Press release

NeuroVive Pharmaceutical AB (publ)
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NeuroVive enrolls first subject in its KL1333 phase Ia/b clinical study

Lund, Sweden, 19 March 2019, **NeuroVive Pharmaceutical AB** *(Nasdaq Stockholm: NVP, OTCQX: NEVPF)* **today announced that the first healthy volunteer in the company's KL1333 phase Ia/b study has been screened and will be enrolled into the study.**

First subject first visit in NeuroVive's KL1333 phase Ia/b study was completed on 18 March 2019. The main aim of this second clinical KL1333 study is to further examine the safety profile of KL1333 and how the drug is metabolized following multiple doses in healthy volunteers and genetic mitochondrial disease patients. In addition, possible efficacy endpoints will be explored.

"This truly is an important project milestone for KL1333. During the past few months we have worked intensely with preparing for study start, including an optimization of the bioanalytical method" said Magnus Hansson, Chief Medical Officer and Vice President Preclinical and Clinical Development at NeuroVive.

"The recent successful share issues have secured the financing needed to take this novel treatment opportunity to its next milestone. We are excited about this study which will take the KL1333 project further towards our goal of offering it to patients with severe genetic mitochondrial disease with few or no treatment options," said Erik Kinnman, NeuroVive's CEO.

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 19 March 2019.

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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, limited mobility of the eyes, vomiting and seizures.

About KL1333
KL1333 is a potent modulator of the cellular levels of NAD^+, a central co-enzyme in the cell's energy metabolism. KL1333 has in preclinical models been demonstrated to increase mitochondrial energy output, reduce lactate accumulation, diminish the formation of free radicals and to have long-term beneficial effects on energy metabolism such as the formation of new mitochondria. It is in clinical development stage intended to document the use for chronic oral treatment in primary genetic mitochondrial disorders such as MELAS, KSS, PEO, Pearson and MERRF. KL1333 is currently being evaluated in clinical phase I studies and has been granted orphan drug designation in both the United States and Europe. KL1333 has been in-licensed from Yungjin Pharm, a Korean pharmaceutical company.

About the KL1333 phase Ia/b study
The primary purpose of the study is to investigate the pharmacokinetics, safety and tolerability of KL1333 in healthy volunteers and thereafter in patients with genetic mitochondrial disease. The study will be conducted in the UK and includes an assessment of a single dose in healthy volunteers, to bridge to the previously conducted single ascending dose study in South Korea, completed in 2018, and an assessment of the effect of food intake. The study will also include a multiple ascending dose part in healthy volunteers, and in patients with genetic mitochondrial disease.

About NeuroVive
NeuroVive Pharmaceutical AB is a leader in mitochondrial medicine, with one project in clinical phase I (KL1333) for genetic mitochondrial diseases and one project in clinical phase II development for the prevention of moderate to severe traumatic brain injury (NeuroSTAT®). The R&D portfolio also consists of projects for genetic mitochondrial disorders, NASH and cancer. The company advances drugs for rare diseases through clinical development into the market with or without partners. 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).