

Pratteln, May 27, 2021

Letter to Our Shareholders

Dear Shareholders,

Having emerged from a challenging 2020, we are pleased to have started 2021 with the strong foundation of a refreshed clinical and operational strategy.

We are committed to pursuing clinical development programs that aim to provide differentiated therapies to patients suffering from rare and severe diseases with high unmet medical needs. Our ambition to deliver a promising new treatment with Puldysa® (idebenone) to patients suffering from DMD was shattered following the disappointing interim analysis of the large Phase 3 SIDEROS trial, last October, and the resulting closure of the program. Moreover, it forced us to embark on a painful restructuring in which we had to say goodbye to the majority of our colleagues but allowed us to cut costs and align our Company with a focus on our future ambitions.

Our strategy is now focused on vamorolone, which we believe can provide significant value to patients, caregivers, and ultimately shareholders. It represents the key foundation for the future of Santhera, which continues to focus on Duchenne muscular dystrophy (DMD) and other rare diseases.

Vamorolone is currently being developed jointly by ReveraGen and Santhera for early stage DMD patients requiring an anti-inflammatory, muscle strengthening treatment with a differentiated safety and favorable tolerability profile to make it suitable for longer term administration. Recent encouraging data leads us to conclude that vamorolone could emerge as a foundational therapy in DMD for all patients irrespective of gene mutation and as a promising alternative to existing corticosteroids, the current standard of care in children and adolescent patients with DMD. Based on the collective clinical experience with vamorolone, we look forward with optimism to the readout of the 6-month top-line data from the pivotal VISION-DMD study, the next value enhancing inflection point. Subject to a positive outcome later this quarter, we will push ahead with the filing of a New Drug Application (NDA) with the US FDA and will step up preparations for market entry.

In terms of pursuing a vamorolone-lead strategy, securing financing was of prime urgency in 2020 and will remain a key priority in 2021. Various financing alternatives have already been put in place and have provided sufficient liquidity for a short period beyond the upcoming development milestone. This month, we completed the restructuring of our outstanding CHF 60 million 5% Convertible Bonds due 2022 (SAN17) with the conversion of 74.7% into new Senior Unsecured Convertible Bonds due 2024 (SAN21), thereby reducing our near-term debt obligation by approximately CHF 45 million and freeing up financials resources to fund operations. We are very pleased and grateful to you as Santhera's shareholders for already having approved, at the Extraordinary General Meeting held on March 18, a substantial capital increase, which was a prerequisite for said bond restructuring and is essential for fundraising post clinical readout.

For lonodelestat, our second clinical development candidate, data from a Phase 1b study were shared in March, which confirmed elastase inhibition in patients with cystic fibrosis (CF) and established an effective dose and a favorable tolerability profile. On this basis, the design of the further clinical study program for lonodelestat in cystic fibrosis has started alongside an evaluation of its potential in other chronic inflammatory conditions of the lung.

On the business development side, the Company is pursuing a proactive portfolio management strategy through out-licensing agreements for both vamorolone (in non-DMD indications and geographies outside the US and Europe) and lonodelestat (in non-CF indications). A further diversification of these platform-type pipeline products may offer an additional source of non-dilutive capital streams in the mid-term.

Subject to positive results from the pivotal VISION-DMD study, we are confident in our ability to successfully raise additional funds to advance the pipeline, prepare for commercialization and fund our operations. Preparing for the next growth phase of Santhera, as previously announced, the Board of Directors (BoD) proposes various capital increases to Santhera's shareholders at the forthcoming Annual General Meeting (AGM) on June 22, 2021. These provide the Company sufficient flexibility to raise additional capital to fund ongoing development activities, increase pre-commercialization activities and expand the organization in view of a US market launch of vamorolone in late 2022 upon FDA approval. The AGM agenda with further explanations on the motions by the BoD is enclosed and can also be viewed on the Santhera website [here](#).

We want to take a moment to recognize the huge efforts of Santhera employees over the last year, in both good times as well as battling their way through difficult phases without ever losing sight of our goals and our passion for delivering therapies to patients in need. We particularly also want to acknowledge the ongoing support of patients and their caretakers, our scientific and clinical partners, advisors and shareholders, without whom we couldn't continue our important research.

In just a year, we have completely transformed Santhera. We have new management, a new pipeline, a smaller, restructured company with lower fixed costs, new shareholders as well as a new and stronger balance sheet. Only very few stones have been left unturned. We have survived market downturns, loss of our lead program and the pandemic's disruption and now feel the new energy and are optimistic about our future. We understand the pain caused to shareholders and employees in 2020 but are also proud to have a future. We are ready to launch 'Santhera 2.0' on the back of positive topline results for vamorolone. We hope you also see the new potential that lies ahead of us—and hope that we can count on your support as a shareholder for many more years to come.

Sincerely,



Elmar Schnee
Chairman



Dario Eklund
Chief Executive Officer