

## Press release

NeuroVive Pharmaceutical AB (publ)  
556595-6538



### NeuroVive signs collaboration agreement with Karolinska Institutet

**Lund, Sweden, January 10, 2017 - NeuroVive Pharmaceutical AB (Nasdaq Stockholm: NVP, OTCQX: NEVPF), the mitochondrial medicine company, today announced that a collaboration agreement has been signed with Karolinska Institutet, Stockholm, Sweden, regarding development of the company's compound NV556 for the treatment of mitochondrial myopathy.**

Under the collaboration agreement, the team led by Prof. Håkan Westerblad at Karolinska Institutet will study NeuroVive's cyclophilin inhibitor compound NV556 and its effects in experimental models of mitochondrial myopathy.

The Karolinska Institutet research team previously published results<sup>1)</sup> where they show that another cyclophilin inhibitor, ciclosporin, shows mitochondrial protective effects mediated through inhibition of cyclophilin D, resulting in prevention of muscle fiber weakness in an experimental model of mitochondrial myopathy. They have also shown that patients with mitochondrial myopathy have increased levels of cyclophilin D, the target for NeuroVive's compound NV556. NV556 is expected to have higher specificity and tolerability than ciclosporin, which may facilitate better optimization of dosage.

"We are now adding mitochondrial myopathy to our genetic mitochondrial disease portfolio, which is very exciting. We are also very glad to be able to explore this area together with Håkan Westerblad's skilled team at Karolinska Institutet and are looking forward to our continued collaboration", said Magnus Hansson, Chief Medical Officer at NeuroVive.

This collaboration expands NeuroVive's presence in the genetic mitochondrial disease field, which is a NeuroVive key focus area and where NeuroVive also develops its NVP015 compounds in mitochondrial respiratory chain diseases. The NV556 compound that is studied under this collaboration is targeting cyclophilin D, and has therefore a different and complementary mode of action than the NVP015 compounds which target the respiratory chain supporting cell energy production.

#### Notes to editors

##### About Mitochondrial myopathy

Mitochondrial myopathies are a group of neuromuscular diseases caused by damage to the mitochondria, the small energy factories found inside almost all the cells in the body. Some of the more common mitochondrial myopathies include Kearns-Sayre syndrome, myoclonus epilepsy with ragged-red fibers, and mitochondrial encephalomyopathy with lactic acidosis and stroke-like episodes. The symptoms of mitochondrial myopathies include muscle weakness, exercise intolerance and fatigue, and are often accompanied by other symptoms of genetic mitochondrial disorders such as heart failure or rhythm disturbances, dementia, movement disorders, stroke-like episodes, deafness, blindness, droopy eyelids, limited mobility of the eyes, vomiting, and seizures. The prognosis for these disorders ranges in severity from progressive weakness to death.<sup>2)</sup> There is a high unmet medical need of new and effective treatment options for mitochondrial myopathy.

1) Cyclophilin D, a target for counteracting skeletal muscle dysfunction in mitochondrial myopathy. Westerblad H. et al. *Human Molecular Genetics*, 2015, Vol.24, No 23; 6580-6587.

2) [http://www.ninds.nih.gov/disorders/mitochondrial\\_myopathy/mitochondrial\\_myopathy.htm](http://www.ninds.nih.gov/disorders/mitochondrial_myopathy/mitochondrial_myopathy.htm)

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### About NeuroVive

NeuroVive Pharmaceutical AB is a leader in mitochondrial medicine. The company is committed to the discovery and development of medicines that preserve mitochondrial integrity and function in areas of unmet medical need. The company's strategy is to take 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 enhances the value of its projects in an organization that includes strong international partnerships and a network of mitochondrial research institutions, as well as expertise with capacities within drug development and production.

NeuroVive has a project in early clinical phase II development for the prevention of moderate to severe traumatic brain injury (NeuroSTAT®). NeuroSTAT has orphan drug designation in Europe and in the US. 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.

NeuroVive is listed on Nasdaq Stockholm, Sweden (ticker: NVP). The share is also traded on the OTCQX Best Market in the US (OTC: NEVPF).

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