# The board of directors' report referred to in Chapter 13, Section 6 and Chapter 14, Section 8 of the Swedish Companies Act

The board of directors of NeuroVive Pharmaceutical AB (publ), reg. no. 556595-6538 (the "**Company**"), submits the following report as referred to in Chapter 13, Section 6 and Chapter 14, Section 8 of the Swedish Companies Act.

After the submission of the annual report for the financial year 2016, which includes the latest adopted balance sheet and profit and loss account, no events of material significance for the Company's financial position have occurred, except from what is stated in the Company's interim report for the period 1 January 2017 to 30 September 2017, **Appendix 1** and attached press releases, **Appendix 2-6** 

Place: Lund		
Date: 15 February 2018		
David Laskow-Pooley	Marcus Keep	
	_ Jan Törnell	

# Continued positive trend for NeuroSTAT®

# **Business operations**

# Significant events July-September 2017

- NeuroVive received positive feedback from the European Medicines Agency (EMA) regarding the NeuroSTAT development plan.
- NeuroVive signed a private placement agreement with Esousa Holdings LLC totalling SEK 4.5 million excluding transaction costs.
- NeuroVive hosted a Mitochondria Day at IVA in Stockholm to raise awareness of mitochondrial disorders.

# Important events after the end of the period

- NeuroVive received a positive opinion from the the European Medicines Agency's (EMA) Committee on Orphan Drugs (COMP) on granting orphan drug designation for KL1333.
- Greg Batcheller, NeuroVive's Chairman of the Board for the past 17 years, resigned on 6 November. David Laskow-Pooley was elected new Chairman.
- On November 3, NeuroVive conducted a directed new share issue to Floyd Associates Europe Limited totaling SEK 5.3 million excluding transaction costs.
- NeuroVive and Lund University were granted funding by the Swedish Foundation for Strategic Research (SSF) for collaboration on liver cancer research.
- NeuroVive signed a collaboration agreement with the University of Florida regarding the development of TBI biomarkers.
- NeuroVive presented the results from preclinical TBI trials related to the NeuroSTAT project at the 2017 Nordic Neurotrauma Conference.

- NeuroVive presented its innovative metabolic regulators for the non-alcoholic steatohepatitis (NASH) liver disease at the Annual Meeting of the American Association for the Study of Liver Diseases (AASLD) held on 20-24 October 2017, in Washington DC.
- NeuroVive's partner, the Children's Hospital of Philadelphia (CHOP), was awarded funding by the US National Institutions of Health (NIH) to study NeuroVive's NVP015 compounds as countermeasures against chemical threats.

# **Financial information**

# Third quarter (July-September 2017)

- Net revenues were SEK 0 (0) and other operating income was SEK 397,000 (16,000)
- Loss before tax was SEK 13,179,000 (loss: 34,290,000)
- Loss per share\* was SEK 0.26 (loss: 0.86)
- Diluted loss per share\*\* was SEK 0.26 (loss: 0.86)

# First nine months (January-September 2017)

- Net revenues were SEK 27,000 (0) and other operating income was SEK 550,000 (90,000)
- Loss before tax was SEK 56,824,000 (loss: 57,265,000)
- Loss per share\* was SEK 1.04 (loss: 1.42)
- Diluted loss per share\*\* was SEK 1.04 (loss: 1.42)
- \* Profit/loss for the period divided by average number of shares before dilution at the end of the period.
- \*\* Profit/loss for the period divided by average number of shares after dilution at the end of the period



# Comments from our CEO, Erik Kinnman

The third quarter of 2017 was both eventful and encouraging. The first clinical trial in our in-licensed project for genetic mitochondrial diseases, KL1333, is progressing, strengthened by a positive opinion on orphan drug designation by the European Medicines Agency's Committee for Orphan Medicinal Products (COMP). Furthermore, we received positive feedback from EMA to support our continued clinical development plans for our NeuroSTAT project, and additional clinical results from the project were presented at the Nordic Neurotrauma Conference in Lund, Sweden.

#### Encouraging developments in our NeuroSTAT project

In September, NeuroVive received positive feedback from EMA regarding the NeuroSTAT development plan. The support for using an imaging endpoint and a relatively homogenous subpopulation of TBI patients was particularly encouraging and will facilitate a more limited phase II clinical trial size. In October, NeuroVive signed a collaboration agreement with the University of Florida's McKnight Brain Institute to study the usefulness of TBI biomarkers by analyzing data from the recent NeuroSTAT clinical TBI study, CHIC, ahead of our upcoming NeuroSTAT clinical phase II exploratory efficacy study. The CHIC trial results were presented in November at the Nordic Neurotrauma Conference in Lund, Sweden.

# Orphan Drug Designation for KL1333

After the end of the period, we received positive news relating to our KL1333 project. The European Medicines Agency's Committee for Orphan Medicinal Products (COMP) has issued a favorable opinion on the classification of orphan drug designation for KL1333. The final decision will be made by the European Commission. Orphan drug designation is an important milestone for KL1333 as it verifies the quality of the project and contributes to a faster and less expensive route to the market. The access to scientific advice and reduced fees from the EMA are both very valuable, and the orphan medicinal product status that may follow will also support the commercialization of KL1333.

# Success in NASH and liver cancer projects

Out-licensing activities for NV556, NeuroVive's most advanced non-alcoholic steatohepatitis (NASH) project, have been intensified. Results from our second NASH project, NVP022, were presented at the annual meeting of the American Association for the Study of Liver Diseases, AASLD. In October, after the end of the period, we were happy to announce that our joint project with Lund Uni-



versity on hepatocellular liver cancer (HCC) research had been granted funding of SEK 2.5 million from the Swedish Foundation for Strategic Research (SSF).

# New applications for NVP015

Before the summer, NVP015, NeuroVive's preclinical project for genetic mitochondrial diseases, was granted close to SEK 1 million in funding from Vinnova for continued development. NVP015 is aimed at treating acute energy crises in patients suffering from genetic mitochondrial diseases. In October, NeuroVive's research partner, the Children's Hospital of Pennsylvania (CHOP), was awarded a grant from the National Institutes of Health (NIH) program, CounterACT, to study the potential of compounds from the NVP015 program in supporting mitochondrial function and preventing organ failure following immediate exposure to toxic chemicals.

In September, during the international week for raising awareness about mitochondrial diseases, NeuroVive hosted a mitochondrial research day in Stockholm. This is a part of the company's goal to contribute to an increased focus on genetic mitochondrial diseases and future therapies.

On 6 November, Greg Batcheller, NeuroVive's Chairman of the Board resigned. Greg has been NeuroVive's Chairman since the company was founded 17 years ago. His efforts have been very important for the company's development. We look forward to continuing working with the Board and its new Chairman David Laskow-Pooley to achieve continued research and development progress and reinforced business opportunities.

# Erik Kinnman

CEO, NeuroVive Pharmaceutical AB 21 November 2017



# **Operations**

NeuroVive is focused on the research and development of targeted drug candidates that maintain mitochondrial integrity and function for indications with a high unmet medical need. NeuroVive creates value in its projects by working in partnerships with leading research institutions in mitochondrial medicine as well as experts with resources in drug development and production. The drug development process is comprehensive and carefully regulated. By collaborating with various partners, NeuroVive strives to make this process as cost-efficient and successful as possible.

# Business model that creates value in therapies for rare and common diseases

NeuroVive is focused on research and development in mitochondrial medicine with the aim of helping patients for whom few, or no, treatment options are currently available.

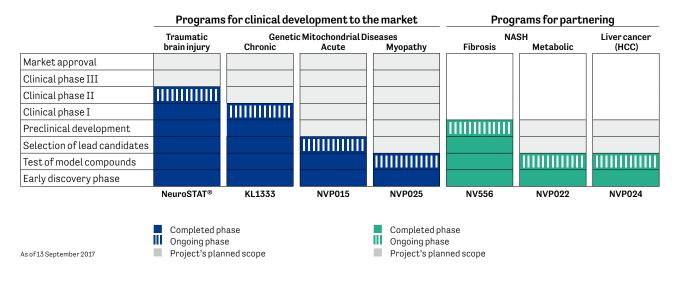
The Company has a two-sided business model. The first component comprises proprietary drug development for rare diseases with a major unmet medical need, from preclinical and clinical development to marketing authorization. The other component comprises projects for common diseases with high commercial potential, where the Company develops drug candidates for out-licensing at the preclinical phase.

The business model enables a diversified portfolio with opportunities for the Company to build value by bringing orphan drugs to market relatively fast, with lower cost and less risk. At the same time, innovation in common diseases is industrialized and value is created through cooperation with a capital and research-intensive partner.

# PROJECTS FOR CLINICAL DEVELOPMENT Traumatic brain injury (TBI)

Traumatic brain injury (TBI) is caused by external violence to the head resulting in immediate damage to nerve cells. The damage continues to worsen for several days after the trauma which, in many cases, has a significantly adverse effect on the overall injury and carries a risk for severe and lifelong impairment. At present, there are no approved treatments for the prevention of these secondary injuries. In the US, some 2.2 million people are affected annually, causing more than 50,000 deaths and 280,000 hospitalizations. The direct and indirect costs associated with TBI are an estimated USD 60 billion, and many patients suffer moderate to severe functional disabilities requiring intensive care and various forms of support (www.nih.gov). The hope is that better preventive therapies for secondary brain damage, such as NeuroSTAT, will lead to higher survival rates, and significantly improve quality of life and neurological function of patients post-TBI.

# **Project overview**





#### **NeuroSTAT®**

Results from the Copenhagen Head Injury Ciclosporin (CHIC) Phase II clinical trial, in which NeuroSTAT was assessed after clinical stabilization, show that appropriate dose-dependent concentration levels can be measured in blood and the target organ, namely the central nervous system (CNS). The registered safety profile was as expected. Thus, the primary objective of CHIC, which was to demonstrate the safety and elucidate the pharmacokinetics of NeuroSTAT at two different dose levels (5 and 10 mg/kg/day) in patients with severe TBI, was reached. Results from the detailed analyses were presented at the Nordic Neurotrauma conference in Lund on 15 November.

In the experimental TBI studies performed in cooperation with the University of Pennsylvania, the extent of brain damage measured by MRI decreased significantly (35%) after treatment with NeuroSTAT. In addition, these studies demonstrated positive changes in the energy metabolism of the brain and improved mitochondrial function, as well as reduced production of free radicals. Results from the studies were presented at the Neurotrauma 2017 conference, co-hosted annually by the National Neurotrauma Society and the American Association of Neurological Surgeons/CNS Joint Section on Neurotrauma and Critical Care.

The combined outcome of results from the clinical and preclinical studies will enable NeuroVive to proceed to the next stage of the clinical development program. The Company is currently focusing all TBI project resources on preparation for the next clinical trial with NeuroSTAT for TBI.

The preliminary development plan has been discussed with the European Medicines Agency (EMA). The Company received a positive recommendation for the planned development program, including the selection of efficacy variables and study population of the next trial. This enables a limited sample size for the study population in the planned exploratory Phase II clinical trial to evaluate efficacy.

After the end of the reporting period, NeuroVive signed a cooperation agreement with the University of Florida regarding the development of TBI biomarkers. The research will be conducted at the University of Florida's McKnight Brain Institute, which is focused on neurotrauma, neuroproteomics and biomarkers research, under the supervision of Dr. Kevin K.W. Wang, Ph.D. and Associate Professor of Psychiatry, Neuroscience and Physiological Science at the University of Florida College

of Medicine, and member of the McKnight Brain Institute. The research will evaluate the use of blood and cerebrospinal fluid-based biomarkers in drug development for TBI and samples taken during NeuroVive's CHIC study will be used in these studies. Biomarkers are considered highly significant for the diagnosis, prognosis and evaluation of treatment efficacy for TBI. Results from the biomarker analysis will help to further optimize NeuroVive's forthcoming Phase II clinical trial to evaluate the efficacy of NeuroSTAT.

#### Genetic mitochondrial disorders

Genetic mitochondrial disorders are congenital metabolic diseases that affect the ability of cells to convert energy. The disorders can manifest differently depending on the organs affected by the mitochondrial genetic mutations and are described as syndromes, depending on the organs affected and how the signs and symptoms occur.

An estimated 12 in every 100,000 people suffer from a mito-chondrial disease. Mitochondrial disorders usually present in early childhood. All projects in this area (KL1333, NVP015, and NVP025) may qualify for orphan drug designation in the US and Europe prior to clinical development, enabling a faster, less costly, and less risky route to market, as well as a higher market price. In 2016, the orphan drug market amounted to USD 114 billion and the average annual cost for the treatment of a single patient in the US in the same year was an estimated USD 140,443 (just over SEK 1.3 million). 1)

1) Evaluate Pharma Orphan Drug Report 2017

# KL1333

In May 2017, the KL1333 clinical development project was in-licensed from the South Korean pharmaceutical company Yungjin Pharm Corporation Ltd. The KL1333 compound has been developed for the treatment of rare genetic mitochondrial disorders, such as MELAS and Kearns-Sayre syndrome, for which there is no drugs currently available.

A Phase I clinical trial of KL1333 has been ongoing in South Korea since June. The trial is a double-blind, place-bo-controlled, single-dose Phase I dose-escalation trial to evaluate the pharmacokinetics, safety and tolerability of KL1333 in healthy volunteers. The Phase I trial in South Korea is led exclusively by Yungjin Pharm, with expert support from NeuroVive. In 2018, NeuroVive is planning to commence a complementary European and/or US-based Phase I trial built on the ongoing trial in South Korea.

Under the agreement, NeuroVive has exclusive rights to the global development and commercialization of KL1333,



except in South Korea and Japan for which Yungjin Pharm has retained all commercialization and marketing rights. NeuroVive paid an initial fee of USD 1 million to Yungjin Pharm upon signing the agreement, an additional fee of USD 1 million will be due one year after signing plus another USD 1 million after a successful Phase I clinical trial. Further payments will be made in conjunction with the successful achievement of various clinical milestones and milestones linked to marketing authorization, pricing and reimbursement. Both companies will develop KL1333 within their own territories, primarily for the treatment of genetic mitochondrial disorders, while cooperating to achieve the clinical milestones as efficiently as possible. This will take place through regular exchanges of results, plans and expert contacts. The documentation produced in the various regions will be used to support further development and for contact with regulatory agencies regardless of where this takes place.

After the end of the period, NeuroVive received a favourable opinion from the European Medicines Agency EMA's Committee for Orphan Drugs (COMP) for Orphan Drug Designation within the EU for KL1333. The statement by the COMP is the basis for a formal decision taken by the European Commission. Orphan Drug Designation has a significant impact on the development of the project, among other things through free scientific advice and reduced application fees from the EMA. Orphan medicinal product status, which may follow when the drug is granted marketing approval, would be very valuable for the project's continued commercialization, for instance through a 10-year market exclusivity within the EU.

# About KL1333

KL1333 is a powerful regulator of cellular NAD+ levels, a coenzyme central to cellular energy metabolism. In preclinical studies, KL1333 has been shown to increase mitochondrial energy production, reduce lactate accumulation, prevent the formation of free radicals, and have long-lasting positive effects on energy metabolism and the formation of new mitochondria. The drug candidate is intended for chronic oral treatment of symptoms, and to prevent the effects, of genetic mitochondrial disorders such as MELAS, KSS, CPEO, PEO, Pearson and MERRF. The NVP015 project is intended to support acute episodes of energy crisis in genetic mitochondrial disorders with complex I dysfunction, and its mechanism of action and function therefore complement KL1333. NVP025 is intended to protect the mitochondria in skeletal muscles from improper calcium handling, and the subsequent muscular dystrophy has a completely different mechanism of action and presents an additional opportunity for treating patients with genetic mitochondrial disorders.

# NVP015 - for complex I dysfunction

Results from experimental studies on the novel series of prodrugs developed by researchers at NeuroVive and Isomerase demonstrate that these compounds show good stability in the bloodstream and uptake by target organs such as muscle tissue. The prodrugs release the energy substrate succinate (succinic acid) and experiments with labeled compounds show that delivered succinate undergoes further mitochondrial energy metabolism, which is an important milestone for the project. The most promising compounds from this series are currently undergoing further testing in various experimental models and the selection of an optimized candidate for continued preclinical development is expected by the end of 2017.

In cooperation with Marni J. Falk, M.D. at the Children's Hospital of Philadelphia (CHOP), compounds from NVP015 are being evaluated in various advanced experimental models of disease, and energy metabolism and disease progression in models of mitochondrial complex I dysfunction are being studied. Marni J. Falk is an Attending Physician and Director of the Mitochondrial Disease Clinical Center at CHOP, a major center for children and adults with mitochondrial disorders, and a leader in this field of research. Dr. Falk's experience ranges from early-phase research to clinical development, with expertise across the entire drug development spectrum. CHOP is one of the largest children's hospitals in the world and one of the highest-ranked children's hospitals in the US.

NVP015 also holds potential for other applications. After the reporting period, it was announced that researchers at CHOP, under the supervision of Dr. Todd Kilbaugh, will examine how NVP015 can support mitochondrial function and contribute to recovery, and prevent organ failure resulting from exposure to toxic chemicals, with a research grant from the NIH Countermeasures Against Chemical Threats (CounterACT) program. By bypassing the first complex of the mitochondrial respiratory chain, which is often affected by chemical toxicity, NVP015 is an ideal candidate to explore as a pharmacological treatment option for exposure to certain toxic chemicals, such as pesticides and chemical agents.

# 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 central to effective energy conversion does not function normally. This has been observed in Leigh syndrome and MELAS, two very serious diseases with symptoms including muscle weakness, epilepsy and other severe neurological



manifestations. The NVP015 project is based on a concept developed by NeuroVive's CSO Associate Professor Eskil Elmér and his colleagues, whereby the body's own energy substrate, succinate, is made available inside the cell using a prodrug technology. A prodrug is an inactive drug that is only activated when it enters the body through the transformation of its chemical structure. Results from the NVP015 project were published in the prestigious Nature Communications¹) journal in August 2016.

1) Ehinger JK et al. (2016) Nat. Commun.7:12317

# NVP025 - Mitochondrial myopathies

In January 2017, a partnership agreement was signed with Karolinska University Hospital in Stockholm regarding the development of a therapeutic option for mitochondrial myopathies. Under the partnership agreement, the research team at Karolinska University Hospital, headed by Professor Håkan Westerblad, will be using Neuro-Vive's cyclophilin inhibitor NV556 as a model compound and studying its effects in experimental models of mitochondrial myopathy. The research team at Karolinska University Hospital has previously published results<sup>1)</sup> showing that another cyclophilin inhibitor, cyclosporine, exhibits mitochondrial protective effects by inhibiting cyclophilin D and thus preventing muscle fiber weakness in an experimental model of mitochondrial myopathy. They have also demonstrated that patients with mitochondrial myopathies have elevated levels of cyclophilin D, a target molecule for NeuroVive's NV556 compound. A compound such as NV556 is expected to have higher specificity and tolerability than cyclosporine, which should facilitate efficacious dosing and reduce the risk of undesired effects. If the ongoing experiments are positive, the Company expects to select a drug candidate in 2018.

### About mitochondrial myopathies

Mitochondrial myopathies are a group of neuromuscular diseases caused by damage to the mitochondria. Some of the most common mitochondrial myopathies are Kearns-Sayre syndrome, MERRF syndrome (myoclonic epilepsy with ragged red fibers) and MELAS (mitochondrial encephalomyopathy, 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 medi-

cal need for new and effective treatment options for mitochondrial myopathies.

- 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.
- http://www.ninds.nih.gov/disorders/mitochondrial\_ myopathy/mitochondrial\_myopathy.htm

# PROJECTS FOR OUT-LICENSING

# Non-alcoholic steatohepatitis (NASH)

NASH – non-alcoholic steatohepatitis – is a progressive disease that can develop into liver cirrhosis or liver cancer. Liver damage in NASH is caused by fat accumulation, inflammation and fibrosis development in the liver. NASH is a form of NAFLD (non-alcoholic fatty liver disease), which is one of the most common conditions worldwide. An estimated 20% of the global population suffers from NAFLD, and about onethird of the US population. There is a strong association between NASH and a variety of metabolic syndromes like diabetes and obesity. Approximately 3-5% of Americans (about 15 million people) suffer from NASH and there are currently no registered drugs for the treatment of this condition.1)

1) Vernon G. et al. Aliment Pharmacol Ther. 2011;34(3): 274-85

# NV556 and NVP022

The antifibrotic effects of the NV556 compound have been shown in two preclinical models of NASH, the STAM™ model and the MCD model. In the STAM model, long-term treatment also showed that NV556 is well tolerated and significantly suppresses liver weight gain, which is an indicator of lower tumor burden and a potential risk factor for developing liver cancer. In these experiments, NV556 tended to reduce both the number and size of surface liver tumors. In cooperation with Professor Massimo Pinzani, MD, PhD, FRCP at University College London (UCL), NV566's mechanism of action is being studied more closely in an in vitro model for liver fibrosis. The results show that NV556 has an inhibitory effect on collagen production, which plays an important role in the fibrosis development. Out-licensing activities for NV556 commenced during the period, and an agreement will hopefully be in place by mid-2018.

In addition to NV556, NeuroVive is also developing a new class of compounds with a different mechanism of action, NVP022, that may serve as complementary treatment for NASH. The NVP022 project is based on NeuroVive's core expertise in mitochondrial energy regulation, combined with the expertise of its partner company, Isomerase,



in innovative chemistry. NVP022 compounds are known as "mild" uncouplers, which means they do not have the same maximum effect on energy conversion as the uncoupling agent Dinitrophenol (DNP), which was previously used for weight loss, but overdosing is associated with the risk of significant adverse effects. In addition, the new compounds are primarily designed to deliver the uncoupling agent to the liver. This reduces the effects on other organs and enables a wide therapeutic window.

After the reporting period, a poster with preclinical data from NeuroVive's NVP022 NASH project was presented at the annual Liver Meeting of the American Association for the Study of Liver Diseases (AASLD) in Washington D.C. The data was considered so promising that the abstract was selected as a Presidential Poster of Distinction, which means that review scores placed the presentation within the top 10% of all posters.

The preclinical results from the first generation of compounds showed mild uncoupling predominantly in liver cells in test tube experiments and that uncoupling agents were delivered to the liver in animal experiments. A selection of candidates for continued preclinical development is expected during the first half of 2018.

# Hepatocellular carcinoma (HCC)

Liver cancer is often diagnosed at a late stage of the disease and mortality rates are high. There are two major types of liver cancer: hepatocellular carcinoma (HCC) and intrahepatic bile duct cancer. Infections with various hepatitis viruses can increase the risk of liver cancer. Patients with liver cirrhosis caused by alcoholism or fatty liver (non-alcoholic steatohepatitis, NASH) are at greater risk of developing liver cancer. Although liver cancer is less common in northern Europe and the US, HCC is still the sixth most-common type of cancer and the third most-common cause of cancer-related deaths worldwide. While surgery and chemotherapy are important starting points for the treatment of liver tumors, there is a major unmet medical need for more, and more effective, complementary therapies. 3)

- 1) Altekruse SF, McGlynn KA, Reichman ME: Hepatocellular Carcinoma Incidence, Mortality, and Survival Trends in the United States From 1975 to 2005. J Clin Oncol 27(9):1485-91, 2009
- 2) Forner A, Llovet JM, Bruix J: Hepatocellular carcinoma, Lancet 379 (9822):1245-55,2012.
- http://www.cancerresearchuk.org/helath-professional/ cancer-statistics/statistics-by-cancer-type/liver-cancer/ Incidence#heading-Nine

#### NVP024

In partnership with Isomerase, NeuroVive's research team has demonstrated that the Company's sanglifehrin-based compounds show powerful anticancer effects in preclinical models of HCC. The results to date show that a novel model compound, in which the anticancer effect is optimized, exhibits an inhibitory effect at a 500-fold lower concentration than the established cancer drug Sorafenib (approved for the treatment of advanced HCC). In addition, this class of compounds demonstrates anticancer activity in a preclinical experimental (in vivo) model of HCC, after both oral and intraperitoneal administration. The compounds show minimal toxicity in healthy cells and are well-tolerated in vivo. Continued preclinical development is ongoing with the aim of selecting a candidate for continued preclinical development during 2018.

After the reporting period, NeuroVive and Lund University were granted funding by the Swedish Foundation for Strategic Research (SSF) to study the role of cyclophilins in hepatocellular cancer (HCC). The grant from SSF will enable NeuroVive and Lund University to recruit an industrial PhD student to conduct research within the framework of the NVP024 project, with the aim of finding a new form of treatment for HCC.



# **Financial information**

#### Revenues

The consolidated turnover during the third quarter of 2017 was SEK 0 (0). Other operating revenues for the third quarter of 2017 were SEK 397,000 (16,000). The consolidated turnover for the first six months was SEK 27,000 (0) and the operating revenues amounted SEK 550,000 (90,000).

#### Results of operations

The operating loss for the third quarter was SEK 12,793,000 (34,190,000). The operating loss for the first nine months was SEK 56,168,000 (57,247,000). The net loss before tax for the third quarter amounted to SEK 13,179,000 (34,290,000). The net loss before tax for the first nine months was SEK 56,824,000 (57,265,000).

The operating loss was affected by external expenses, which for the first nine months were SEK 34,505,000 (24,308,000). During the first nine months, expenses related to development projects have affected the result with SEK 22,827,000 (8,204,000) whereof SEK 11,779,000 relates to project in clinical phase. Projects from clinical phase are from April 1st reported directly in the Income Statement.\* Personnel expenses during the first nine months amount to SEK 9,968,000 (11,332,000). Other operating expenses amount to, SEK 11,117,000 (20,888,000). Whereof SEK 10,981,000 relates to disposal of subsidiary. The remaining portion of other operating expenses pertains to exchange-rate losses.

The company has sold its shares in the Asian subsidiary and, together with its collaboration partner Foundation Asia Pacific Ltd., reacquired the Hong Kong-based subsidiary, NeuroVive Pharmaceutical Asia Ltd., which holds the Asian territorial licensing rights for NeuroSTAT and the agreements with the Chinese pharmaceutical company Sihuan Pharmaceutical and Sanofi Korea. The Hong Kong company is owned by NeuroVive Pharmaceutical AB 82.47% and Foundation Asia Pacific Ltd. 17.5%. Other assets, previously licensed to NeuroVive's Asian company, were transferred to NeuroVive Pharmaceutical AB. In addition to license assets, the Company received approximately SEK 5 million before administrative expenses.

\* For information on accounting principles for intangible assets, see page 50 of the Annual Report 2016 and changed assessment and positions, page 9 of this report.

## Financial position

The equity/assets ratio was 94 (95) % as of 30 September 2017, and equity was SEK 115,936,000 (182,385,000) compared to beginning of the year. The equity includes funds from the rights issue of SEK 4,500,000 completed July 21 reduced with transaction costs of SEK 360,000 and funds from option programs TO2 and TO3 of SEK 32,000. Cash and cash equivalents amounted to SEK 35,436,000 (112,889,000) as of 30 September 2017, a decrease of SEK 57,815,000 from the beginning of the year. Total assets as of 30 September 2017 were SEK 123,851,000 (192,978,000). The Board has taken steps to ensure the business's need for funding are addressed and actively works with solutions to execute the company's communicated business plan.

#### Cash flow and investments

Operating cash flow for the third quarter was SEK -14,929,000 (-10,137,000). Operating cash flow from the first nine months was SEK -48,192,000 (-45,595,000). The cash flow effect related to investments in intangibles equals SEK -3,250,000 (-9,828,000) for the first nine months. The disposal of shares in the Asian subsidiary has affected cash flow by SEK -11,035,000. Cash flow for the third quarter equals SEK -11,866,000 (-20,127,000). Cash flow for the first nine months equals SEK -58,080,000 (14,960,000).

# Transactions with related parties

Transactions between the company and its subsidiaries, which are related parties to the company, have been eliminated on consolidation, and accordingly, no disclosures are made regarding these transactions. In addition, there are Board fees and remunerations that follow from employment contracts with related parties.

Apart from remuneration to senior managers including remuneration for consulting services, no purchases or sales between the group and related parties occurred. Disclosures regarding transactions between the group and other related parties are stated below.

(SEK 000)		1 Jan. 2016 30 Sep. 2016
Stanbridge bvba (owned by Gregory Batcheller, Executive Chairman)	620	458
Ankor Consultants bvba (owned by Arne Ferstad, Board member)	-	94
Total transactions with related parties	620	552



# Segment information

Financial information reported to the chief operating decision maker (CEO) as the basis for allocating resources and judging the group's profit or loss is not divided into different operating segments. Accordingly, the group consists of a single operating segment.

### Financial instruments

NeuroVive holds unlisted securities, classified as "financial assets available for sale." These assets are measured at fair value through other comprehensive income on an ongoing basis. However, when the fair value of these securities cannot be reliably measured, they are recognized at cost. Other financial assets are classified as "loans and receivables," which are measured at amortized cost. The carrying amount of this category is estimated to correspond to fair value.

### **Human resources**

The average number of employees of the group for the period January to September was 10 (17), of which 5 (8) are women.

# Parental company

In connection with the sale of the Asian subsidiary and the reacquisition of the Hong Kong company, a positive result from shares in Group companies amounted to SEK 7,652,000. Company earnings after tax for the first six months amounts to SEK -24,620,000 (-20,128,000). Most of the Group's operations are conducted within the parent company. Accordingly, no further specific information regarding the parent company is presented.

# Changed assessment and position

During the period, the Company's Board of Directors has chosen to change its assessment and position regarding the timing of capitalization of development costs. The new assessment is in line with the company's new strategy and the history of earlier completed development projects. The new assessment means that all development work is expensed continuously until the product has received market approval.

The development for the NeuroSTAT / TBI project is proceeding according to plan, and is under preparation for transition to FAS IIb. It has therefore not been estimated that there is an impairment need for historically capitalized development costs for this project. Book value amounts to SEK 51,941,000 thousand.

# Risks and uncertainty factors

A research company such as NeuroVive Pharmaceutical AB (publ) is subject to high operational and financial risks because the projects the company conducts are in different developmental phases, where a number of parameters influence the likelihood of commercial success. Briefly, operations are associated with risks relating to factors including drug development, competition, technological progress, patents, regulatory requirements, capital requirements, currencies and interest rates. The Board of Directors works continuously to secure the business operation's need for financing. A way to spread risks is to out-license projects directed towards larger indications already in the pre-clinical phase, while orphan indication projects are developed by the company up until market registration. With exception that the Board has taken steps to ensure the business's need for funding are addressed and actively works with solutions to execute the company's communicated business plan, no significant changes in relation to risk or uncertainties occurred during the current period.

In March 2013, CicloMulsion AG commenced arbitration pertaining to certain obligations of the Company under a 2004 License Agreement, including payment of royalties. As previously reported, on May 25, 2016, the Tribunal rendered a partial award which has been appealed by each party. The hearing at the competent Swedish court in Skåne and Blekinge will take place end November 2017 and a decision is to be expected in the first half of 2018. The arbitration proceeding remains suspended.

For more detail of risks and uncertainty factors, refer to the Statutory Administration Report in the Annual Report 2015 and the prospectus published 14 April 2016 for the share issue in April/May 2016.

# Incentive programs/share warrants

Currently there is no incentive program.

# **Audit review**

This Interim Report has been subject to review by the company's auditors in accordance with the Standard on Review Engagements (ISRE) 2410, Review of Interim Financial Information Performed by the Independent Auditor of the Entity.



# Upcoming financial statements

Year-End Report 2017 20 February 2018
Interim Report January-March 22 May 2018
Interim Report January-June 21 August 2018
Interim Report January-September 22 November 2018
Year-End Report 2018 21 February 2019

The Interim Reports and the Annual Year Report are available at www.neurovive.com.

# **Annual General Meeting 2018**

NeuroVives Annual General Meeting will be held at Medicon Village, Scheelevägen 2, in Lund on 26 April 2018 at 4 pm.

The Nomination Committee for the 2018 AGM comprises: Michael Vickers – nominated by Maas Biolab LLC / Marcus Keep; Andreas Inghammar – nominated by Eskil Elmer; Tomas Hagström – nominated by Greg Batcheller.

Shareholders wishing to make proposals on the above matters can contact the Committee by email at: valberedningen@neurovive.com

or by post at:
NeuroVive Pharmaceutical AB
FAO: Nomination Committee
Medicon Village
223 81 Lund
Sweden.

In order for the Nomination Committee to consider the proposals received with due care, proposals should be received by the Nomination Committee by no later than 1 February 2018.

# Principles of preparation of the Interim Report

NeuroVive prepares its consolidated accounts in accordance with International Financial Reporting Standards (IFRS) issued by the International Accounting Standards Board (IASB) and interpretation statements from the IFRS Interpretations Committee, as endorsed by the EU for application within the EU. This Interim Report has been prepared in accordance with IAS 34 Interim Financial Reporting.

The parent company applies the Swedish Annual Accounts Act and RFR's (the Swedish Financial Reporting Board) recommendation RFR 2 Accounting for Legal Entities. Application of RFR 2 implies that, as far as possible, the parent company applies all IFRS endorsed by the EU within the limits of the Swedish Annual Accounts Act and the Swedish Pension Obligations Vesting Act, and considering the relationship between accounting and taxation.

The group and parent company have applied the same accounting principles as described in the Annual Report for 2016 on pages 48-61. However, a changed assessment has been made regarding the capitalization of development expenses as described above under the heading Changed assessments and positions.

New and revised standards and interpretation statements applicable from 1 January 2017 onwards did not have any effect on the group or parent company's results of operations or financial position.



# Consolidated Statement of Comprehensive Income

		1 Jul, 2017	1 Jul, 2016	1 Jan, 2017	1 Jan, 2016	1 Jan, 2016
(SEK 000)	Note	30 Sep, 2017	30 Sep, 2016		30 Sep, 2016	31 Dec, 2016
Net sales	···•		-	27	-	14
Other operating income		397	16	550	90	104
Or and the suppose		397	16	576	90	118
Operating expenses Other external expenses		-9,757	-9,172	-34,505	-24,308	-34,168
Other external expenses  Personnel cost		-2,963	-9,172 -4,050	-9,968	-11,332	-15,276
Depreciation and write-down of tangible and intangible assets		-2,963	-4,030 -278	-1,155	-11,332	-1,121
Depreciation and write-down or tangible and intangible assets		-412	-278	-1,100	-808	-1,121
Other operating expenses		-57	-20,705	-11,117	-20,888	-21,663
		-13,190	-34,206	-56,745	-57,337	-72,228
Operating income		-12,793	-34,190	-56,168	-57,247	-72,110
Profit/loss from financial items						
Result from shares in associated company		_	_	_	_	28
Financial income	,	11	21	121	176	432
Financial costs	••••	-397	-121	-776	-194	-195
		-386	-99	-655	-17	265
Profit/loss before tax		-13,179	-34,290	-56,824	-57,265	-71,845
Income tax	2					
Profit/loss for the period		-13,179	-34,290	-56,824	-57,265	-71,845
Other comprehensive income		•	•			
Items that may be reclassified to profit or loss						
Translation differences on foreign subsidiaries		30	745	19	1,282	1,782
Total comprehensive income for the period		-13,149	-33,545	-56,805	-55,983	-70,063
Loss for the period attributable to:						
Parent company shareholders		-13,098	-33,919	-51,950	-56,090	-70,240
Non-controlling interests	••••	-81	-371	-4,874	-1,175	-1,605
		-13,179	-34,290	-56,824	-57,265	-71,845
Total comprehensive income for the period	<del>-</del>					
Parent company shareholders		-13,016	-33,528	-51,955	-55,548	-69,271
Non-controlling interests	····	-133	-17	-4,850	-435	-792
		-13,149	-33,545	-56,805	-55,983	-70,063
Earnings per share before and after dilution(SEK) based on average number of shares		-0.26	-0.86	-1.04	-1.42	-1.67



# Consolidated Statement of Financial Position

(SEK 000)	Note	30 Sep, 2017	30 Sep, 2016	31 Dec, 2016
ASSETS				
Non-current assets				
Intangible assets	1			
Development costs		51,941	47,681	51,255
Patents		19,972	14,703	17,979
Other Intangible assets		1,781	1,956	1,917
		73,694	64,339	71,151
Tangible assets				
Equipment		187	290	274
		187	290	274
Financial assets				
Other long-term securities		13,102	13,101	13,102
Other long-term receivables		-	119	118
	-	13,102	13,220	13,220
Total non-current assets		86,983	77,849	84,645
Total foil cuit cite assess	_	00,000	77,040	
Current assets				
Other receivables		1,118	1,574	1,650
Prepaid expenses and accrued income		314	665	1,171
Cash and cash equivalents		35,436	112,889	93,251
	_	36,868	115,128	96,072
TOTAL ASSETS		123,851	192,978	180,717
EQUITY AND LIABILITIES				
Equity attributable to the shareholders of the parent company				
Share capital	•	2,528	2,473	2,473
Additional paid in capital		422,607	418,339	418,339
Translation reserve	***************************************	775	352	780
Retained earnings		-314,962	-251,995	-266,146
Total equity attributable to the shareholders of the parent		110,948	169,169	155,446
Non-controlling interests		4,988	13,216	12,858
Total equity		115,936	182,385	168,304
Chaut taum liabilities				
Short-term liabilities		0.770	7.040	
Accounts payable		2,336	3,642	6,000
Other liabilities	-	847	734	483
Accrued expenses and deferred income		4,732	6,217	5,930
		7,915	10,593	12,413
Total liabilities	-	7,915	10,593	12,413
TOTAL EQUITY AND LIABILITIES		123,851	192,978	180,717
TOTAL LQUITT AND LIABILITIES		120,001	132,370	100,717



# Consolidated Statement of Changes in Equity

(SEK 000)	Equity attribu				company	N.	
	Share-	Additional		Datained		Non-	Tota
			Translation	Retained	Takal	controlling	
Onening helence 1 January 2017	capital	capital		earnings	Total	interests	equity
Opening balance, 1 January 2017	2,473	418,339	780	-266,146	155,446	12,858	168,304
Comprehensive profit/loss for the period				F1 0F0	E1 0E0	4.074	FC 00/
Profit/loss for the period	-	-	-	-51,950	-51,950	-4,874	-56,824
Other comprehensive income					F		10
Translation differences	•	-	-5	-	-5 -5	24	19
Other comprehensive profit/loss for the period, net after tax	-		-5 - <b>5</b>			24	19
Total comprehensive profit/loss	<u>-</u>	<u>-</u>	-5	-51,950	-51,955	-4,850	-56,805
Transactions with shareholders		4.000			4 707		4 707
Share issue	55	4,268	-	=	4,323	-	4,323
Shareholder contribution	-		_			114	114
Change of ownership in share issue			-	3,134	3,134	-3,134	
Total transactions with shareholders	55	4,268		3,134	7,457	-3,020	4,437
Closing balance, 30 September 2017	2,528	422,607	775	-314,962	110,948	4,988	115,936
Opening balance, 1 January 2016	1,537	335,687	-190	-195,906	141,128	13,651	154,779
Comprehensive profit/loss for the period	1,007		100	100,000		10,001	
Profit/loss for the period	-		-	-56,090	-56,090	-1,175	-57,265
Other comprehensive income		•••••••••••••••••••••••••••••••••••••••	•	00,000		2,2.0	
Translation differences			542		542	740	1,282
Other comprehensive profit/loss for the period, net after		_	542		542	740	1,282
tax			· · · -		V.=		_,
Total comprehensive profit/loss	_	-	542	-56,090	-55,548	-435	-55,983
Transactions with shareholders							
New share issue	936	82,652	-	-	83,588	-	83,588
Issue through non-controlling interest	-	-	-	-	-	-	
Total transactions with shareholders	936	82,652	_	-	83,588	-	83,588
Closing balance, 30 September 2016	2,473	418,339	352	-251,995	169,169	13,216	182,384
Opening balance, 1 January 2016	1,537	335,687	-190	-195,906	141,128	13,651	154,779
Comprehensive profit/loss for the period		······································			•		
Profit/loss for the period	-	-	-	-70,240	-70,240	-1,605	-71,845
Other comprehensive income		•	•	•	-	•	
Translation differences	=	-	970	_	970	812	1,782
Other comprehensive profit/loss for the period, net after tax	<del>-</del>	-	970	-	970	812	1,782
Total comprehensive profit/loss	-	-	970	-70,240	-69,270	-793	-70,063
Transactions with shareholders		-			-		
New share issue	936	82,652	-	-	83,588	-	83,588
Total transactions with shareholders	936	82,652		-	83,588	-	83,588
Closing balance, 31 December 2016	2,473	418,339	780	-266,146	155,446	12,858	168,304



# Consolidated Statement of Cash Flows

Adjustments for non-cash items: Depreciation 412 278 1,155 808 1,12 Currency differences on intercompany items -289 12 -279 25 4 Impaired Value -20,618 - 20,618 - 20,618 21,3 Disposal of Business -7 10,991 7 Result from shares in associated company -7 10,991 77 Result from shares in associated company -7 10,991 77 Result from shares in associated company -7 10,991 77 Result from shares in associated company -7 10,991 77 Result from shares in associated company -7 10,991 77 Result from shares in associated company -7 10,991 77 Result from shares in associated company -7 10,991 776 194 - 12 Ret cash from operating activities before changes in working capital -7 13,055 13,375 44,967 35,807 49,53  Changes in working capital -7 1,010 785 524 - 1 Increase/decrease of other current assets -7 1,210 785 524 - 1 Increase/decrease of other short-term liabilities -7 1,875 3,238 3,225 9,785 7,84  Cash flow from operating activities -7 1,875 3,238 3,225 9,785 7,84  Cash flow from operating activities -7 1,222 3,690 3,250 9,828 18,15  Acquisition of tangible assets -7 10 40 108 13  Disposal to ftangible assets -7 10 40 108 13  Disposal to ftangible assets -7 10 40 108 13  Disposal business -7 1,222 3,690 4,250 9,828 18,15  Encrease in other financial assets -7 1,222 3,991 14,325 16,780 25,13  Financing activities -7 1,222 3,991 14,325 16,780 25,13  Financing activities -7 1,222 3,991 14,325 77,332 77,332  Financing activities -7 1,222 3,991 14,325 16,780 25,13  Financing activities -7 1,222 3,991 14,325 77,332 77,332 77,332  Financing activities -7 1,220 3,991 14,325 77,332 77,332 77,332  Cash flow from investing activities 4,285 7,360 4,360 4,960 5,18  Cash flow from financing activities 1,280 93,251 96,662 96,66  Effect of exchange rate changes on cash 318 736 265 1,267 1,76	(SEK 000)	1 Jul, 2017	1 Jul, 2016	1 Jan, 2017	1 Jan, 2016	1 Jan, 2016
Operating income   -12,792	Cash flow from operating activities	30 Зер, 2017	30 Зер, 2016	30 Sep, 2017	30 Зер, 2016	31 Dec, 2016
Depreciation	Operating income	-12,792	-34,190	-56,169	-57,247	-72,110
Currency differences on intercompany items   -289   12   -279   25   4	Adjustments for non-cash items:					
Impaired Value	Depreciation	412	278	1,155	808	1,121
Disposal of Business   7   10,981   7   Result from shares in associated company   2   2   1   1   1   1   1   1   1   1	Currency differences on intercompany items	-289	12	-279	25	48
Result from shares in associated company	Impaired Value	-	20,618	-	20,618	21,135
Interest received	Disposal of Business	-	7	10,981	7	7
Interest paid   -397   -121   -776   -194   -12     -176   -194   -12     -176   -194   -12     -176   -194   -12     -176   -194   -12     -176   -194   -12     -176   -194   -12   -176   -194   -12   -13,055   -13,375   -44,967   -35,807   -49,53   -49,53   -49,53   -44,967   -35,807   -49,53	Result from shares in associated company	-	-	-	-	28
Net cash from operating activities before changes in working capital   -13,055   -13,375   -44,967   -35,807   -49,53	Interest received	11	21	121	176	363
Changes in working capital  Increase/decrease of other current assets 271 2,101 785 524 -1 Increase/decrease of other short-term liabilities 2,145 1,137 4,010 -10,309 -7,82 Changes in working capital -1,875 3,238 -3,225 9,785 -7,84  Cash flow from operating activities -14,929 -10,137 -48,192 -45,592 -57,37  Investing activities  Acquisition of intangible assets -1,222 -3,690 -3,250 -9,828 -18,15  Acquisition of tangible assets -110 -40 -108 -13  Disposal business -110,35 -1	Interest paid	-397	-121	-776	-194	-126
Increase   decrease of other current assets   271   2,101   785   524   -1     Increase   decrease of other short-term liabilities   -2,145   1,137   -4,010   -10,309   -7,82     Changes in working capital   -1,875   3,238   -3,225   -9,785   -7,84     Cash flow from operating activities   -14,929   -10,137   -48,192   -45,592   -57,37     Investing activities   -1,222   -3,690   -3,250   -9,828   -18,15     Acquisition of intangible assets   -1,222   -3,690   -3,250   -9,828   -18,15     Acquisition of tangible assets   -10   -40   -108   -13     Disposal business   -1,1035   -     Increase in other financial assets   -6,291   -   -6,844   -6,84     Cash flow from investing activities   -1,222   -9,991   -14,325   -16,780   -25,13     Financing activities   -1,222   -9,991   -14,325   -16,780   -25,13     Cash flow from financing activities   -1,124   -114   -     Cash flow from financing activities   -1,126   -2,127   -58,080   14,960   -5,18     Cash flow for the period   -11,866   -20,127   -58,080   14,960   -5,18     Cash and cash equivalents at the beginning of the period   -11,866   -20,127   -58,080   14,960   -5,18     Cash and cash equivalents at the beginning of the period   -132,280   93,251   96,662   96,662     Effect of exchange rate changes on cash   -1,267   -1,768   -2,176   -1,768	Net cash from operating activities before changes in working capital	-13,055	-13,375	-44,967	-35,807	-49,534
Cash flow from operating activities   -2,145   1,137   -4,010   -10,309   -7,82	Changes in working capital			······································	······································	
Changes in working capital         -1,875         3,238         -3,225         -9,785         -7,84           Cash flow from operating activities         -14,929         -10,137         -48,192         -45,592         -57,37           Investing activities         -1,222         -3,690         -3,250         -9,828         -18,15           Acquisition of intangible assets         -10         -40         -108         -13           Disposal business         -11,035         -         -           Increase in other financial assets         -6,291         -6,844         -6,84           Cash flow from investing activities         -1,222         -9,991         -14,325         -16,780         -25,13           Financing activities         4,171         -4,323         77,332         77,332           Shareholder contribution subsidary         114         -114         -           Cash flow from financing activities         4,285         -4,437         77,332         77,33           Cash flow for the period         -11,866         -20,127         -58,080         14,960         -5,18           Cash and cash equivalents at the beginning of the period         -132,280         93,251         96,662         96,662           Effect of exchange rate changes on ca	Increase/decrease of other current assets	271	2,101	785	524	-19
Cash flow from operating activities	Increase/decrease of other short-term liabilities	-2,145	1,137	-4,010	-10,309	-7,824
Investing activities	Changes in working capital	-1,875	3,238	-3,225	-9,785	-7,843
Acquisition of intangible assets -1,222 -3,690 -3,250 -9,828 -18,150 -10 -10 -10 -10 -10 -10 -10 -10 -10 -1	Cash flow from operating activities	-14,929	-10,137	-48,192	-45,592	-57,377
Acquisition of tangible assets10 -40 -108 -13 Disposal business11,035 - Increase in other financial assets6,2916,844 -6,84  Cash flow from investing activities -1,222 -9,991 -14,325 -16,780 -25,13  Financing activities  New share issue - 4,171 - 4,323 -77,332 -77,332  Shareholder contribution subsidary - 114 114  Cash flow from financing activities - 4,285 - 4,437 -77,332 -77,333  Cash flow for the period -11,866 -20,127 -58,080 -14,960 -5,18  Cash and cash equivalents at the beginning of the period - 132,280 -93,251 -96,662 -96,666  Effect of exchange rate changes on cash - 318 -736 -265 -1,267 -1,766	Investing activities		•	•	•••••••••••••••••••••••••••••••••••••••	
Disposal business	Acquisition of intangible assets	-1,222	-3,690	-3,250	-9,828	-18,152
The crease in other financial assets	Acquisition of tangible assets	-	-10	-40	-108	-139
Cash flow from investing activities         -1,222         -9,991         -14,325         -16,780         -25,13           Financing activities           New share issue         4,171         -         4,323         77,332	Disposal business	-	-	-11,035	-	-
Financing activities  New share issue	Increase in other financial assets	-	-6,291	-	-6,844	-6,844
New share issue         4,171         -         4,323         77,332         77,332           Shareholder contribution subsidary         114         -         114         -           Cash flow from financing activities         4,285         -         4,437         77,332         77,332           Cash flow for the period         -11,866         -20,127         -58,080         14,960         -5,18           Cash and cash equivalents at the beginning of the period         -         132,280         93,251         96,662         96,662           Effect of exchange rate changes on cash         318         736         265         1,267         1,766	Cash flow from investing activities	-1,222	-9,991	-14,325	-16,780	-25,135
Shareholder contribution subsidary         114         -         114         -         114         -           Cash flow from financing activities         4,285         -         4,437         77,332         77,332           Cash flow for the period         -11,866         -20,127         -58,080         14,960         -5,18           Cash and cash equivalents at the beginning of the period         -         132,280         93,251         96,662         96,662           Effect of exchange rate changes on cash         318         736         265         1,267         1,766	Financing activities				······································	
Cash flow from financing activities         4,285         -         4,437         77,332         77,332           Cash flow for the period         -11,866         -20,127         -58,080         14,960         -5,18           Cash and cash equivalents at the beginning of the period         -         132,280         93,251         96,662         96,662           Effect of exchange rate changes on cash         318         736         265         1,267         1,76	New share issue	4,171	-	4,323	77,332	77,332
Cash flow for the period         -11,866         -20,127         -58,080         14,960         -5,18           Cash and cash equivalents at the beginning of the period         -         132,280         93,251         96,662         96,662           Effect of exchange rate changes on cash         318         736         265         1,267         1,766	Shareholder contribution subsidary	114	-	114	-	-
Cash and cash equivalents at the beginning of the period - 132,280 93,251 96,662 96,66 Effect of exchange rate changes on cash 318 736 265 1,267 1,76	Cash flow from financing activities	4,285	-	4,437	77,332	77,332
Effect of exchange rate changes on cash 318 736 265 1,267 1,76	Cash flow for the period	-11,866	-20,127	-58,080	14,960	-5,180
	Cash and cash equivalents at the beginning of the period	-	132,280	93,251	96,662	96,662
Cash and cash equivalents at end of period -11,548 112,889 35,436 112,889 93,25	Effect of exchange rate changes on cash	318	736	265	1,267	1,769
	Cash and cash equivalents at end of period	-11,548	112,889	35,436	112,889	93,251



# Parent Company Income Statement

(SEK 000)	1 Jul, 2017	1 Jul, 2016	1 Jan, 2017	1 Jan, 2016	1 Jan, 2016
Note	30 Sep, 2017	30 Sep, 2016	30 Sep, 2017	30 Sep, 2016	31 Dec, 2016
Net sales	-	-	27	9	30
Other operating income	397	16	550	90	104
	397	16	576	99	134
Operating expenses			-	-	
Other external expenses	-9,313	-8,782	-33,993	-22,501	-31,521
Personnel cost	-2,964	-3,250	-9,742	-9,160	-12,495
Depreciation and write-down of tangible and intangible assets	-412	-250	-1,144	-724	-1,006
Other operating expenses	-35	-20,711	-141	-20,895	-21,660
	-12,723	-32,994	-45,020	-53,280	-66,683
Operating income	-12,326	-32,978	-44,443	-53,180	-66,548
Profit/loss from financial items				······································	
Result from shares in group company	=	_	7,652	-20,870	-20,880
Result from shares in associated company	-	-20,870	_	_	29
Interest income and other similar profit items	5	33	85	116	288
Interest expenses and other similar loss items	-398	-94	-631	-102	-7
	-393	-20,931	7,106	-20,856	-20,570
Profit/loss before tax	-12,719	-53,908	-37,337	-74,037	-87,118
Income tax 2	-				
Profit/loss for the period	-12,719	-53,908	-37,337	-74,037	-87,118

# Statement of Comprehensive Income, Parent Company

(SEK 000)		1 Jul, 2017	1 Jul, 2016	1 Jan, 2017	1 Jan, 2016	1 Jan, 2016
	Note	30 Sep, 2017	30 Sep, 2016	30 Sep, 2017	30 Sep, 2016	31 Dec, 2016
Profit/loss for the period		-12,719	-53,908	-37,337	-74,037	-87,118
Other comprehensive income		-	-	-	-	
Total comprehensive profit/loss for the period		-12,719	-53,908	-37,337	-74,037	-87,118



# Parent Company Balance Sheet

(SEK 000)	Note	30 Sep, 2017	30 Sep, 2016	31 Dec, 2016
ASSETS				
Non-current assets		•		
Intangible assets	1	-	•	
Development costs		51,706	47,446	51,020
Patents		19,972	14,704	17,979
Other intangible assets		1,781	1,915	1,881
		73,460	64,065	70,881
Tangible assets				
Equipment		187	229	221
		187	229	221
Financial assets				
Other long-term placement		13,102	13,102	13,102
Shares in subsidiaries	3	23,625	20,870	20,870
		36,727	33,972	33,972
Total non-current assets		110,373	98,265	105,074
Current assets				
Short term receivables				
Receivables from group companies				7
Other receivables		1,115	1,567	1,643
Prepaid expenses and accrued income		315	246	515
	•	1,430	1,813	2,165
Cash and bank balances		35,318	95,010	75,954
Total current assets	-	36,747	96,823	78,119
TOTAL ASSETS	_	147,120	195,088	183,193
EQUITY AND LIABILITIES				
Equity				
Restricted equity				
Share capital		2,528	2,473	2,473
Statutory reserve		1,856	1,856	1,856
Development expenditure reserve		10,779	5,611	9,924
		15,163	9,940	14,253
Unrestricted equity				
Share premium reserve		4,268	393,648	82,653
Retained earnings		157,114	-144,249	162,434
Profit/loss for the period		-37,337	-74,037	-87,118
	-	124,045	175,362	157,969
Total equity	-	139,208	185,302	172,222
Short-term liabilities				
Accounts payable	-	2,336	3,110	5,582
Other liabilities		847	729	473
Accrued expenses and deferred income		4,729	5,947	4,916
		7,912	9,786	10,971
TOTAL EQUITY AND LIABILITIES		147,120	195,088	183,193
-				



# **Notes**

# Note 1 — Intangible assets

(SEK 000)	Development costs	Patents*	Other	Total
ACCUMULATED COST				
Opening balance 1 Jan. 2017	51,255	24,349	2,899	78,503
Additions	686	3,020	_	3,706
Impaired value	-	-	-35	-35
Closing balance 30 Sep. 2017	51,941	27,369	2,864	82,174
ACCUMULATED DEPRECIATION			······································	
Opening balance 1 Jan. 2017	-	-6,370	-982	-7,352
Depreciation for the period	-	-1,027	-101	-1,128
Closing balance 30 Sep. 2017	-	-7,397	-1,083	-8,480
Residual value 30 Sep. 2017	51,941	19,972	1,781	73,694

(SEK 000)	Development costs	Patents*	Other	Total
ACCUMULATED COST				
Opening balance 1 Jan. 2016	59,803	18,193	2,899	80,895
Additions	12,487	6,156	-	18,643
Impaired value	-21,035	-	-	-21,035
Closing balance 31 Dec. 2016	51,255	24,349	2,899	78,503
ACCUMULATED DEPRECIATION				
Opening balance 1 Jan. 2016	-	-5,170	-821	-5,991
Depreciation for the period	-	-1,200	-161	-1,361
Closing balance 31 Dec. 2016	-	-6,370	-982	-7,352
Residual value 31 Dec. 2016	51,255	17,979	1,917	71,151

# Note 2 - Tax

The group's total loss carry-forwards amount to SEK 345,556,000 as of 30 September 2017 (284,764,000). The parent company's total loss carry-forwards amount to SEK 319,384,000 as of 30 September 2017 (260,946,000). Because the company is loss making, management cannot judge when deductible loss carry-forwards will be utilized.

# Note 3 - Shares and participations in group companies

These shares are the holding of 82.47% in the subsidiary NeuroVive Pharmaceutical Asia Ltd., domiciled in Hong Kong.



# **Affirmation**

This Interim Report gives a true and fair view of the parent company and group's operations, financial position and results of operations, and states the significant risks and uncertainty factors facing the parent company and group companies.

Lund, Sweden, 21 November 2017

This Interim Report is published in Swedish and English. In the event of any difference between the English version and the Swedish original, the Swedish version shall prevail.

David Laskow-Pooley	David Bejker	Marcus Keep
Chairman of the Board	Board member	Board member
Jan Törnell	Erik Kinnman	
Board member	Chief Executive Officer	

For more information concerning this report, please contact CEO Erik Kinnman. Telephone: +46 (0)46-275 62 20

The information was submitted for publication, through the agency of the contact person set out above, at 08:30 a.m. CET on 21 November 2017.

# **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 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 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®) and one project entering clinical

Phase I (KL1333). 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).

# NeuroVive Pharmaceutical AB (publ)

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www.neurovive.com



# Auditor's review report

# NeuroVive Pharmaceutical AB (publ) Corp.Id.No 556595-6538

#### Introduction

We have performed a review of the condensed interim financial statements (the interim report) for NeuroVive Pharmaceutical AB (publ) at September 30, 2017 and the nine months' period then ended. The Board of Directors and the President are responsible for the preparation and presentation of this interim report in accordance with IAS 34 and the Swedish Annual Accounts Act. Our responsibility is to express a conclusion on this interim report based on our review.

# Scope of review

We conducted our review in accordance with the Standard on Review Engagements ISRE 2410 Review of Interim Financial Information Performed by the Independent Auditor of the Entity. A review consists of making inquiries, primarily of persons responsible for financial and accounting matters, and applying analytical and other review procedures. A review is substantially less in scope than an audit conducted in accordance with the International Standards on Auditing and other generally accepted auditing practices.

The procedures performed in a review do not enable us to obtain a level of assurance that would make us aware of all significant matters that might be identified in an audit. Therefore, the conclusion expressed based on a review does not give the same level of assurance as a conclusion expressed based on an audit.

#### Conclusion

Based on our review, nothing has come to our attention that causes us to believe that the interim report, in all material aspects, is not prepared for the Group in accordance with IAS 34 and the Swedish Annual Accounts Act and for the Parent company in accordance with the Swedish Annual Accounts Act.

# Information of particular importance

As described on page 8 under the financial position, the Board of Directors conducts active work to ensure continued business financing needs.

This special enlightenment does not affect our conclusion as stated above.

Helsingborg, November 21th, 2017

Mazars SET Revisionsbyrå AB

# Bengt Ekenberg

Authorized Public Accountant

NeuroVive Pharmaceutical AB (publ) 556595-6538



# Amended number of shares and votes in NeuroVive Pharmaceutical AB (publ)

Lund, Sweden, 30 November 2017 - NeuroVive Pharmaceutical AB (Nasdaq Stockholm: NVP, OTCQX: NEVPF), the mitochondrial medicine company, today announced an increase of the number of shares and votes in NeuroVive of 1,760,000 as a result of a directed share issue in NeuroVive Pharmaceutical AB, resolved by the Board of Directors on 2 November 2017.

Today, the last trading day of the month, there are in total 52,326,197 shares and votes in NeuroVive Pharmaceutical AB.

# For more information please contact:

Daniel Schale, Director of Communications, NeuroVive +46 (0)46-275 62 21, ir@neurovive.com

# **NeuroVive Pharmaceutical AB (publ)**

Medicon Village, SE-223 81 Lund, Sweden Tel: +46 (0)46 275 62 20 (switchboard) info@neurovive.com, www.neurovive.com

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 above, at 08:30 a.m. CET on 30 November 2017.

# **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).

NeuroVive Pharmaceutical AB (publ) 556595-6538



# NeuroVive's genetic mitochondrial disease program NVP015 reaches significant milestone

Lund, Sweden, 30 November 2017 – NeuroVive Pharmaceutical AB (Nasdaq Stockholm: NVP, OTCQX: NEVPF), the mitochondrial medicine company, today announced that molecules in its NVP015 program have demonstrated positive effects in experimental models of genetic mitochondrial disease and that a lead compound has been selected. The lead was selected based on tolerability, plasma stability and organ delivery, specifically to the brain, and will now be taken into further in vivo experimental efficacy studies and preclinical development.

The objective of NeuroVive's NVP015 program is to develop a treatment option for patients with genetic mitochondrial diseases, such as Leigh syndrome and MELAS, when acute cellular energy support is needed, such as during an ordinary infection. These patients are at risk of further deterioration in their disease and symptoms during such energy crisis.

"We are very pleased with the progress in the NVP015 program which, together with our KL1333 program, shows our strong commitment to the development of novel treatments for genetic mitochondrial diseases," commented Magnus Hansson, M.D., Ph.D., Chief Medical Officer and VP of Preclinical and Clinical Development at NeuroVive.

NeuroVive collaborates with Dr. Marni Falk's research lab at the Children's Hospital of Philadelphia (CHOP) to evaluate the NVP015 compound class. Top line data from these studies demonstrates that the compounds exert beneficial effects in experimental *C. elegans* models of genetic mitochondrial disease. Approximately 50 per cent of genetic mitochondrial disease patients have a complex I dysfunction (isolated or in combination with other defects) in the mitochondria. Complex I dysfunction occurs when the first of the five complexes in the mitochondrial respiratory chain fails to function normally, resulting in the mitochondria being unable to generate enough cellular energy, ATP.

"This is a breakthrough which takes us a step closer to our goal of developing a therapy for acute energy crisis situations in genetic mitochondrial disease patients. Currently, patients with these types of genetic mitochondrial disorders can only be offered symptomatic treatment alternatives, whereas the aim of the NVP015 program is to create a drug that can bypass the dysfunction, helping the mitochondria to function properly," said Eskil Elmér, M.D., Ph.D., Chief Scientific Officer and VP of Discovery at NeuroVive.

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# For more information please contact:

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NeuroVive Pharmaceutical AB (publ) - the mitochondrial medicine company. The company is listed on Nasdaq Stockholm, Small Cap, under the ticker symbol NVP. The share is also traded on the OTC Markets Group Inc market in the US. NeuroVive Pharmaceutical (OTC: NEVPF) trades on the OTCQX Best Market. Investors can find Real-Time quotes and market information for the company at <a href="https://www.otcmarkets.com/stock/NEVPF/quote">www.otcmarkets.com/stock/NEVPF/quote</a>

NeuroVive Pharmaceutical AB (publ) 556595-6538



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#### **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 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 program is based on a concept instigated by NeuroVive's Chief Scientific Officer Dr. Eskil Elmér and his colleagues by 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 program were published in Nature Communications<sup>1</sup> in 2016. The NVP015 program is focused on enabling a systemic therapy to counteract acute energy crises in patients with genetic mitochondrial disease. NeuroVive has received a grant from the Swedish innovation agency Vinnova's 2017 Swelife call for the NVP015 development. NeuroVive collaborates with Dr. Marni Falk's research lab at the Children's Hospital of Philadelphia (CHOP) to evaluate the NVP015 compound class. Mitochondrial complex I dysfunction is also recognized in several other conditions and in October 2017, Dr. Todd Kilbaugh, also at CHOP, received a grant from the NIH program Countermeasures Against Chemical Threats (CounterACT) to study the NVP015 as a novel treatment against chemical threats.

# **About Mitochondrial Disorders**

Approximately 12 in every 100,000 people suffer from a genetic mitochondrial disorder. Genetic mitochondrial disorders are congenital metabolic diseases that affect cellular energy conversion. The disorders can manifest differently depending on which organs are affected by the gene defects and are viewed as syndromes, depending on the organs affected and the signs and symptoms. The disease usually presents in early childhood, such as Leigh syndrome, a rare genetic neurometabolic disorder, characterized by the degeneration of the central nervous system. Another example of mitochondrial disease is MELAS, which can cause mitochondrial myopathy, stroke like episodes and brain damage with epilepsy. Candidate drugs from NeuroVive's genetic mitochondrial disease programs would qualify for orphan drug designation in the US and Europe during clinical development, enabling a faster and less costly route to market, and a higher sales price. In 2016, the orphan drug market amounted to USD 114 billion and in the same year, the average annual cost for the treatment of a single patient was an estimated USD 140,443 (approx. 1.3 million SEK)<sup>2</sup>.

## **About NeuroVive**

NeuroVive Pharmaceutical AB is a leader in mitochondrial medicine, with one program in clinical phase II development for the prevention of moderate to severe traumatic brain injury (NeuroSTAT®) and one program 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 programs 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).

- 1) Ehinger JK et al. (2016) Nat. Commun.7:12317
- 2) Evaluate Pharma Orphan Drug Report 2017

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NeuroVive Pharmaceutical AB (publ) 556595-6538



# NeuroVive reports promising progress in its clinical project for genetic mitochondrial diseases, KL1333

**Lund, Sweden, 11 December 2017 – NeuroVive Pharmaceutical AB** (Nasdaq Stockholm: NVP, OTCQX: NEVPF) and Yungjin Pharm today announced that the phase I clinical study in Korea within the companies' joint project KL1333, an investigational treatment for genetic mitochondrial disorders, including MELAS (Mitochondrial Myopathy, Encephalopahty, Lactic Acidosis and Stroke-like episodes), is proceeding according to plan.

The first part of the study has been successful. The pharmacokinetic data was in line with expectations and no adverse safety signals were detected. The study's remaining higher dose-cohorts have now been approved by the Korean medicinal authority, the Ministry of Food and Drug Safety (MFDS).

"We are very excited about the progress of the first KL1333 clinical study. Along with the recent positive opinion on European orphan drug designation, it brings us one step closer to initiating our own clinical phase lb study, and to our ultimate goal of providing a treatment opportunity to patients with different genetic mitochondrial disorders, where there is a high medical need and in most cases no specific treatments available," commented Magnus Hansson, M.D., Ph.D., Chief Medical Officer and VP of Preclinical and Clinical Development at NeuroVive.

The study is a double-blind, placebo-controlled, single-dose, dose-escalation phase I clinical study to investigate the pharmacokinetics, safety and tolerability of KL1333 in healthy volunteers. The first part of the study included dose levels of 25 mg and 50 mg. With both doses having been deemed safe, the MFDS has approved dose escalation and an updated study design. A multiple-dose, phase Ib study, sponsored by NeuroVive, is planned to be initiated in Europe and/or the U.S. in 2018.

"The results from the first part of the study are positive and the approval from the MFDS is of crucial importance for KL1333 and the continuation of its global development program. In close collaboration with our partner NeuroVive, we have a great possibility to develop KL1333 into a novel treatment opportunity for patients with genetic mitochondrial disorders, such as MELAS," said Soo-Hyun Lew, M.D. Executive Director of Development at Yungjin Pharm.

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 11:00 a.m. CET on 11 December 2017.

# For more information please contact:

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### About KL1333

KL1333 is a potent modulator of the cellular levels of NAD+, a central coenzyme 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, CPEO, PEO, Pearson and MERRF. Its mode of action is complementary to that of NVP015, which is intended to alleviate acute episodes of energy crises in genetic mitochondrial disorders with dysfunction in complex I and to NVP025, intended to protect the mitochondria in skeletal muscle from dysfunctional calcium handling and consequential muscle wasting.

## **About Yungjin Pharm**

Yungjin Pharm Co. Ltd., established in 1952, has been playing a major role as a forerunner in the Korean pharmaceutical industry for half a century. With the inspiring mission statement, "To relieve the suffering of mankind from diseases with our innovative, effective and safe pharmaceutical products", they have shown a successful contribution not only within Korea, but also through global expansion. As a result, they have received a total of 25 awards including the President Award for Superior Product Development, the Prime Minister Award, Industry Award and many more. These accomplishments demonstrate their sustainability and commitment to the development of innovative products and business excellence in both overseas and domestic segments. The company is listed on the South Korean stock market, KOSPI (KRX 003520).

NeuroVive Pharmaceutical AB (publ) 556595-6538



# NeuroVive's KL1333 program granted orphan drug designation by the European Commission

Lund, Sweden, 13 December 2017 – NeuroVive Pharmaceutical AB (Nasdaq Stockholm: NVP, OTCQX: NEVPF), announced today that the European Commission has granted orphan drug designation (ODD) for the company's KL1333 program for oral treatment of the genetic mitochondrial disease Mitochondrial Myopathy, Encephalopathy, Lactic acidosis and Stroke-like episodes (MELAS).

The decision by the Commission confirms the opinion previously issued by the European Medicine Agency's (EMA) Committee for Orphan Medicinal Products (COMP), which was communicated by NeuroVive on 8 November 2017. The orphan drug designation for KL1333, together with NeuroVive's status as a micro, small and medium-sized enterprise (SME), offers NeuroVive access to free scientific advice (protocol assistance) from the EMA, as well as fee reductions and further administrative assistance from EMA's SME office. If approved for orphan drug status when authorized for marketing, KL1333 would benefit from ten years of market exclusivity within the European Union.

KL1333 is being developed jointly by NeuroVive and the Korean pharmaceutical company Yungjin Pharm. KL1333 has shown promising results in preclinical models and a clinical phase I study is currently being conducted in Korea. NeuroVive plans to initiate the next clinical phase I study in 2018.

# For more information, please contact:

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NeuroVive Pharmaceutical AB (publ) - the mitochondrial medicine company. The company is listed on Nasdaq Stockholm, Small Cap, under the ticker symbol NVP. The share is also traded on the OTC Markets Group Inc market in the US. NeuroVive Pharmaceutical (OTC: NEVPF) trades on the OTCQX Best Market. Investors can find Real-Time quotes and market information for the company at www.otcmarkets.com/stock/NEVPF/quote

NeuroVive Pharmaceutical AB (publ) 556595-6538



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NeuroVive Pharmaceutical AB (publ) 556595-6538



# NeuroVive reports breakthrough in mitochondrial myopathy project

**Lund, Sweden, 30 January 2018, NeuroVive Pharmaceutical AB** (Nasdaq Stockholm: NVP, OTCQX: NEVPF) today announced a breakthrough in the company's project NVP025 for developing treatment of mitochondrial myopathy. In an experimental study carried out in collaboration with researchers at Karolinska Instutet in Stockholm, Sweden, the project's model substance has shown favourable effects which may counter disease progression in mitochondrial myopathy.

Professor Håkan Westerblad and his research group at Karolinska Institutet have evaluated the effects of NeuroVive's cyclophilin inhibitors in an experimental model of mitochondrial myopathy. At the end of the study's treatment duration, survival rate was 94 % in the group receiving treatment, compared to 50 % in the control group. Furthermore, muscle function was better in the treated group compared to the control group.

"The results from the study is an important milestone in NeuroVive's NVP025 project. The study shows that our cyclophilin inhibitors can be further developed towards the goal of offering mitochondrial myopathy patients a novel treatment option. We are now taking the project into its next phase, where we will optimize a candidate drug suitable for further development for patients with different types of muscle disorders," says Magnus Hansson, M.D., Ph.D, Chief Medical Officer and head of preclinical and clinical development at NeuroVive.

The researchers at Karolinska Institutet have previously shown that cyclophilin D, which is the target molecule in the NVP025 project, is present at abnormally high levels in the muscles of mitochondrial myopathy patients, and that inhibiting cyclophilin D with cyclosporin A counters the muscle weakness present in the experimental models.<sup>1)</sup> Cyclophilin inhibitors have also shown favourable effects in experimental models of other muscle disorders, such as Duchenne muscular dystrophy.<sup>2)</sup>

"The effects of NeuroVive's cyclophilin inhibitor are very exciting and are in line with results from previous studies, where cyclophilin inhibitors have been shown to counter disruption of mitochondrial function and muscle weakness in different models of muscular disorders. We are looking forward to continuing the positive collaboration with NeuroVive and taking the project additional steps forward in development," says Håkan Westerblad, Professor, Karolinska Institutet.

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 01:00 p.m. CET on 30 January 2018.

# For more information please contact:

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NeuroVive Pharmaceutical AB (publ) 556595-6538



# **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, MERRF syndrome (myoclonus epilepsy with ragged-red fibers) and mitochondrial MELAS (mitochondrial myopathy, encephalopathy, 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. There is a high unmet medical need of new and effective treatment options for mitochondrial myopathy.

#### **About Karolinska Institutet**

Karolinska Institutet is one of the world's leading medical universities. Its vision is to significantly contribute to the improvement of human health. Karolinska Institutet accounts for the single largest share of all academic medical research conducted in Sweden and offers the country's broadest range of education in medicine and health sciences. The Nobel Assembly at Karolinska Institutet selects the Nobel laureates in Physiology or Medicine.

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- 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) Alisporivir rescues defective mitochondrial respiration in Duchenne muscular dystrophy. <u>Schiavone M. Et al. Pharmacol Res.</u> 2017 Nov;125(Pt B):122-131.