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Santhera Signs Distribution and Supply Agreement for Raxone[®] with Ewopharma Covering Eastern Europe and The Baltics

Liestal, Switzerland, January 18, 2016 – Santhera Pharmaceuticals (SIX: SANN) announces that it has signed an agreement with Ewopharma to launch Raxone[®] for the treatment of Leber's hereditary optic neuropathy (LHON) in a number of Eastern European countries (Bulgaria, Croatia, Czech Republic, Hungary, Poland, Romania, Slovakia and Slovenia) and the Baltics (Estonia, Latvia and Lithuania). LHON is a rare, heritable, mitochondrial disease that leads to rapid, profound and usually permanent blindness in otherwise healthy patients. Raxone is the first treatment option for LHON and the first approved therapy for a mitochondrial disease.

"The agreement with Ewopharma represents an important achievement for Santhera and underscores our commitment to making Raxone available to patients across all EU member states," said **Giovanni Stropoli**, Santhera's Chief Commercial Officer for Europe. "We are confident that Ewopharma's expertise in the field of rare diseases, coupled with their understanding of the region, will greatly benefit patients with this devastating disease."

"As an organization historically present in Eastern European and Baltic countries with a specific focus on niche indications and diseases with high unmet medical need, we are very excited to have an opportunity to team up with Santhera to offer for the first time a treatment to patients with LHON," said **Alain Staub**, CEO and Chairman of the Board of Ewopharma. "We will make every effort to introduce Raxone as quickly as possible in these countries."

About Leber's Hereditary Optic Neuropathy and the Therapeutic Use of Raxone

LHON is a heritable genetic disease causing blindness. The disease presents predominantly in young, otherwise healthy adult males as rapid, painless loss of central vision, usually leading to permanent bilateral blindness within a few months of the onset of symptoms. About 95% of patients harbor one of three pathogenic mutations of the mitochondrial DNA, which cause a defect in the complex I subunit of the mitochondrial respiratory chain. This defect leads to decreased cellular energy (ATP) production, increased reactive oxygen species (ROS) production and retinal ganglion cell dysfunction, which cause progressive loss of visual acuity and blindness.

Raxone (idebenone), a synthetic short-chain benzoquinone and a cofactor for the enzyme NAD(P)H:quinone oxidoreductase (NQO1), circumvents the complex I defect, reduces and scavenges ROS, restores cellular energy levels in retinal ganglion cells and promotes recovery of visual acuity.

Raxone is an oral medication, authorized at a daily dose of 900 mg (given as 2 tablets three times a day with food), for the treatment of visual impairment in adolescent and adult patients with LHON. Treatment should be initiated and supervised by a physician with experience in LHON. Efficacy data

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come from Santhera's randomized, placebo-controlled RHODOS trial and from the open label Expanded Access Program, which together have demonstrated that vision loss can be mitigated or reversed in patients treated with Raxone.

About Santhera

Santhera Pharmaceuticals (SIX: SANN) is a Swiss specialty pharmaceutical company focused on the development and commercialization of innovative pharmaceutical products for the treatment of orphan mitochondrial and neuromuscular diseases. Santhera's lead product Raxone® is authorized in the European Union for the treatment of Leber's hereditary optic neuropathy (LHON). Santhera develops Raxone® in two additional indications, Duchenne muscular dystrophy (DMD) and primary progressive multiple sclerosis (ppMS), and omigapil for congenital muscular dystrophy (CMD), all areas of high unmet medical need. For further information, please visit the Company's website www.santhera.com.

About Ewopharma

Ewopharma AG, headquartered in Schaffhausen (Switzerland), is a pharmaceutical marketing company with over 270 employees and focused on the markets of Central Eastern Europe, where it has 15 local offices. With more than 50 years presence in the region, Ewopharma has gained extensive knowledge of these countries, which has enabled Ewopharma to achieve a privileged position in the area. Ewopharma activities cover all facets of the registration and marketing of ethical and over the counter pharmaceuticals. The company offers its services to partner companies either as a package or tailored to their specific needs. Ewopharma is also a member of the RX-Alliance, a professional cooperation network of successful pharmaceutical companies active in selected European markets. Further information is available at www.ewopharma.com.

Raxone® and Catena® are trademarks of Santhera Pharmaceuticals.

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