Press release

NeuroVive Pharmaceutical AB (publ)
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NeuroVive out-licenses targeted LHON therapy to BridgeBio Pharma's new subsidiary Fortify Therapeutics

Lund, Sweden and Palo Alto, Calif., June 18, 2018 *– NeuroVive Pharmaceutical AB (Nasdaq Stockholm: NVP, OTCQX: NEVPF) and BridgeBio Pharma today jointly announced that BridgeBio has entered into an exclusive licensing agreement for a subset of succinate prodrug chemistry under NeuroVive's NVP015 program. BridgeBio also announced that it has launched a subsidiary company Fortify Therapeutics to further develop this chemistry for local treatment of Leber's Hereditary Optic Neuropathy (LHON), with an initial financial commitment of $20 million USD. NeuroVive's NVP015 program for other mitochondrial disorders will continue without any changes in focus or timelines.*

LHON is caused by mitochondrial DNA mutations in subunits of NADH dehydrogenase (complex I), leading to reduced oxidative phosphorylation and energy production in retinal cells. The disease predominantly affects young adults, and results in sudden onset of progressive and severe vision loss. The licensed succinate prodrugs have the potential to overcome the disease by bypassing the dysfunctional metabolic pathway, providing an alternate source of energy to the retinal cells.

"As a targeted treatment for a genetic disease, the LHON program is a clear fit with the BridgeBio model," said Neil Kumar, Ph.D., CEO of BridgeBio. "We have been impressed with the ability of these compounds to rescue specific genetic mitochondrial deficiencies, and we have assembled a team of international experts to further develop a subset of the NVP015 chemistry to address this devastating disease."

Fortify Therapeutics will develop selected lead compounds derived from NeuroVive's novel NVP015 succinate prodrug program into drug candidates for the localized treatment of LHON. These compounds have been selected because they have properties that make them suitable for delivery to the eye.

The licensing agreement for this particular subset of the NVP015 program has a total deal value of approximately $60 million USD, which includes limited initial funding for research, and later milestone payments and a single digit royalty stream, that are dependent on successful development and market approval.

"The agreement with BridgeBio is important to both NeuroVive and our innovative NVP015 program, as it validates the quality of the program, our business development model and potential in a variety of mitochondrial disorders," commented NeuroVive CEO Erik Kinnman, M.D., Ph.D. "We will work closely with BridgeBio to further develop this chemistry subset and make the LHON program successful. It is important to note that our intentions for the NVP015 program are unchanged, and we are progressing towards experimental proof-of-principle during 2018."

This information is information that NeuroVive Pharmaceutical AB (publ) is obliged to make public pursuant to the EU Market Abuse Regulation. The information was submitted for publication, through the agency of the contact person set out below, at 08:30 a.m. CET on 18 June 2018.

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About LHON
Leber's Hereditary Optic Neuropathy (LHON) is a disease caused by mitochondrial DNA mutations in subunits of NADH dehydrogenase (complex I), a component of the electron transport chain. This results in dysfunctional oxidative phosphorylation and ATP production, leading to degeneration of the retinal ganglion cells and loss of central vision. LHON most commonly affects males in their second or third decade of life. The prevalence of LHON in Europe is between 1:30,000-1:50,000.

About NVP015
One of the most common causes of mitochondrial diseases relates to Complex I dysfunction, i.e. when energy conversion in the first of the five protein complexes in the mitochondrion that are essential for effective energy conversion does not function normally. This is apparent in disorders including Leigh's Syndrome and MELAS, both of which are very serious diseases with symptoms such as muscle weakness, epileptic fits and other severe neurological manifestations. The NVP015 project is based on a NeuroVive innovation in which the body's own energy substrate, succinate, is made available in the cell via a prodrug technology. A prodrug is an inactive drug that is activated first when it enters the body by the transformation of its chemical structure. Results from the NVP015 project were published in the prestigious Nature Communications journal in August 2016.

About BridgeBio Pharma
BridgeBio is a privately held clinical-stage biotech company developing novel, genetically targeted therapies to improve the lives of patients. The BridgeBio approach combines a traditional focus on drug development with a unique corporate model, allowing rapid translation of early stage science into medicines that treat disease at its source. Founded in 2015 by a team of industry veterans, the company, based in Palo Alto, CA, has built a robust portfolio of nineteen transformative assets, each housed in its own subsidiary, ranging from pre-clinical to late stage development in multiple therapeutic areas including oncology, cardiology, neurology, dermatology and endocrinology. The company's focus on scientific excellence and rapid execution aims to translate today's discoveries into tomorrow's medicines.

About NeuroVive
NeuroVive Pharmaceutical AB is a leader in mitochondrial medicine, with one project in clinical phase II development for the prevention of moderate to severe traumatic brain injury (NeuroSTAT®) and one project in clinical phase I (KL1333) for genetic mitochondrial diseases. The R&D portfolio consists of several late stage research programs in areas ranging from genetic mitochondrial disorders to cancer and metabolic diseases such as NASH. The company's strategy is to advance drugs for rare diseases through clinical development and into the market. The strategy for projects within larger indications outside the core focus area is out-licensing in the preclinical phase. NeuroVive is listed on Nasdaq Stockholm, Sweden (ticker: NVP). The share is also traded on the OTCQX Best Market in the US (OTC: NEVPF).