

Santhera Announces Closing of Licensing Transaction with Chiesi Group for Raxone® in LHON

Pratteln, Switzerland, August 2, 2019 – Santhera Pharmaceuticals (SIX: SANN) announces the closing of the licensing transaction with Chiesi Farmaceutici, an international research-focused healthcare group (Chiesi Group). As per the agreement, Chiesi Group has in-licensed Raxone® for the treatment of Leber’s hereditary optic neuropathy (LHON) and the initial payment of EUR 44 million of the total consideration of up to EUR 93 million became due.

“We are pleased to have closed the transaction so quickly and efficiently. We will continue to work closely with Chiesi Group in the coming months to ensure a smooth transition and a seamless supply of Raxone to LHON patients,” said **Thomas Meier, PhD, Chief Executive Officer of Santhera**. “As previously announced, the agreement with Chiesi Group enables us to focus on business areas core to our long-term growth strategy through advancing our clinical-stage neuromuscular and pulmonary programs.”

With the closing, Santhera has licensed its rights for the development, commercialization and distribution of Raxone for the treatment of LHON and any other potential ophthalmological indications to Chiesi Group for all territories worldwide except the US and Canada. In an interim phase, Santhera will provide support services to Chiesi Group to enable a seamless handover of the business and will continue to commercialize Raxone for LHON in France.

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

Leber’s hereditary optic neuropathy (LHON) is a heritable genetic disease causing profound vision loss and 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. Current data demonstrate that a majority of patients benefit from treatment and are protected from progression of visual acuity loss or experience a clinically relevant recovery of visual acuity.

Raxone for the treatment of LHON was granted orphan drug status in the EU, US, Switzerland and South Korea.

About Santhera

Santhera Pharmaceuticals (SIX: SANN) is a Swiss specialty pharmaceutical company focused on the development and commercialization of innovative medicines for rare neuromuscular and pulmonary diseases with high unmet medical need. Santhera is building a leading Duchenne muscular dystrophy (DMD) franchise. A marketing authorization application for Puldysa® (idebenone) is currently under review by the European Medicines Agency. Santhera has an option to license vamorolone, a first-in-class dissociative steroid currently investigated in a pivotal study in patients with DMD to replace standard corticosteroids. The clinical stage pipeline also includes POL6014 to treat cystic fibrosis (CF) and other neutrophilic pulmonary diseases, as well as omigapil and an exploratory gene therapy approach targeting congenital muscular dystrophies. Santhera out-licensed ex-North American rights to its first approved product, Raxone® (idebenone), for the treatment of Leber's hereditary optic neuropathy (LHON) to Chiesi Group. For further information, please visit www.santhera.com.

Raxone® and Puldysa® are trademarks of Santhera Pharmaceuticals.

About Chiesi Group

Based in Parma, Italy, Chiesi Farmaceutici is an international research-oriented group with over 80 years' experience in the pharmaceutical sector, and is present in 27 countries. The group researches, develops and commercialises innovative medicines in the respiratory disease, special care and rare disease therapeutic areas. The Group's Research & Development centre is based in Parma (Italy) and integrated with 6 other important research and development groups in France, the USA, the UK and Sweden, to promote its pre-clinical, clinical and registration programmes. The Group employs around 5,700 people. www.chiesi.com.

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