launch Raxone® in eleven countries in Eastern Europe (Bulgaria, Croatia, Czech Republic, Hungary, Poland, Romania, Slovakia and Slovenia) and the Baltics (Estonia, Latvia and Lithuania) and which will expire in February 2022. Since we have out-licensed Raxone® to Chiesi Group, we have to provide the full benefit of our exclusive distribution arrangement with Ewopharma with respect to Raxone® to a member of the Chiesi Group. Following the expiration of the exclusive distribution arrangement with Ewopharma, Raxone® will be distributed by Chiesi Group in these eleven countries.

In March 2017, we entered into an exclusive distribution agreement with Pharmathen Hellas S.A., Athens, Greece (now Innovis Hellas S.A., "**Innovis**"), to launch Raxone® in Greece and Cyprus. In the same year, we entered into an exclusive distribution agreement with Megapharm Ltd., Raanana, Israel ("**Megapharm**"), to launch Raxone® in Israel and the Palestinian Authority. In addition, we have entered into exclusive distribution agreements with other distributors with regard to other jurisdictions. We have assigned our contracts with Innovis, Megapharm and other distributors, to the extent they relate to Raxone®, to Chiesi Group. Megapharm formally holds the marketing authorization for Raxone® in Israel and will be required to transfer such marketing authorization to Chiesi Group upon termination or expiry of the distribution agreement.

c. Clinical development, manufacturing and other contracts

As part of our ordinary course of business, we have entered into several contracts with third-party service providers for, *e.g.*, clinical or technical development services and the manufacturing of API and finished drug products. Payments to our service providers account for a significant portion of our development expenses.

d. Financing Arrangements

CHF 60 million Senior Unsecured Convertible Bonds due 2022

On February 17, 2017, the Company issued an aggregate of CHF 60 million Senior Unsecured Convertible Bonds due 2022, which are listed on the SIX Swiss Exchange (ISIN CH0353955195, Ticker Symbol SAN17) and governed by Swiss law (the "**2017/22 Bonds**"). The nominal value of each Bond is CHF 5,000.

Upon completion of the Exchange Offer, the remaining aggregate principal amount of the 2017/22 Bonds is CHF 15,155,000. To the extent that the Company does not repurchase or redeem these 2017/22 Bonds, such 2017/22 Bonds continue to be outstanding and will become due for redemption on February 17, 2022.

The 2017/22 Bonds carry interest at 5% *per annum* payable semi-annually on February 17 and August 17. The 2017/22 Bonds are senior and unsecured obligations of the Company and rank pari passu with all its other non-subordinated debt and will become due for redemption on February 17, 2022, unless previously converted, redeemed or purchased and canceled under their terms and conditions. We may repurchase the 2017/22 Bonds at any time and may also have the repurchased 2017/22 Bonds canceled.

The 2017/22 Bonds are convertible between May 17, 2017 and their date of maturity at a conversion price of currently CHF 64.80. The prevailing conversion price is subject to anti-dilution and other adjustments in certain events, as further set forth in the terms and conditions of the 2017/22 Bonds.

The Company has the right to redeem all, but not only some of the outstanding 2017/22 Bonds at their principal amount, plus accrued interest, if any, at any time if the volume-weighted average price ("**VWAP**") of the Shares is at least 160% of the prevailing conversion price on at least 20 out of 30 consecutive trading days ending not earlier than 5 trading days prior to the Company's redemption notice.

If a person or persons acting in concert acquire more than 50% of all voting rights (whether exercisable or not) of the Company, via a public tender offer, a merger or otherwise, or if the legal or beneficial ownership of all or substantially all of the assets of the Group is acquired by a third party, the holders of the Bond have the right to convert their 2017/22 Bonds at an increased conversion price or to seek redemption of their 2017/22 Bonds, each during a specified period and all as further specified in the terms and conditions of the 2017/22 Bonds.

The terms and conditions of the 2017/22 Bonds provide for, among other things, a negative pledge with regard to (i) marketable debt instruments and (ii) financial indebtedness exceeding CHF 10 million in the aggregate, subject to certain exceptions. In addition, the 2017/22 Bonds may be declared immediately due and payable in certain events of default, such as (i) a default with regard to payments or the delivery of Shares under the 2017/22 Bonds (subject to grace periods); (ii) material breaches of the terms and conditions of the 2017/22 Bonds which are not remedied in a timely manner; (iii) a cross-default with respect to financial indebtedness whose nominal value is at least CHF 5 million in the aggregate; (iv) if the Company or a material subsidiary is insolvent, bankrupt, unable to pay its debt, stops or suspends payment of material debt, proposes or applies for a stay of execution, enters into a postponement of payments, a general assignment or arrangement or composition with creditors, or a moratorium or postponement of payments is declared; (v) if the Company or a material subsidiary alters its commercial structure through bankruptcy, liquidation or disposal of all assets, the Company changes its objects or commercial activities or merges with a third party, if such action has or may have a material adverse effect on the Company's capacity to meet its obligations in connection with the 2017/22 Bonds; or (vi) a dissolution of the Company or a the Company's merger into another company where the surviving company does not assume all of the Company's liabilities.

Equity-linked financing arrangement with Highbridge

In July 2020, the Company and certain of the Company's subsidiaries entered into a subscription agreement with Highbridge Tactical Credit Master Fund, L.P. ("**Highbridge**"), a fund managed by Highbridge

Capital Management LLC, providing for the issuance of an aggregate principal amount of originally up to CHF 20.0 million in senior secured exchangeable notes with a principal amount of CHF 5,000 each (the "**Exchangeable Notes**"), issued by Santhera Pharmaceuticals (Schweiz) AG ("**Santhera Schweiz**"), subject to certain conditions and in tranches, and exchangeable for Shares. As consideration for the commitment and utilization of the original financing, Highbridge received 300,000 Shares.

Under the Exchangeable Notes, Santhera Schweiz agreed to pay a fixed interest, which the Company could pay in cash at a rate of 12% *per annum* or in Exchangeable Notes at a rate of 13% *per annum*. Subject to certain restrictions, Highbridge could elect to exchange Exchangeable Notes for Shares at any time, at an exchange price of 90% of the VWAP of the Shares on the exchange date, subject to a floor price. The Exchangeable Notes would have become due for redemption at their principal value on January 13, 2022, if not exchanged or redeemed prior to that date. The Exchangeable Notes were guaranteed by the Issuer and certain subsidiaries of the Issuer and secured by a comprehensive security package, including security over all shares in SPCH and other subsidiaries of the Issuer as well as over the Group's material intellectual property and other assets of the Group. Of the original aggregate principal amount of up to CHF 20 million, Exchangeable Notes in the aggregate principal amount of CHF 7.5 million were issued and fully exchanged by October 2020. The unused amount of up to CHF 12.5 million could no longer be drawn down by Santhera Schweiz following the termination of the development program with regard to Puldysa®.

In November 2020, the Company, Santhera Schweiz and Highbridge agreed to replace the commitment with respect to such amount by a commitment with respect to new Exchangeable Notes in the aggregate principal amount of up to CHF 10.0 million (the "**Further Notes**") and changed some of the terms of them, including a lowering of the floor price. Santhera Schweiz has issued Further Notes in the full aggregate principal amount of CHF 10.0 million to Highbridge; such Further Notes have been fully exchanged. The Further Notes were secured in the same way as the Exchangeable Notes.

In February 2021, the Company, Santhera Schweiz and Highbridge amended their agreement to provide for the issuance of additional Exchangeable Notes in the aggregate principal amount of up to CHF 12.0 million (the "**New Money Notes**"), subject to certain conditions, and changed some of the terms, including a further lowering of the floor price and an extension of the maturity to July 13, 2022. The New Money Notes are also secured in the same way as the Exchangeable Notes. In connection with this amendment, the parties agreed that Highbridge shall receive an amendment and commitment fee in 984,769 warrants, each of which will be exerciseable for one Share at an exercise price of CHF 2.7418 as from its issuance until March 15, 2026.

2. Distribution and marketing

Our product candidates (if approved) will have different prescriber bases: primarily neurologists in the case of vamorolone, and primarily pulmonologists in the case of lonodelestat in CF. Due to limited synergy potential for marketing and sales, we may have to build separate sales channels for each of our products.

Effective August 2019, we outlicensed our rights in Raxone®, including relating to development, commercialization and distribution, to Chiesi Group. See "*—Material agreements—License and collaboration agreements—License agreement with Chiesi Group*" beginning on page 79. In France, we continue to sell Raxone® via our own distributor under a transitory regime agreed with Chiesi Group, but we do not expect sales of Raxone® in France to continue from August 2021 onwards.

Starting in fall 2020, as a result of the discontinuation of the development of our late-stage product candidate, Puldysa®, and the resulting need for cost savings, we had to significantly reduce our operations and had to take many other cash-preserving and cost-saving measures. For instance, we ceased our commercial capabilities in the EU, which has led to a reduction of our headcount of approximately 50%.

We have a U.S. subsidiary in the Boston metropolitan area. Our U.S. team currently manages our patient advocacy interactions, prepares for market entry in the U.S. and is the source of our U.S. regulatory and medical affairs expertise. A full roll-out of U.S. marketing and sales activities will only be possible upon

receipt of a marketing authorization of vamorolone in DMD from the FDA. If and when we should receive marketing authorization for vamorolone in DMD in the EU, we will consider expanding our sales force and may also develop further in-house marketing, sales and distribution capabilities to commercialize the product.

3. Supply and manufacturing

We have no internal manufacturing capabilities and rely on third parties for the manufacture, formulation, packaging, storage and distribution of Raxone®, vamorolone and our other product candidates. See also risk factor "*We have no manufacturing capabilities or capacity of our own and rely on third parties for production of our compounds and finished drug products.* " beginning on page 30.

4. Competition

The pharmaceutical industry is rapidly evolving and highly competitive. Companies can expect to face significant competition from biotechnology and pharmaceutical companies, in particular when "first-in-class" products are introduced and new markets are opened. After the innovator company has successfully developed an underserved market by creating awareness of a new therapeutic agent, other companies are quick to introduce competitive products. Competition generally comes from new and existing therapies developed and marketed by large and small pharmaceutical and biotechnology companies. It is the nature of the competitive landscape that a marketer of a product has difficulty in predicting the future basis upon which it will compete with new products marketed by others.

For overviews of therapeutic approaches that compete or may compete with our own product and product candidates see "*—Our lead product candidate: vamorolone for the treatment* of DMD*—Treatment options and competing therapeutic approaches*" beginning on page 66, "*—Our product outlicensed to Chiesi Group: Raxone®—Competing therapeutic approaches*" beginning on page 74, "*—Our early stage pipeline—Lonodelestat as phase Ib product candidate in cystic fibrosis—Market opportunity, treatment options and competing therapeutic approaches*" beginning on page 75, "*—Our early stage pipeline—Activities regarding congenital muscle dystroph—Market opportunity, treatment options and competing therapeutic approaches*" beginning on page 76, and "*—Our early stage pipeline—Potential gene therapy for LAMA2-MD*" beginning on page 77.

In addition to alternative therapeutic approaches, we face competition by off-label uses in particular of idebenone (the active ingredient in Raxone®). For more information see risk factor "*We may be required to refund to the French Social Security part of our revenue generated from the sale of Raxone® in France since January 1, 2016. If we are required to make such a refund in cash, our financial situation, results of operations and prospects may be materially adversely affected.*" beginning on page 22. Also, pharmacies have been compounding idebenone, *i.e.*, they made a so-called pharmacy or magistral preparation of it. For more information see risk factor "*Pharmacies have been compounding idebenone. Future compounding may adversely affect sales by Chiesi Group of Raxone®.*" beginning on page 25.

5. Market exclusivity and intellectual property

Our success depends in part on our ability to obtain various layers of protection for market exclusivity through a combination of orphan drug, patent protection and documents/files protection, as available.

It is our strategy to develop and commercialize product candidates in indications qualifying for orphan drug designation in order to obtain marketing exclusivity. Orphan drug designations are available in some jurisdictions, including the U.S. and the EU, for drugs with relatively small patient populations. Generally, if a product candidate with an orphan drug designation in a particular indication subsequently receives the first marketing authorization, then the product is entitled to a period of marketing exclusivity, *i.e.*, no other marketing authorizations will be granted for the same drug for the same indication during the exclusivity period. The applicable period is 7 years in the U.S. and 10 years in the EU (to be reduced to 6 years if a drug no longer meets the criteria or is sufficiently profitable) after receipt of marketing authorization. Orphan drug exclusivity may be lost if the respective regulatory authority determines that the

request for designation was materially defective, if the manufacturer is unable to assure sufficient quantity of the drug to meet patient needs, or for other reasons. To date, our licensor ReveraGen has received orphan drug designation for vamorolone by both the FDA and the EMA. We ourselves have obtained orphan drug designations for our marketed product, Raxone® (which is outlicensed to Chiesi Group), in the EU (maximum duration until fall 2025), the U.S. and South Korea, and for lonodelestat in Alpha-1 antitrypsin deficiency, Primary ciliary dyskinesia and CF in the EU.

With respect to patent protection, we are, depending on availability and strategic importance, pursuing patents for composition of matter and method of use patents as well as new formulations for drug production.

There are to our knowledge no composition of matter patents or patent applications with respect to vamorolone, which we have in-licensed from ReveraGen. ReveraGen has been granted method of use patents for the use of vamorolone in DMD and approximately 20 other indications in the U.S., the EU, China, Japan and other jurisdictions. The main patents relating thereto are due to expire in 2029. In addition, as the invention was made with support from the U.S. Army Medical Research and Materiel Command (USAMRMC) as well as the National Institutes of Health, the U.S. government has reserved certain rights to vamorolone. Relatedly, ReveraGen has to comply with certain formalities, including in particular the filing of certain information with governmental databases.

Polyphor holds composition of matter patents with respect to lonodelestat, which we have in-licensed and sublicensed, as applicable, from Polyphor on an exclusive basis, in the EU (expiring in 2025, with potential market protection until 2030 based on a supplementary protection certificate for 5 years after launch of commercial sale in the EU) and the U.S. (expiring in 2025, with the potential of a patent term adjustment for several years after launch of commercial sale in the U.S.). As we continue development we anticipate the ability to develop additional intellectual property.

Raxone®, our marketed product which is outlicensed to Chiesi Group, is not patent protected.

We rely on trade secrets, including proprietary know-how that is not patentable or that we elect not to patent, processes for which patents are difficult to enforce and any other elements of our technology and development processes that involve proprietary know-how, information or technology that is not covered by patents. In addition, we rely on our employees, advisors, third party contractors, consultants, collaboration partners as well as on CROs to develop and manufacture our product and product candidates.

We hold registered the trademarks Santhera and Raxone® in the EU, the U.S., and certain other countries. We hold registered trademarks or have filed trademark applications, which are pending, with respect to the logos of Santhera and Raxone® in the EU, the U.S., and certain other countries. We have outlicensed the trademark Raxone® to Chiesi Group under our outlicensing arrangement with Chiesi Group.

X. INFORMATION ON THE ISSUER

A. Name, registered office, incorporation

The Issuer, Santhera Pharmaceutical Holding AG, is a stock corporation (*Aktiengesellschaft*), incorporated under the laws of Switzerland and registered since July 16, 2002, in the commercial register of the Canton of Basel-Landschaft (register number CHE-105.388.338). The current domicile of the Issuer is Pratteln and its registered office is at Hohenrainstrasse 24, 4133 Pratteln, Switzerland.

B. Articles, business purpose

The Issuer's current articles of association (the "**Articles**") as at the date of this Prospectus are dated April 30, 2021.

The Issuer is a holding company and does not carry out operative activities. The Issuer's business purpose according to the Articles is to acquire, hold, permanently manage, dispose of and finance participations in and outside of Switzerland. The Issuer may establish branches and subsidiaries in and outside of Switzerland, may provide security for other group companies and enter into guaranty obligations, and may conduct any business that seems appropriate to foster or is related to its purpose. The Issuer may acquire, manage, exploit commercially and sell real estate and intellectual property in and outside of Switzerland and finance other companies.

C. Major shareholders

As at April 30, 2021, the shareholders of the Issuer holding at least 3% of its share capital registered in the Commercial Register of the Canton of Basel-Landschaft were, according to their latest disclosure notices pursuant to art. 120 et seq. FMIA, JP Morgan Chase & Co. (with which Highbridge is affiliated) (44.220% (including rights to receive Shares under the Bond), whereof 2.312% in Shares), Idorsia Pharmaceuticals Ltd (22.94%, whereof 8.08% in Shares), Fabrice Evangelista (14.83%, whereof 0.08% in Shares), and Ernesto Bertarelli and Donata Guichard-Bertarelli (beneficial owners of WDI Invest L.P.) (4.67%). Percentages indicated in this section relate to the share capital recorded in the commercial register at the time the respective reporting obligation was triggered. Current percentages may therefore be lower.

D. Administrative and management bodies

1. Board of directors

The ultimate responsible body for management and supervision of the Issuer is the board of directors (the "**Board**"). The members of the Board are:

Name	***Function***
Elmar Schnee	Chairman of the Board, member of the compensation committee
Philipp Gutzwiler	Member of the Board, member of the audit committee
Patrick Vink	Member of the Board, chairman of the compensation committee and member of the scientific committee
Dr. Thomas Meier	Member of the Board, chairman of the scientific committee
Martin Gertsch	Member of the Board, chairman of the audit committee

The business address of the members of the Board is Santhera Pharmaceuticals Holding AG, Hohenrainstrasse 24, 4133 Pratteln, Switzerland.

2. Executive management

The members of the Issuer's executive management (the "**Executive Management**") are:

Name	*Function*
Dario Eklund	Chief Executive Officer
Andrew Smith	Chief Financial Officer
Günther Metz	Head Business Development, Executive Vice President
Oliver Strub	Group General Counsel & Secretary to the Board, Executive Vice President

The business address of the members of the Executive Management is Santhera Pharmaceuticals Holding AG, Hohenrainstrasse 24, 4133 Pratteln, Switzerland.

E. Auditors

The Issuer's auditors are appointed on an annual basis by the Issuer's shareholder at the shareholders' meeting. The current auditors are Ernst & Young AG, Aeschengraben 27, 4051 Basel, Switzerland, who have acted as the Issuer's auditors for more than a decade. Ernst & Young AG is supervised and regulated by the Swiss Federal Audit Oversight Authority.

F. Notices to shareholders and Bondholders

In accordance with the Articles, unless personal notification is mandated by law, notices to shareholders are validly made by publication in the Swiss Official Gazette of Commerce (*Schweizerisches Handelsamtsblatt*). In contrast, notices in respect of the Bonds will be published in accordance with the Terms of the Bonds (Section VIII). Announcements concerning information with significant relevance for the price of the Shares or the Bonds will be made as required and by the means provided by the SIX Swiss Exchange ad-hoc publicity regulations.

G. Legal and regulatory proceedings

As of the date of this Prospectus, there are no pending or threatened court, arbitral or administrative proceedings that are of material importance to Santhera's assets and liabilities or profits and losses, other than the filings pending before medical product authorities and similar authorities and other proceedings discussed elsewhere in this Prospectus.

XI. INFORMATION ON THE ISSUER'S SHARE CAPITAL AND THE SHARES

A. Capital

1. Capital structure

The share capital of the Issuer as of the date of this Prospectus amounts to CHF 26,201,136 and is divided into 26,201,136 fully paid-up registered shares (*Namenaktien*), each with a par value of CHF 1 (each a "**Share**"), which form one single class of shares. The General Meeting of Shareholders may at any time convert registered shares into bearer shares (*Inhaberaktien*) and bearer shares into registered shares.

The Issuer's current authorized and conditional share capital as of the date of this Prospectus is disclosed in article 3a ("Genehmigtes Aktienkapital"), article 3b (""Bedingtes Aktienkapital für Mitarbeiterbeteiligungen") and article 3c ("Bedingtes Aktienkapital für Finanzierungen, Zusammenschlüsse und Unternehmensübernahmen") of the Articles (see "*ABOUT THIS PROSPECTUS—Documents incorporated by reference*").

2. Treasury shares

As of December 31, 2020, the Issuer held 1,301,075 Shares in treasury. As of May 3, 2021, the Issuer held 2,543,239 Shares in treasury, including the 233,194 Shares that will be delivered to holders of the 2017/22 Bonds who have accepted the Exchange Offer.

3. Asset transfer and contributions in kind

The Issuer's past contributions in kinds are disclosed in article 35 ("Sacheinlagen") of the Articles (see "*ABOUT THIS PROSPECTUS—Documents incorporated by reference*").

On the occasion of the authorized capital increase of September 29, 2009, pursuant to the authorization resolution of April 21, 2009, and pursuant to an agreement on contributions in kind dated September 25, 2009, the Issuer took over participations at an aggregate price of CHF 105,973, in consideration of which 105,973 Shares were issued.

B. The Shares

1. General description

The Shares are fully paid-up registered shares (*Namenaktien*) with a par value of CHF 1 each. By decision of the shareholders' meeting, registered shares may be converted into bearer shares (*Inhaberaktien*) and vice versa.

The Shares rank *pari passu* in all respects with each other, including in respect of entitlements to dividends, liquidation proceeds and pre-emptive rights.

Only persons registered in the share register are recognized as shareholders by the Issuer.

2. Form of Shares

The Shares are issued in the form of uncertificated securities (*Wertrechte*) within the meaning of article 973c CO and are maintained as book-entry securities (*Bucheffekten*) within the meaning of the Swiss Federal Intermediated Securities Act ("**FISA**"; *Bucheffektengesetz*).

3. Transfer restrictions

The Shares are freely transferable and the Issuer recognizes acquirers of Shares as shareholders with voting rights and records them as such in the share register, provided that they declare that they acquired the Shares in their own name and for their own account. Registration in the share register is made upon request. The Board may record persons (including individuals or entities acting in concert) who do not explicitly declare in their registration request that they hold Shares for their own account and who have entered into a nominee agreement with the Issuer ("**Nominees**") in the share register with the right to vote up to 2% of the share capital (as recorded in the commercial register). Shares held by a Nominee in excess of 2% of the share capital may be recorded with voting rights if the respective Nominee discloses the names, addresses and number of Shares of persons for whose account it holds such Shares. Since January 1, 2018, the Board has granted no exceptions to these rules.

4. Voting at shareholders' meetings

Subject to the restrictions described above, each Share carries one vote at the Issuer's shareholders' meetings. Voting rights may be exercised only after a shareholder has been recorded in the Issuer's share register as a shareholder with voting rights. Only shareholders, usufructuaries or nominees recorded with voting rights in the Issuer's share register may exercise a shareholder's voting and related rights on its behalf.

5. Opting out of mandatory tender offer rules

Under the Federal Act on Financial Market Infrastructures and Market Conduct in Securities and Derivatives Trading of June 19, 2015 (the **FMIA**), any person who acquires shares of a company whose shares are listed on a Swiss stock exchange, whether directly or indirectly or acting in concert with third parties, and, as a result, exceeds the threshold of 33 1/3% of the voting rights (whether exercisable or not) of such company, must submit a public tender offer to acquire 100% of the listed equity securities of such company, subject to certain exceptions. The FMIA allows companies to waive this requirement or raise the relevant threshold to up to 49% ("opting-out" and "opting-up", respectively) in their articles of association.

The Articles do not contain an opting-out or an opting-up provision.

6. Listing

The existing Shares are listed on the SIX Swiss Exchange in accordance with the International Reporting Standard.

7. Share price information

Information on past performance of the Shares is available free of charge for the entire term of the bond at: http://www.santhera.com/investors-and-media/investor-toolbox.

8. Security number, ISIN, common code and ticker symbol of the Shares

Swiss security number:	002714864
ISIN:	CH0027148649
Common Code	026905214
SIX Swiss Exchange ticker symbol	SANN

XII. GENERAL INFORMATION

A. Authorization

The issue of the Bonds and the issuance and delivery of Shares by the Issuer upon conversion of the Bonds was authorized by the resolutions of the Board passed on March 24, 2021 and May 3, 2021.

B. Issue of the Bonds and Exchange Offer

On March 25, 2021, the Company announced its Exchange Offer in respect of the 2017/22 Bonds. The press release and the "notice of a repurchase offer" containing the details of the Exchange Offer are incorporated in this Prospectus by reference. The holders of the 2017/22 Bonds who have accepted the Exchange Offer will, for each of their 2017/22 Bond, receive one Bond and 26 Shares on the Exchange Date. The entire issuance of the Bonds is used for the settlement of the Exchange Offer.

C. Source of Shares to be delivered upon conversion

The Shares potentially to be delivered under the 2021/24 Bonds will be Shares issued from the conditional capital or authorized capital, in an ordinary capital increase, or existing Shares of the Issuer, in each case with the same entitlements as the other outstanding Shares, except that (i) the Shares so delivered will not be entitled to any dividend or other distribution declared, paid or made by reference to a record date prior to the relevant conversion date and (ii) the voting rights may not be exercised unless the recipient of the Shares is registered with voting rights in the Issuer's share register.

D. Use of net proceeds

The Bonds are issued to holders of 2017/22 Bonds who have accepted the Exchange Offer. In exchange for the issuance of a new Bond together with 26 (twenty-six) Shares, the Issuer receives 1 (one) 2017/22 Bond back. The Issuer will not receive any cash proceeds from the offering of the Bonds.

E. Listing agent

In accordance with Article 58a of the Listing Rules of the SIX Swiss Exchange, Basler Kantonalbank has been appointed by the Issuer as representative to lodge the listing application with regard to the Bonds with the SIX Swiss Exchange.

F. Recent material developments

Other than as disclosed in this Prospectus, there has been no material change in the assets and liabilities, financial position or profits and losses of the Issuer since June 30, 2020.

G. Responsibility statement

The Issuer accepts responsibility for all information contained in this Prospectus and hereby confirms that to the best of its knowledge the information stated herein is correct and no material facts or circumstances have been omitted.

XIII. SWISS TAXATION

The following discussion is a summary of certain material Swiss tax considerations. The discussion is based on legislation as of the date of this Prospectus. It does not aim to be a comprehensive description of all the Swiss tax considerations that may be relevant for a decision to invest in Bonds. The tax treatment for each investor depends on the particular situation. All investors are advised to consult with their professional tax advisors as to the respective Swiss tax consequences of the purchase, ownership, disposition, lapse, exercise or redemption of Bonds in the light of their particular circumstances.

A. Taxation of the Bonds

1. Swiss federal withholding tax

The Bonds are classified as a "classical" convertible bond ("*klassische Wandelanleihe*") as described in Circular No. 15 issued by the Swiss Federal Tax Administration on October 3, 2017. Each payment of interest on the Bonds as well as payments which qualify as interest for Swiss withholding tax purposes (such as a potential issue discount or repayment premium, but not the repayment of principal) will be subject to deduction of 35% Swiss federal withholding tax (*Verrechnungssteuer*) by the Issuer.

A holder of a Bond who resides in Switzerland and who is the beneficial owner of a taxable payment on the Bond at the time such payment is due and, in the case of a holder who is an individual holding the Bond privately, duly reports the gross taxable payment in his or her tax return, and, in the case of a holder who is a legal entity, or who is an individual holding the Bond as part of a business situated in Switzerland for which he or she is required to keep accounting books, includes such payment as earnings in the income statement, is entitled to a full refund of or a full tax credit for the Swiss federal withholding tax, provided that certain other conditions are met.

A holder of a Bond who is resident outside Switzerland and who during the taxation year has not engaged in a trade or business carried on through a permanent establishment or fixed place of business in Switzerland and at the time a taxable payment on the Bond is due is the beneficial owner of the taxable payment may be able to claim a full or partial refund of the Swiss federal withholding tax by virtue of the provisions of a double taxation treaty, if any, between Switzerland and the country of residence of the holder.

2. Swiss stamp duties

The issuance and sale of the Bonds on the Issue Date will not be subject to Swiss federal securities turnover tax (*Umsatzabgabe*) (primary market).

Secondary market dealings in Bonds may be subject to the Swiss securities turnover tax at a rate of up to 0.15% of the purchase price of the Bonds, but only if a securities dealer in Switzerland or Liechtenstein, as defined in the Swiss Federal Act on Stamp Duties (*Bundesgesetz über die Stempelabgaben*), is a party or an intermediary to the transaction and no exemption applies. An exemption applies, *inter alia*, for each party to a transaction in Bonds that is not resident in Switzerland or Liechtenstein.

3. Swiss income taxation on principal or interest

a. Bonds held by non-Swiss holders

Payments of interest and repayment of principal by the Issuer to, and gain realized on the sale or redemption of Bonds by, a holder of a Bond who is not a resident of Switzerland and who during the current taxation year has not engaged in a trade or business through a permanent establishment in Switzerland to which such Bond is attributable will not be subject to any Swiss federal, cantonal or communal income tax in respect of such Bonds.

b. Bonds held as private assets by Swiss holders

Individuals who are resident in Switzerland and who hold Bonds as private assets are required to include all payments of interest (including discount, if any) on such Bonds in their personal income tax return for the relevant tax period and will be taxable on any net taxable income for such tax period.

The Bonds should be classified as "classical" convertible bonds (*klassische Wandelanleihe)* as described in Circular No. 15 issued by the Swiss Federal Tax Administration on October 3, 2017, for Swiss federal income tax purposes. In most cantons, the tax treatment for cantonal and municipal income tax will correspond to the Swiss federal tax treatment. Consequently, a capital gain is a tax-free private capital gain. Conversely, a loss on the Bonds is a non-tax-deductible private capital loss.

c. Bonds held as Swiss business assets (including by private persons classified as professional securities dealers)

Individuals who hold Bonds as part of a business in Switzerland and Swiss resident corporate taxpayers and corporate taxpayers resident abroad holding Bonds as part of a permanent establishment in Switzerland are required to recognize the payments of interest and any gain realized on the sale or redemption of Bonds and any loss on the Bonds in their income statement for the respective tax period and will be taxable on any net taxable earnings for such period. The same taxation treatment also applies to Swiss resident individuals who, for income tax purposes, are classified as "professional securities dealers" for reasons of, *inter alia*, frequent dealings or leveraged investments in shares or other securities.

B. Taxation of the Shares

1. Non-resident shareholders

Shareholders who are not resident in Switzerland for tax purposes and who, during the respective taxation year, have not engaged in a trade or business carried on through a permanent establishment or fixed place of business located in Switzerland for tax purposes who are not subject to corporate or individual income taxation in Switzerland for any other reason (all such shareholders, "**Non-Resident Shareholders**"), will not be subject to any Swiss federal, cantonal and communal income tax on dividends (or repayments of nominal value) paid to them with respect to Shares.

2. Resident private shareholders and domestic commercial shareholders

Shareholders who are Swiss residents (all such shareholders, "**Resident Private Shareholders**") and who hold Shares as private assets are required to include dividends (but not repayments of the nominal value of qualifying additional paid-in capital (*Kapitaleinlagereserven*) of Shares) in their personal income tax returns and are subject to Swiss federal, cantonal and communal income tax on any net taxable income for the relevant taxation period. Capital gains resulting from the sale or other disposition of Shares are not subject to Swiss federal, cantonal and communal income tax and, conversely, capital losses are not tax-deductible for Resident Private Shareholders.

Corporate and individual shareholders who hold Shares as part of a trade or business carried on in Switzerland or through a permanent establishment or fixed place of business located in Switzerland for tax purposes are required to recognize dividends (and repayment of nominal value) received on Shares and capital gains or losses realized on the sale or other disposition of Shares in their income statement for the respective taxation period and are subject to Swiss federal, cantonal and communal individual or corporate income tax, as the case may be, on any net taxable earnings for such taxation period. The same taxation treatment also applies to Swiss-resident private individuals who, for income tax purposes, are classified as "professional securities dealers" for reasons including frequent dealing, or leveraged investments, in shares and other securities (all shareholders referred to in this paragraph, "**Domestic Commercial Shareholders**"). Domestic Commercial Shareholders who are corporate taxpayers may be eligible for dividend relief (*Beteiligungsabzug*) in respect of dividends (and repayments of nominal value on

Shares) if Shares held by them as part of a Swiss business have an aggregate market value of at least CHF 1 million.

3. Swiss federal withholding tax

Dividends (if any) paid on Shares that are not a repayment of the nominal value of qualifying equity reserves from capital contributions (*Kapitaleinlagereserven*) of Shares are, with their gross amount, subject to Swiss federal withholding tax (*Verrechnungssteuer*) at a rate of 35%. The Issuer is required to withhold the Swiss federal withholding tax from such dividends and remit it to the Swiss Federal Tax Administration.

The Swiss federal withholding tax on a dividend will be refundable in full to a Resident Private Shareholder and to a Domestic Commercial Shareholder who, in each case, among other things, as a condition to a refund, duly reports the dividend in his or her individual income tax return as income or recognizes the dividend in his or her income statement as earnings, as applicable.

A Non-Resident Shareholder may be entitled to a partial refund of the Swiss federal withholding tax on a dividend if such Non-Resident Shareholder's country of residence for tax purposes has entered into a bilateral treaty for the avoidance of double taxation with Switzerland and the conditions of such treaty are met. Such shareholders should be aware that the procedures for claiming treaty benefits (and the time required for obtaining a refund) might differ from country to country.

4. Swiss federal stamp taxes

The issuance of Shares upon Conversion is subject to 1% Swiss issuance stamp duty (*Emissionsabgabe*).

A transfer of Shares in a secondary market transaction where a bank or another securities dealer in Switzerland (as defined in the Swiss Federal Stamp Tax Act) acts as an intermediary or is a party to the transaction may be subject to Swiss securities transfer tax (*Umsatzabgabe*) at an aggregate rate of up to 0.15% of the consideration paid for such Shares.

C. Automatic exchange of information

On November 19, 2014, Switzerland signed the Multilateral Competent Authority Agreement, which is based on article 6 of the OECD/Council of Europe administrative assistance convention and is intended to ensure the uniform implementation of automatic exchange of information (the "**AEOI**"). The Federal Act on the International Automatic Exchange of Information in Tax Matters (the "**AEOI Act**") entered into force on January 1, 2017. The AEOI Act is the legal basis for the implementation of the AEOI standard in Switzerland.

The AEOI is being introduced in Switzerland through bilateral agreements or multilateral agreements. The agreements have been, and will be, concluded on the basis of guaranteed reciprocity, compliance with the principle of specialty (*i.e.*, the information exchanged may only be used to assess and levy taxes (and for criminal tax proceedings)) and adequate data protection.

Based on such multilateral agreements and bilateral agreements and the implementing laws of Switzerland, Switzerland collects and exchanges data in respect of financial assets, including, as the case may be, Bonds or Shares, held in, and income derived thereon and credited to, accounts or deposits with a paying agent in Switzerland for the benefit of individuals resident in a EU member state or in a treaty state.

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