



Q3

Interim report January - September 2018

KL1333 approved for phase I clinical trial

Important events January-September 2018

- NeuroVive's genetic mitochondrial disease drug candidate KL1333 mechanism of action published in scientific journal
- NeuroVive receives positive FDA feedback on its NeuroSTAT TBI development plan
- NeuroVive's NVP015 program will be supported through award of major research grant to Children's Hospital of Philadelphia
- NeuroVive and Yungjin reports positive KL1333 phase I clinical study results
- KL1333 receives FDA Orphan Drug Designation for treatment of mitochondrial diseases.
- NeuroVive initiates collaboration with leading US TBI research organization TRACK-TBI
- NeuroVive out-licenses targeted LHON therapy to BridgeBio Pharma's new subsidiary Fortify Therapeutics.
- The company reported positive efficacy data in an experimental model, entailing a breakthrough for the NVP025 mitochondrial myopathy project.
- NeuroVive conducts a preferential rights issue.

Important events after the reporting period

- NeuroVive receives KL1333 clinical trial regulatory approval from the UK regulatory authority
- NeuroVive reports first NeuroSTAT clinical efficacy signal in TBI
- NeuroVive presents first preclinical NV354 efficacy results in a model for mitochondrial disease
- NeuroVive has been awarded SEK 1.5 million as a first tranche of total SEK 5 million in funding from Vinnova, Sweden's innovation agency, and the Swelife call, for intensified development in the NVP015 project, the goal of which is to advance the candidate compound NV354 to clinical studies.

Financials third quarter (July-September 2018)

- Net revenues: SEK 0 (0)
- Other operating income: SEK 0 (397,000)
- Loss before tax: SEK -14,982,000 (-13,179,000)
- Loss per share:* SEK -0.20 (-0.26)
- Diluted loss per share:** SEK -0.20 (-0.26)

Financials first nine months (January-September 2018)

- Net revenues: SEK 0 (27,000)
- Other operating income: SEK 1,452,000 (550,000)
- Loss before tax: SEK -53,516,000 (-56,824,000)
- Loss per share:* SEK -0.72 (-1.04)
- Diluted loss per share:** SEK -0.72 (-1.04)

* 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 CEO, Erik Kinnman

The third quarter of this year was yet another quarter with several significant advances for NeuroVive's research and development. In our traumatic brain injury program, we received positive results from biomarker measurements and in our KL1333 project, the UK Medicines and Healthcare Products Regulatory Agency (MHRA) approved our application for a Phase I clinical trial in patients and healthy volunteers. Another major success during the quarter was that our NVP015 project was accelerated by the award of a large research grant to the Children's Hospital of Philadelphia.

NeuroSTAT – encouraging results from biomarker measurements

Biomarkers in patient samples from the CHIC trial have been analyzed to determine endpoints for our clinical traumatic brain injury (TBI) program. The biomarkers, reflecting various aspects of the ongoing brain cell damage caused by TBI, were measured in the cerebrospinal fluid of patients with severe brain injuries. The results are highly positive and indicate that NeuroSTAT inhibits the secondary injury cascades resulting from brain trauma. The outcome of the trial further strengthens our positive view of NeuroSTAT's possibilities and we are now continuing to prepare for a larger Phase II efficacy trial.

KL1333 trial receives regulatory approval

The important KL1333 project for the treatment of mitochondrial genetic disorders took a major step forward when our application for a Phase I trial in patients and healthy volunteers was approved by the UK Medicines and Healthcare Products Regulatory Agency (MHRA). The trial will be conducted in the UK. The MHRA's approval is not only a key milestone, it is also a stamp of quality for the planning and design of our trial. The trial will bring us one step closer to our goal of developing a new therapy for patients with severe mitochondrial genetic disorders who currently lack treatment options.

NVP015 project supported by research grant to CHOP

Some more good news this quarter was that our academic partner, the Children's Hospital of Philadelphia (CHOP), was granted a three-year research grant of USD 4 million by the US Department of Defense Congressionally Directed Medical Research Program (CDMRP) for academic studies focusing on NV354, an alternative energy source for the treatment of mitochondrial genetic disorders. The aim of the development program is to select a candidate drug for an Investigational New Drug (IND) application to the US Food and Drug Administration. An approved IND application is an important process before initiating Phase I clinical trials in the US.



Intense business development

During the quarter, our business development was focused on three areas: continued intense efforts to identify a suitable partner for our liver fibrosis niched NASH project, financing of the NeuroSTAT project's planned Phase II trial and strengthened relationships with BridgeBio/Fortify in regard to the out-licensed NVP015 project, focused on targeted treatment of the eye disease Leber hereditary optic neuropathy (LHON). Overall, the third quarter of the year was positive for NeuroVive, with advanced positions in several key areas.

Financing for continued success

For NeuroVive, as for a majority of companies developing new drugs, financing of the business is constantly in focus. During the year we have conducted a successful rights issue, which provided the company with SEK 64.2 million after transaction costs. In addition, we have received minor external funding for individual projects and out-licensed chemistry from NVP015 for LHON.

For continued success in our clinical projects, we will need additional financing in the first half of 2019. The Board therefore actively works with various funding options.

Erik Kinnman

CEO, NeuroVive Pharmaceutical AB
November 22, 2018



NeuroVive's research and development

NeuroVive has a leading position in mitochondrial research and development. The company's objective is to design pharmaceuticals that preserve the integrity and function of mitochondria for indications where there is a great medical need. Drug development is a comprehensive and carefully regulated process. By working with different partners, NeuroVive strives to make this process as flexible, cost-effective and successful as possible.

Two principle paths

NeuroVive's aim is to help patients who currently have few or no treatment options. The company's business model consists of two parts: the first one involves the development of drugs for rare diseases, and the second one includes the development of mitochondrial drugs for commonly occurring diseases.

Development to the market. Development of drugs for rare diseases is done through preclinical and clinical development to market. NeuroVive's ambition is that these drugs shall be classified as orphan drugs.

Out-licensing in pre-clinical phase. For commonly occurring diseases with high commercial potential, where the clinical studies are very extensive and costly, NeuroVive's objective is to out-license the projects in late preclinical phase.

Value creation with limited cost and risk

The diversified project portfolio allows the company to take orphan drugs to the market comparatively quickly, and at a limited cost and risk. At the same time, innovation in common diseases can be industrialized and value created through out-licensing and partnerships.

	Development to the market with/without partner					Outlicensed	Outlicensing at the preclinical stage		
	Brain Injury	Mitochondrial Respiratory Chain Diseases					NASH		Liver cancer
	TBI	Chronic	Acute	Myopathy	LHON		Fibrosis	Metabolic	HCC
Market									
Phase III									
Phase II									
Phase I									
Preclinical									
Lead selection									
Discovery									
	NeuroSTAT®	KL1333	NV354	NVP025	NVP015		NV556	NVP022	NVP024

A photograph showing emergency responders in reflective gear at night, attending to a person on the ground. The scene is illuminated by emergency lights, creating a sense of urgency.

More than 50 million people suffer a traumatic brain injury each year.

The global healthcare costs are estimated to 400 billion dollars.

NeuroVive's program for traumatic brain injury (TBI)

Traumatic brain injury (TBI) is caused by external force to the head resulting in immediate damage to nerve cells. The damage continues to worsen for several days after the acute trauma. The most common causes for TBI are trips and falls, traffic accidents, and assault and battery.¹⁾ With more than 50 million new cases occurring each year, TBI is estimated to cost the global economy nearly 400 billion dollars annually in direct and indirect healthcare costs.²⁾ A large number of patients suffer moderate to severe functional disabilities requiring intensive care and various forms of support.

NeuroSTAT – candidate drug in clinical phase II study

Treatment objective

The aim for NeuroStat is to prevent the emergence of neurological and functional secondary brain damage after a traumatic injury, and thereby establish a therapy that will lead to increased survival, improved quality of life and preserved neurological function.

Project status: clinical phase II

NeuroVive's candidate drug for TBI treatment, NeuroSTAT, has been evaluated in a Phase II clinical study (Copenhagen Head Injury Ciclosporin-CHIC) at Copenhagen University Hospital in Denmark. The study, which ended in May 2017, studied safety, tolerability and pharmacokinetics, i.e. the effect of two different doses of the active ingredient ciclosporin on circulation in the body and passage to the brain in patients with severe traumatic brain injury.

The protective effects in traumatic brain injury and the relationship between efficacy and drug concentrations in the brain, were also assessed in an experimen-

tal study at the University of Pennsylvania (Penn). The NeuroSTAT candidate drug has orphan drug designation in both Europe and the US.

Objectives for 2018

- Publication/presentation of results from the CHIC study and from the collaboration with Penn. ✓
- Results from evaluation of biomarkers prior to the upcoming NeuroSTAT development program. ✓
- Secure financing for upcoming Phase II efficacy study.
- Meeting with the FDA prior to development programs in the US. ✓

Third quarter news

- NeuroVive received positive FDA feedback on its NeuroSTAT TBI development plan

News after the end of the period

- NeuroVive reports first NeuroSTAT clinical efficacy signal in TBI

1) www.internetmedicin.se/page.aspx?id=1178

2) Maas A Et al. Traumatic brain injury: integrated approaches to improve prevention, clinical care, and research. The Lancet Neurology. 2017 Nov; 16(12):987.



Genetic mitochondrial diseases can lead to severe symptoms, such as mental retardation, dementia, deafness and blindness.

NeuroVive's program for genetic mitochondrial diseases

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

KL1333 – candidate drug in clinical phase I

Treatment objective

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 longterm beneficial effects on energy metabolism. The candidate drug is intended for oral treatment in primary genetic mitochondrial disorders such as MELAS, KSS, PEO, Pearson and MERRF.

The candidate drug in the KL1333 project has already been granted orphan drug designation in Europe and the United States, and there is also potential for obtaining orphan drug designation for the future drug candidates in the NVP015 and NVP025 projects. Orphan drug designation allows for a faster and less costly route to the market, as well as a higher market price for the drug.

Project status: clinical phase I

KL1333 is currently being evaluated in clinical phase I-studies and has been granted orphan drug designation in both the United States and Europe.

Original objectives for 2018

- Results from clinical single-dose Phase Ia study of KL1333 in South Korea, sponsored by Yungjin Pharm. ✓
- Start-up of NeuroVive's Phase Ib clinical study of KL1333 in Europe.

Updated objectives

- Start-up of NeuroVive's Phase Ib clinical study of KL1333 in Europe during the first six months of 2019 with further optimized bio-analysis methods.

Third quarter news

- KL1333 mechanism of action published in scientific journal

News after the end of the period

- NeuroVive receives KL1333 clinical trial regulatory approval from the UK regulatory authority

NVP025 – selection of lead candidate

Treatment objective

NVP025 is focused on chronic treatment of mitochondrial myopathies by utilizing a potent cyclophilin inhibitor from NeuroVive's Sangamide class of compounds. Mitochondrial myopathies manifest in the MELAS, PEO, KSS, and MERRF syndromes.

The goal is to develop a treatment that protects the mitochondria in the muscles from disturbed calcium handling and subsequent muscular dystrophy, thus preventing the muscle weakness associated with these diseases. In collaboration with the Karolinska Institute in Stockholm, NeuroVive has demonstrated that cyclophilin inhibitors can slow the disease progression and increase survival rates in an experimental mitochondrial myopathy model.

Project status: selection of lead candidate

The company will conduct follow-up dose-response studies in 2018 to be able to choose the optimized drug candidate and route of administration.

Original objectives for 2018

- Results from experimental studies of mitochondrial myopathies, carried out at the Karolinska Institute. ✓
- Selection of candidate compound.

Updated objectives

- Perform dose-response studies for selection of candidate substance and route of administration during 2019.

NVP015/NV354 – candidate drug in pre-clinical development

Treatment objective

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

Project status: pre-clinical development

Within the project a lead compound, NV354, has been selected for further development in the program based on tolerability, oral bioavailability, plasma stability and organ delivery, specifically to the brain.

Objectives for 2018

- Final results from the collaborative project with Dr. Marni Falk at CHOP for studies of NVP015 compounds in experimental disease models. ✓
- Initial results from CHOP studies of NeuroVive's NVP015 compounds as a therapeutic option for chemical threats. ✓
- Continued experimental in vivo efficacy studies of the selected NVP015 compound, NV354. ✓

Third quarter news

- NeuroVive's NVP015 program will be supported through award of major research grant to Children's Hospital of Philadelphia.


News after the end of the period

- NeuroVive presents first preclinical NV354 efficacy results in a model for mitochondrial disease.
- NeuroVive has been awarded SEK 1.5 million as a first tranche of total SEK 5 million in funding from Vinovna, Sweden's innovation agency, and the Swelife call, for intensified development in the NVP015 project, the goal of which is to advance the candidate compound NV354 to clinical studies.

Out-licensing of project for local treatment of LHON

On June 18, 2018, NeuroVive out-licensed molecules from NVP015 for a targeted treatment of Leber's Hereditary Optic Neuropathy (LHON) to BridgeBio Pharma's new subsidiary Fortify Therapeutics. Fortify's ambition is to further develop the in-licensed NVP015 chemistry in order to establish a therapy for LHON.

LHON is caused by mitochondrial DNA mutations in subunits of complex I, and affects primarily retinal cells and visual nerve, and results in severe vision loss. The disease predominantly affects young adult men between 20 to 40 years of age. The prevalence of LHON in Europe is between 1:30 000–1:50 000.



The fatty liver disease NASH has a strong link to diabetes and obesity.

The global NASH market is expected to exceed 25 billion dollars 2026.

NeuroVive's program for NASH

Non-alcoholic fatty liver disease (NAFLD) affects 20-25 percent of the global population. When fat deposits in the liver are combined with inflammation and scar tissue (fibrosis), the disease has progressed to non-alcoholic steatohepatitis (NASH) – a condition that may lead to liver cirrhosis or hepatocellular carcinoma (liver cancer). An estimated 20 percent of people with NAFLD have NASH and there is a strong link between NASH and other metabolic syndromes such as diabetes and obesity. There are no approved drugs for treating NASH at the present time, but with forthcoming treatments, the NASH market is expected to exceed USD 25 billion, globally, by 2026.¹⁾

NV556 – candidate drug in pre-clinical development

Treatment objective

NV556 is a candidate drug with a directly acting anti-fibrotic mechanism of action targeting NASH patients who have progressed from the initial metabolic stage characterized by fat storage and inflammation. NV556 is a potent cyclophilin inhibitor derived from NeuroVive's Sangamide class of compounds.

Project status: pre-clinical

As preclinical results have been analyzed it became clear that the greatest potential for the project is within the subgroup of NASH patients with liver fibrosis, meaning at a later stage of disease progression. This makes NV556 best suited as a complement to NASH therapy focused on the early metabolic stage of the disease. Furthermore, it provides an opportunity to

develop projects targeting other types of fibrotic disease. The company has therefore reinforced the message about the liver fibrosis effect of NASH and the appropriate market segment, and it has partly redirected the out-licensing activities for NV556 aimed at reaching an agreement with a suitable partner for this niched NASH product.

Original objectives for 2018

- Out-licensing and/or partnering.

Updated objectives

- Out-licensing and/or partnership within liver fibrosis and NASH with an opportunity to expand treatment options to other types of fibrotic disease during the first six months of 2019.

1) Global Data, OpportunityAnalyzer: NASH – Opportunity Analysis and Forecasts to 2026

NVP022 – evaluation of model compounds

Treatment objective

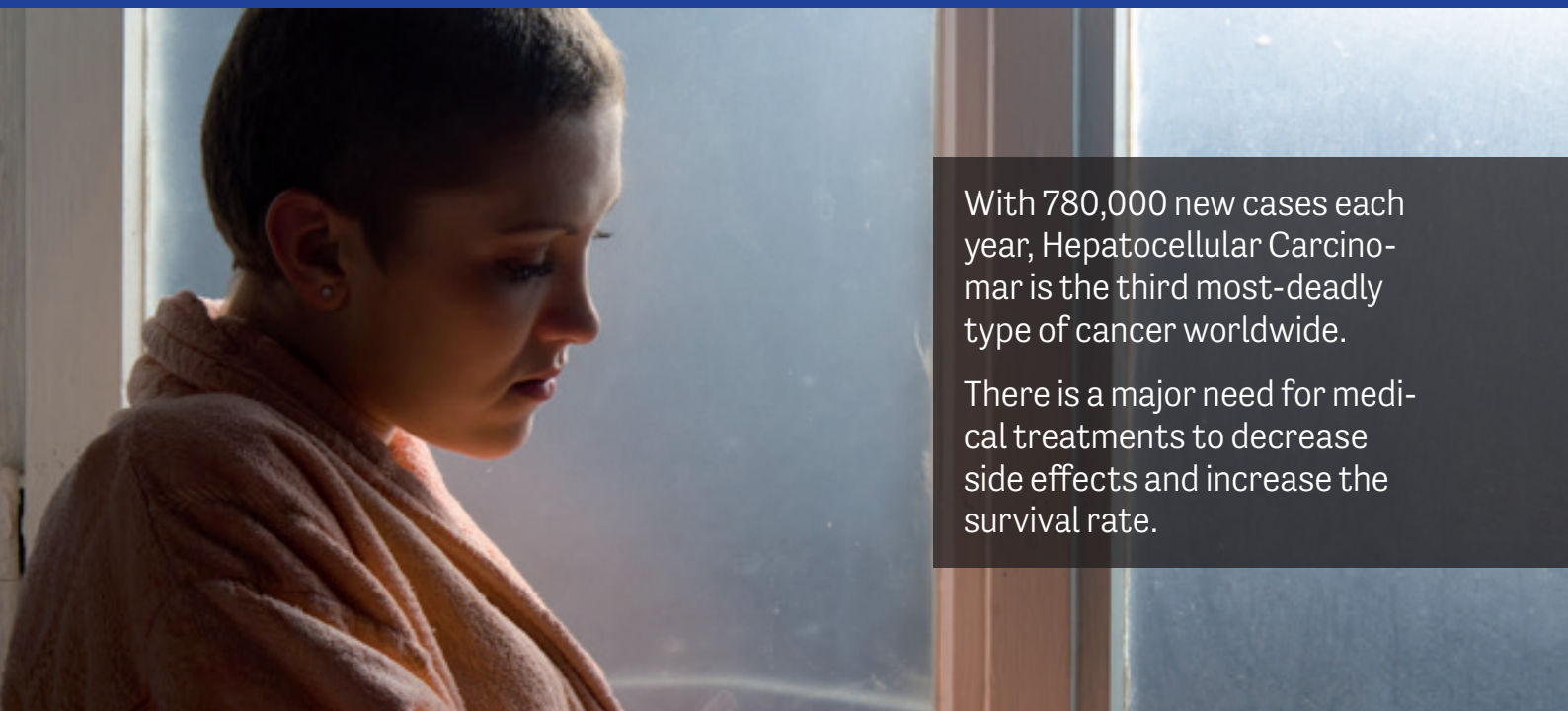
NVP022 targets the metabolic components of NASH by using mild, liver-targeted protonophores to uncouple energy-linked functions and increase energy expenditure in the liver. This removes excess fat storage and thereby counteracts the pathophysiological processes in NASH.

Project status: evaluation of model compounds

NeuroVive has evaluated various substances within the project in 2018 and has selected a candidate substance, NV422, based on favorable pharmacokinetic capacity and good tolerability.

Objectives for 2018

- Proven effect of an NVP022 compound in a preclinical NASH model
- Selection of candidate compound ✓
- Initiation of out-licensing activities



With 780,000 new cases each year, Hepatocellular Carcinoma is the third most-deadly type of cancer worldwide.

There is a major need for medical treatments to decrease side effects and increase the survival rate.

NeuroVive's program for liver cancer

Hepatocellular Carcinoma (HCC) is the sixth most common type of cancer, with about 780,000 new cases diagnosed annually, and the third most deadly type of cancer worldwide. In Europe, HCC is the 14th most common type of cancer, with 63,500 new cases diagnosed each year. While surgery, chemotherapy and radiotherapy are important starting points for the treatment of liver tumors, there is a major medical need for more and effective complementary medical treatments to decrease side effects and increase the survival rate for people with liver cancer.^{1) 2) 3) 4)}

NVP024 – evaluation of model compounds

Treatment objective

NVP024 is focused on the anticancer properties of a sub-set of the company's sanglifehrin-based compounds. Together with international partners, NeuroVive's research team has demonstrated that these compounds show powerful anticancer effects in preclinical models of HCC.

Project status: evaluation of model compounds

Additional confirmatory tests are ongoing, for instance within the framework of a PhD project at Lund University, funded by the Foundation for Strategic Research.

Objectives for 2018

- Confirmatory tests in complementary preclinical experimental models.
- Initial results from the industrial PhD student collaboration with Lund University. ✓

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.

3) Sandhu DS1, Tharayil VS, Lai JP, Roberts LR. Treatment options for hepatocellular carcinoma. *Expert Rev Gastroenterol Hepatol*. 2008 Feb;2(1):81-92. doi:10.1586/17474124.2.1.81.

4) <http://www.cancerresearchuk.org/health-professional/cancer-statistics/statistics-by-cancer-type/liver-cancer/incidence#heading-Nine>

Financial information

Revenues

The consolidated turnover during the third quarter of 2018 was SEK 0 (0). Other operating revenues for the third quarter of 2018 were SEK 0 (397,000). The consolidated turnover for the first nine months was SEK 0 (27,000) and the operating revenues amounted SEK

1,452,000 (550,000). Other operating income includes research contributions from BridgeBio of SEK 876,000 and research contribution from Vinnova of 452,000 SEK.

Results of operations

The operating loss for the third quarter was SEK 15,075,000 (12,793,000). The operating loss for the first nine months was SEK 53,230,000 (56,168,000). The net loss before tax for the third quarter amounted to SEK 14,982,000 (13,179). The net loss before tax for the first nine months was SEK 53,516,000 (56,824,000).

The operating loss was affected by external expenses, which for the first nine months were SEK 41,437,000 (34,505,000). During the first nine months, expenses

related to development projects have affected the result with SEK 30,284,000 (22,827,000) whereof SEK 18,214,000 (11,779,000) relates to project in clinical phase. Personnel expenses during the first nine months amount to SEK 11,039,000 (9,968,000). Other operating expenses amount to, SEK 765,000 (11,117,000) and pertains to exchange-rate losses. Previous year SEK 10,981,000 related to disposal of subsidiary and the remaining portion of other operating expenses pertained to exchange-rate losses.

Financial position

The equity/assets ratio was 90 (94) percent as of 30 September 2018, and equity was SEK 116,510,000 (115,936,000) compared to beginning of the year. The equity includes funds from the in April completed rights issue, which provided the company with SEK 64,176,000 after deduction of issue costs and compensation for guarantee commitments of SEK 14,324,000. Cash and cash equivalents amounted to SEK 38,371,000 (35,436,000) as of 30 September 2018,

an increase of SEK 9,379 from the beginning of the year. Total assets as of 30 September 2018 were SEK 129,692,000 (123,851,000).

The Group needs additional financing in the first half of 2019 to ensure continued operations. 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 -12,536,000 (-14,929,000). Operating cash flow from the first nine months was SEK -52,075,000 (-48,192,000). The cash flow effect related to investments in intangibles equals SEK -2,645,000

(-3,250,000) for the first nine months. Cash flow for the third quarter equals SEK -13,524,000 (-11,866,000). Cash flow for the first nine months equals SEK 9,375,000 (-58,080,000).

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 to the Board fees and remuneration resulting from employment contracts, no related party transactions have taken place. The principles for these Board fees are stated in the Annual Report 2017. During the period, no compensation has been paid under the agreement, in relation to mitochondrial energy regulation projects, with the Research Group at Lund University, which includes CSO Eskil Elmér and CMO Magnus Hansson.

Disclosures regarding transactions between the group and other related parties are stated below.

	1 Jan. 2018	1 Jan. 2017
(SEK 000)	30 Sep. 2018	30 Sep. 2017
Stanbridge bvba (owned by Gregory Batcheller, until Nov 6, 2017, Executive Chairman)	-	560
Total transactions with related parties	-	560

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. These assets should be measured at fair value and are classified as "financial assets measured at fair value through other comprehensive income."

The holding corresponds to 10% in one of NeuroVive's R&D partner companies. No information is available for measuring the holding at present value, and NeuroVive makes the assessment that there are no circum-

stances to indicate that fair value should deviate materially from cost. For this reason, the holding continues to be recognized at cost.

Other financial assets belong to the category "loan receivables and accounts receivable" which are valued at accrued acquisition value. 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 2018 was 10 (10), of which 4 (5) are women.

Parental company

Company earnings after tax for the first nine months amounts to SEK -53,511,000 (-7,337,000). Most of the Group's operations are conducted within the parent

company. Accordingly, no further specific information regarding the parent company is presented.

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. During the current period, the Board has been actively working to ensure the business needs for financing. No other significant changes in relation to risk or uncertainties occurred during the current period.

In 2004, NeuroVive entered into a License Agreement with CicloMulsion AG under which NeuroVive licensed the rights to use and develop products based on a certain pharmaceutical technology.

In March 2013, CicloMulsion AG commenced an arbitration seeking declaratory relief aimed at establishing the company's rights to royalties, which CicloMulsion AG claims that NeuroVive is obliged to pay under the terms of the License Agreement. CicloMulsion AG also

made other claims in relation to NeuroVive's obligations under the License Agreement.

A partial award issued in 2016 was set aside by the Court of Appeal for Skåne and Blekinge with the exception of the question for which the Tribunal had reserved its decision. NeuroVive has appealed parts of the ruling to the Supreme Court. This proceeding is pending.

The former tribunal was replaced by a new tribunal following a request for the release of the arbitrators filed by CicloMulsion and the constitution of a new Arbitral Tribunal has been finalized. The scope and the timeline of the further proceeding is currently under discussion.

The ongoing dispute with CicloMulsion AG may result in future payment obligations, which could have a negative impact on the company's earnings and financial position. The Company has not reserved for any future payment obligations as the amount at this time cannot be calculated.

NeuroVive is not involved in any other disputes.

For more detail of risks and uncertainty factors, refer to the Statutory Administration Report in the Annual Report 2017 and the prospectus published 9 April 2018 for the preferential rights issue in April 2018.

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 2018	21 February 2019
Interim Report Jan-Mar 2019	21 May 2019
Interim Report Jan-Jun 2019	21 August 2019
Interim Report Jan-Sep 2019	20 November 2019
Year-End Report 2019	19 February 2020
The interim reports and the Annual Year Report are available at www.neurovive.com	

Annual General Meeting 2019

NeuroVives Annual General Meeting will be held at Medicon Village, Scheelevägen 2, in Lund on 25 April 2019 at 4 pm.

The Nomination Committee for the 2019 AGM comprises:

- **Kristina Ingvar** – for Rothesay Limited/
John Fällström
- **Michael Vickers** – for Maas Biolab LLC /Marcus Keep
- **Fredrik Olsson** – for Baulos Capital Belgium SA

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 2019.

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 2017 on pages 54-67.

IFRS 9 Financial instruments specifies how an entity should classify, measure and recognize financial assets and financial liabilities. IFRS 9 introduces a new approach for recognizing credit losses based on expected credit losses, which may entail earlier recognition of credit losses. Considering that NeuroVive's revenue generation has been limited to date, the need for impairment is also limited and no quantitative impact has thus arisen. IFRS 9 also introduces new rules for hedge accounting. Since NeuroVive does not apply hedge accounting, the company is not affected by these changes.

Financial instruments are classified in accordance with IFRS 9, based on the company's business model. Neuro-

Vive classifies and measures its financial instruments based on the business model for managing the asset and the asset's contractual cash flow characteristics. On this basis, NeuroVive will continue to apply the previous method of classification, whereby all financial assets, with the exception of holdings in unlisted securities, are measured at amortized cost, in the category now known as "Financial assets measured at amortized cost." As in preceding years, the unlisted securities will be measured at fair value through other comprehensive income, and the new name of the category will be "Financial assets measured at fair value through other comprehensive income." As in preceding periods, all financial liabilities will be measured at amortized cost. IFRS 9 came into effect on January 1, 2018 and has not therefore had any quantitative impact on NeuroVive.

IFRS 15 "Revenue from contracts with customers". IFRS 15 treats how the accounting of revenues shall be made. Revenue shall according to IFRS 15 be accounted when the customer receives the control over the sold goods or service and the customer has opportunity to use the goods or services. The Group estimate that the new standard has no major impact on the financial statements, but will lead to increased disclosure requirements. IFRS 15 came into effect on January 1, 2018. Since the Group's inflows are still limited, the introduction has not resulted in any quantitative impact or need for additional disclosures on historical inflows.

IFRS 16, Lease Agreement, replaces IAS 17 and will apply as of January 1, 2019. The standard requires that assets and liabilities attributable to all leases, with some exceptions, be reported in the balance sheet. Evaluation and assessment of the consequences of the implementation has not yet been completed.

Consolidated Statement of Comprehensive Income

(SEK 000)	Note	1 Jul, 2018 30 Sep, 2018	1 Jul, 2017 30 Sep, 2017	1 Jan, 2018 30 Sep, 2018	1 Jan, 2017 30 Sep, 2017	1 Jan, 2017 31 Dec, 2017
Net sales		-	-	-	27	27
Other operating income		-	397	1,452	550	248
		-	397	1,452	576	275
<i>Operating expenses</i>						
Other external expenses		-11,217	-9,757	-41,437	-34,505	-46,415
Personnel cost		-3,249	-2,963	-11,039	-9,968	-12,417
Depreciation and write-down of tangible and intangible assets		-498	-412	-1,441	-1,155	-1,595
Other operating expenses		-111	-57	-765	-11,117	-10,936
		-15,075	-13,190	-54,682	-56,745	-71,363
Operating income		-15,075	-12,793	-53,230	-56,168	-71,088
<i>Profit/loss from financial items</i>						
Result from shares in associated company		-	-	-	-	56
Financial income		94	11	320	121	65
Financial costs		-1	-397	-606	-776	-636
		93	-386	-286	-655	-515
Profit/loss before tax		-14,982	-13,179	-53,516	-56,824	-71,603
Income tax	2		-			
Profit/loss for the period		-14,982	-13,179	-53,516	-56,824	-71,603
<i>Other comprehensive income</i>						
Items that may be reclassified to profit or loss						
Translation differences on foreign subsidiaries			30	4	19	1
Total comprehensive income for the period		-14,983	-13,149	-53,512	-56,805	-71,602
<i>Loss for the period attributable to:</i>						
Parent company shareholders		-14,982	-13,098	-53,515	-51,950	-66,728
Non-controlling interests		-	-81	-1	-4,874	-4,875
		-14,982	-13,179	-53,516	-56,824	-71,603
<i>Total comprehensive income for the period</i>						
Parent company shareholders		-14,917	-13,016	-53,742	-51,955	-66,895
Non-controlling interests		-66	-133	230	-4,850	-4,707
		-14,983	-13,149	-53,512	-56,805	-71,602
Earnings per share before and after dilution(SEK)						
based on average number of shares		-0.20	-0.26	-0.72	-1.04	-1.33

Consolidated Statement of Financial Position

(SEK 000)	Note	30 Sep, 2018	30 Sep, 2017	31 Dec, 2017
ASSETS				
Non-current assets				
<i>Intangible assets</i>	1			
Development costs		51,941	51,941	51,941
Patents		22,597	19,972	20,627
Other Intangible assets		1,647	1,781	1,747
		76,185	73,694	74,315
<i>Tangible assets</i>				
Equipment		167	187	162
		167	187	162
<i>Financial assets</i>				
Other long-term securities		13,102	13,102	13,102
		13,102	13,102	13,102
Total non-current assets		89,454	86,983	87,579
<i>Current assets</i>				
Other receivables		1,263	1,118	1,568
Prepaid expenses and accrued income		604	314	1,967
Cash and cash equivalents		38,371	35,436	28,992
		40,238	36,868	32,527
TOTAL ASSETS		129,692	123,851	120,106
EQUITY AND LIABILITIES				
<i>Equity attributable to the shareholders of the parent company</i>				
Share capital		4,579	2,528	2,616
Additional paid in capital		489,440	422,607	427,226
Translation reserve		386	775	613
Retained earnings		-383,255	-314,962	-329,740
Total equity attributable to the shareholders of the parent		111,149	110,948	100,716
Non-controlling interests		5,361	4,988	5,131
Total equity		116,510	115,936	105,846
<i>Short-term liabilities</i>				
Accounts payable		3,698	2,336	7,525
Other liabilities		852	847	863
Accrued expenses and deferred income		8,632	4,732	5,871
		13,182	7,915	14,260
Total liabilities		13,182	7,915	14,260
TOTAL EQUITY AND LIABILITIES		129,692	123,851	120,106

Consolidated Statement of Changes in Equity

(SEK 000)	Equity attributable to the shareholders of the parent company					Non-controlling interests	Total equity
	Share-capital	Additional paid in capital	Translation reserve	Retained earnings	Total		
Opening balance, 1 January 2018	2,616	427,226	613	-329,740	100,716	5,131	105,846
Comprehensive profit/loss for the period							
Profit/loss for the period	-	-		-53,515	-53,515	-1	-53,516
Other comprehensive income							
Translation differences	-	-	-227	-	-227	231	4
Other comprehensive profit/loss for the period, net after tax	-	-	-227	-	-227	231	4
Total comprehensive profit/loss	-	-	-227	-53,515	-53,742	230	-53,512
Transactions with shareholders							
Rights Issue*	1,963	62,214	-	-	64,176	-	64,176
Total transactions with shareholders	1,963	62,214	-	-	64,176	-	64,176
Closing balance, 30 September 2018	4,579	489,440	386	-383,255	111,149	5,361	116,510

Opening balance, 1 January 2017	2,473	418,339	780	-266,146	155,446	12,858	168,304
Comprehensive profit/loss for the period							
Profit/loss for the period	-	-		-51,950	-51,950	-4,874	-56,824
Other comprehensive income							
Translation differences	-	-	-5	-	-5	24	19
Other comprehensive profit/loss for the period, net after tax	-	-	-5	-	-5	24	19
Total comprehensive profit/loss	-	-	-5	-51,950	-51,955	-4,850	-56,805
Transactions with shareholders							
Share issue	55	4,268	-	-	4,323	-	4,323
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 2017	2,473	418,339	780	-266,146	155,446	12,858	168,304
Comprehensive profit/loss for the period							
Profit/loss for the period	-	-		-66,728	-66,728	-4,875	-71,603
Other comprehensive income							
Translation differences	-	-	-167	-	-167	168	1
Other comprehensive profit/loss for the period, net after tax	-	-	-167	-	-167	168	1
Total comprehensive profit/loss	-	-	-167	-66,728	-66,895	-4,707	-71,602
Transactions with shareholders							
Share issue	143	8,887	-	-	9,030	-	9,030
Shareholder contribution	-	-	-	-	-	114	114
Change of ownership in share issue	-	-	-	3,134	3,134	-3,134	-
Total transactions with shareholders	143	8,887	-	3,134	12,164	-3,020	9,144
Closing balance, 31 December 2017	2,616	427,226	613	-329,740	100,716	5,131	105,846

*Total equity includes funds from the in April completed preferential rights issue with SEK 78,500,000 less transaction costs and compensation for guarantee commitments, SEK 14,326,000.

Consolidated Statement of Cash Flows

(SEK 000)	1 Jul, 2018 30 Sep, 2018	1 Jul, 2017 30 Sep, 2017	1 Jan, 2018 30 Sep, 2018	1 Jan, 2017 30 Sep, 2017	1 Jan, 2017 31 Dec, 2017
<i>Cash flow from operating activities</i>					
Operating income	-15,075	-12,792	-53,230	-56,169	-71,088
<i>Adjustments for non-cash items:</i>					
Depreciation	498	412	1,441	1,155	1,595
Currency differences on intercompany items	-	-289	-	-279	-35
Disposal of Business	-	-	-	10,981	10,936
Result from shares in associated company	-	-	-	-	56
Interest received	94	11	320	121	65
Interest paid	-0	-397	-606	-776	-149
Net cash from operating activities before changes in working capital	-14,484	-13,055	-52,075	-44,967	-58,620
<i>Changes in working capital</i>					
Increase/decrease of other current assets	1,495	271	1,669	785	-1,273
Increase/decrease of other short-term liabilities	453	-2,145	-1,669	-4,010	1,769
Changes in working capital	1,948	-1,875	0	-3,225	496
Cash flow from operating activities	-12,536	-14,929	-52,075	-48,192	-58,124
<i>Investing activities</i>					
Acquisition of intangible assets	-944	-1,222	-2,645	-3,250	-4,204
Acquisition of tangible assets	-45	-	-82	-40	-40
Disposal business	-	-	-	-11,035	-11,035
Cash flow from investing activities	-989	-1,222	-2,727	-14,325	-15,279
<i>Financing activities</i>					
New share issue	-	4,171	64,176	4,323	9,031
Shareholder contribution subsidiary	-	114	-	114	114
Cash flow from financing activities	-	4,285	64,176	4,437	9,145
Cash flow for the period	-13,524	-11,866	9,375	-58,080	-64,258
Cash and cash equivalents at the beginning of the period	51,896	46,984	28,992	93,251	93,251
Effect of exchange rate changes on cash	-1	318	5	265	-
Cash and cash equivalents at end of period	38,371	35,436	38,371	35,436	28,992

Parent Company Income Statement

(SEK 000)		1 Jul, 2018	1 Jul, 2017	1 Jan, 2018	1 Jan, 2017	1 Jan, 2017
	Note	30 Sep, 2018	30 Sep, 2017	30 Sep, 2018	30 Sep, 2017	31 Dec, 2017
Net sales		-	-	-	27	27
Other operating income		-0	397	1,452	550	248
		-0	397	1,452	576	275
<i>Operating expenses</i>						
Other external expenses		-11,218	-9,313	-41,430	-33,993	-45,857
Personnel cost		-3,249	-2,964	-11,039	-9,742	-12,190
Depreciation and write-down of tangible and intangible assets		-498	-412	-1,441	-1,144	-1,584
Other operating expenses		-110	-35	-765	-141	
		-15,076	-12,723	-54,675	-45,020	-59,631
Operating income		-15,076	-12,326	-53,223	-44,443	-59,357
<i>Profit/loss from financial items</i>						
Result from shares in group company		-		-	7,652	7,652
Result from shares in associated company		-		-	-	56
Interest income and other similar profit items		94	5	314	85	29
Interest expenses and other similar loss items		-	-398	-602	-631	-490
		94	-393	-288	7,106	7,247
Profit/loss before tax		-14,982	-12,719	-53,511	-37,337	-52,109
Income tax	2	-	-	-		-
Profit/loss for the period		-14,982	-12,719	-53,511	-37,337	-52,109

Statement of Comprehensive Income, Parent Company

(SEK 000)		1 Jul, 2018	1 Jul, 2017	1 Jan, 2018	1 Jan, 2017	1 Jan, 2017
	Note	30 Sep, 2018	30 Sep, 2017	30 Sep, 2018	30 Sep, 2017	31 Dec, 2017
Profit/loss for the period		-14,982	-12,719	-53,511	-37,337	-52,109
Other comprehensive income		-	-	-	-	-
Total comprehensive profit/loss for the period		-14,982	-12,719	-53,511	-37,337	-52,109

Parent Company Balance Sheet

(SEK 000)	Note	30 Sep, 2018	30 Sep, 2017	31 Dec, 2017
ASSETS				
Non-current assets				
<i>Intangible assets</i>	1			
Development costs		51,706	51,706	51,706
Patents		22,597	19,972	20,627
Other intangible assets		1,647	1,781	1,747
		75,950	73,460	74,080
<i>Tangible assets</i>				
Equipment		167	187	162
		167	187	162
<i>Financial assets</i>				
Other long-term placement		13,101	13,102	13,102
Shares in subsidiaries	3	23,625	23,625	23,625
		36,726	36,727	36,727
Total non-current assets		112,843	110,373	110,969
Current assets				
<i>Short term receivables</i>				
Receivables from group companies		-	-	-
Other receivables		1,260	1,115	1,566
Prepaid expenses and accrued income		604	315	1,967
		1,864	1,430	3,533
<i>Cash and bank balances</i>		38,281	35,318	28,883
Total current assets		40,145	36,747	32,416
TOTAL ASSETS		152,988	147,120	143,385
EQUITY AND LIABILITIES				
Equity				
<i>Restricted equity</i>				
Share capital		4,579	2,528	2,616
Statutory reserve		1,856	1,856	1,856
Development expenditure reserve		10,610	10,779	10,610
		17,045	15,163	15,082
<i>Unrestricted equity</i>				
Share premium reserve		71,101	4,268	8,887
Retained earnings		105,173	157,114	157,283
Profit/loss for the period		-53,511	-37,337	-52,109
		122,763	124,045	114,061
Total equity		139,808	139,208	129,143
Short-term liabilities				
Accounts payable		3,698	2,336	7,525
Other liabilities		852	847	863
Accrued expenses and deferred income		8,630	4,729	5,854
		13,180	7,912	14,242
TOTAL EQUITY AND LIABILITIES		152,988	147,120	143,386

Notes

Note 1 – Intangible assets

(SEK 000)	Development costs	Patents	Other	Total
ACCUMULATED COST				
Opening balance 1 Jan. 2018	51,941	28,405	2,864	83,210
Additions	-	3,233	-	3,233
Closing balance 30 Sep. 2018	51,941	31,638	2,864	86,443
ACCUMULATED DEPRECIATION				
Opening balance 1 Jan. 2018	-	-7,778	-1,117	-8,895
Depreciation for the period	-	-1,263	-100	-1,363
Closing balance 30 Sep. 2018	-	-9,041	-1,217	-10,258
Residual value 30 Sep. 2018	51,941	22,597	1,647	76,185

(SEK 000)	Development costs	Patents	Other	Total
ACCUMULATED COST				
Opening balance 1 Jan. 2017	51,255	24,349	2,899	78,503
Additions	686	4,056	-	4,742
Impaired value	-	-	-35	-35
Closing balance 31 Dec. 2017	51,941	28,405	2,864	83,210
ACCUMULATED DEPRECIATION				
Opening balance 1 Jan. 2017	-	-6,370	-982	-7,352
Depreciation for the period	-	-1,408	-135	-1,543
Closing balance 31 Dec. 2017	-	-7,778	-1,117	-8,895
Residual value 31 Dec. 2017	51,941	20,627	1,747	74,315

Note 2 – Tax

The group's total loss carry-forwards amount to SEK 428,981,000 as of 30 September 2018 (345,556,000). The parent company's total loss carry-forwards amount to SEK 402,798,000 as of 30 September 2018 (319,384,000). 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.



David Laskow-Pooley



David Beijer



Denise Goode

Board of Directors and CEO



Jan Törnell



Erik Kinnman

Board of Directors' declaration

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, 22 November 2018

David Laskow-Pooley
Chairman of the Board

David Beijer
Board member

Denise Goode
Board member

Jan Törnell
Board member

Erik Kinnman
Chief Executive Officer

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.

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

This information is information that NeuroVive Pharmaceutical AB (publ) is obliged to make public pursuant to the EU Market Abuse Regulation and the Securities Markets Act. The information was submitted for publication, through the agency of the contact person set out above, at 08:30 a.m. CET on 22 November 2018.

Auditor's review report

To the board of 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, 2018 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 10 under the financial position, the Group needs additional financing during the first half of 2019 to ensure continued operations. The Board of Directors has taken steps that these needs are addressed, and conducts active work with solutions to execute the company's communicated business plan.

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

Helsingborg, November 22nd, 2018
Mazars SET Revisionsbyrå AB

Bengt Ekenberg
Authorized Public Accountant



NeuroVive is a leading company in mitochondrial medicine focusing on indications with great medical needs.

About NeuroVive

NeuroVive Pharmaceutical AB is a leader in mitochondrial medicine, with one project in clinical phase II development for the prevention of moderate to severe traumatic brain injury (NeuroSTAT®) and one project in clinical phase I (KL1333) for genetic mitochondrial diseases. The R&D portfolio also consists of projects for genetic mitochondrial disorders, cancer and NASH. The company advances drugs for rare diseases through clinical development into the market. For projects for common indications the goal is out-licensing in the preclinical phase. A subset of compounds under NeuroVive's NVP015 program has been licenced to Fortify Therapeutics, a BridgeBio company, for local treatment development of Leber's Hereditary Optic Neuropathy (LHON).

What is mitochondrial medicine?

Mitochondrial medicine is an area spanning from cell protection in acute and chronic medical conditions to the regulation of energy production and cancer development. Mitochondria are found inside the cells and can be considered as the cells' power plants. They give us the amount of energy we need to move, grow and think.

NeuroVive's project portfolio includes a large number of cyclophilin inhibitors that serve as organ protection by increasing the mitochondrial resistance to various types of stress and by reducing the development of fibrosis. NeuroVive is also working with several molecules in the project portfolio, focused on the regulation of mitochondrial energy production, in both genetic mitochondrial disorders and prevalent metabolic diseases such as NASH. NeuroVive also has a liver cancer project in which the company is working with a completely new treatment strategy for these types of diseases.

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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Glossary

Active compound. A pharmaceutical active ingredient in a pharmaceutical product.

Alpers Disease. Mitochondrial disease. Also known as Alpers-Huttenlocher's disease. Usually appear in children under four years of age, first as difficult-to-treat epilepsy followed by brain injury, and usually also affecting the liver, the gastrointestinal tract and the peripheral nerves. The disease is progressive and results in increasing dementia, visual impairments and paralysis. There is no cure, but treatment efforts are focused on relieving the symptoms, preventing medical complications and providing support.

Blood-brain barrier. The blood-brain barrier consists of very closely joined capillary walls in the blood vessels of the brain that reduce the availability of certain bloodborne substances to access brain tissue (nerve cells).

Candidate drug. A particular compound which is selected during the preclinical phase. The candidate drug is subsequently tested in humans in clinical studies.

Cell proliferation. When cells grow, and divide, i.e the number of cells are increased keeping the size of the cell intact. This results in an expansion of the tissue and consequently an increase of the size of the organ/tumor.

CHIC. Copenhagen Head Injury Ciclosporin study, phase IIa study of NeuroSTAT.

CHOP. The Children's Hospital of Philadelphia.

Ciclosporin. A natural active compound (cyclical molecule) produced by the fungus *Tolypocladium inflatum*. Ciclosporin is now produced by artificial or chemical methods. Ciclosporin is a well-known substance that has been demonstrated to potentially protect brain in animal models of brain injury, where ciclosporin has transited the blood-brain barrier and entered the brain.

Clinical study. The examination of healthy or unhealthy humans to study the safety and efficacy of a pharmaceutical or treatment method. Clinical trials are divided into different phases, termed phase I, phase II, phase III. Phase II is usually divided into an early phase (phase IIa) and a later phase (phase IIb). See also "phase (I, II and III)".

COMP. EMA's Committee for Orphan Medicinal Products.

CRO. Contract research organization.

Cyclophilin D. The mitochondria target of ciclosporin and other cyclophilin inhibitors present in virtually all cells of the body.

EMA. The European Medicines Agency.

Energy metabolites. Digestion products from foodstuffs which reflects cell energy status and function of the mitochondria.

Experimental model. A model of a disease or other injury to resemble a similar condition or disease in humans.

FDA. The United States Federal Food and Drug Administration.

HCC. Hepatocellular carcinoma, liver cancer.

Indication. A disease condition requiring treatment, such as traumatic brain injury or fatty liver; NASH.

In vivo/in vitro. In vivo are scientific studies in animal models. In vitro are scientific studies carried out outside of the living body, for example in cells in test tubes.

KSS. Mitochondrial disease, Kearns-Sayre's syndrome. The disease debuts before the age of 20 and is characterized by eye related symptoms with pigment retention in the retina and paralysis of the outer eye muscles, as well as the effects on the cardiac retinal system and the cerebellum with disorders in the coordination of muscle movements (ataxia).

Leigh syndrome.

Leigh syndrome is a serious condition with characteristic changes to the brain that usually affects small children. This disease is caused by faults in energy-producing mitochondria and is also known as subacute (fast onset) necrotizing (tissue destroying) encephalomyopathy (a disease of the brain and muscles).

LHON. Mitochondrial disease, Leber Hereditary Optic Neuropathy. Affects the retina and the optic nerve, but in rare cases symptoms can be found in other parts of the central nervous system. There is no cure, but treatments are focused primarily on compensating for the visual impairment.

Liver fibrosis/cirrhosis. Liver fibrosis is the formation of fibrous tissue (scar tissue) in the liver as a result of, for example, infection. May lead to liver cirrhosis.

MELAS. MELAS is an acronym of mitochondrial encephalomyopathy (brain and muscle disease) with lactic acidosis (increased lactic acid levels in the blood) and stroke-like episodes.

MERRF. Mitochondrial disease. The most prominent symptoms of MERRF (Myoclonic epilepsy with ragged-red fibers) are epilepsy, muscle twitches and difficulty coordinating muscle movements, but the disease affects many functions.

Mitochondria. That part of each cell that provides effective energy production in the form of conversion of oxygen and nutrients in the body into chemical energy.

Mitochondrial medicine. Field of research and development of pharmaceuticals that protect the mitochondria.

Mitochondrial myopathy. Genetic mitochondrial disease which affects the muscles.

NAFLD. Non-Alcoholic Fatty Liver Disease.

NASH. Non-alcoholic steatohepatitis, inflammatory fatty liver disease.

NIH. The National Institutes of Health, the American equivalent of the Swedish Research Council.

ODD. Orphan Drug Designation. Facilitates development and commercialization, and may, upon receiving marketing authorization, provide orphan drug status with seven or ten years of market exclusivity (in the US and Europe, respectively).

Pearson syndrome. Mitochondrial disease. Appears early, in infants, with symptoms from several different tissues, mainly from the bone marrow, resulting in severe blood deficiency, as well as from the pancreas. Children with Pearson's syndrome who survive past adolescence later in life develop Kearns-Sayre's syndrome or other types of mitochondrial diseases.

Penn. University of Pennsylvania.

PEO/CPEO. Mitochondrial disease. Progressive External Ophthalmoplegia/Chronic Progressive External Ophthalmoplegia.

Pharmacokinetics. Describes how the body affects a specific drug after administration.

Phase (I, II and III). The various stages of trials on the efficacy of a pharmaceutical in humans. See also "clinical trial." Phase I examines the safety on healthy human subjects, phase II examines efficacy in patients with the relevant disease and phase III is a large-scale trial that verifies previously achieved results. In the development of new pharmaceuticals, different doses are trialed and safety is evaluated in patients with relevant disease, phase II is often divided between phase IIa and phase IIb.

Preclinical. That stage of drug development that occurs before a candidate drug is trialed on humans.

Protonophores. Substance which carries protons across the mitochondrial membrane leading to increased energy expenditure.

Sangamides. Compound class of cyclophilin-D inhibitors.

TBI. Traumatic Brain Injury. An injury to the brain where some nerve cells are subjected to immediate damage. The injury then continues to exacerbate several days after the incident, which significantly impacts the final extent of damage.

ToxPhos®. NeuroVive's registered trademark for the Company's mitochondrial toxicity test.