

## First subjects enrolled in the KL1333 study

## Important events January-March 2019

- NeuroVive enrolls first subject in its KL1333 phase Ia/b clinical study.
- NeuroVive is supplied with approximately MSEK 99.0 in share issue proceeds.
- NeuroVive receives SEK 28.2 Million in a directed new share issue.
- NeuroVive enters commercial partnership with Oroboros Instruments on mitochondrial medicine research compounds.

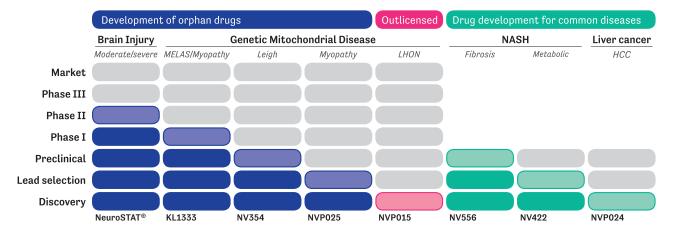
## Important events after the reporting period

- The Supreme Court had delivered its ruling concerning arbitration between NeuroVive and CicloMulsion AG. NeuroVive appealed to the Supreme Court on certain points. The Supreme Court has rejected the appeal.
- The US Food and Drug Administration, FDA, has approved NeuroVive's IND (Investigational New Drug) application, enabling clinical studies in the US with the company's drug candidate NeuroSTAT.

## Financial information January-March 2019

- Net revenues: KSEK 0 (0)
- Other operating income: KSEK 0 (174)
- Loss before tax: KSEK -13,822 (-13,053)
- Loss per share:\* SEK -0,12 (-0,25)
- Diluted loss per share:\*\* SEK -0,12 (-0,25)

## Project portfolio



- 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

2019 is a very important year for NeuroVive's three most significant projects — KL1333, NeuroSTAT and NV354. After the two successful share issues in early 2019, we are financially equipped to deliver on this year's targets for these projects. We made great steps forward in 2018, and in 2019 we will continue to build value in NeuroVive through progress in our projects and through continuing our intensive business development.



# KL1333: First test subjects enrolled for the clinical Phase Ia/b study

At the end of 2018, our application for a Phase Ia/b study with patients and healthy volunteers was approved by MHRA, the UK supervisory authority, and the initial visit by the first test subjects was screened in March this year. As a result of reaching this milestone, this highly important study was initiated according to plan and we have come a step closer to the ultimate goal of developing drugs for the serious mitochondrial disorders MELAS, PEO, KSS and Pearson syndrome. We expect to present the initial results of the study towards the end of 2019.

## NeuroSTAT: strengthened by clinical effect signal

In 2018, we obtained very positive results from analyses of biomarkers from patient samples in the CHIC trial. The results indicate that NeuroSTAT inhibits the secondary injury cascades resulting from brain trauma. In mid-March, we presented this exciting biomarker data, and our strategy of using biomarkers for effective clinical development in traumatic brain injury, at The 13th World Congress on Brain Injury in Toronto, Canada. Both the important biomarkers and the opportunity to use them in clinical studies have aroused great interest in NeuroSTAT, and our ambition now is to ensure external, undiluted partial financing for a Phase II efficacy study.

# NV354: alternative energy source for genetic mitochondrial diseases

NV354 is also an important project for us. The initial experimental results are very positive; we are continuing preclinical development and expect to present further experimental data this year with the goal of starting clinical trials in 2020.

# New share issues ensure continued clinical development

I would like to thank our current and new shareholders who took part in the preferential rights issue for your confidence and invaluable support. I would also like to welcome the Swedish and international investors who took part in the private placements that generated proceeds for NeuroVive of SEK 26 million after issue expenses in March. The largest of these new investors, Nyenburgh Investment Partners, is known for being a knowledgeable long-term owner.

NeuroVive has entered an exciting phase in its development, and the funds provided will ensure the completion of crucial value-creating activities in our projects in the clinical phase — and above all for NV354 in the preclinical phase — in 2019.

## Intense business development

NeuroVive is conducting intense business development, and we are open for establishing partnerships in all our current projects. At present, we are actively seeking interesting partners who can contribute capital and know-how to the projects that have come furthest in their development: KL1333, NeuroSTAT and NV354.

In parallel, we are looking for external partners for NV556, which is being developed for the treatment of liver fibrosis in connection with NASH. We are conducting interesting dialogues with several potential partners, however an agreement is not likely to be reached by the end of the first half of this year.

#### Erik Kinnman

CEO, NeuroVive Pharmaceutical AB May 21, 2019



# KL1333 - for treatment of genetic mitochondrial diseases

The project has just started a clinical phase Ia/b study in the UK

Initial results are expected towards the end of 2019.

Discovery

Lead selection

Preclinical

Phase I

Phase II

Phase III

Market

## About genetic mitochondrial diseases

Genetic mitochondrial diseases are metabolic diseases that affect the ability of cells to convert energy. The disorders can manifest differently depending on the organs affected by the genetic defects and are viewed as clinical syndromes. An estimated 12 in every 100,000 people suffer from a genetic mitochondrial disease. Genetic 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.

## Activities in the first quarter

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

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

## Objectives for 2019

- Start clinical phase Ia/b study in Europe during first half of 2019.
- Present initial results from the clinical phase Ia/b study.
- Prepare for phase II efficacy studies.



# NeuroSTAT - for treatment of traumatic brain injury

The project is in preparation for clinical phase II efficacy study

NeuroVive is seeking external, non-dilutive funding

Discovery

Lead selection

Preclinical

Phase I

Phase II

Phase III

Market

## **About 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. 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. A large number of patients suffer moderate to severe functional disabilities requiring intensive care and various forms of lifelong support.

## Activities in the first quarter

NeuroVive participated in *The 13th World Congress* on *Brain Injury* in Toronto, Canada, 13 – 16 March. NeuroVive's team presented the exciting NeuroSTAT biomarker data from the CHIC study, indicating drug effect, and the company's approach to utilize biomarkers for effective clinical development in Traumatic Brain Injury.

#### Important events after the reporting period

The US Food and Drug Administration, FDA, has approved NeuroVive's IND (Investigational New Drug) application, enabling clinical studies in the US with the company's drug candidate NeuroSTAT.

## Objectives for 2019

- Secure external non-dilutive financing for upcoming Phase II efficacy study.
- Receive approval of IND application for clinical development in the US.
- Preparation for a clinical phase II efficacy study. This study requires external financing.

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

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



# NV354 - alternative energy source in genetic mitochondrial disease

The project is in preparation for clinical phase

Further experimental data will be presented during 2019

Discovery

Lead selection

Preclinical

Phase I

Phase II

Phase III

Market

## **Energy conversion in mitochondrial diseases**

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.

## Activities in the first quarter

The NV354 project has progressed according to plan in the quarter with continued preparations for the important toxicology studies. Likewise, the scaling up of compound production has continued according to the original plan.

## **Objectives for 2019**

- Present further results from preclinical in vivo dose-response studies.
- Scale up compound production.
- Initiate toxicology studies.
- Run experimental studies in cooperation with CHOP, financed by the US Department of Defense grants.

## Other projects

#### NV556 - FOR TREATMENT OF NASH

## 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 Neuro-Vive's Sangamide class of compounds. The anti-fibrotic effect can also be developed for other diseases involving liver fibrosis, such as Primary Biliary Cholangitis (PBC) and Primary Sclerosing Cholangitis (PSC).

## Project status: pre-clinical development

Preclinical results have shown 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 goal is to reach an agreement with a suitable partner for this niched NASH product.

## **NVP025 - FOR TREATMENT OF MITOCHONDRIAL MYOPATHIES**

#### 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.

#### NV422 - FOR TREATMENT OF NASH

## **Treatment objective**

NV422 targets the metabolic components of the fatty liver disease 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 candidate compound

NeuroVive has evaluated various substances within the project in 2018 and has selected a candidate substance, NV422, based on favorable pharmacokinetic profile and good tolerability. This compound is now going through confirmatory experimental studies.

## **NVP024 – FOR TREATMENT OF LIVER CANCER**

## **Treatment objective**

NVP024 is focused on the anticancer properties of a subset of the company's Sangamide compounds. Together with international partners, NeuroVive's research team has demonstrated that these compounds show powerful anticancer effects in preclinical models of hepatocellular carcinoma (liver cancer).

## 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 Foun-dation for Strategic Research.

## **Out-licensed projects**

## 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 local 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.

## **Financial information**

#### Revenues

The consolidated turnover during the first quarter of 2019 was KSEK 0 (0). Other operating revenues for the first quarter were KSEK 0 (174).

## Results of operations

The operating loss for the first quarter was KSEK 13,809 (12,993). The net loss before tax for the first quarter amounted to KSEK 13,822 (13,053).

The operating loss was affected by other external expenses, which for the full were KSEK 9,630 (9,222). During the first quarter, expenses related to development projects, as a part of external expenses, have affected the result with KSEK 6,128 (3,987) whereof KSEK 2,415 (1,745) relates to project in clinical phase. Personnel expenses during the first quarter amount to KSEK 3,480 (3,433). Other operating expenses amount to, KSEK 133 (50) and pertains to exchange-rate losses.

## Financial position

The equity/assets ratio was 93 (86) percent as of 31 March 2019, and equity was KSEK 191,199 (92,797) compared to beginning of the year. The equity includes funds from the in February completed rights issue, which provided the company with KSEK 81,998 after deduction of issue costs and compensation for guarantee commitments of KSEK 17,035 and funds from the in March completed directed issue with KSEK 26,009 after deduction of issue costs of KSEK 2,203. Cash and cash equivalents amounted to KSEK 113,339 (15,757) as of 31 March 2019, an increase of KSEK 87,388 from the beginning of the year. Total assets as of 31 March 2019 were KSEK 204,646 (108,242).

## Cash flow and investments

Operating cash flow for the first quarter was KSEK -13,255 (-12,596). The cash flow effect related to investments in intangibles equals KSEK -260 (-624) for the first quarter. Cash flow for the first quarter equals KSEK 87,385 (-13,240).

## 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.

During the the period no purchases or sales between the group and related parties occurred. No compensation has been paid during the period 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.

## 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 Neuro-Vive makes the assessment that there are no circumstances 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 March 2019 was 8 (9), of which 4 (4) are women.

#### Parental company

Company earnings after tax for the first quarter amounts to KSEK 13,814 (-13,045). 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 Company has performed a Preferential Rights Issue and a Directed Rights Issue. The result from the rights issues brings the company approximately MSEK 108, after transaction costs, which is expected to finance the company's activities in the coming year.

The Supreme Court delivered its ruling concerning arbitration between NeuroVive and CicloMulsion AG on April 30. For further information, see below.

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 Scania and Blekinge Court of Appeal with the exception of the question for which the Tribunal had reserved its decision. NeuroVive appealed parts of the ruling to the Supreme Court. On April 30th, 2019, the Supreme Court announced that the appeal had been rejected. This means that the partial award is ultimately and completely set aside.

The former arbitral tribunal was replaced by a Newly Composed Arbitral Tribunal following a request for the release of the arbitrators filed by CicloMulsion. The constitution of the Newly Composed Arbitral Tribunal has been finalized. After the decision of the Supreme Court, the Newly Composed Arbitral Tribunal will proceed with the arbitration proceedings with the aim of announcing an award in 2020 and the parties will submit respective briefs in course of this year.

Through the ruling from the Supreme Court, NeuroVive has also been ordered to compensate CicloMulsion's court costs of SEK 531,899 and EUR 20,187 for the Supreme Court.

The ongoing dispute with CicloMulsion AG may result in further 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, if any, at this time cannot be calculated.

For further Information see Prospectus 2019 and Annual Report 2018.

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 2018 and the prospectus published January 22, 2019 for the preferential rights issue carried out in February 2019.

## Incentive programs/share warrants

Currently there is no incentive program.

#### **Audit review**

This Interim Report has not been subject to review by the company's auditors.

## Upcoming financial statements

Interim Report Jan-Jun 2019 Interim Report Jan-Sep 2019 Year-End Report 2019 August 21, 2019 November 20, 2019 February 19, 2020

The interim reports and the Annual Year Report are available at www.neurovive.com

## 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 accounting principles described in the Annual Report for 2018 on pages 56-69.

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. NeuroVive 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, are reported in the balance sheet. NeuroVive has lease contracts for office premises that will be reported in the balance sheet as of January 1, 2019. IFRS 16 will be applied retroactively without recalculation of comparative figures, so called simplified approach. For further information see page 56 in the Annual Report 2018.

# Consolidated Statement of Comprehensive Income

|  |      | 1 Jan, 2019  | 1 Jan, 2018  | 1 Jan, 2018  |
|--|------|--------------|--------------|--------------|
| (SEK 000)  | Note | 31 Mar, 2019 | 31 Mar, 2018 | 31 Dec, 2018 |
| Net sales  |      | -            | -            | 5            |
| Other operating income   |      | -            | 174          | 2,461        |
|  |      | _            | 174          | 2,466        |
| Operating expenses   |      |              |              |              |
| Other external expenses  |      | -9,630       | -9,222       | -55,812      |
| Personnel cost   |      | -3,480       | -3,433       | -14,454      |
| Depreciation and write-down of tangible and intangible assets                |      | -567         | -462         | -4,771       |
| Other operating expenses   |      | -133         | -50          | -789         |
|  |      | -13,809      | -13,167      | -75,826      |
|  |      |              |              |              |
| Operating income   |      | -13,809      | -12,993      | -73,360      |
| Profit/loss from financial items   |      |              |              |              |
| Result from shares in associated company                                     |      | -            | -            | 66           |
| Financial income   |      | -            | 9            | 407          |
| Financial costs  |      | -13          | -70          | -607         |
|  |      | -13          | -61          | -134         |
| Profit/loss before tax   |      | -13,822      | -13,053      | -73,494      |
| Income tax   | 2    | _            | _            | -            |
| Profit/loss for the period   |      | -13,822      | -13,053      | -73,494      |
| Other comprehensive income   |      |              |              |              |
| Items that may be reclassified to profit or loss                             |      |              |              |              |
| Translation differences on foreign subsidiaries                              |      | 2            | 4            | 4            |
| Total comprehensive income for the period                                    |      | -13,821      | -13,049      | -73,490      |
| Loss for the period attributable to:   |      |              |              |              |
| Parent company shareholders  |      | -13,822      | -13,052      | -68,373      |
| Non-controlling interests  |      | _            | -1           | -5,121       |
|  |      | -13,822      | -13,053      | -73,494      |
| Total comprehensive income for the period                                    |      |              |              |              |
| Parent company shareholders  |      | -13,820      | -13,278      | -68,370      |
| Non-controlling interests  |      | -            | 229          | -5,120       |
|  |      | -13,820      | -13,049      | -73,490      |
| Earnings per share before and after dilution(SEK) based on average number of |      |              |              |              |
| shares   |      | -0.12        | -0.25        | -0.87        |

# Consolidated Statement of Financial Position

| (SEK 000)   | Note | 31 Mar, 2019 | 31 Mar, 2018 | 31 Dec, 2018 |
|---|------|--------------|--------------|--------------|
| ASSETS  |      |              |              |              |
| Non-current assets  |      |              |              |              |
| Intangible assets   | 1    |              |              |              |
| Development costs   |      | 51,706       | 51,941       | 51,706       |
| Patents   |      | 20,057       | 21,459       | 20,121       |
| Other Intangible assets                                       |      | 1,579        | 1,714        | 1,613        |
|   |      | 73,343       | 75,114       | 73,440       |
| Tangible assets   |      |              |              |              |
| Equipment   |      | 113          | 157          | 140          |
| Rigth of use asset leases                                     |      | 945          | -            | -            |
|   |      | 1,057        | 157          | 140          |
| Financial assets  |      |              |              |              |
| Other long-term securities                                    |      | 13,101       | 13,102       | 13,101       |
|   |      | 13,101       | 13,102       | 13,101       |
|   |      |              |              | •            |
| Total non-current assets                                      |      | 87,501       | 88,373       | 86,681       |
|   |      |              |              |              |
| Current assets  |      | 7.000        | 1 710        | 4 470        |
| Other receivables   |      | 3,080        | 1,312        | 1,432        |
| Prepaid expenses and accrued income                           |      | 726          | 2,800        | 1,244        |
| Cash and cash equivalents                                     |      | 113,339      | 15,757       | 25,951       |
|   |      | 117,145      | 19,869       | 28,627       |
| TOTAL ASSETS  |      | 204,646      | 108,242      | 115,308      |
|   |      |              |              |              |
| EQUITY AND LIABILITIES  |      |              |              |              |
| Equity attributable to the shareholders of the parent company |      |              |              |              |
| Share capital   |      | 9,298        | 2,616        | 4,585        |
| Additional paid in capital                                    |      | 593,207      | 427,226      | 489,913      |
| Translation reserve   |      | 618          | 386          | 616          |
| Retained earnings   |      | -411,935     | -342,791     | -398,113     |
| Total equity attributable to the shareholders of the parent   |      | 191,188      | 87,437       | 97,001       |
| Non-controlling interests                                     |      | 11           | 5,360        | 11           |
| Total equity  |      | 191,199      | 92,797       | 97,012       |
|   |      |              | ,            |              |
| Long-term liabilities   |      |              |              |              |
| Other longtrem liabilities                                    |      | 687          | -            | -            |
|   |      | 687          | -            | -            |
| Short-term liabilities  |      |              |              |              |
| Accounts payable  |      | 5,120        | 5,350        | 10,162       |
| Other liabilities   |      | 703          | 5,041        | 808          |
| Accrued expenses and deferred income                          |      | 6,937        | 5,041        | 7,326        |
| Acci ded expenses and deferred income                         |      | 12,760       | 15,445       | 18,296       |
|   |      | 12,700       | 10,770       | 10,200       |
| Total liabilities   |      | 13,447       | 15,445       | 18,296       |
| TOTAL FOURTY AND LIABILITIES                                  |      | 004.040      | 400.040      | 445 700      |
| TOTAL EQUITY AND LIABILITIES                                  |      | 204,646      | 108,242      | 115,308      |

# Consolidated Statement of Changes in Equity

|  | Equity attrib |            | e shareholder | s of the pare | nt company |             |         |
|--|---------------|------------|---------------|---------------|------------|-------------|---------|
|  |               | Additional |               |               |            | Non-        |         |
|  | Share-        | paid in    | Translation   | Retained      |            | controlling | Total   |
| (SEK 000)                                | capital       | capital    | reserve       | earnings      | Total      | interests   | equity  |
| Opening balance, 1 January 2019          | 4,585         | 489,913    | 616           | -398,113      | 97,002     | 11          | 97,012  |
| Comprehensive profit/loss for the period |               |            |               |               |            |             |         |
| Profit/loss for the period               | _             | -          | -             | -13,822       | -13,822    | _           | -13,822 |
| Other comprehensive income               |               |            |               |               |            |             |         |
| Translation differences                  | -             | -          | 2             | -             | 2          | -           | 2       |
| Other comprehensive profit/loss for the  |               |            |               |               |            |             |         |
| period, net after tax                    | -             | -          | 2             | -             | 2          | -           | 2       |
| Total comprehensive profit/loss          | -             | -          | 2             | -13,822       | -13,820    | -           | -13,820 |
| Transactions with shareholders           | -             | -          | -             | -             | -          | -           | -       |
| Rights Issue*                            | 4,713         | 103,294    | -             | -             | 108,007    | -           | 108,007 |
| Total transactions with shareholders     | 4,713         | 103,294    | -             | -             | 108,007    | -           | 108,007 |
| Closing balance, 31 March 2019           | 9,298         | 593,207    | 618           | -411,935      | 191,188    | 11          | 191,199 |
|  |               |            |               |               |            |             |         |
| 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               | -             | -          |               | -13,052       | -13,052    | -1          | -13,053 |
| Other comprehensive income               |               |            |               |               |            |             |         |
| Translation differences                  | -             | -          | -227          | -             | -227       | 230         | 4       |
| Other comprehensive profit/loss for the  |               |            |               |               |            |             |         |
| period, net after tax                    | -             | -          | -             | -             | -          | -           | -       |
| Total comprehensive profit/loss          | -             | -          | -227          | -13,052       | -13,278    | 229         | -13,049 |
| Transactions with shareholders           |               |            |               |               |            |             |         |
| Total transactions with shareholders     | -             | -          | -             | -             | -          | -           | -       |
| Closing balance, 31 March 2018           | 2,616         | 427,226    | 386           | -342,792      | 87,437     | 5,360       | 92,797  |
|  |               |            |               |               |            |             |         |
| 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               | -             | -          |               | -68,373       | -68,373    | -5,121      | -73,494 |
| Other comprehensive income               |               |            |               |               |            |             |         |
| Translation differences                  | _             | -          | 3             | -             | 3          | 11          | 4       |
| Other comprehensive profit/loss for the  |               |            |               |               |            |             |         |
| period, net after tax                    |               | -          | 3             | -             | 3          | 1           | 4       |
| Total comprehensive profit/loss          |               | -          | 3             | -68,373       | -68,370    | -5,120      | -73,490 |
| Transactions with shareholders           |               |            |               |               |            |             |         |
| Rights Issue                             | 1,969         | 62,687     | -             | -             | 64,656     | -           | 64,656  |
| Total transactions with shareholders     | 1,969         | 62,687     | -             | -             | 64,656     | -           | 64,656  |
| Closing balance, 31 December 2018        | 4,585         | 489,913    | 616           | -398,113      | 97,002     | 11          | 97,012  |

<sup>\*</sup>Total equity includes funds from the in February 11, 2019 completed rights issue with KSEK 99,033 less expenses and guranties KSEK 17,035 and the directed rights issue completed in March 7, 2019 with KSEK 28,212 less expenses KSEK 2,202.

# **Consolidated Statement of Cash Flows**

| (SEK 000)  | 1 Jan, 2019  | 1 Jan, 2018  | 1 Jan, 2018  |
|--|--------------|--------------|--------------|
| Cash flow from operating activities                                  | 31 Mar, 2019 | 31 Mar, 2018 | 31 Dec, 2018 |
| Operating income   | -13,809      | -12,993      | -73,360      |
| operating meaning  | 20,000       | 12,000       | . 0,000      |
| Adjustments for non-cash items:                                      |              |              |              |
| Depreciation   | 567          | 462          | 1,914        |
| Impaired Value   | -            | -            | 3,324        |
| Result from shares in associated company                             | -            | _            | 66           |
| Interest received  | -            | 9            | 407          |
| Interest paid  | -13          | -70          | -606         |
| Net cash from operating activities before changes in working capital | -13,255      | -12,592      | -68,255      |
| Changes in working capital   |              |              |              |
| Increase/decrease of other current assets                            | -1,129       | -577         | 859          |
| Increase/decrease of other short-term liabilities                    | -5,978       | 572          | 3,567        |
| Changes in working capital   | -7,107       | -5           | 4,426        |
| Cash flow from operating activities                                  | -20,362      | -12,596      | -63,829      |
| Investing activities   |              |              |              |
| Acquisition of intangible assets                                     | -260         | -624         | -3,791       |
| Acquisition of tangible assets                                       | -            | -19          | -82          |
| Increase in other financial assets                                   | -            | -            | 1            |
| Cash flow from investing activities                                  | -260         | -643         | -3,872       |
| Financing activities   |              |              |              |
| New share issue  | 108,007      | -            | 64,656       |
| Cash flow from financing activities                                  | 108,007      | -            | 64,656       |
| Cash flow for the period   | 87,385       | -13,240      | -3,046       |
| Cash and cash equivalents at the beginning of the period             | 25,951       | 28,992       | 28,992       |
| Effect of exchange rate changes on cash                              | 3            | 5            | 5            |
| Cash and cash equivalents at end of period                           | 113,339      | 15,757       | 25,951       |

# Parent Company Income Statement

| (SEK 000)   | Note | 1 Jan, 2019<br>31 Mar, 2019 | 1 Jan, 2018<br>31 Mar, 2018 | 1 Jan, 2018<br>31 Dec, 2018 |
|---|------|-----------------------------|-----------------------------|-----------------------------|
| Net sales   |      | -                           | -                           | 5                           |
| Other operating income  |      | -                           | 174                         | 2,461                       |
|   |      |                             | 174                         | 2,466                       |
| Operating expenses  |      |                             |                             |                             |
| Other external expenses                                       |      | -9,721                      | -9,217                      | -55,777                     |
| Personnel cost  |      | -3,480                      | -3,433                      | -14,454                     |
| Depreciation and write-down of tangible and intangible assets |      | -481                        | -462                        | -4,536                      |
| Other operating expenses                                      |      | -133                        | -50                         | -789                        |
|   |      | -13,814                     | -13,162                     | -75,556                     |
| Operating income  |      | -13,814                     | -12,987                     | -73,090                     |
| Profit/loss from financial items                              |      |                             |                             |                             |
| Result from shares in associated company                      |      | -                           | -                           | 66                          |
| Interest income and other similar profit items                |      | -                           | 9                           | 400                         |
| Interest expenses and other similar loss items                |      | -                           | -66                         | -602                        |
|   |      | -                           | -57                         | -136                        |
| Profit/loss before tax  |      | -13,814                     | -13,045                     | -73,226                     |
| Income tax  | 2    | -                           | -                           | -                           |
| Profit/loss for the period                                    |      | -13,814                     | -13,045                     | -73,226                     |

# Statement of Comprehensive Income, Parent Company

| (SEK 000)                                      |      | 1 Jan, 2019  | 1 Jan, 2018  | 1 Jan, 2018  |
|--|------|--------------|--------------|--------------|
|  | Note | 31 Mar, 2019 | 31 Mar, 2018 | 31 Dec, 2018 |
|  |      |              |              |              |
| Profit/loss for the period                     |      | -13,814      | -13,045      | -73,226      |
| Other comprehensive income                     |      | -            | -            | -            |
| Total comprehensive profit/loss for the period |      | -13,814      | -13,045      | -73,226      |

# Parent Company Balance Sheet

| (SEK 000)                            | Note | 31 Mar, 2019 | 31 Mar, 2018 | 31 Dec, 2018 |
|--------------------------------------|------|--------------|--------------|--------------|
| ASSETS                               |      |              |              | ·            |
| Non-current assets                   |      |              |              |              |
| Intangible assets                    | 1    |              |              |              |
| Development costs                    |      | 51,706       | 51,706       | 51,706       |
| Patents                              |      | 20,057       | 21,459       | 20,121       |
| Other intangible assets              |      | 1,579        | 1,714        | 1,613        |
|                                      |      | 73,343       | 74,879       | 73,440       |
| Tangible assets                      |      |              |              |              |
| Equipment                            |      | 113          | 157          | 140          |
|                                      |      | 113          | 157          | 140          |
| Financial assets                     |      |              |              |              |
| Other long-term placement            |      | 13,101       | 13,102       | 13,100       |
| Shares in subsidiaries               | 3    | 23,625       | 23,625       | 23,625       |
|                                      |      | 36,726       | 36,727       | 36,725       |
| Total non-current assets             |      | 110,182      | 111,763      | 110,305      |
| Current assets                       |      |              |              |              |
| Short term receivables               |      |              |              |              |
| Other receivables                    |      | 3,077        | 1,309        | 1,430        |
| Prepaid expenses and accrued income  |      | 726          | 2,800        | 1,244        |
|                                      |      | 3,803        | 4,109        | 2,674        |
| Cash and bank balances               |      | 113,277      | 15,651       | 25,871       |
| Total current assets                 |      | 117,081      | 19,760       | 28,545       |
| TOTAL ASSETS                         |      | 227,263      | 131,522      | 138,849      |
| EQUITY AND LIABILITIES               |      |              |              |              |
| Equity                               |      |              |              |              |
| Restricted equity                    |      |              |              |              |
| Share capital                        |      | 9,298        | 2,616        | 4,585        |
| Statutory reserve                    |      | 1,856        | 1,856        | 1,856        |
| Development expenditure reserve      |      | 10,610       | 10,610       | 10,610       |
|                                      |      | 21,764       | 15,082       | 17,051       |
| Unrestricted equity                  |      | -            |              |              |
| Share premium reserve                |      | 103,294      | 8,887        | 62,687       |
| Retained earnings                    |      | 103,523      | 105,173      | 114,061      |
| Profit/loss for the period           |      | -13,814      | -13,045      | -73,226      |
|                                      |      | 193,003      | 101,016      | 103,522      |
| Total equity                         |      | 214,767      | 116,098      | 120,573      |
| Short-term liabilities               |      |              |              |              |
| Accounts payable                     |      | 5,120        | 5,350        | 10,162       |
| Other liabilities                    |      | 439          | 5,041        | 808          |
| Accrued expenses and deferred income |      | 6,937        | 5,033        | 7,307        |
| F                                    |      | 12,496       | 15,424       | 18,277       |
| TOTAL EQUITY AND LIABILITIES         |      | 227,263      | 131,522      | 138,850      |
| 10 ME EQUIT I AND ELADIEITED         |      | 221,200      | 101,022      | 100,000      |

## **Notes**

## Note 1-Intangible assets

| (SEK 000)                    | Development costs | Patents | Other  | Total   |
|------------------------------|-------------------|---------|--------|---------|
| ACCUMULATED COST             |                   |         |        |         |
| Opening balance 1 Jan. 2019  | 51,706            | 29,107  | 2,864  | 83,677  |
| Additions                    | -                 | 357     | -      | 357     |
| Closing balance 31 Mar. 2019 | 51,706            | 29,464  | 2,864  | 84,034  |
| ACCUMULATED DEPRECIATION     |                   |         |        |         |
| Opening balance 1 Jan. 2019  | -                 | -8,986  | -1,251 | -10,237 |
| Depreciation for the period  | -                 | -421    | -34    | -455    |
| Closing balance 31 Mar. 2019 | -                 | -9,407  | -1,285 | -10,692 |
| Residual value 31 Mar. 2019  | 51,706            | 20,057  | 1,579  | 73,343  |

| (SEK 000)                    | Development costs | Patents | Other  | Total   |
|------------------------------|-------------------|---------|--------|---------|
| ACCUMULATED COST             |                   |         |        |         |
| Opening balance 1 Jan. 2018  | 51,941            | 28,405  | 2,864  | 83,210  |
| Additions                    | -                 | 3,791   | -      | 3,791   |
| Impaired value               | -235              | -3,089  | -      | -3,324  |
| Closing balance 31 Dec. 2018 | 51,706            | 29,107  | 2,864  | 83,677  |
| ACCUMULATED DEPRECIATION     |                   |         |        |         |
| Opening balance 1 Jan. 2018  | -                 | -7,778  | -1,117 | -8,895  |
| Depreciation for the period  | -                 | -1,675  | -134   | -1,809  |
| Closing balance 31 Dec. 2018 | -                 | -8,986  | -1,251 | -10,704 |
| Residual value 31 Dec. 2018  | 51,706            | 20,121  | 1,613  | 73,440  |

## Note 2 - Tax

The group's total loss carry-forwards amounts to KSEK 481,524 as of 31 March 2019 (374,195). The parent company's total loss carry-forwards amounts to SEK 455,749 as of 31 March 2019 (340,008). 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.

## Interim report January-March 2019



## The declaration of the Board of Directors and the CEO

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, May 21, 2019

David Laskow-PooleyDavid BejkerDenise GoodeChairman of the BoardBoard memberBoard member

Magnus PerssonJan TörnellErik KinnmanBoard memberBoard memberChief 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

The information was submitted for publication, through the agency of the contact person set out above, at 08:30 a.m. CET on May 21, 2019.

## Glossary

 $\begin{tabular}{ll} \bf Active\ compound.\ A\ pharmaceutical\ active\ ingredient\ in\ a\ pharmaceutical\ product. \end{tabular}$ 

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 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 potently 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.

 $\label{lem:cyclophilin D.} \textbf{ The mitochondria target of ciclosporin and other cyclophilin inhibtors present in virtually all cells of the body.}$ 

**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 strokelike 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

 $\mbox{\bf NIH}.$  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.



## **About NeuroVive**

NeuroVive Pharmaceutical AB is a leader in mitochondrial medicine, with one project in clinical phase I (KL1333) for genetic mitochondrial diseases and one project in preparation for a clinical phase II efficacy study for the prevention of moderate to severe traumatic brain injury (NeuroSTAT®). The R&D portfolio also consists of projects for genetic mitochondrial disorders, NASH and cancer.

The company advances drugs for rare diseases through clinical development into the market with our without partners. For projects for common indications the goal is out-licensing in the preclinical phase. A subset of compounds under NeuroVive's NVP015 program was in 2018 licenced to Fortify Therapeutics, a Bridge-Bio company, for local treatment of Leber's Hereditary Optic Neuropathy (LHON). NeuroVive is listed on Nasdaq Stockholm, Sweden (ticker: NVP). The share is also traded on the OTCQX Best Market in the US (OTC: NEVPF).

## 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 cell proliferation. 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, involving cyclophilin inhibition, for this disease.

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

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