

NeuroVive out-licenses targeted LHON therapy to BridgeBio

Important events January-June 2018

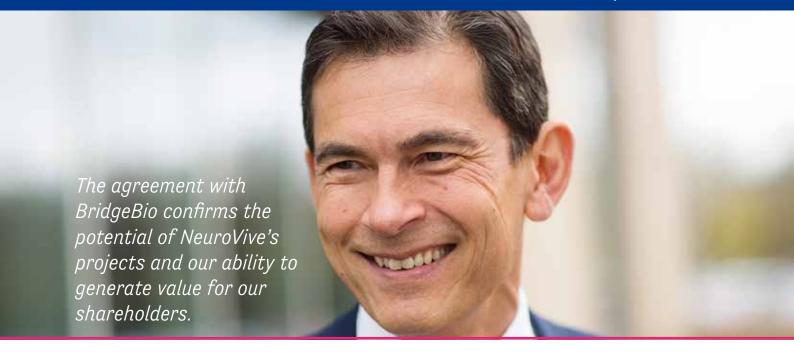
- NeuroVive out-licenses targeted LHON therapy to BridgeBio Pharma's new subsidiary Fortify Therapeutics. The agreement has a total potential value of approximately \$60 million USD including any royalties.
- NeuroVive conducts a preferential rights issue.
- NeuroVive and Yungjin reports positive KL1333 phase I clinical study results paving the way for further clinical development.
- KL1333 receives FDA Orphan Drug Designation for treatment of mitochondrial diseases.
- NeuroVive initiates collaboration with leading US TBI research organization TRACK-TBI, a network of world-leading TBI clinicians and researchers.
- Annual General Meeting in Lund, April 27.

Financials second quarter (April-June 2018)

- Net revenues: SEK 0 (0)
- Other operating income: SEK 1,278,000 (88,000)
- Loss before tax: SEK -25,481,000 (-22,256,000)
- Loss per share:* SEK -0,40 (-0,45)
- Diluted loss per share:** SEK -0,40 (-0,45)

Financials first six months (January-June 2018)

- Net revenues: SEK 0 (27,000)
- Other operating income: SEK 1,452,000 (152,000)
- Loss before tax: SEK -38,534,000 (-43,646,000)
- Loss per share:* SEK -0,61 (-0,79)
- Diluted loss per share:** SEK -0,61 (-0,79)
- * 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 year's second quarter was marked by a number of positive events, which bode well for the future. The first clinical trial with KL1333 showed very positive results. In mid-June, we completed an out-licensing transaction with the US firm BridgeBio Pharma with a potential total value for us of USD 60 million, including any royalties. Together with the successful rights issue we conducted in April, this means we can now continue the development of our promising pharmaceuticals projects with full force!

KL1333 - successful clinical trial

There have been many positive events within the KL1333 project during the quarter. In April, we reported that KL1333 has been granted orphan drug designation by the FDA, which offers the program access to regulatory and scientific advice from the FDA. The orphan drug designation will also allow the pursuit of focused development with a swift approval process. In addition, an orphan drug designation offers an opportunity for market exclusivity for seven years in the US in conjunction with market approval. The orphan drug designation is also a validation of the quality of the KL1333 documentation and is an important project milestone. KL1333 already has orphan drug designation in the EU. In May, Yungjin, our partner in the KL1333 project, concluded the first clinical safety trial with healthy volunteers with highly positive results. No serious side effects occurred, only mild gastrointestinal symptoms at high doses. The trial also demonstrated good pharmacokinetic results, how the drug is distributed in the body. The results are very promising and we are now preparing a dose escalation trial in Europe using both healthy volunteers and patients. This study is expected to start during the second half of this year.

NVP015 - agreement confirms potential

The agreement with BridgeBio, which entails a very positive expansion of the NVP015 program, pertains to the development of drugs for the treatment of the LHON mitochondrial eye disorder. The agreement validates the scientific quality of the NVP015 program while confirming the practical functionality of

NeuroVive's business development activities. Longterm potential revenue amounts to USD 60 million. Bridgebio represents a highly reputed partner with extensive experience in similar deals and drug development, and I am convinced that other areas of the NVP015 project and our other research programs will now be viewed in a new light. Opportunities to license other parts of the NVP015 project have been boosted and we have begun discussions with a number of interested parties regarding the parts of NVP015 that target the treatment of other mitochondrial disorders.

It is important to note that our own development ambitions for the NVP015 program remain unchanged. The next stage of the pre-clinical work is ongoing experimental proof-of-principle studies with the lead candidate NV354, the results of which are expected in the second half of 2018.

Activity is also high in our other projects

With respect to NV556, out-licensing discussions and activities continues with the aim of selecting a partner, who strategically fits our objectives, by the end of 2018/first half of 2019. Our ambition for NeuroSTAT is to seek non-dilutive funding, alternatively a partner for co-financing, by the end of the year and thereafter to initiate clinical efficacy trials during 2019.

Erik Kinnman

CEO, NeuroVive Pharmaceutical AB 21 August 2018

NeuroVive's research and development



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

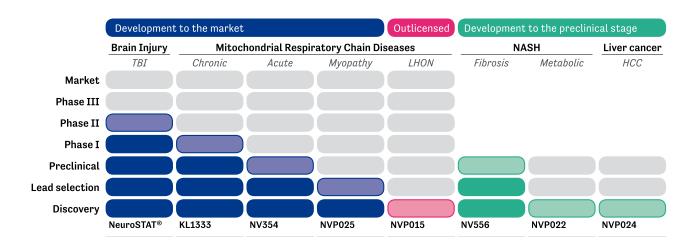
NeuroVive's aim is to help patients who currently have few or no treatment options. The company's business model consists of two parts: the first one involves the development of drugs for rare diseases, and the second one includes the development of mitochondrial drugs for commonly occurring diseases.

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

Outlicensing in pre-clinical phase. For commonly occurring diseases with high commercial potential, where the clinical studies are very extensive and costly, NeuoVive's objective is to outlicense 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 outlicensing and partnerships.



NeuroVive's program for traumatic brain injury (TBI)



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

Objectives for 2018

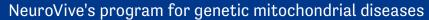
- Publication of results from the CHIC study and from the collaboration with Penn.
- Results from evaluation of biomarkers prior to the upcoming NeuroSTAT development program.
- Secure financing for upcoming Phase II efficacy study.
- Meeting with the FDA prior to development programs in the US.

Second quarter news

 NeuroVive commenced a collaboration with TRACK-TBI, a network of world-class traumatic brain injury (TBI) researchers.

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

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





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, droopy eyelids, 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.

Objectives for 2018

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

Second quarter news

- NeuroVive and Yungjin reported positive KL1333 phase I clinical study results, paving the way for further clinical development.
- The FDA granted orphan drug designation for KL1333 in the US for the treatment of mitochondrial diseases.

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 expects to select an optimized lead candidate compound in 2018.

Objectives for 2018

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

NVP015 - candidate drug in pre-clinical development

Treatment objective

NVP015 is focused on enabling a systemic therapy to prevent acute energy crises in patients with mitochondrial genetic disorders. One of the leading causes of mitochondrial genetic disorders is loss of function in the first of the five protein complexes in the mitochondria, complex I, which is essential for efficient energy conversion. This has been observed in Leigh syndrome, MELAS and Alpers syndrome, all very serious diseases with symptoms including muscle weakness, epilepsy and other severe neurological effects.

Project status: pre-clinical development

A compound, NV354, has been selected for continued development on the basis of tolerability, plasma stability and organ delivery, to the brain and other organs, and is currently undergoing additional experimental in vivo efficacy studies.

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.

Second quarter news

 Out-licensing of a subset of succinate prodrug under NeuroVive's NVP015 program to BridgeBio / Fortify, see below.

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'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.



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 antifibrotic 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

NV556 has undergone extensive preclinical development, has favorable drug-like properties and confirmed antifibrotic effect in several experimental models. The company has accelerated and continued its NV556 out-licensing activities with the aim of reaching a partnership agreement.

Objectives for 2018

• Out-licensing and/or partnering.

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.

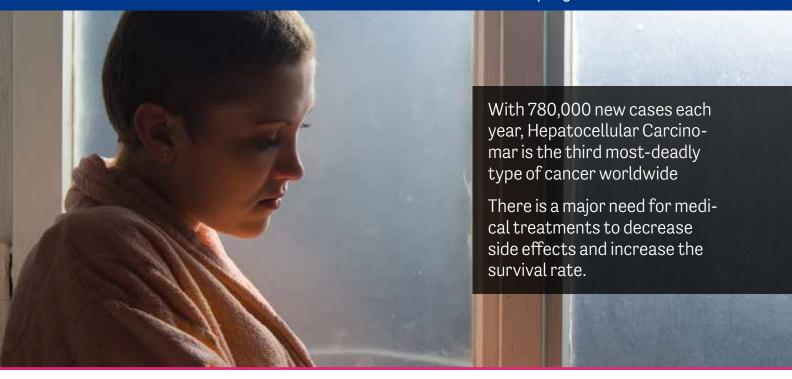
Project status: evaluation of model compounds

NeuroVive is currently evaluating compounds with the expectation to select a lead compound in 2018.

Objectives for 2018

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

NeuroVive's program for liver cancer



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. $^{1/2(3)}$

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.

 $^{4) \\ \}text{ http://www.cancerresearchuk.org/health-professional/cancer-statistics/statistics-by-cancer-type/liver-cancer/incidence\#heading-Nine} \\$

Financial information

Revenues

The consolidated turnover during the second quarter of 2018 was SEK 0 (0). Other operating revenues for the second quarter of 2018 were SEK 1,278,000 (88,000). The consolidated turnover for the first six months was SEK 0 (27,000) and the operating revenues

amounted SEK 1,452,000 (152,000). Other operating income includes research contributions from Bridge-Bio of SEK 876,000 and research contribution from Vinnova of 576,000 SEK.

Results of operations

The operating loss for the second quarter was SEK 25,162,000 (22,145,000). The operating loss for the first six months was SEK 38,155,000 (43,376,000). The net loss before tax for the second quarter amounted to SEK 25,481,000 (22,256,000). The net loss before tax for the first six months was SEK 38,534,000 (43,646,000).

The operating loss was affected by external expenses, which for the first six months were SEK 39,607,000 (43,555,000). During the first six months, expenses

related to development projects have affected the result with SEK 23,023,000 (16,342,000) whereof SEK 14,382,000 (9,537,000) relates to project in clinical phase. Personnel expenses during the first six months amount to SEK 7,791,000 (7,005,000). Other operating expenses amount to, SEK 654,000 (11,060,000) 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 91 (92) percent as of 30 June 2018, and equity was SEK 131,494,000 (105,846,000) compared to beginning of the year. Cash and cash equivalents amounted to SEK 51,896,000 (46,984,000) as of 30 June 2018, an increase of SEK

22,904 from the beginning of the year. Total assets as of 30 June 2018 were SEK 144,850,000 (134,969,000). The company completed a preferential rights issue in April providing SEK 64,176,000 after deduction of issue costs and compensation for guarantee commitments.

Cash flow and investments

Operating cash flow for the second quarter was SEK -26,943,000 (-19,959,000). Operating cash flow from the first six months was SEK -39,540,000 (-33,263,000). The cash flow effect related to investments in intangibles equals SEK -1,701,000 (-2,028,000) for the first six months. A preferential rights issue was

conducted during the quarter with a positive effect on the cash flow of SEK 64,176,000. Cash flow for the second quarter equals SEK 36,138,000 (-20,531,000). Cash flow for the first six months equals SEK 22,898,000 (-46,214,000).

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.

Apart from remuneration to senior managers including remuneration for consulting services, no purchases or sales between the group and related parties occurred. Transactions with related parties affecting profit/loss

for the period are stated below. 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.

| | 1 Jan. 2018 | 1 Jan. 2017 |
|-----------------------------------|--------------|--------------|
| (SEK 000) | 30 Jun. 2018 | 30 Jun. 2017 |
| Stanbridge bvba (owned by Gregory | | |
| Batcheller, until Nov 6, 2017, | | |
| Executive Chairman) | - | 508 |
| Total transactions with related | | |
| parties | | 508 |

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 percent 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

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 June 2018 was 10 (10), of which 4 (5) are women.

Parent company

Parent company earnings after tax for the first six months amounts to SEK -38,653,000 (-24,620,000). 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. The fully subscribed rights issue, which was completed in April, provided the company with SEK 64,176,000 after deduction of issue costs and compensation for guarantee commitments. 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 secured 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.

On May 25, 2016, the Tribunal rendered a partial award stating, among other things, that NeuroVive has certain payment obligations under the terms of the License Agreement,

for a period of 15 years following the initial launch in the respective country of such products covered by the License Agreement. The Tribunal reserved its decision on CicloMulsion's request to establish the obligation to pay royalties based on sales in countries where patents never existed but where it is alleged that know-how had been transferred, with the intention to consider the issue in a final arbitration. Other claims by CicloMulsion AG were rejected.

The arbitration award was contested by both parties to the Court of Appeal for Skåne and Blekinge and in its ruling, the Court of Appeal ordered all parts of the arbitration award to be set aside, with the exception

of the question for which the Tribunal had reserved its decision. In its ruling, the Court of Appeal also stated that because the case covers issues that are of such importance to the correct application of law, it allows an appeal to be made to the Supreme Court. This means that it is not necessary to seek leave to appeal to have the case heard by the Supreme Court. Neuro-Vive has appealed parts of the ruling to the Supreme Court. CicloMulsion has filed a submission to answer to the appeal. Hereafter, the Supreme Court has so far not dealt with the case further.

After CicloMulsion submitted a request for the release of the arbitrators from their appointments, and in response to this the arbitrators submitted their resignation, the arbitrators were released from their appointment by a decision taken by the Arbitration Institute of the Stockholm Chamber of Commerce (SCC). The constitution of a new Arbitral Tribunal has been finalized, but the scope and the timeline of the further proceeding are as yet unclear.

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.

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 not been subject to review by the company's auditors.

Upcoming financial statements

Interim Report January-September 22 November 2018
Year-End Report 2018 21 February 2019
The interim reports and the Annual Year Report are available at www.neurovive.com

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. 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.

Consolidated Statement of Comprehensive Income

| (0.000) | | 1 Apr, 2018 | 1 Apr, 2017 | 1 Jan, 2018 | 1 Jan, 2017 | 1 Jan, 2017 |
|--|------|--------------|-------------------|-------------------|--------------|--------------|
| (SEK 000) | Note | 30 Jun, 2018 | 30 Jun, 2017 | 30 Jun, 2018 | 30 Jun, 2017 | 31 Dec, 2017 |
| Net sales | | 1 070 | - 00 | 1 450 | 27 | 27 248 |
| Other operating income | | 1,278 | 88 88 | 1,452 | 152 179 | |
| Operating expenses | | 1,278 | 88 | 1,452 | 1/9 | 275 |
| Other external expenses | | 00.007 | 10.015 | 70.000 | 04.740 | 46 415 |
| Other external expenses Personnel cost | | -20,997 | -18,015 -3,629 | -30,220 -7,791 | -24,748 | -46,415 |
| Depreciation and write-down of tangible and | | -4,358 | -3,629 | -7,791 | -7,005 | -12,417 |
| intangible assets | | -481 | -538 | -942 | -742 | -1,595 |
| | | -401 -604 | -51 | -942 -654 | -11,060 | -10,936 |
| Other operating expenses | | | | | | , |
| | | -26,440 | -22,233 | -39,607 | -43,555 | -71,363 |
| Operating income | | -25,162 | -22,145 | -38,155 | -43,376 | -71,088 |
| Profit/loss from financial items | | | | | | |
| Result from shares in associated company | | - | - | - | - | 56 |
| Financial income | | 217 | 75 | 227 | 110 | 65 |
| Financial costs | | -536 | -186 | -606 | -379 | -636 |
| | | -319 | -111 | -379 | -269 | -515 |
| | | | | | | |
| Profit/loss before tax | | -25,481 | -22,256 | -38,534 | -43,646 | -71,603 |
| Income tax | 2 | | - | | | |
| Profit/loss for the period | | -25,481 | -22,256 | -38,534 | -43,646 | -71,603 |
| Other comprehensive income | | | | | | |
| Items that may be reclassified to profit or loss | | | | | | |
| Translation differences on foreign subsidiaries | | 1 | -29 | 5 | -11 | 1 |
| Total comprehensive income for the period | | -25,480 | -22,285 | -38,529 | -43,657 | -71,602 |
| Loss for the period attributable to: | | | | | | |
| Parent company shareholders | | -25,481 | -22,262 | -38,533 | -38,853 | -66,728 |
| Non-controlling interests | | - | 6 | -1 | -4,793 | -4,875 |
| | | -25,481 | -22,256 | -38,534 | -43,646 | -71,603 |
| Total comprehensive income for the period | | | | | | |
| Parent company shareholders | | -25,547 | -22,357 | -38,825 | -38,940 | -66,895 |
| Non-controlling interests | | 67 | 72 | 296 | -4,717 | -4,707 |
| | | -25,480 | -22,285 | -38,529 | -43,657 | -71,602 |
| Earnings per share before and after dilution(SEK | 7) | | | | | |
| based on average number of shares | .) | -0.40 | -0.45 | -0.61 | -0.79 | -1.33 |
| based on average number of shares | | -0.40 | -0.43 | -0.61 | -0.79 | -1.33 |

Consolidated Statement of Financial Position

| (SEK 000) | Note | 30 Jun 2018 | 30 Jun 2017 | 31 Dec 2017 |
|---|------|-------------|-------------|-------------|
| ASSETS | | | | |
| Non-current assets | | | | |
| Intangible assets | 1 | | | |
| Development costs | | 51,941 | 51,941 | 51,941 |
| Patents | | 22,721 | 19,253 | 20,627 |
| Other Intangible assets | | 1,680 | 1,815 | 1,747 |
| | | 76,342 | 73,009 | 74,315 |
| Tangible assets | | | | |
| Equipment | | 149 | 172 | 162 |
| | | 149 | 172 | 162 |
| Financial assets | | | | |
| Interpretation of the parent company hare capital dittional paid in capital capital dittional paid in capital | | 13,102 | 13,102 | 13,102 |
| | | 13,102 | 13,102 | 13,102 |
| Total non-current assets | | 89,593 | 86,283 | 87,579 |
| Current assets | | | | |
| Other receivables | | 2,979 | 1,037 | 1,568 |
| Prepaid expenses and accrued income | | 382 | 666 | 1,967 |
| Cash and cash equivalents | | 51,896 | 46,984 | 28,992 |
| | | 55,257 | 48,687 | 32,527 |
| TOTAL ASSETS | | 144,850 | 134,969 | 120,106 |
| EQUITY AND LIABILITIES | | | | |
| Equity attributable to the shareholders of the parent company | | | | |
| Share capital | | 4,579 | 2,474 | 2,616 |
| Additional paid in capital | | 489,440 | 418,489 | 427,226 |
| Translation reserve | | 321 | 693 | 613 |
| Retained earnings | | -368,273 | -301,865 | -329,740 |
| Total equity attributable to the shareholders of the parent | | 126,067 | 119,792 | 100,716 |
| Non-controlling interests | | 5,427 | 5,007 | 5,131 |
| Total equity | | 131,494 | 124,799 | 105,846 |
| Short-term liabilities | | | | |
| | | 5,263 | 3,657 | 7,525 |
| Other liabilities | | 871 | 1,911 | 863 |
| Accrued expenses and deferred income | | 7,222 | 4,602 | 5,871 |
| | | 13,356 | 10,171 | 14,260 |
| Total liabilities | | 13,356 | 10,171 | 14,260 |
| TOTAL FOLIVTY AND LYADYLYTYFO | | 444.050 | 474.000 | 100 100 |
| TOTAL EQUITY AND LIABILITIES | | 144,850 | 134,969 | 120,106 |

Consolidated Statement of Changes in Equity

| | Equity attribu | table to the | shareholders | s of the paren | t company | | |
|--|----------------|--------------|--------------|----------------|-----------|-------------|---------|
| (SEK 000) | | Additional | | | | Non- | |
| | Share- | paid in | Translation | Retained | | controlling | Total |
| | capital | capital | reserve | earnings | Total | interests | equity |
| 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 | - | - | | - | - | -1 | -38,534 |
| Other comprehensive income | | | | | | | |
| Translation differences | - | - | -292 | - | -292 | 297 | 5 |
| Other comprehensive profit/loss for the | | | | | | | |
| period, net after tax | - | - | -292 | - | -292 | 297 | 5 |
| Total comprehensive profit/loss | - | - | -292 | -38,533 | -38,825 | 296 | -38,529 |
| 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 June 2018 | 4,579 | 489,440 | 321 | -368,273 | 126,067 | 5,427 | 131,494 |

| 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 | - | - | | -38,853 | -38,853 | -4,793 | -43,646 |
| Other comprehensive income | | | | | | | |
| Translation differences | - | - | -87 | - | -87 | 76 | -11 |
| Other comprehensive profit/loss for the | | | | | | | |
| period, net after tax | - | - | -87 | - | -87 | 76 | -11 |
| Total comprehensive profit/loss | - | - | -87 | -38,853 | -38,940 | -4,717 | -43,657 |
| Transactions with shareholders | | | | | | | |
| Share issue | 1 | 150 | - | - | 152 | - | 152 |
| Change of ownership in share issue | | | - | 3,134 | 3,134 | -3,134 | - |
| Total transactions with shareholders | 1 | 150 | - | 3,134 | 3,286 | -3,134 | 152 |
| Closing balance, 30 June 2017 | 2,474 | 418,489 | 693 | -301,865 | 119,792 | 5,007 | 124,799 |

| 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 Apr, 2018 | 1 Apr, 2017 | 1 Jan, 2018 | 1 Jan, 2017 | 1 Jan, 2017 |
|--|--------------|--------------|--------------|--------------|--------------|
| | 30 Jun, 2018 | 30 Jun, 2017 | 30 Jun, 2018 | 30 Jun, 2017 | 31 Dec, 2017 |
| Cash flow from operating activities | | | | | |
| Operating income | -25,162 | -22,145 | -38,155 | -43,377 | -71,088 |
| Adjustments for non-cash items: | | | | | |
| Depreciation | 481 | 538 | 942 | 742 | 1,595 |
| Currency differences on intercompany items | - | -111 | - | 10 | -35 |
| Disposal of Business | - | - | - | 10,981 | 10,936 |
| Result from shares in associated company | | - | - | | 56 |
| Interest received | 217 | 75 | 227 | 110 | 65 |
| Interest paid | -536 | -186 | -606 | -379 | -149 |
| Net cash from operating activities before changes in | | | | | |
| working capital | -25,000 | -21,829 | -37,592 | -31,913 | -58,620 |
| Changes in working capital | | | | | |
| Increase/decrease of other current assets | 750 | -176 | 174 | 515 | -1,273 |
| Increase/decrease of other short-term liabilities | -2,693 | 2,046 | -2,122 | -1,865 | 1,769 |
| Changes in working capital | -1,943 | 1,870 | -1,948 | -1,350 | 496 |
| Cash flow from operating activities | -26,943 | -19,959 | -39,540 | -33,263 | -58,124 |
| Investing activities | | | | | |
| Acquisition of intangible assets | -1,078 | -573 | -1,701 | -2,028 | -4,204 |
| Acquisition of tangible assets | -17 | | -37 | -40 | -40 |
| Disposal business | _ | - | _ | -11,035 | -11,035 |
| Cash flow from investing activities | -1,095 | -573 | -1,738 | -13,103 | -15,279 |
| Financing activities | | | | | |
| New share issue | 64,176 | | 64,176 | 152 | 9,031 |
| Shareholder contribution subsidary | | | | _ | 114 |
| Cash flow from financing activities | 64,176 | | 64,176 | 152 | 9,145 |
| Cash flow for the period | 36,138 | -20,380 | 22,898 | -46,214 | -64,258 |
| Cash and cash equivalents at the beginning of the period | 15,757 | 67,289 | 28,992 | 93,251 | 93,251 |
| Effect of exchange rate changes on cash | 10,707 | 75 | 6 | -53 | |
| Cash and cash equivalents at end of period | 51,896 | 46,984 | 51,896 | 46,984 | 28,992 |

Parent Company Income Statement

| (SEK 000) | | 1 Apr, 2018 | 1 Apr, 2017 | 1 Jan, 2018 | 1 Jan, 2017 | 1 Jan, 2017 |
|--|------|--------------|--------------|--------------|--------------|--------------|
| N | Note | 30 Jun, 2018 | 30 Jun, 2017 | 30 Jun, 2018 | 30 Jun, 2017 | 31 Dec, 2017 |
| Net sales | | - | - | - | 27 | 27 |
| Other operating income | | 1,278 | 89 | 1,452 | 152 | 248 |
| | | 1,278 | 89 | 1,452 | 179 | 275 |
| Operating expenses | | | | | | |
| Other external expenses | | -20,995 | -18,013 | -30,212 | -24,680 | -45,857 |
| Personnel cost | | -4,358 | -3,628 | -7,791 | -6,778 | -12,190 |
| Depreciation and write-down of tangible and | | | | | | |
| intangible assets | | -481 | -539 | -942 | -732 | -1,584 |
| Other operating expenses | | -604 | -78 | -654 | -106 | |
| | | -26,437 | -22,258 | -39,599 | -32,297 | -59,631 |
| | | | | | | |
| Operating income | | -25,159 | -22,169 | -38,147 | -32,118 | -59,357 |
| Profit/loss from financial items | | | | | | |
| Result from shares in group company | | - | | - | 7,652 | 7,652 |
| Result from shares in associated company | | - | | - | - | 56 |
| Interest income and other similar profit items | | 212 | 65 | 220 | 80 | 29 |
| Interest expenses and other similar loss items | | -536 | -185 | -602 | -232 | -490 |
| | | -324 | -120 | -382 | 7,498 | 7,247 |
| Profit/loss before tax | | -25,484 | -22,289 | -38,529 | -24,620 | -52,109 |
| Income tax | 2 | | - | - | | - |
| Profit/loss for the period | | -25,484 | -22,289 | -38,529 | -24,620 | -52,109 |

Statement of Comprehensive Income, Parent Company

| (SEK 000) | | 1 Apr, 2018 | 1 Apr, 2017 | 1 Jan, 2018 | 1 Jan, 2017 | 1 Jan, 2017 |
|--|------|--------------|--------------|--------------|--------------|--------------|
| | Note | 30 Jun, 2018 | 30 Jun, 2017 | 30 Jun, 2018 | 30 Jun, 2017 | 31 Dec, 2017 |
| | | | | | | |
| Profit/loss for the period | | -25,484 | -22,289 | -38,529 | -24,620 | -52,109 |
| Other comprehensive income | | - | - | - | - | - |
| Total comprehensive profit/loss for the period | | -25,484 | -22,289 | -38,529 | -24,620 | -52,109 |

Parent Company Balance Sheet

| (SEK 000) | Note | 30 Jun 2018 | 30 Jun 2017 | 31 Dec 2017 |
|---------------------------------------|------|-------------|-------------|-------------|
| ASSETS | | | | |
| Non-current assets | | | | |
| Intangible assets | 1 | | | |
| Development costs | | 51,706 | 51,706 | 51,706 |
| Patents | | 22,721 | 19,253 | 20,627 |
| Other intangible assets | | 1,680 | 1,815 | 1,747 |
| | | 76,107 | 72,774 | 74,080 |
| Tangible assets | | | | |
| Equipment | | 149 | 172 | 162 |
| | | 149 | 172 | 162 |
| Financial assets | | | | |
| Other long-term placement | | 13,102 | 13,102 | 13,102 |
| Shares in subsidiaries | 3 | 23,625 | 23,099 | 23,625 |
| | | 36,727 | 36,201 | 36,727 |
| | | | | 28,883 |
| Total non-current assets | | 112,983 | 109,147 | 110,969 |
| Current assets | | | | |
| Short term receivables | | | | |
| Receivables from group companies | | - | 5,187 | - |
| Other receivables | | 2,977 | 1,015 | 1,566 |
| Prepaid expenses and accrued income | | 382 | 666 | 1,967 |
| | | 3,359 | 6,868 | 3,533 |
| Cash and bank balances | | 51,786 | 40,808 | 28,883 |
| Total current assets | | 55,145 | 47,676 | 32,416 |
| TOTAL ASSETS | | 168,128 | 156,823 | 143,385 |
| | | | | |
| EQUITY AND LIABILITIES | | | | |
| Equity | | | | |
| Restricted equity | | | | |
| Share capital | | 4,579 | 2,474 | 2,616 |
| Statutory reserve | | 1,856 | 1,856 | 1,856 |
| Development expenditure reserve | | 10,610 | 10,779 | 10,610 |
| | | 17,045 | 15,109 | 15,082 |
| Unrestricted equity | | | | |
| Share premium reserve | | 71,101 | - | 8,887 |
| Retained earnings | | 105,173 | 157,264 | 157,283 |
| Profit/loss for the period | | -38,529 | -24,620 | -52,109 |
| | | 137,745 | 132,644 | 114,061 |
| Total equity | | 154,790 | 147,753 | 129,143 |
| Short-term liabilities | | | | |
| Accounts payable | | 5,263 | 3,657 | 7,525 |
| Other liabilities | | 871 | 813 | 863 |
| Accrued expenses and deferred income | | 7,204 | 4,600 | 5,854 |
| And and expenses and deterred meditie | | 13,338 | 9,070 | 14,242 |
| | | | | |
| TOTAL EQUITY AND LIABILITIES | | 168,128 | 156,823 | 143,385 |

Notes

Not 1 - Immateriella tillgångar

| (SEK 000) | Development costs | Patents | Other | Total |
|------------------------------|-------------------|---------|--------|--------|
| ACCUMULATED COST | | | | |
| Opening balance 1 Jan. 2018 | 51,941 | 28,405 | 2,864 | 83,210 |
| Additions | - | 2,919 | - | 2,919 |
| Closing balance 30 Jun. 2018 | 51,941 | 31,324 | 2,864 | 86,129 |
| ACCUMULATED DEPRECIATION | | | | |
| Opening balance 1 Jan. 2018 | - | -7,778 | -1,117 | -8,895 |
| Depreciation for the period | - | -825 | -67 | -892 |
| Closing balance 30 Jun. 2018 | - | -8,603 | -1,184 | -9,787 |
| Residual value 30 Jun. 2018 | 51,941 | 22,721 | 1,680 | 76,342 |

| (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 | -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 413,468,000 as of 30 June 2018 (332,390,000). The parent company's total loss carry-forwards amount to SEK 387,284,000 as of 30 June 2018 (306,679,000). 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 percent in the subsidiary NeuroVive Pharmaceutical Asia Ltd., domiciled in Hong Kong.

Interim report January - June 2018



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, 21 August 2018

David Laskow-PooleyDavid BejkerDenise GoodeChairman of the BoardBoard memberBoard member

Jan Törnell Erik Kinnman

Board member 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. The information was submitted for publication, through the agency of the contact person set out above, at 08:30 a.m. CET on 21 August 2018.



About NeuroVive

NeuroVive Pharmaceutical AB has two orphan drug projects in clinical development, and several research projects in the pipeline. The company is a leader in mitochondrial medicine and focused on the research and development of drug candidates that maintain mitochondrial integrity and function for indications with a major unmet medical need.

The company's strategy is to take drugs for rare diseases through clinical development and to the market. For the company's projects focused on common indications with high commercial potential, the strategy is to out-license at preclinical stage.

A subset of succinate prodrug under NeuroVive's NVP015 program has been out-licensed to Fortify Therapeutics, a BridgeBio company, for the development of a local treatment 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 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.

Cyclophilin D. 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

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.

 $\textbf{ToxPhos}^{\textbf{o}}. \ \text{NeuroVive's registered trademark for the Company's mitochondrial toxicity test}.$