Important information
This presentation (the “Presentation”) has been prepared by NeuroVive Pharmaceutical AB (publ), 556595-6538 ("NeuroVive" or the “Company”).
The Presentation is governed by Swedish law. The courts of Sweden have exclusive jurisdiction to settle any dispute arising out of or in connection with this Presentation.
This Presentation does not constitute an offer of financial instruments to the public or an admission of such financial instruments to trading on a regulated market requiring an approved prospectus under the Swedish Financial Instruments Trading Act (1991:980) and, accordingly, this Presentation does not constitute a prospectus for these purposes and have not been, and will not be, approved or registered by the Swedish Financial Supervisory Authority (Sw: Finansinspektionen) under the Swedish Financial Instruments Trading Act.

Forward-looking statements
The Presentation contains certain forward-looking statements that reflect NeuroVive’s current views or expectations with respect to future events and financial and operational development. The words “intend”, “estimate”, “expect”, “may”, “plan”, “anticipate” or similar expressions regarding indications or predictions of future developments or trends and which are not based on historical facts constitute forward-looking information.
Although NeuroVive believes that these statements are based on reasonable assumptions and expectations, NeuroVive cannot give any assurances that such statements will materialize. Forward-looking statements are in its nature involved with both known and unknown risks and uncertainties, since they are depending on future events and circumstances. Forward-looking statements do not constitute any representations and warranties of future development and the outcome could differ materially from the information set out in the forward-looking statements.
The forward-looking statements included in this Presentation apply only to the date of this Presentation. NeuroVive undertakes no obligation to publicly update or revise any forward-looking statements, whether as a result of new information, future events or similar circumstances other than as required by applicable law.
Corporate Highlights

• Swedish Specialty Pharmaceutical company founded in 2000
  • Frontier mitochondrial medicine research

• Two projects in clinical development
  • Traumatic Brain Injury in Phase II
  • Genetic Mitochondrial Diseases in Phase I

• Several preclinical assets
  • Genetic Mitochondrial Diseases
  • Direct antifibrotic NASH project

• Listed on Nasdaq Stockholm (NVP.ST) since 2008
  • Market cap: ~325 MSEK (~36 MUSD)
  • Average trading volume: ~700,000
  • OTCQX (NEVPF:US)
Mitochondria are the Powerhouse of Cells

1. Produces the energy that cells need
2. Protects cells and controls cell death

- A non-functioning or damaged mitochondrion leads to cell death and organ damage

NeuroVive develops compounds that protect and enhance mitochondrial function
# New Compelling and Diversified Portfolio with High Opportunities

## Development to the Market with/without Partner vs. Outlicensed

<table>
<thead>
<tr>
<th>Brain Injury</th>
<th>Mitochondrial Respiratory Chain Diseases</th>
<th>NASH</th>
<th>Liver Cancer</th>
</tr>
</thead>
<tbody>
<tr>
<td>TBI</td>
<td>Chronic, Acute, Myopathy, LHON</td>
<td>Fibrosis, Metabolic, HCC</td>
<td></td>
</tr>
</tbody>
</table>

## Market

<table>
<thead>
<tr>
<th>Phase III</th>
</tr>
</thead>
<tbody>
<tr>
<td>Market</td>
</tr>
</tbody>
</table>

## Phase II

<table>
<thead>
<tr>
<th>Phase II</th>
</tr>
</thead>
<tbody>
<tr>
<td>Market</td>
</tr>
</tbody>
</table>

## Phase I

<table>
<thead>
<tr>
<th>Phase I</th>
</tr>
</thead>
<tbody>
<tr>
<td>Market</td>
</tr>
</tbody>
</table>

## Preclinical

<table>
<thead>
<tr>
<th>Preclinical</th>
</tr>
</thead>
<tbody>
<tr>
<td>Market</td>
</tr>
</tbody>
</table>

## Lead Selection

<table>
<thead>
<tr>
<th>Lead Selection</th>
</tr>
</thead>
<tbody>
<tr>
<td>Market</td>
</tr>
</tbody>
</table>

## Discovery

<table>
<thead>
<tr>
<th>Discovery</th>
</tr>
</thead>
<tbody>
<tr>
<td>Market</td>
</tr>
</tbody>
</table>

## Mechanism of Action

<table>
<thead>
<tr>
<th>Mechanism of Action</th>
<th>NeuroSTAT®</th>
<th>KL1333</th>
<th>NV354</th>
<th>NVP025</th>
<th>NVP015</th>
<th>NV556</th>
<th>NVP022</th>
<th>NVP024</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cyclophilin D inhibition</td>
<td>NeuroSTAT®</td>
<td>KL1333</td>
<td>NV354</td>
<td>NVP025</td>
<td>NVP015</td>
<td>NV556</td>
<td>NVP022</td>
<td>NVP024</td>
</tr>
<tr>
<td>NAD+ modulation</td>
<td>NeuroSTAT®</td>
<td>KL1333</td>
<td>NV354</td>
<td>NVP025</td>
<td>NVP015</td>
<td>NV556</td>
<td>NVP022</td>
<td>NVP024</td>
</tr>
<tr>
<td>Succinate prodrug</td>
<td>NeuroSTAT®</td>
<td>KL1333</td>
<td>NV354</td>
<td>NVP025</td>
<td>NVP015</td>
<td>NV556</td>
<td>NVP022</td>
<td>NVP024</td>
</tr>
<tr>
<td>Cyclophilin D inhibition</td>
<td>NeuroSTAT®</td>
<td>KL1333</td>
<td>NV354</td>
<td>NVP025</td>
<td>NVP015</td>
<td>NV556</td>
<td>NVP022</td>
<td>NVP024</td>
</tr>
<tr>
<td>Succinate prodrug</td>
<td>NeuroSTAT®</td>
<td>KL1333</td>
<td>NV354</td>
<td>NVP025</td>
<td>NVP015</td>
<td>NV556</td>
<td>NVP022</td>
<td>NVP024</td>
</tr>
<tr>
<td>Cyclophilin inhibition</td>
<td>NeuroSTAT®</td>
<td>KL1333</td>
<td>NV354</td>
<td>NVP025</td>
<td>NVP015</td>
<td>NV556</td>
<td>NVP022</td>
<td>NVP024</td>
</tr>
<tr>
<td>Respiratory chain uncoupling</td>
<td>NeuroSTAT®</td>
<td>KL1333</td>
<td>NV354</td>
<td>NVP025</td>
<td>NVP015</td>
<td>NV556</td>
<td>NVP022</td>
<td>NVP024</td>
</tr>
<tr>
<td>Undisclosed</td>
<td>NeuroSTAT®</td>
<td>KL1333</td>
<td>NV354</td>
<td>NVP025</td>
<td>NVP015</td>
<td>NV556</td>
<td>NVP022</td>
<td>NVP024</td>
</tr>
</tbody>
</table>

## Partner

<table>
<thead>
<tr>
<th>Partner</th>
</tr>
</thead>
</table>
## Corporate Timelines

<table>
<thead>
<tr>
<th>Indication - lead candidate/project</th>
<th>Event</th>
<th>When</th>
</tr>
</thead>
<tbody>
<tr>
<td>Genetic mitochondrial disease - KL1333</td>
<td>Initiation of Ph Ib (MAD) study EU</td>
<td>Q4 2018</td>
</tr>
<tr>
<td>Genetic mitochondrial disease - NV354</td>
<td><em>In vivo</em> preclinical PoP results</td>
<td>Q4 2018</td>
</tr>
<tr>
<td>Mitochondrial myopathy - NVP025</td>
<td>Lead candidate drug selection</td>
<td>Q4 2018</td>
</tr>
<tr>
<td>Traumatic Brain Injury - NeuroSTAT®</td>
<td>Initiation Ph IIb efficacy study</td>
<td>2019</td>
</tr>
<tr>
<td>NASH - NV556 (for out-licensing)</td>
<td>Out-licensing</td>
<td>H1 2019</td>
</tr>
<tr>
<td>NASH - NVP022 (for out-licensing)</td>
<td>Lead candidate drug selection</td>
<td>Q4 2018</td>
</tr>
<tr>
<td>HCC - NVP024 (for out-licensing)</td>
<td>Lead candidate drug selection</td>
<td>Q4 2018</td>
</tr>
</tbody>
</table>
New Two-Pronged Corporate Strategy

Frontier mitochondrial medicine research in areas of high unmet medical need project starting point

1. Develop orphan drug projects to the market with or without partner
2. Complete outlicensing of common disease projects at the preclinical stage

Purpose
- Risk diversification
- Near-term revenues
- Mid- to long-term value creation
**NeuroVive’s Management Team**

**Erik Kinnman,**
**CEO**
Born: 1958  
**Education:** Holds an Executive MBA from the Stockholm School of Economics and has comprehensive scientific qualifications from the Karolinska Institutet, which has rendered him a Ph.D. and an Associate Professor. Moreover, Erik Kinnman is an M.D., board certified in Neurology and Pain Management.  
**Previous experience:** Has held a number of senior leadership positions in biopharmaceutical companies such as AstraZeneca and Sobi.

---

**Eskil Elmér,**
**CSO, VP Discovery**
Born: 1970  
**Education:** Associate professor of experimental neurology at Lund University.  
**Previous experience:** Dr. Elmér is patentee and co-founder of both Maas Biolab, LLC and NeuroVive Pharmaceutical AB, and CSO of NeuroVive, with overall charge of the company’s pre-clinical research. In addition, Eskil Elmér is a practicing physician in the department of clinical neurophysiology at Skåne University Hospital in Lund.

---

**Catharina Johansson,**
**CFO, VP IR & Comms.**
Born: 1967  
**Education:** Holds a M.Sc. in Business and Economics.  
**Previous experience:** Her previous experience includes serving as interim CFO for medical device company Cellavision, which is listed on Nasdaq Stockholm, and Accounting Manager for Bong and Alfa Laval Europe.

---

**Magnus Hansson,**
**CMO, VP Preclin. & Clincal Dev.**
Born: 1976  
**Education:** Holds a PhD in Experimental brain research from Lund University.  
**Previous experience:** He has previously been serving as a Senior Scientist in NeuroVive since 2008 and as a consultant physician and associate professor in medical imaging and physiology at Skåne University Hospital.

---

**Mark Farmery,**
**VP Business Dev.**
Born: 1969  
**Education:** Dr. Farmery received his BSc in Biomedical Sciences (Microbiology) from the University of Bradford and his PhD in Biochemistry and Molecular Microbiology from the University of Leeds.  
**Previous experience:** More than 15 years’ experience in biopharma business development from Karolinska Institutet Innovations AB, AstraZeneca and Karo Bio AB.
Network operational model to ensure fit for purpose organization

External experts in opportunistic therapeutic areas
- Prof. Ramon Diaz-Arrastia, Penn (TBI)
- Prof. Massimo Pinzani, UCL (NASH)
- Prof. Philippe Gallay, Scripps (NASH)
- Prof. Marni Falk, CHOP (Mito.)
- Prof. Patrick Chinnery, Cambridge (Mito.)

Resources for development and production just in time
- SMC Laboratories, Japan (STAM model)
- Physiogenex, Frankrike (MCD model)
- Chempartner, Kina (DMPK, Chemical synthesis)
- Engitix Ltd, England (3D liver model)
- Fresenius Kabi, Österrike (GMP Manufacturing)
- Wuxi-AppTec, Kina (DMPK, Chemical synthesis)

Chas Hude, patent lawyers

Commercial organization when products have matured
NeuroVive's Board of Directors

David Laskow-Pooley, Chairman of the Board
Born: 1954
Education: BSc Pharmacy (1st), Pharmaceutical/Chemical engineering specialty and QP., Sunderland School of Pharmacy.
Previous experience: CEO and Board Member of London Pharma Ltd, and Board member of CubiteL Ltd TapImmune Inc (USA), and Pharmafor Ltd.

Denise Goode
Born: 1958
Education: BSc in Zoology/Animal Biology from University of Manchester. Chartered Accountant.

David Bejker
Born: 1975
Education: M.Sc. (Econ.), Stockholm School of Economics.
Previous experience: Has many years of industry experience both from an investor perspective, through employment in HealthCap, and also operating as a business developer of Affibody AB during 2003 to 2005.

Jan Törnell
Born: 1960
Education: MD and PhD in physiology, University of Gothenburg
Previous experience: 20 years’ experience from different senior positions within the pharmaceutical industry, both in Sweden and internationally. Jan held the position as Vice President in AstraZeneca Oncology & Infection 2009-2011 and Vice President Translational Science 2006-2008. He was Director at AstraZeneca Discovery 1999-2005 and Astra 1996-1999.
Genetic Mitochondrial Diseases
Genetic Mitochondrial Diseases are Rare Syndromes

- Includes MELAS, KSS, CPEO, PEO, Pearson, MERRF, Leigh, LHON and Alper’s syndromes
  - Caused by rare and heritable mutations in the mitochondrial respiratory chain
- Chronic deterioration and reduced life expectancy
  - Diverse symptoms from multiple organs
- Prevalence 12.5 per 100,000 population
  => Orphan indications
- High unmet medical need
  - One approved treatment in EU
    - LHON (idebenone, Santhera)
Symptoms and Signs of Genetic Mitochondrial Diseases in Various Organs

**Nervous system**
Seizures, tremors, mental retardation, deafness, dementia, stroke before the age of 40, balance disorders, effects on the peripheral nervous system

**Heart**
Cardiomyopathy (heart failure, conduction block)

**Liver**
Liver failure is rare except in infants experiencing loss of mitochondrial DNA

**Kidney**
Fanconi syndrome (the loss of key metabolites in the urine)

**Eyes**
Drooping eyelids (ptosis), inability to move the eyes sideways (external ophthalmoplegia), blindness (retinitis pigmentosa)

**Muscles**
Muscle weakness, exercise intolerance, seizures

**Digestive system**
Regurgitation, vomiting, chronic diarrhea, intestinal obstruction

**Pancreas**
Diabetes
Strong Portfolio of Complementary MRCD Programs

KL1333 (NAD\(^+\) modulator) - Disease modifier

NV354 (succinate prodrug) - Alternative energy source

NVP025 (cyclophilin inhibitor) - Muscle cell protection
Dysfunctions in Genetic Mitochondrial Diseases

- ATP
- Free radicals
- Altered balance of key metabolic co-factors
- Lactate

KL1333 increases energy production and production of new mitochondria

KL1333 increases NAD+ levels

1. Restores aerobic energy production
2. Increases antioxidant defense
3. Builds up new mitochondria
KL1333 for mitochondrial disorders

“ping-pong mechanism”

- Orally available small molecule (MW<250)
- Potent NQO1 substrate
- Direct effects:
  - Increases NAD+/NADH ratio
  - Bypass mechanism - ATP production
- Secondary effects
  - Mitochondrial biogenesis and longevity pathways
    - SIRT1/AMPK/PGC-1α activation
  - Increased antioxidant/GSH status

KL1333 and NQO1 – NAD⁺ and ATP are increased.
KL1333 Development Program

- FTIM Phase Ia study completed
  - Only mild GI side-effects at high doses
  - No SAEs
  - Favorable PK

- Orphan Drug Designation EMA and FDA
- EU/US Phase Ia/b in UK 2018-2019
  - Healthy volunteers
  - Mitochondrial disease patients
- Targeting adult and pediatric patients
  - MELAS and associated disease m.3243A>G mutation
    - prevalence ≈ 2/100,000
  - Mitochondrial DNA deletion syndromes (PEO, KSS)
    - prevalence ≈ 3/100,000

- Possible market approval by 2024
- Peak Yearly sales 1.8 BUSD*

* Monocl Strategy Services 2017
NVP354: Alternative Energy Source in Genetic Mitochondrial Diseases

- Energy regulating substance
- Overcomes complex I deficiencies by delivering cell-permeable prodrug substrate to complex II
- NV354 preclinical studies ongoing
  - In-vivo results H2 2018/2019
  - Project supported by Vinnova (Swelife) and US Department of Defence grants (4MUSD)
- Clinical development 2020
  - Local eye therapy: LHON (Fortify/BridgeBio)
    - Prevalence 2-3/100,000
  - Oral treatment: Leigh disease
    - Incidence 2-3/100,000 children
  - Possible market approval
    - 2025E: US & EU5
    - 2027E: JP & CH
    - Peak sales 1.3 MUSD (2031E1)

1 Monocl Strategy Services 2017
Traumatic Brain Injury
Traumatic Brain Injury (TBI)

• Brain continues to deteriorate for several days after external trauma
  • TBI leads to significant neurological disabilities and high burden of disease

• >3 million patients in US and EU
  • 15% moderate to severe damage

• TBI cost to the global economy approximately 400 BUSD annually*
  • TBI direct health care cost per patient 400 kUSD
  • Only symptomatic treatment existing
  • One project in advanced clinical development
  • 23 BUSD treatment market 2024

* Lancet Neurol. 2017 Dec;16(12):987-1048
Prevention of Secondary Brain Injury after TBI - NeuroSTAT®

- Orphan drug designation in the EU and the US
- Open phase IIa safety & pharmacokinetic study (CHIC)
  - Safe and tolerable, reaches the brain
  - Biochemical and biomarker results indicate treatment effect
- 35% reduction of brain injury volume showed in an experimental study at University of Pennsylvania
- Phase IIb clinical efficacy study planned for 2019
  - Primarily non-dilutive soft money financing
  - Project to be further partnered
  - Existing agreement with Chinese co Sihuan and Sanofi in Korea
- NeuroSTAT possible market approval year
  - 2025E*: US & EU
  - 2027E*: JP & CH
- Peak sales 1.6 BUSD*

*Monocl Strategy Services 2017
NeuroSTAT® development program

• Phase IIb PoP 2019
  • Placebo-controlled randomized study
  • Novel biomarker/imaging design
  • Targets a homogeneous subpopulation
  • 70-80 patient study
  • 17 high end clinical sites in 5 countries (US & EU)
  • 6 month follow up

• Phase III H1 2021
  • Study design and size based on IIb
NASH
NASH - Non-Alcoholic Steatohepatitis

- 30% of the US population have nonalcoholic fatty liver disease (NAFLD)
- 3-5% of the US population (15M people) have NASH
- Strong association NASH and diabetes/obesity
  - Buildup of fat and inflammation in the liver
  - Worsening fibrosis is major variable associated with poor outcomes and death
  - May lead to liver cirrhosis and hepatocellular carcinoma
- No registered drugs on the market\(^1\)
- 25 BUSD global market estimated by 2026\(^1\)

\(^1\) Global Data, OpportunityAnalyzer: NASH - Opportunity Analysis and Forecasts to 2026
NASH Out-licensing Projects

- NV556 prevents fibrosis development in experimental NASH models
  - Direct anti-fibrotic effect through cyclophilin B inhibition
  - Cytoprotective action through mitochondrial stabilization

- Extensive preclinical development documentation
  - Orally available, liver targeted, once-daily
  - Good safety profile
  - Manufacturing has been scaled to kg levels

- Peak Yearly Sales 1.7 BUSD*

- Outlicensing possible H1 2019

- NVP022 is targeted to metabolic component
  - Mild uncoupling novel molecule

---

1 Monocl Strategy Services 2017
Hepatocellular Carcinoma
Hepatocellular Carcinoma (HCC) - NVP024

- **HCC 6th most prevalent cancer**
  - Most common causes: NASH, alcohol, viral hepatitis infections
- **745,000 deaths/year**
- **Existing treatments have limited effect on survival and symptomatic progression**

- **New generation sanglifehrin-based compounds**
  - Potent inhibitory effects on hepatocellular carcinoma (HCC) cells
  - Anti-cancer activity in an experimental model of HCC
  - High liver exposure
  - Not toxic to normal cells and well tolerated *in vivo*

- **Further preclinical studies ongoing**

- Lead candidate drug selection 2018
Financial Situation
### Financial Position and Use of Proceeds

<table>
<thead>
<tr>
<th>30 June 2018</th>
<th>MUSD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intangible assets</td>
<td>8.4</td>
</tr>
<tr>
<td>Liquid assets</td>
<td>5.8</td>
</tr>
<tr>
<td>Short-term debts</td>
<td>1.5</td>
</tr>
<tr>
<td>Burn rate 2017*</td>
<td>7.0</td>
</tr>
</tbody>
</table>

* Cash flow from operating and investment activities

- **78.5 MSEK in over-subscribed rights issue will fund:**
  - KL1333 phase Ib study 2018
  - Initiation of NeuroSTAT phase IIb study 2018
  - Preclinical portfolio development
  - Warrants 37.3 MSEK Nov. 2018 @3.80 SEK
## Shareholders

<table>
<thead>
<tr>
<th>Shareholder</th>
<th>Votes and capital (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Avanza Pension Försäkrings AB</td>
<td>14.66</td>
</tr>
<tr>
<td>EuroClear Bank S.A/N.V, W8-IMY (registered holding on behalf of Maas Biolab, LLC and Marcus Keep and others with US domicile)</td>
<td>4.90</td>
</tr>
<tr>
<td>Nordnet Pensionförsäkring AB</td>
<td>3.79</td>
</tr>
<tr>
<td>Baulos Capital Belgium SA (prev. Private Placement SPRL)</td>
<td>3.28</td>
</tr>
<tr>
<td>Rothesay Limited</td>
<td>2.40</td>
</tr>
<tr>
<td>Danske Bank International S.A.</td>
<td>2.29</td>
</tr>
<tr>
<td>Handelsbanken Liv</td>
<td>2.19</td>
</tr>
<tr>
<td>Ekman, Tobias</td>
<td>1.09</td>
</tr>
<tr>
<td>Försäkrings AB, Skandia</td>
<td>0.88</td>
</tr>
<tr>
<td>Swedbank försäkring AB</td>
<td>0.74</td>
</tr>
<tr>
<td>Other owners (app. 9,000 shareholders)</td>
<td>63.78</td>
</tr>
<tr>
<td><strong>Total (Source: Euroclear 20180928)</strong></td>
<td><strong>100.00</strong></td>
</tr>
</tbody>
</table>
Investment Case - High Value portfolio with Near Term Inflection Points

1. New strong diversified portfolio with near term value triggers and growth potential
   - KL1333 for orphan mitochondrial diseases to move into Ph Ib
   - Outlicensing deal for NV556 (NASH) end 2018/H1 2019
   - NeuroSTAT for TBI ready for Ph IIb to be partially financed with non-dilutive funds
   - NVP015 experimental PoP

2. New two-pronged corporate strategy beginning to materialize
   - World leader in mitochondrial medicine
   - Strong patent portfolio and broad technology base

3. New members in management and board
   - Cost-effective collaborative semi-virtual organization