Press release
NeuroVive Pharmaceutical AB (publ)
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NeuroVive presents first preclinical NV354 efficacy results in a model for mitochondrial disease

Lund, Sweden, 18 October 2018 – **NeuroVive Pharmaceutical AB** *(Nasdaq Stockholm: NVP, OTCQX: NEVPF)* **today announced that positive experimental results for NV354, NeuroVive's preclinical lead candidate in the NVP015 program for mitochondrial diseases, will be presented by Magnus Hansson, NeuroVive's Chief Medical Officer and Vice President Preclinical and Clinical Development, at the Cold Spring Harbor Laboratory (CSHL) Meeting – The Evolving Concept of Mitochondria: From Symbiotic Origins to Therapeutic Opportunities, on Long Island, New York, October 18-21, 2018.**

The presentation at the CSHL Meeting on October 19 will include *in vivo* efficacy data from an advanced experimental model for acute energy crisis involving complex I dysfunction. The initial results show that NV354 restores tissue succinate levels and reduces lactate levels. Further NV354 efficacy studies are ongoing using both short- and long-term treatment regimens.

A parallel evaluation of NV354 drug properties in a separate set of experiments demonstrated high oral bioavailability and efficient brain delivery. Based on these data, NeuroVive will broaden the further preclinical development of NV354 as a chronic therapy for conditions associated with genetic mitochondrial disease, in addition to a therapy for acute energy crisis. The focus on severe pediatric conditions, such as Leigh syndrome, involving dysfunction of mitochondrial respiratory complex I, will continue.

"This is indeed very promising first efficacy data in a model highly relevant to patients with mitochondrial disease. We expect additional ongoing efficacy studies to provide further guidance for future clinical development, including the extensive studies planned at the Children's Hospital of Philadelphia," said Magnus Hansson, NeuroVive's Chief Medical Officer and Vice President Preclinical and Clinical Development.

"The most exciting finding from the ongoing studies is that through its drug properties, NV354 demonstrates potential to treat not only acute, but also chronic conditions in patients with mitochondrial disease, which expands therapeutic opportunities as well as commercial potential," said NeuroVive's CEO Erik Kinnman.

The information was submitted for publication, through the agency of the contact person set out below, at 08:30 a.m. CEST on 18 October 2018

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About NV354
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 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. Within the project a lead compound, NV354, has been selected for further development in the program based on tolerability, oral bioavailability, plasma stability and organ delivery, specifically to the brain. In 2017 NeuroVive received a research grant from the Swedish innovation agency, Vinnova, for developing the succinate prodrugs as a new treatment for genetic mitochondrial diseases.

About genetic mitochondrial diseases
Genetic mitochondrial diseases are metabolic diseases that affect the ability of cells to convert energy. The disorders can manifest differently depending on the organs affected by the genetic defects and are viewed as syndromes. An estimated 12 in every 100,000 people suffer from a mitochondrial disease. Mitochondrial diseases often present in early childhood and lead to severe symptoms, such as mental retardation, heart failure and rhythm disturbances, dementia, movement disorders, stroke-like episodes, deafness, blindness, droopy eyelids, limited mobility of the eyes, vomiting and seizures.

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 also consists of projects for genetic mitochondrial disorders, cancer and NASH. The company advances drugs for rare diseases through clinical development into the market. For projects for common indications the goal is out-licensing in the preclinical phase. A subset of compounds under NeuroVive's NVP015 program has been licenced to Fortify Therapeutics, a BridgeBio company, for local treatment development of Leber's Hereditary Optic Neuropathy (LHON). NeuroVive is listed on Nasdaq Stockholm, Sweden (ticker: NVP). The share is also traded on the OTCQX Best Market in the US (OTC: NEVPF).