Santhera Pharmaceuticals Holding AG
Hohenrainstrasse 24, 4133 Pratteln, Switzerland

Phone: +41 61 906 89 50 | Fax: +41 61 906 89 51
www.santhera.com

Santhera Obtains Worldwide Exclusive License from Polyphor to Develop and Commercialize Clinical Stage Candidate for Cystic Fibrosis and Other Pulmonary Diseases

Pratteln, Switzerland, February 15, 2018 – Santhera Pharmaceuticals (SIX: SANN) announces that it has entered into a license agreement with Polyphor Ltd. for POL6014, a clinical stage selective inhibitor of human neutrophil elastase with the potential to treat cystic fibrosis (CF) and other neutrophilic pulmonary diseases such as non-cystic fibrosis bronchiectasis (NCFB), alpha-1 antitrypsin deficiency (AATD) and primary ciliary dyskinesia (PCD).

Santhera will assume the global development, regulatory filings and commercialization of POL6014. The development program has been advanced with financial support by the Cystic Fibrosis Foundation Therapeutics Inc. (CFFT), USA, to Polyphor. With POL6014, Santhera is expanding its product pipeline in pulmonary diseases where the Company is already developing its lead product idebenone for respiratory complication in Duchenne muscular dystrophy.

Under the agreement, Santhera obtains the worldwide, exclusive rights to develop and commercialize POL6014, an innovative macrocycle elastase inhibitor, and analogs for an initial payment of CHF 6.5 million, payable in Santhera shares at an agreed valuation of CHF 27.2053 per share and additional cash payments of up to CHF 121 million contingent to future development, regulatory and particularly sales milestones. In addition, Polyphor is entitled to tiered royalty payments from Santhera´s future net sales of POL6014 and to undisclosed milestone payments and royalties provided that Santhera advances the development and market entry of POL6014 in other pulmonary diseases. Santhera expects to issue the 238,924 shares (3.8% of its currently issued shares) required for the initial payment to Polyphor out of its existing authorized share capital.

“With the licensing of POL6014 Santhera is broadening its clinical stage pipeline in rare diseases with the option to address multiple pulmonary indications,” said **Thomas Meier**, PhD, CEO of Santhera. “There is still a high unmet medical need in treating the chronic inflammation in patients with cystic fibrosis and other neutrophilic pulmonary diseases. In a first step, we intend to execute a multiple ascending dose (MAD) tolerability trial during the second half of 2018 which has already been planned by Polyphor. In parallel, we will initiate discussions with EU and U.S. regulators around the development program for POL6014 in CF and other indications.”

About POL6014

POL6014 is a highly potent and selective inhibitor of human neutrophil elastase (hNE) and was shown to reach high concentrations in the lung when administered by inhalation via an optimized eFlow® nebulizer (PARI Pharma GmbH). A first-in-man Phase 1 study in healthy volunteers and a Single Ascending Dose (SAD) safety and tolerability Phase 1 study in CF patients have successfully been completed with POL6014. The drug candidate was well tolerated and safe with a favorable pharmacokinetic profile and strong elastase inhibition as previously shown in animal models. In addition, POL6014 may show therapeutic benefit for a range of neutrophilic pulmonary diseases with high medical need such as non-cystic fibrosis bronchiectasis, alpha-1 antitrypsin deficiency or primary ciliary dyskinesia.

About Cystic Fibrosis

Cystic fibrosis (CF) is a rare, hereditary, life-threatening, progressive disease affecting approximately 70,000 patients in the U.S. and Europe and is characterized by persistent lung infection and chronic inflammation thereby limiting the ability to breathe over time. Activated or necrotic neutrophils liberate human neutrophil elastase (hNE) in the lung that causes damage to structural, cellular and soluble components of the pulmonary microenvironment. High levels of hNE play a central role in the pathophysiology of CF and correlate with disease severity as measured by functional lung parameters. Inhibition of hNE is expected to stop or slow the damage to lung tissue and may help to improve the overall quality of life for individuals with CF.

About Santhera

Santhera Pharmaceuticals (SIX: SANN) is a Swiss specialty pharmaceutical company focused on the development and commercialization of innovative medicines for orphan and other diseases with high unmet medical needs. The portfolio comprises clinical stage and marketed treatments for neuro-ophthalmologic, neuromuscular and pulmonary diseases. The most advanced pipeline product, idebenone, is in clinical Phase III for the treatment of Duchenne muscular dystrophy (DMD). Santhera's Raxone® (idebenone) is authorized in the European Union, Norway, Iceland, Liechtenstein and Israel for the treatment of Leber's hereditary optic neuropathy (LHON) and currently commercialized in 20 countries. For further information, please visit www.santhera.com.

Raxone® is a trademark of Santhera Pharmaceuticals.

For further information please contact:
public-relations@santhera.com or

Eva Kalias, VioConsult
Phone: +41 78 671 98 86
kalias@vioconsult.com

For Investors:
investor-relations@santhera.com or

Christoph Rentsch, Chief Financial Officer
Europe: +41 61 906 89 65
christoph.rentsch@santhera.com

Hans Vitzthum, LifeSci Advisors
US: +1 212 915 2568
hans@lifesciadvisors.com

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