# ABLIVA

# Year-End Report

January – December 2021

2021 was a defining year for Abliva, focused on portfolio delivering, de-risking our assets and building relationships as we prepared for success in 2022.

Ellen Donnelly, CEO

Delivering mitochondrial health

# 2021 summary

## KL1333 shows signs of efficacy in patients and has an approved IND for a Phase 2/3 study

## Regulatory feedback for NV354 enables clinical development

### Important events during 2021

**KL1333**

- Data from the Phase 1a/b clinical study of KL1333 were released in May and confirmed the safety and pharmacokinetic profile of the drug. In addition, in a cohort of eight patients, there were signs of efficacy across well-established relevant clinical endpoints including two patient-reported fatigue endpoints and a functional endpoint.
- The study to create a primary mitochondrial disease fatigue endpoint for use in the KL1333 Phase 2/3 study was completed in August.
- The first chronic toxicology study with KL1333 (six months) was completed in September.
- The US Food and Drug Administration approved Abliva's Investigational New Drug (IND) application for KL1333 in November, enabling the start of a registrational Phase 2/3 study with first patients due to be recruited in 2022.

**NV354**

- In September, favorable feedback was received from UK pharmaceutical regulators (MHRA) on the NV354 preclinical data package.

**Financials**

- A directed issue of SEK 80m in two tranches was carried out in March.
- Abliva resolved on a directed issue of convertibles amounting to SEK 26m, subject to the approval of an extraordinary general meeting, in December.

**Other**

- Dr. Ellen K. Donnelly, Ph.D. was appointed new CEO of Abliva.

### Important events after the reporting period

- An extraordinary general meeting was held on 14 January 2022. The general meeting approved the Board of Directors' resolution from 20 December 2021 on a directed issue of convertible bonds amounting to SEK 26 million.

### Financial information

**October-December 2021***

- Net revenues: SEK 48,000 (112,000)
- Other operating income: SEK 126,000 (1,629,000)
- Loss before tax: SEK 36,871,000 (13,067,000)
- Loss per share before dilution: SEK 0.09 (0.03)
- Diluted loss per share: SEK 0.09 (0.03)

**January-December 2021***

- Net revenues: SEK 115,000 (216,000)
- Other operating income: SEK 0,000 (1,648,000)
- Loss before tax: SEK 123,494,000 (59,994)
- Loss per share before dilution: SEK 0.33 (0.24)
- Diluted loss per share: SEK 0.33 (0.24)

* APM Alternative perfomance measures, see definition on page 20.

# KL1333 Ends the Year with FDA Approval for Commencement of Phase 2/3 Study

**2021 ended on a high note with the approval of the KL1333 Investigational New Drug (IND) application by the FDA. This milestone serves as both a capstone of 2021, confirming the strength of the KL1333 package, and a launch pad into 2022, providing us with the approval to start our Phase 2/3 study in the United States. The approval came at the end of an important foundational year for the company where the KL1333 program was de-risked with efficacy in primary mitochondrial disease patients and chronic toxicology in preparation for the global, Phase 2/3 study in 2022. In addition, the NV354 program had a successful review by MHRA and will look to also enter the clinic in 2022.**

**The Value of IND Approval for KL1333**

The approval of the IND for KL1333 at the end of November was a major milestone for the company for many reasons. First, this represented the most extensive review of the KL1333 preclinical and clinical package by a regulatory body to date. We took the package to the US FDA first as we knew the program would get a thorough review. During their review of the KL1333 package the FDA also looked at the Phase 2/3 study design, and their valuable input was used to finalize the protocol for future submissions to other countries. In this one milestone, then, we received confirmation that the studies done over the past five years were appropriate and, that the clinical study we expect to run over the next three years, is also well founded. One important milestone validated 8 years of work!

**De-Risking the KL1333 Program**

Although the IND approval was the highlight of the year, I would be remiss not to call out a few of the other important milestones for this program. The Phase 1b data from mitochondrial disease patients provided important evidence that KL1333 is driving an effect in patients, and this effect not only impacts important and relevant disease manifestations, but is also dependent upon the exposure (i.e patients with more KL1333 in their blood get more effect). Other highlights included increasing our confidence in the safety profile of the drug with the chronic toxicology studies and the drug-drug interaction study, and validating our fatigue endpoint so it is specific to the primary mitochondrial disease condition. Finally, the year ended with a productive trip to Seoul, South Korea to meet and strengthen our collaboration with our partner on KL1333, Yungjin Pharm.

*"2021 was a defining year for Abliva, focused on portfolio delivering, de-risking our assets and building relationships as we prepared for success in 2022"*

**NV354 Takes One Step Closer to the Clinic**

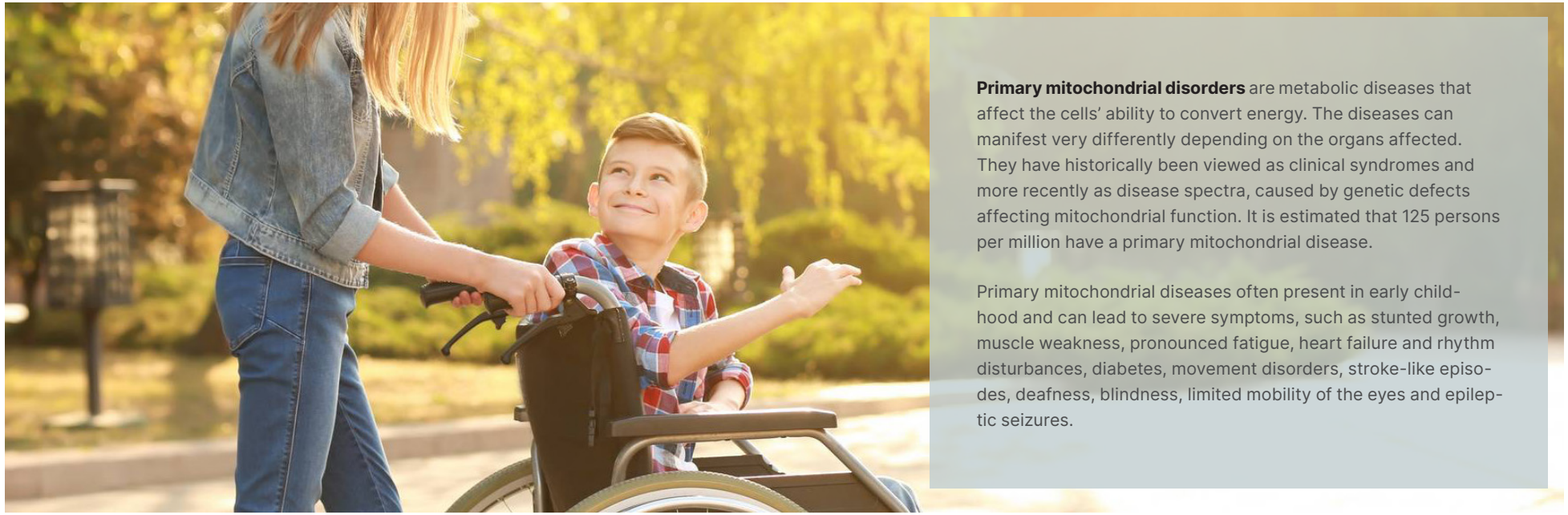
NV354 also made good strides in 2021 with the completion of the preclinical data package and a successful meeting with the MHRA. We look forward to progressing this program in 2022.

**Establishing Abliva as a Leader in Mitochondrial Disease Drug Development**

A key focus at Abliva is to spread our story globally and grow our network of patients, physicians, investors and collaborators across the world. This important work is largely foundational with the impact to be seen later. In 2021 we opened the US subsidiary to assist in this work and strengthened our US network. We collaborated with a number of patient organizations and worked with patients on the validation of our fatigue endpoints, and gave recognized talks at a number of important meetings including the Mitochondrial Medicine meeting in Cambridge in November. At the end of the year we focused on physicians and sites as we work to select our network of sites for the upcoming study. We have learned and grown from these interactions and we look forward to seeing their impact in 2022.

2021 was a defining year for Abliva, focused on portfolio delivering, de-risking our assets and building relationships as we prepared for success in 2022.

**Ellen Donnelly**
CEO



**Primary mitochondrial disorders** are metabolic diseases that affect the cells' ability to convert energy. The diseases can manifest very differently depending on the organs affected. They have historically been viewed as clinical syndromes and more recently as disease spectra, caused by genetic defects affecting mitochondrial function. It is estimated that 125 persons per million have a primary mitochondrial disease.

Primary mitochondrial diseases often present in early childhood and can lead to severe symptoms, such as stunted growth, muscle weakness, pronounced fatigue, heart failure and rhythm disturbances, diabetes, movement disorders, stroke-like episodes, deafness, blindness, limited mobility of the eyes and epileptic seizures.


*Orphan drug designation in the US and Europe

PMD stands for Primary Mitochondrial Diseases, often referred to as 'mito disease'.
mtDNA disorders are disorders caused by mutation(s) in mitochondrial DNA (as opposed to nuclear DNA).

# Strategic focus: Primary Mitochondrial Diseases

**Abliva is focused on becoming the leading biopharmaceutical company in mitochondrial medicine, developing therapeutics for primary mitochondrial diseases, orphan indications of high unmet medical need. The company intends to build a fully integrated research, development, and commercial organization, developing innovative therapeutics and taking them directly to the patients.**

**Building the Premier Mitochondrial Medicine Company**
Abliva's long-term goal is to become the leading global biopharmaceutical company focused on the discovery of therapeutics for mitochondrial diseases. Abliva has the foundation to do this with a clear strategy, a strong portfolio of assets, a research organization and a team that has over two decades of experience in mitochondrial medicine as well as decades of experience in drug development.

Over the next five years we will focus on the delivery of our portfolio to the market. We will augment our strong research and development capabilities and build a commercial organization. We will bring new innovative therapeutics to the clinic and fuel our pipeline with new candidates from discovery. We will attract and retain talented colleagues with a passion for drug development. We will build a strong network of experts that will complement, enhance and support our efforts across development that will include patients, physicians, researchers, regulators, payers and technical experts. We will generate future revenues through two paths: sales revenue for the drugs Abliva intends to bring to market, and revenue from out-licensing assets (through milestone payments and royalties).

**Addressing Primary Mitochondrial Diseases (PMD)**
Mitochondria function as the powerhouses of our cells and are crucial for the cells' energy metabolism. PMD are rare orphan diseases where the energy metabolism in the cells is impaired, causing deterioration that leads to multifaceted disorders and great suffering for patients. The symptoms worsen over time and, in many cases, the diseases lead to premature mortality. Mitochondrial medicine has become an area of increasing focus for the pharmaceutical industry as there are currently no effective treatment options. Through Abliva's research and development, we have an opportunity to improve the quality of life for these patients.

**Delivering a Portfolio of First-in-Class Therapies**
Abliva's in-house R&D capabilities have been instrumental in creating and delivering a portfolio that includes several projects with mechanisms of action suitable for a wide range of PMDs.

KL1333 restores the balance of the coenzymes NAD+ and NADH, creating new mitochondria and improved energy levels. KL1333 has completed a number of key Phase 1 studies to prepare the asset for registrational Phase 2/3 study start in 2022. KL1333 is protected by both a composition of matter patent as well as Orphan Drug Designation (ODD) in the U.S. and in Europe. The commercial opportunity is significant with even conservative estimates exceeding USD 1 billion per year in annual sales.

NV354, an energy replacement therapy, is a pro-drug of succinate. The drug was invented in the Abliva laboratories at Lund University and is supported by a strong group of patents. NV354 is being developed for the PMD Leigh Syndrome initially with potential to expand to other PMD indications that have a dysfunctional complex I in the electron transport chain.

Additionally, Abliva has additional efforts ongoing in discovery that are focused on the regulation and stabilization of the mitochondrion's energy production.

**Leveraging Opportunities in Rare Diseases**
Abliva is continually working to take advantage of the opportunities afforded to companies working in the rare disease space. The company requested, and was granted, orphan drug designation (ODD) for KL1333 in both the US and EU. ODD is a regulatory designation that provides sponsors with a number of advantages including more regulatory assistance and scientific advice during the development process, lower development costs, attractive pricing, and market exclusivity (10 years in the EU and 7 years in the US). The outlook for reaching the market is also better than for traditional medicines[1,2].

In addition, we have sought advice from pharmaceutical regulators in the US, UK and Europe. This advice has been extremely important to the company, as is clearly demonstrated with the advice from the FDA that led us to move to a single, registrational Phase 2/3 study (versus the traditional sequential Phase 2 followed by Phase 3 design), allowing us to get to market more quickly.

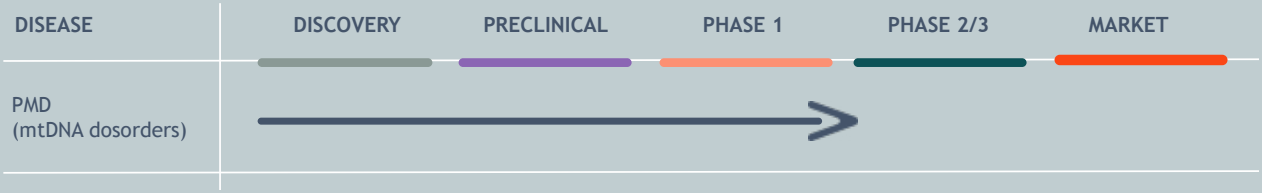
**Building a World Class Organization**
The key to the success of any company is the people who work there, and the leadership at Abliva is committed to attracting and retaining a group of bright, innovative scientists, clinicians, and drug development experts. We will continue to support development opportunities for our colleagues and ensure that they have the tools and resources available to deliver on our goals. We will continue to complement our core team with a network of specialists, physicians, advisors and others who will bring their expertise to our programs.

**Accessing Capital to Finance the Vision**
Abliva is a public company traded on NASDAQ Stockholm (ABLI, Small cap). The company appreciates the continued commitment of our shareholders and looks to attract new investors as we advance our portfolio and build the company. The investment of Hadean Ventures was the first step to bringing specialist investors into the company and the company aims to continue to attract new specialist and institutional investors across Sweden, Europe and America as the financial needs of the company increase with the KL1333 registrational study, the progression of the portfolio, and the build of a commercial organization.

1) Jayasundra et al. Orphanet J of Rare Dis. Estimating the clinical cost of drug development for orphan versus non-orphan drugs. 2019.
2) EvaluatePharma, Orphan Drug Report 2019.

# KL1333 Blockbuster candidate ready for registrational Phase 2/3 study

## IND approval enables recruitment of first patient to registrational Phase 2/3 study in 2022
## Phase 1a/b study: Positive safety results and signs of efficacy
## Orphan drug designation in both the United States and Europe




**Events in the fourth quarter**

- The US Food and Drug Administration approved Abliva's Investigational New Drug (IND) application for KL1333, enabling the start of a registrational Phase 2/3 study with first patients due to be recruited in 2022.

**Objectives for 2021**

- Complete the Phase 1a/b study and report results. ✓
- Complete the drug-drug interaction study and report results. ✓
- Preparatory activities for the Phase 2/3 study:
  – conduct a patient registry study ✓
  – conduct a validation study of endpoints ✓
  – initiate chronic toxicology studies ✓
- Initiate registrational Phase 2/3 study.

**Objectives for 2022**

- Initiate patient recruitment to registrational Phase 2/3 study.
- Initiate dosing in first patient in registrational Phase 2/3 study.

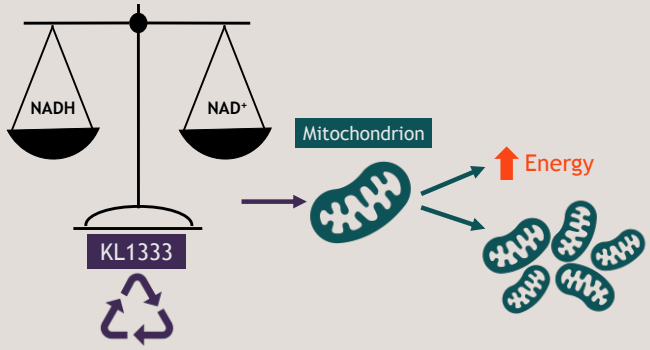
### DISEASE AREA

KL1333 is being developed as a treatment for a subset of adult primary mitochondrial disease patients suffering from multiple debilitating symptoms, including mitochondrial fatigue and myopathy. Diagnoses can include MELAS-MIDD and KSS-CPEO spectrum disorders as well as MERRF syndrome.

The drug candidate is intended for long-term oral treatment.

### PATH TO MARKET

The recommendation from the FDA to make a coherent, registrational Phase 2/3 study brings significant benefits to the KL1333 project, and Abliva's intention is to apply for market approval during 2025. The prerequisite for starting the study is dependent on financing for the entire study. The number of patients in the target group for treatment with KL1333 is approximately 40,000[1)] in Europe and the US. At typical orphan drug pricing, this translates into a blockbuster opportunity.




KL1333 has the ability to restore the balance of $NAD^+$ and NADH, and thus leads to the formation of new mitochondria and improved energy levels.

# NV354 First-in-class therapeutic heading towards clinical development

## Positive feedback from UK MHRA
## Clinical Phase 1 study planned to commence in 2022


**Events in the fourth quarter**
- During the quarter, the company has continued to assemble the regulatory documentation to support clinical entrance.

**Objectives for 2021**
- Complete preclinical pharmacology and safety studies ✓
- Produce NV354 clinical trial material for clinical studies.

Complete regulatory documentation to support clinical entrance.

**Objectives for 2022**
- Produce NV354 clinical trial material for clinical studies.
- Complete regulatory documentation to support clinical entrance.
- Start clinical Phase 1 study

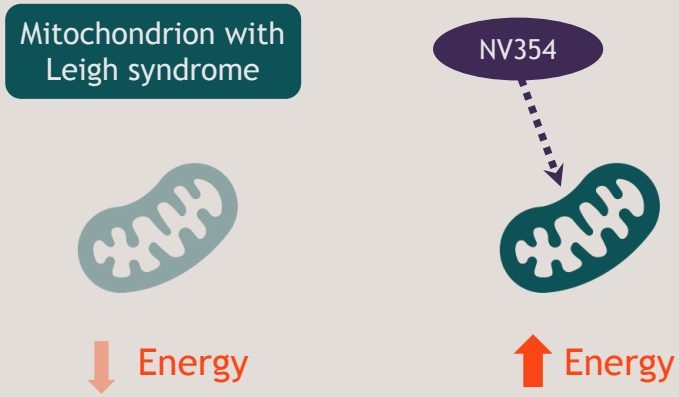
### PRIMARY INDICATION
NV354 is being developed for the treatment of Leigh syndrome, a severe primary mitochondrial disease that usually debuts at one to two years of age. The disease is fatal and children usually die before age 5.

Symptoms include developmental delay, psychomotor regression and hypotonia. There are currently no approved medicines. The drug candidate is intended for long-term oral treatment.

### EXPANSION OPPORTUNITY
The unique mechanism of action and high brain uptake may be utilized to develop NV354 for the treatment of MELAS in children and adolescents with neurological symptoms, and for the treatment of LHON. MELAS is a serious disease with symptoms such as muscle weakness, diabetes, fatigue, epilepsy, other severe neurological effects, and shortened life span. LHON is a disease that causes sudden severe permanent visual impairment and can lead to blindness on both eyes.

### PATH TO MARKET
25 per 1,000,000 children are estimated to be born with Leigh syndrome. MELAS and LHON could also be treated with NV354. There are approximately 25,000 people with LHON in Europe.[1)]

1 Gorman et al., Prevalence of Nuclear and Mitochond- rial DNA Mutations Related to Adult Mitochondrial Disease, 2015




In Leigh syndrome, the first step in energy formation does not work. NV354 restores the energy needed in the diseased mitochondria.

# Non-core asset: NeuroSTAT – For treatment of Traumatic Brain Injury

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.

**Treatment objective**

The aim for NeuroSTAT, targeting the mitochondria, is to counteract 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**

NeuroSTAT has shown favorable properties in a Phase 1b/IIa clinical study and in advanced experimental TBI models at the University of Pennsylvania (Penn). NeuroSTAT has orphan drug designation in Europe and the US as well as an IND approval and Fast Track designation for clinical development in the US.

Abliva continues in preliminary discussions with the TRACK- TBI network regarding a potential collaboration within the scope of the Precision Medicine project[1) 2)] for a Phase 2 study on traumatic brain injury with NeuroSTAT. The study, if authorized by US Department of Defense (DOD), would commence in 2022, contingent upon DOD's approval of earlier steps of the project.

With a potential agreement with TRACK-TBI as a partner, the company will review possible options that may enable developing the NeuroSTAT program further.

1 Precision Medicine grant: TRACK-TBI Precision Medicine is a DOD-funded project run by the leading traumatic brain injury (TBI) clinical trial network TRACK-TBI in the US. The aim of the project is to validate novel imaging and blood -based biomarkers for moderate/severe TBI to enable precision medicine TBI clinical trials with a focus on specific disease pathologies and enriched study populations.

2 The views expressed regarding the Precision Medicine project are those of the company/authors and may not reflect the official policy or position of the Department of the Army, Department of Defense, or the U.S. Government.

## Consolidated Statement of

# Comprehensive Income

### Revenues

The consolidated turnover during the fourth quarter of 2021 was SEK 48,000 (112,000). Other operating revenues for the fourth quarter were SEK 126,000 (1,629,000) and pertains exchange rate gains. During the full year of 2021 the consolidated turnover was 151,000 (216,000) SEK. Other operating revenues for the full year amounted 0,000 (1,628,000) SEK and related in 2020 mainly to Vinnova grant.

### Results of operations

The operating loss for the fourth quarter was SEK 36,869,000 (13,169,000) and for the full year the operating loss amounted SEK 123,482,000 (60,071,000). The net loss before tax for the fourth quarter amounted to SEK 13,067 (13,067,000). For the full year the loss before tax was SEK 123,494,000 (59,994,000).

The operating loss was affected by other external expenses, which for the full year was SEK 103,695,000 (46,072,000). Increased external costs relates primarily to preparing and start-up activities in the KL1333 project including a milestone payment to Yungjin Pharm of USD two million related to the IND-approval from American Food and Drug Administration. Expenses related to development projects, as a part of external expenses, have affected the result with SEK 29,510 (29,510,000) whereof SEK 90,690,000 (22,817,000) relates to project in clinical phase including payment to Yungjin Pharm in total SEK 18,198,000. Personnel expenses during the full year amounts to SEK 16,844,000 (13,035,000) including notice period and severance pay to former CEO of SEK 2,881,000. Other operating expenses amount to, SEK 330,000 (0,000) and pertains to exchange-rate losses.

| (SEK 000) | Note | 1 Oct, 2021<br>31 Dec, 2021 | 1 Oct, 2020<br>31 Dec, 2020 | 1 Jan, 2021<br>31 Dec, 2021 | 1 Jan, 2020<br>31 Dec, 2020 |
|---|---|---|---|---|---|
| Net sales | | 48 | 112 | 151 | 216 |
| Other operating income | | 126 | 1,629 | - | 1,648 |
| | | **174** | **1,740** | **151** | **1,864** |
| **Operating expenses** | | | | | |
| Other external expenses | | -33,314 | -11,466 | -103,695 | -46,072 |
| Personnel cost | | -2,945 | -2,799 | -16,844 | -13,305 |
| Depreciation and write-down of tangible and intangible assets | | -784 | -644 | -2,764 | -2,558 |
| Other operating expenses | | -0 | 0 | -330 | - |
| | | **-37,043** | **-14,909** | **-123,633** | **-61,935** |
| **Operating income** | | **-36,869** | **-13,169** | **-123,482** | **-60,071** |
| **Profit/loss from financial items** | | | | | |
| Result from other securities and receivables related to non current assets | | - | 107 | - | 107 |
| Financial costs | | -1 | -5 | -12 | -30 |
| | | **-1** | **102** | **-12** | **77** |
| **Profit/loss before tax** | | **-36,871** | **-13,067** | **-123,494** | **-59,994** |
| Income tax | 2 | -4 | - | -4 | - |
| **Profit/loss for the period** | | **-36,875** | **-13,067** | **-123,498** | **-59,994** |
| **Other comprehensive income** | | | | | |
| *Items that may be reclassified to profit or loss* | | | | | |
| Translation differences on foreign subsidiaries | | 61 | -2 | 71 | -3 |
| **Total comprehensive income for the period** | | **-36,814** | **-13,069** | **-123,427** | **-59,997** |
| **Loss for the period attributable to:** | | | | | |
| Parent company shareholders | | -36,870 | -13,063 | -123,492 | -59,989 |
| Non-controlling interests | | -5 | -4 | -6 | -5 |
| | | **-36,875** | **-13,067** | **-123,498** | **-59,994** |
| **Total comprehensive income for the period** | | | | | |
| Parent company shareholders | | -36,809 | -13,065 | -123,420 | -59,992 |
| Non-controlling interests | | -5 | -4 | -7 | -5 |
| | | **-36,814** | **-13,069** | **-123,427** | **-59,997** |
| Earnings per share before and after dilution(SEK) based on average number of shares | | -0.09 | -0.03 | -0.33 | -0.24 |
| Average number of shares before and after dilution | | 403,006,798 | 296,340,132 | 370,168,023 | 250,321,204 |

Consolidated Statement of

# Financial Position

## Financial position

The equity/assets ratio was 70 (90) percent as of 31 December 2021, and equity was SEK 41,528,000 (88,656,000). The equity includes funds from the in April and May completed two tranches directed share issue, which provided the company with SEK 75,868,000 after deduction of issue costs of SEK 4,132,000. Short term Liabilties amounted SEK 17,390,000 (10,209,000) as of 31 December 2021, the increase is mainly related to prepatory activities in the KL1333 phase 2/3 study. Cash and cash equivalents amounted to SEK 22,339,000 (61,643,000) as of 31 September 2021, an decrease of SEK 39,304,000 from the beginning of the year. Total assets as of 31 December 2021 were SEK 58,914,000 (98,957,000).

The board has initiated a process to ensure adequate funding to enable execution of the company's strategy with the start of a registrationable Phase 2/3 study in the company's KL1333 project during 2022. The prerequisite for starting the study is dependent on financing for the entire study.

## Financial instruments

Abliva 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 Abliva's R&D partner companies, which conducts development activities. A prudent assessment is that book value corresponds to the market value.

Other financial assets and liabilities are valued at amortized cost. The carrying amount of these assets and liabilities is estimated to correspond to fair value.

| (SEK 000) | Note | 31 Dec, 2021 | 31 Dec, 2020 |
|---|---|---|---|
| **ASSETS** | | | |
| **Non-current assets** | | | |
| **Intangible assets** | 1 | | |
| Development costs* | | - | - |
| Patents | | 20,293 | 20,971 |
| Other Intangible assets | | 1,210 | 1,344 |
| | | **21,503** | **22,315** |
| **Tangible assets** | | | |
| Equipment | | 60 | 41 |
| Rigth of use asset leases | | - | 343 |
| | | **60** | **384** |
| **Financial assets** | | | |
| Other long-term securities | | 13,101 | 13,101 |
| | | **13,101** | **13,101** |
| **Total non-current assets** | | **34,664** | **35,800** |
| **Current assets** | | | |
| Other receivables | | 912 | 928 |
| Prepaid expenses and accrued income | | 1,003 | 586 |
| Cash and cash equivalents | | 22,339 | 61,643 |
| | | **24,254** | **63,157** |
| **TOTAL ASSETS** | | **58,918** | **98,957** |

*Capitalized Development Costs

The Board of Swedish Accounting Supervision examined the Company's interim report as of September 30, 2020 and the Annual report for 2020 regarding the accounting of capitalized development costs, and referred the case to Finansinspektionen (FI). In October FI announced that they would investigate whether Abliva AB complied with the regulations for accounting in its annual and consolidated accounts for 2020. More specifically, whether Abliva AB had violated the provisions of Article 4 of the European Parliament and Council Regulation (EC) No 1606/2002 of 19 July 2002 on the application of international accounting standards, and the Annual Accounts Act (1995: 1554) regarding the accounting of development expenses as an intangible asset. As an adaptation to the The Board of Swedish Accounting Supervision's view on the handling of capitalized development costs (IAS 38), the Board has made a correction of Opening balances 1 January 2020 in Equity, in accordance with IAS 8 Accounting Policies, Changes in Accounting Estimates and Errors. The total adjustment of SEK 51,706,000 refers to accumulated capitalized development costs related to the NeuroSTAT program processed up to and including 31 March 2017..

Consolidated Statement of

# Financial Position

| (SEK 000) | Note | 31 Dec, 2021 | 31 Dec, 2020 |
|---|---|---|---|
| **EQUITY AND LIABILITIES** | | | |
| **Equity attributable to the shareholders of the parent company** | | | |
| Share capital | | 20,150 | 14,817 |
| Additional paid in capital | | 730,560 | 660,025 |
| Translation reserve | | 688 | 616 |
| Retained earnings* | | -709,879 | -586,802 |
| **Total equity attributable to the shareholders of the parent** | | **41,519** | **88,656** |
| Non-controlling interests | | 9 | 0 |
| **Total equity** | | **41,528** | **88,656** |
| **Long-term liabilities** | | | |
| Other longtrem liabilities | | - | 92 |
| | | **-** | **92** |
| **Short-term liabilities** | | | |
| Accounts payable | | 9,616 | 4,201 |
| Other liabilities | | 277 | 675 |
| Accrued expenses and deferred income | | 7,497 | 5,333 |
| | | **17,390** | **10,209** |
| **Total liabilities** | | **17,390** | **10,392** |
| **TOTAL EQUITY AND LIABILITIES** | | **58,918** | **98,957** |

*Retained earnings

For further information, see page 10, *Capitalized Development Costs.

Consolidated Statement of

# Changes in Equity

| (SEK 000) | Equity attributable to the shareholders of the parent company: Share-capital | Additional paid in capital | Translation reserve | Retained earnings | Total | Non-controlling interests | Total equity |
|---|---|---|---|---|---|---|---|
| **Opening balance, 1 January 2020** | **9,298** | **592,980** | **619** | **-475,107** | **127,791** | **5** | **127,795** |
| Correction of error pertaining to capitalized development costs*** | - | - | - | -51,706 | -51,706 | - | -51,706 |
| **Restated total equity at the beginining of the year** | **9,298** | **592,980** | **619** | **-526,813** | **76,084** | **5** | **76,089** |
| **Comprehensive profit/loss for the period** | | | | | | | |
| Profit/loss for the period | - | - | - | -59,989 | -59,989 | -5 | -59,994 |
| Other comprehensive income | | | | | | | |
| Translation differences | - | - | -3 | - | -3 | - | -3 |
| **Other comprehensive profit/loss for the period, net after tax** | **-** | **-** | **-3** | **-** | **-3** | **-** | **-3** |
| **Total comprehensive profit/loss** | **-** | **-** | **-3** | **-59,989** | **-59,992** | **-5** | **-59,997** |
| **Transactions with shareholders** | | | | | | | |
| Rights Issue* | 5,519 | 67,045 | - | - | 72,564 | - | 72,564 |
| **Total transactions with shareholders** | **5,519** | **67,045** | **-** | **-** | **72,564** | **-** | **72,564** |
| **Closing balance, 31 December 2020** | **14,817** | **660,025** | **616** | **-586,802** | **88,656** | **0** | **88,656** |
| | | | | | | | |
| **Opening balance, 1 January 2021** | **14,817** | **660,025** | **616** | **-586,802** | **88,656** | **0** | **88,656** |
| **Comprehensive profit/loss for the period** | | | | | | | |
| Profit/loss for the period | - | - | - | -123,492 | -123,492 | -6 | -123,498 |
| Other comprehensive income | | | | | | | |
| Translation differences | - | - | 72 | - | 72 | -1 | 71 |
| **Other comprehensive profit/loss for the period, net after tax** | **-** | **-** | **72** | **-** | **72** | **-1** | **71** |
| **Total comprehensive profit/loss** | **-** | **-** | **72** | **-123,492** | **-123,420** | **-7** | **-123,427** |
| **Transactions with shareholders** | | | | | | | |
| Rights Issue* | 5,333 | 70,534 | - | - | 75,868 | - | 75,868 |
| Share-based payment | - | - | - | 415 | 415 | - | 415 |
| Shareholder contribution | - | - | - | - | - | 16 | 16 |
| **Total transactions with shareholders** | **5,333** | **70,534** | **-** | **415** | **76,283** | **16** | **76,299** |
| **Closing balance, 31 December 2021** | **20,150** | **730,560** | **688** | **-709,879** | **41,519** | **9** | **41,528** |

*Total equity includes funds from the April 6, 2021 and May 4th completed directed share issue with SEK 75,868,000 less expenses SEK 4,132,000.

* The adjustment pertains to development costs, for further information, see page 10, *Capitalized Development Costs.

## Consolidated Statement of

# Cash Flows

### Cash flow and investments

Operating cash flow for the fourth quarter was SEK -40,778,000 (-11,154,000). For the full year the operating cash flow amounted SEK -114,075,000 (-67,558,000). The increase is due to increased activities in the KL1333 program as well as the milestone payment under the in-licensing contract with Yungjin Pharm. The cash flow effect related to investments in intangibles equals SEK-1,089,000 (-1,407,000) for the full year. Cash flow for the fourth quarter equals SEK -40,990 (-11,367,000). Cashflow for the full year equals SEK -39,372,000 (3,330,000).

| (SEK 000) | 1 Oct, 2021<br>31 Dec, 2021 | 1 Oct, 2020<br>31 Dec, 2020 | 1 Jan, 2021<br>31 Dec, 2021 | 1 Jan, 2020<br>31 Dec, 2020 |
|---|---|---|---|---|
| **Cash flow from operating activities** | | | | |
| Operating income | -36,869 | -13,169 | -123,482 | -60,071 |
| | | | | |
| **Adjustments for non-cash items:** | | | | |
| Depreciation | 680 | 644 | 2,660 | 2,558 |
| Currency differences on intercompany items | -13 | - | -7 | - |
| Impaired Value | 104 | - | 104 | - |
| Share-based payments | - | - | 415 | - |
| Result from other securities and receivables related to non current assets | - | - | - | 107 |
| Interest received | - | - | - | - |
| Interest paid | -1 | -5 | -12 | -30 |
| Paid taxes | | | | |
| **Net cash from operating activities before changes in working capital** | **-35,688** | **-12,530** | **-120,326** | **-57,436** |
| | | | | |
| **Changes in working capital** | | | | |
| Increase/decrease of other current assets | -83 | -86 | -400 | 86 |
| Increase/decrease of other short-term liabilities | -5,007 | 1,462 | 6,651 | -10,208 |
| **Changes in working capital** | **-5,090** | **1,376** | **6,251** | **-10,122** |
| | | | | |
| **Cash flow from operating activities** | **-40,778** | **-11,154** | **-114,075** | **-67,558** |
| | | | | |
| **Investing activities** | | | | |
| Acquisition of intangible assets | -182 | -320 | -1,024 | -1,407 |
| Acquisition of tangible assets | -w | - | -65 | - |
| **Cash flow from investing activities** | **-182** | **-320** | **-1,089** | **-1,407** |
| | | | | |
| **Financing activities** | | | | |
| Shareholder contribution subsidiary | 2 | - | 16 | - |
| New share issue | -32 | - | 75,868 | 72,564 |
| Amoritization lease | - | - | -92 | -269 |
| **Cash flow from financing activities** | **-30** | **-** | **75,792** | **72,295** |
| | | | | |
| Cash flow for the period | -40,990 | -11,367 | -39,372 | 3,330 |
| Cash and cash equivalents at the beginning of the period | 63,267 | 73,013 | 61,643 | 58,319 |
| Effect of exchange rate changes on cash | 62 | -4 | 68 | -6 |
| **Cash and cash equivalents at end of period** | **22,339** | **61,643** | **22,339** | **61,643** |

Parent Company

# Income Statement

**Parental company**

Company earnings after tax for the fourth quarter amounts to SEK -39,727,000 (-13,043,000). Earnings after tax for the full year amounts to SEK -123,072,000 (-59,961,000). Most of the Group's operations are conducted within the parent company. Accordingly, no further specific information regarding the parent company is presented.

| (SEK 000) | Note | 1 Oct, 2021 31 Dec, 2021 | 1 Oct, 2020 31 Dec, 2020 | 1 Jan, 2021 31 Dec, 2021 | 1 Jan, 2020 31 Dec, 2020 |
|---|---|---|---|---|---|
| Net sales | | 48 | 112 | 151 | 216 |
| Other operating income | | 0 | 1,603 | 0 | 1,648 |
| | | **48** | **1,715** | **151** | **1,864** |
| **Operating expenses** | | | | | |
| Other external expenses | | -36,999 | -11,508 | -107,521 | -46,411 |
| Personnel cost | | -2,203 | -2,799 | -12,952 | -13,305 |
| Depreciation and write-down of tangible and intangible assets | | -698 | -558 | -2,420 | -2,215 |
| Other operating expenses | | 126 | - | -330 | - |
| | | **-39,775** | **-14,865** | **-123,223** | **-61,931** |
| **Operating income** | | **-39,727** | **-13,150** | **-123,072** | **-60,067** |
| **Profit/loss from financial items** | | | | | |
| Result from other securities and receivables related to non current assets | | - | 107 | - | 107 |
| Interest expenses and other similar loss items | | - | 0 | - | -1 |
| | | **-** | **107** | **-** | **106** |
| **Profit/loss before tax** | | **-39,727** | **-13,043** | **-123,072** | **-59,961** |
| Income tax | 2 | - | - | - | - |
| **Profit/loss for the period** | | **-39,727** | **-13,043** | **-123,072** | **-59,961** |

Parent Company

# Statement of Comprehensive Income

| (SEK 000) | Note | 1 Oct, 2021 31 Dec, 2021 | 1 Oct, 2020 31 Dec, 2020 | 1 Jan, 2021 31 Dec, 2021 | 1 Jan, 2020 31 Dec, 2020 |
|---|---|---|---|---|---|
| Profit/loss for the period | | -39,727 | -13,043 | -123,072 | -59,961 |
| Other comprehensive income | | - | - | - | - |
| **Total comprehensive profit/loss for the period** | | **-39,727** | **-13,043** | **-123,072** | **-59,961** |

Parent Company

# Balance Sheet

| (SEK 000) | Note | 31 Dec, 2021 | 31 Dec, 2020 |
|---|---|---|---|
| **ASSETS** | | | |
| **Non-current assets** | | | |
| *Intangible assets* | 1 | | |
| Development costs* | | - | - |
| Patents | | 20,293 | 20,971 |
| Other intangible assets | | 1,210 | 1,344 |
| | | **21,503** | **22,315** |
| **Tangible assets** | | | |
| Equipment | | 60 | 41 |
| | | **60** | **41** |
| **Financial assets** | | | |
| Other long-term placement | | 13,101 | 13,101 |
| Shares in subsidiaries | 3 | 24,557 | 23,625 |
| | | **37,658** | **36,726** |
| **Total non-current assets** | | **59,222** | **59,082** |
| **Current assets** | | | |
| *Short term receivables* | | | |
| Other receivables | | 890 | 926 |
| Prepaid expenses and accrued income | | 1,003 | 585 |
| | | 1,893 | 1,511 |
| Cash and bank balances | | 21,696 | 61,634 |
| **Total current assets** | | **23,589** | **63,145** |
| **TOTAL ASSETS** | | **82,810** | **122,226** |

*Development costs.

The Board of Swedish Accounting Supervision examined the Company's interim report as of September 30, 2020 and the Annual report for 2020 regarding the accounting of capitalized development costs, and referred the case to Finansinspektionen (FI). In October FI announced that they would investigate whether Abliva AB complied with the regulations for accounting in its annual and consolidated accounts for 2020. More specifically, whether Abliva AB had violated the provisions of Article 4 of the European Parliament and Council Regulation (EC) No 1606/2002 of 19 July 2002 on the application of international accounting standards, and the Annual Accounts Act (1995: 1554) regarding the accounting of development expenses as an intangible asset. As an adaptation to the The Board of Swedish Accounting Supervision's view on the handling of capitalized development costs (IAS 38), the Board has made a correction of Opening balances 1 January 2020 in Equity, in accordance with IAS 8 Accounting Policies, Changes in Accounting Estimates and Errors. The total adjustment of SEK 51,706,000 refers to accumulated capitalized development costs related to the NeuroSTAT program processed up to and including 31 March 2017.

Parent Company

# Balance Sheet

| (SEK 000) | Note | 31 Dec, 2021 | 31 Dec, 2020 |
|---|---|---|---|
| **EQUITY AND LIABILITIES** | | | |
| **Equity** | | | |
| *Restricted equity* | | | |
| Share capital | | 20,150 | 14,817 |
| Statutory reserve | | 1,856 | 1,856 |
| Development expenditure reserve** | | 2,613 | 3,821 |
| | | **24,619** | **20,494** |
| **Unrestricted equity** | | | |
| Share premium reserve | | 70,534 | 67,045 |
| Retained earnings* | | 93,017 | 84,725 |
| Profit/loss for the period | | -123,072 | -59,961 |
| | | **40,479** | **91,809** |
| **Total equity** | | **65,098** | **112,302** |
| | | | - |
| **Short-term liabilities** | | | |
| Accounts payable | | 9,616 | 4,201 |
| Liabilities subsidiary | | 1,253 | |
| Other liabilities | | 273 | 406 |
| Accrued expenses and deferred income | | 6,570 | 5,317 |
| | | **17,713** | **9,924** |
| **TOTAL EQUITY AND LIABILITIES** | | **82,810** | **122,226** |

*Retained earnings

For further information, see page 15, *Development costs.

**Development expenditure reserve.

As a consequence of restroactive adjustments related to capitalized development costs, an adjustment of the Development expenditure reserve has been implemented in the Parent company of in total SEK 9,755,000 regarding accumulated capitalized development costs, which means that the new opening balance Development expenditure reserve as of January 1, 2020 amounts to SEK 4,351,000. Opening balance 1 January 2020 before adjustment was 14,106,000. The adjustment has been recorded against retained earnings and the net effect on equity is zero.

# Notes

## Note 1 — Intangible assets

| (SEK 000) | Development costs | Patents | Other | Total |
|---|---|---|---|---|
| **ACCUMULATED COST** | | | | |
| **Opening balance 1 Jan. 2021** | **-** | **33,771** | **2,864** | **36,635** |
| Additions | - | 1,561 | - | 1,561 |
| Impaired value | - | -153 | - | -153 |
| **Closing balance 31 Dec. 2021** | **-** | **35,179** | **2,864** | **38,043** |
| | | | | |
| **ACCUMULATED DEPRECIATION** | | | | |
| **Opening balance 1 Jan. 2021** | **-** | **-12,800** | **-1,519** | **-14,319** |
| Depreciation for the period | - | -2,136 | -135 | -2,271 |
| Impaired value | - | 49 | - | 49 |
| **Closing balance 31 Dec. 2021** | **-** | **-14,887** | **-1,654** | **-16,541** |
| **Residual value 31 Dec. 2021** | **-** | **20,293** | **1,210** | **21,503** |

| (SEK 000) | Development costs | Patents | Other | Total |
|---|---|---|---|---|
| **ACCUMULATED COST** | | | | |
| **Opening balance 1 Jan. 2020** | **51,706** | **32,279** | **2,864** | **86,849** |
| Correction of error pertaining to capitalized development costs* | -51,706 | - | - | -51,706 |
| **Restated Opening Balance 1 Jan 2020** | **-** | **32,279** | **2,864** | **35,143** |
| Additions | - | 1,492 | - | 1,492 |
| **Closing balance 31 Dec. 2020** | **-** | **33,771** | **2,864** | **36,635** |
| | | | | |
| **ACCUMULATED DEPRECIATION** | | | | |
| **Opening balance 1 Jan. 2020** | **-** | **-10,778** | **-1,385** | **-12,163** |
| Correction of error pertaining to capitalized development costs* | - | - | - | - |
| **Restated Opening Balance 1 Jan 2020** | **-** | **-10,778** | **-1,385** | **-12,163** |
| | | | | |
| Depreciation for the period | - | -2,022 | -134 | -2,156 |
| Closing balance 31 Dec. 2020 | - | -12,800 | -1,519 | -14,319 |
| **Residual value 31 Dec. 2020** | **-** | **20,971** | **1,345** | **22,315** |

## Note 2 – Tax

The group's total loss carry-forwards amounts to SEK 746,806,000 as of 31 December 2021 (619,183,000). The parent company's total loss carry-forwards amounts to SEK 720,526,000 as of 31 December 2021 (593,525,000). Because the company is loss making, management cannot judge when deductible loss carry-forwards will be utilized.

## Note 3 – Shares and participations in group companies

These shares are the holding of 82.47% in the subsidiary NeuroVive Pharmaceutical Asia Ltd., domiciled in Hong Kong, the holly owned american subsidiary Abliva Inc., registered in March 2021 and the Swedish subsidiary Abliva Incentive AB, registered in May 2021, holding option program for the CEO.

*Retroactive adjustment of capitalized development costs.

For further information, see page 10, Capitalized Development Costs.

# Other disclosures

### Transactions with related parties

Transactions between the company and its subsidiarie, which are related parties to the company, have been eliminated on consolidation, and accordingly, no disclosures are made regarding these transactions.

| (SEK 000) | 1 Jan.2021-31 Dec. 2021 | 1 Jan.2020-31 Dec. 2020 |
|---|---|---|
| Eskil Elmér, CSO | 5 | 6 |
| Magnus Hansson, CMO | 3 | 4 |
| **Total** | **8** | **10** |

Compensation based on sales 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. During the period no other transactions with related parties have occured.

### 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.

### Human resources

The average number of employees of the group for the period January to December 2021 was 9 (9), of which 7(5) are women.

### Important events during 2021

For further information, see page 2.

### Important events after the reporting period

For further information, see page 2.

### Incentive programs/share warrants

The AGM on May 20, 2021, decided on a four-year incentive stock option program 2021/2025 for the Company's CEO. The incentive stock option program entitles the holder to a new ordinary share in Abliva AB up to a maximum of 4,600,000 ordinary shares. The redemption price amounts to 0.725 öre. The program is vested at 25% per year on June 1, 2022, June 1, 2023, June 1, 2024 and June 1, 2025. Latest redemption date is December 31, 2025.

### Audit review

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

### Upcoming financial statements

| | |
|---|---|
| Annual Report | The week beginning on 28 March 2022 |
| Q1 Report January-March 2022 | May 20, 2022 |
| Q2 Report January-June 2022 | August 19, 2022 |
| Q3 Report January-September 2022 | November 22, 2022 |
| Year-End Report 2022 | February 21, 2023 |

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

### Annual General Meeting 2022

Abliva's Annual General Meeting will be held at Medicon Village, Scheeletorget 1, in Lund, on Wednesday 27 April 2022 at 4 p.m.

## Risks and uncertainty factors

A research company such as Abliva 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..

**Financing**
The Board continuously monitors and evaluates the company's funding need and financial position. The main project KL1333 is about to start a registrational Phase 2/3 study. The prerequisite for starting the study is dependent on financing for the entire study. If the company does not succeed in funding the KL1333 program there is a risk for a delay in the start of the program.

**Impact of COVID-19 on the Company's clinical trials**
COVID-19 may, among other things, lead to delays in the Company's clinical studies, but it is currently difficult to assess all the potential effects that COVID-19 may have on the Company. Due to COVID-19, there is a risk of further delays because healthcare authorities and healthcare providers re-prioritize available resources, care locations and healthcare professionals to better meet the influx of COVID-19 patients. There is  a risk that the start of the upcoming Phase 2/3 study, which is expected to begin in 2022, will be further delayed due to COVID-19. The company's second drug candidate NV354, which is being prepared for a Phase 1 study in 2022, also risks being delayed due to the COVID-19 pandemic.

Abliva is not involved in any disputes.

For more detail of risks and uncertainty factors, refer to the Statutory Administration Report in the Annual Report 2020 and the prospectus published April 30, 2021.

## Principles of preparation of the Interim Report

Abliva 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.

IFRS 2 Share-based Payment applies to incentive programs that are regulated with equity instruments. The fair value of employee stock options is calculated according to Black & Scholes' valuation model at the time the options are granted. The cost, which is distributed over the vesting period of four years, is reported against equity. No costs for social security contributions are expected to occur.

The group and parent company have otherwise applied the accounting principles described in the Annual Report for 2020 on pages 46-61.

# Definitions alternative performance measures

Alternative Performance Measures (APM) are key figures not defined in financial reports prepared according to IFRS.
Of the below key figures, only the key figure Earnings per share before and after dilution is mandatory and defined according to IFRS.
Of the other key figures, net sales, earnings per share before and after dilution, cash flow from operating activities and cash flow for the period are defined according to IFRS.

| The following key figures are used: | Definition | Reason for use |
|---|---|---|
| Net revenues | Revenue from goods and services sold that are part of the company's normal operations | |
| Other operating income | Income from secondary activities in ordinary activities such as grants received | |
| Operating income | Net sales and other revenues minus expenses for other external costs, personnel costs, depreciation and impairment and other expenses | Measures the result in the operations |
| Profit/loss before tax | Operating income after profit/loss from finacial items and allocations | Measures the result in the business after profit/loss from financial items and allocations |
| Earnings per share before dilution(SEK) based on average number of shares | Profit/loss for the period divided by average number of shares before dilution at the end of the period | |
| Earnings per share after dilution(SEK) based on average number of shares | Profit/loss for the period divided by average number of shares after dilution at the end of the period | |
| Cash flow from operating activities | Cash flow from operating activities, including cash flow from working capital, ie changes in current liabilities and current receivables | Measures total cash flow generated in the business |
| Cash flow for the period | The company's total cash flow from operating activities, investment activities and financing activities | Measures total cash flow generated in the business including investment activities and financing activities |
| Average number of shares before and after dilution | Average number of shares before and after dilution | Measures the average number of shares during the period before and after dilution. As the Group's earnings are negative, there is no dilution |
| Equity Ratio % | Equity as a percentage of total assets | Shows how much of the company's assets are financed with equity and shows the company's ability to pay |
| Liquidity Ratio (%) | Current assets divided by current liabilities | Shows on the company's short-term ability to pay |

# 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, 22 February, 2022

**David Laskow-Pooley**
Chairman of the Board

**David Bejker**
Board member

**Roger Franklin**
Board member

**Denise Goode**
Board member

**Jan Törnell**
Board member

**Ellen Donnelly**
Chief Executive Officer


David Laskow-Pooley


David Bejker


Roger Franklin


Denise Goode


Jan Törnell


Ellen Donnelly

For more information concerning this report, please contact CEO Ellen Donnelly. 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 22 February, 2022.

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.

# Glossary

**Active compound.** A pharmaceutical active ingredient in a pharmaceutical product.
**Candidate drug.** A particular compound which is selected during the preclinical phase. The candidate drug is subsequently tested in humans in clinical studies.
**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 1, Phase 2, Phase 3. Phase 2 is usually divided into an early phase (Phase 2a) and a later phase (Phase 2b). See also "phase (1,2 and 3)".
**Drug-drug interaction study.** A clinical study in healthy volunteers to investigate the drug-drug interactions when co-administering a (candidate) drug with other drugs. Drug-drug interactions can lead to changed systemic exposure, resulting in variations in drug response of the co-administered drugs.
**Fatigue.** Extreme tiredness. Often includes muscle fatigue with exercise intolerance.
**FDA.** The United States Federal Food and Drug Administration.
**Hypotonia.** An abnormally low level of tension, important for posture, in the resting muscle.
**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.** (Myoclonic epilepsy with ragged-red fibers). Primary mitochondrial disease with symptoms such as epilepsy, involuntary muscle twitching and difficulty coordinating muscle movements, but the disease can affect many functions. When examined under a microscope, muscle tissue has characteristic changes.
**MHRA.** The UK Medicines and Healthcare products Regulatory Agency.
**MIDD.** Maternally Inherited Diabetes and Deafness
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.** Primary mitochondrial disease which affects the muscles.
**$NAD^+$/NADH.** A coenzyme involved in metabolism. $NAD^+$ and NADH have central roles in cell- and mitochondrial metabolism and energy production.
**NAFLD.** Non-Alcoholic Fatty Liver Disease.
**NASH.** Non-alcoholic steatohepatitis, inflammatory fatty liver disease.
**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).
**PEO/CPEO.** Mitochondrial disease. Progressive External Ophthalmoplegia/Chronic Progressive External Ophthalmoplegia.
**Pharmacokinetics.** Describes how the body affects a specific drug after administration.
**Phase (1,2 and 3).** The various stages of trials on the efficacy of a pharmaceutical in humans. See also "clinical trial." Phase 1 examines the safety on healthy human subjects, Phase 2 examines efficacy in patients with the relevant disease and Phase 3 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 2 is often divided between Phase 2a and Phase 2b.
**Preclinical.** That stage of drug development that occurs before a candidate drug is trialed on humans.
**Primary mitochondrial diseases.** Metabolic diseases that affect the ability of cells to convert energy. An estimated 12 in every 100,000 people affected. Often present in early childhood and lead to severe symptoms, such as mental retardation, heart failure and rhythm disturbances, dementia, movement disorders, severe diabetes, stroke-like episodes, deafness, blindness, limited mobility of the eyes, vomiting and seizures.
**Psychomotor regression.** When the development of the ability to perform will-driven movements is initially normal but deteriorates during infancy or early childhood.
**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 Abliva

Abliva develops medicines for the treatment of primary mitochondrial diseases. These rare and often very severe diseases occur when the cell's energy provider, the mitochondria, do not function properly. The company is focused on two projects. KL1333, a powerful $NAD^+$ regulator, is in clinical development and has been granted orphan drug designation in Europe and the US. NV354, an energy replacement (succinate) therapy, will enter the clinic in 2022. Abliva is based in Lund, Sweden.

**What is primary mitochondrial disease?**

Primary mitochondrial disorders are metabolic diseases that affect the cells' ability to convert energy. The diseases can manifest very differently depending on the organs affected. They have historically been viewed as clinical syndromes and more recently as disease spectra, caused by genetic defects affecting mitochondrial function. It is estimated that 125 persons per million have a primary mitochondrial disease.

Abliva's discovery projects focus on deeper understanding of the mechanisms for our unique chemistry platforms, and the development of next-generation compounds for primary mitochondrial diseases.

**Stock exchange**

Abliva is listed on Nasdaq Stockholm, Sweden (ticker: ABLI).

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