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ValuationLAB issues research report on Santhera

Liestal, Switzerland, June 24, 2014 – Santhera Pharmaceuticals (SIX: SANN) today announced that the independent life science research company *ValuationLAB AG* has issued a research report on the Company which estimates a current “risk-adjusted NPV (net present value) for its late-stage development pipeline of CHF 114 per share”.

The valuation is based on the Company's lead compound Raxone[®]/Catena[®] (INN: idebenone) as the key value driver in its first two indications: Leber's Hereditary Optic Neuropathy (LHON) and Duchenne Muscular Dystrophy (DMD).

LHON is a rare genetic disease that without treatment usually leads to blindness. Santhera filed a marketing authorization application (MAA) for Raxone[®]/Catena[®] for LHON in the EU in May 2014 and will now prepare to file for approval in the US. Santhera already holds a temporary authorization for Raxone[®] in France and ValuationLAB anticipates that Raxone[®]/Catena[®] will become the first-ever drug approved for the treatment of LHON in the EU and North America, estimating that peak sales could amount to CHF 70 million.

Based on the recent announcement that its pivotal phase III DELOS trial demonstrated that treatment with Raxone[®]/Catena[®] delays respiratory function loss in patients not using steroids, ValuationLAB expects Santhera to prioritize a new drug application (NDA) for DMD in the US and apply for a variation on the LHON label once approved in the EU. Respiratory function loss is the main cause of death in DMD and represents an urgent and unmet medical need. Once approved in this indication, ValuationLAB estimates that Raxone[®]/Catena[®] could be used in approximately 40% of all DMD patients and generate peak sales of CHF 560 million in the EU and North America.

LHON and DMD are orphan indications with no approved therapies and a large overlap of specialists (neurologists) facilitating a fast market entry and ValuationLab assumes EU and US approvals and product sales starting in late 2015/early 2016.

ValuationLab's current NPV estimation includes no contribution from Raxone[®]/Catena[®] in the third target indication, Primary Progressive Multiple Sclerosis (PPMS) or for other clinical stage pipeline projects that could provide substantial upside when developed successfully. Santhera is investigating the efficacy of Raxone[®]/Catena[®] in patients with PPMS in a phase I/II trial run by the US National Institutes of Health (NIH). With no approved treatment for PPMS, ValuationLAB estimates that peak sales for Raxone[®]/Catena[®] in this indication could exceed CHF 1.5 billion.

Thomas Meier, Chief Executive Officer of Santhera, commented: “This first analyst report follows closely after the announcement of exciting results for our lead compound which sparked tremen-

dous renewed investor interest. We hope that additional analysts will now start to actively track our company, providing useful guidance to the investment community.”

The full analyst research report is available on www.valuationlab.com and on Santhera's website at www.santhera.com under *Investors/Analyst Reports*.

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About ValuationLAB and the report

ValuationLAB AG is an independent life science research boutique with no securities or banking services. The company does not hold any positions in the securities mentioned in its reports. Its financial analyses are based on the *Directives on the Independence of Financial Research* issued by the Swiss Bankers Association in January 2008. The research report on Santhera has been commissioned by Santhera Pharmaceuticals Holding AG and prepared and issued by ValuationLAB AG. The analyst views and expectations were made independently and as such do not necessarily reflect those of Santhera.

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 develops Catena[®]/Raxone[®] as treatment for patients with Leber's Hereditary Optic Neuropathy (LHON), Duchenne Muscular Dystrophy (DMD) and Primary Progressive Multiple Sclerosis (PPMS), all areas of high unmet medical need for which no therapies are currently available. For further information, please visit the Company's website www.santhera.com.

Raxone[®] and Catena[®] are trademarks of Santhera Pharmaceuticals.

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