



## Interim Report

January - June 2021

A photograph of three young women with long hair, smiling and looking towards the camera. They are outdoors, with a bright, warm light source (likely the sun) behind them, creating a soft, golden glow and some lens flare. The woman in the foreground is wearing a white lace top, while the others are in darker clothing.

The first signs of efficacy seen  
in patients in our Phase 1b  
study strengthens KL1333.

Ellen Donnelly, CEO

Delivering mitochondrial health

# Second quarter summary

Phase 1a/b study: **positive placebo-controlled safety data for KL1333 and the first evidence of efficacy**

## Important events second quarter (Apr - Jun 2021)

- Data from the Phase 1a/b clinical study of KL1333 was released and confirmed the safety and pharmacokinetic profile of the drug. In addition, in a cohort of eight patients (six dosed with KL1333, two with placebo), there were signs of efficacy across well-established relevant clinical endpoints including two patient-reported fatigue endpoints and a functional endpoint.
- Extraordinary General Meeting was held on 29 April 2021.
- The directed share issue, approved by the Extraordinary General Meeting, was completed. The Company raised approx. SEK 76 million after deduction of issue costs.
- Annual General Meeting was held on 20 May 2021.

## Important events after the reporting period

- The validation study of fatigue as an endpoint, for the KL1333 Phase 2/3 study, was completed.



## Financial information

### April-June 2021\*

- Net revenues: SEK 18,000 (97,000)
- Other operating income: SEK 0,000 (34,000)
- Loss before tax: SEK 30,314,000 (20,312,000)
- Loss per share: SEK 0.08 (0.09)
- Diluted loss per share: SEK 0.08 (0.09)

### January-June 2021\*

- Net revenues: SEK 18,000 (105,000)
- Other operating income: SEK 0,000 (34,000)
- Loss before tax: SEK 51,770,000 (36,849,000)
- Loss per share: SEK 0.13 (0.18)
- Diluted loss per share: SEK 0.13 (0.18)

\* APM Alternative performance measures, see definition on page 20.



# Registrational study towards regulatory approval

The second quarter (Q2) was a defining quarter for the company with the release of important safety and efficacy data for KL1333 in both healthy volunteers and Primary Mitochondrial Disease (PMD) patients. The completion of this Phase 1a/b study, and a second study examining the effects of KL1333 when administered with common therapeutics, are important as we work to assemble key documentation to enable the KL1333 Phase 2/3 study start. The KL1333 program has been de-risked substantially in Q2 and we remain optimistic that we will be able to complete a large round of financing to enable the start of the study later this year.

## Efficacy and safety in patients

The release of data from our Phase 1a/b study in May was a key milestone for the company, providing the first indications of efficacy for KL1333 in PMD patients. In this PMD patient cohort, patients dosed with KL1333 had both functional improvements and improvements in their fatigue over the ten days of treatment, something that was not observed in the patients dosed with placebo. This data was strengthened by the fact that we saw a correlation between exposure and efficacy, with patients with higher levels of KL1333 in their blood doing better in the assessments. In addition, we were pleased to see evidence that KL1333 is modulating lactate and pyruvate, key molecules in the target pathway.

Another important study for KL1333 read out in the second quarter. This study, called a drug-drug interaction study, was requested by the FDA to confirm that KL1333 does not interact with medicines commonly used in PMD patients. We were pleased that the study showed no concerning drug-drug interactions, a fact that should facilitate patient recruitment in our upcoming study.

The data from these two studies is critically important for the company as they provide strong data on both the efficacy and safety of KL1333 in PMD patients, something that will increase

confidence of patients as they consider entering our upcoming study.

## Using Patient Data to finalize Phase 2/3 trial Design

The data from the Phase 1a/b study indicated, in two independent endpoints, that KL1333 treatment may improve fatigue in PMD patients. It also provided signs that KL1333 may improve muscle function and endurance in these patients. This is important information as we will now expand our endpoints in the Phase 2/3 study to evaluate muscle function in addition to fatigue, providing two important ways to help this patient population – and two ‘shots on goal’ in the upcoming study.

*“The data from these two studies is critically important for the company as they provide strong data on both the efficacy and safety of KL1333 in PMD patients, something that will increase confidence of patients as they consider entering our upcoming study”*

The work has continued during the summer with an FDA (U.S. Food and Drug Administration) meeting to discuss our primary endpoint. We also completed our fatigue endpoint validation study and an important long-term toxicological study. These

activities all further de-risk our program as we prepare to file our clinical trial application.

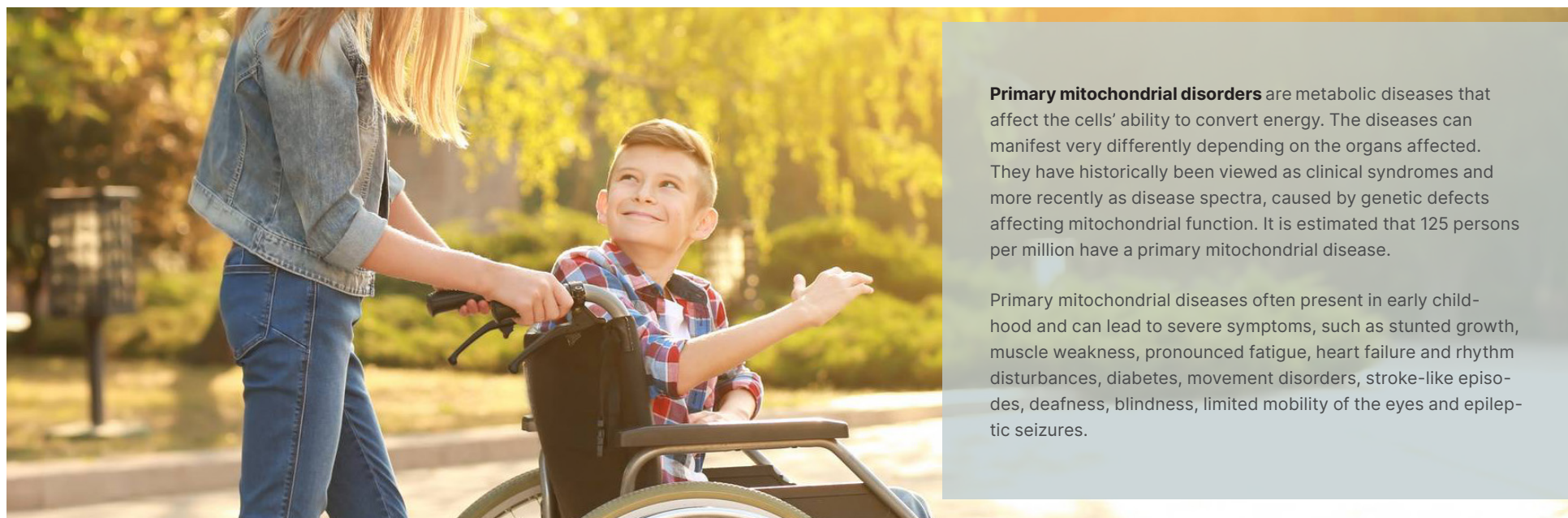
## Financing the KL1333 Study

In the second quarter we held an extraordinary general meeting and successfully closed the directed share issue initiated in March. This financing was critical as it provided the funds necessary to commence study start up activities. We are continuing to reach out to international investors with a view to finance the Phase 2/3 program.

## Going Forward: Pushing to the Phase 2/3 clinical study

The first half of the year was successful in that we demonstrated that we can deliver on our two most important tasks – we can raise money and we can deliver on our clinical commitments. During the second half of the year, we will work to do this again, but we will do it on a much larger scale. We remain excited about the days ahead and look forward to dosing our first patient in this large global study.

**Ellen Donnelly**  
CEO



PROJECT (partner)	DISEASE	DISCOVERY	PRECLINICAL	PHASE 1	PHASE 2/3	MARKET
KL1333* (Yungjin)	PMD (MELAS-MIDD, KSS-CPEO, MERRF)					
NV354	PMD (Leigh syndrome)					
Early programs	PMD					

\*Orphan drug designation in the US and Europe

# Strategic Focus: Primary Mitochondrial Diseases (PMD)

**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 will 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 for patients. 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<sup>+</sup> 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 late 2021. 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 reg-

ulatory 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<sup>1,2</sup>.

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 2020 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 in the company in 2020 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.

# KL1333 Blockbuster candidate heading to registrational Phase 2/3 study

Phase 1a/b study: Positive safety results and signs of efficacy  
 Registrational Phase 2/3 study planned to start during late 2021  
 Orphan drug designation in both the United States and Europe



## Events in the second quarter

- Data from the Phase 1a/b clinical study was released and confirmed the safety and pharmacokinetic profile of the drug. In addition, in a cohort of eight patients (six dosed with KL1333, two with placebo), there were signs of efficacy across well-established relevant clinical endpoints including two patient-reported fatigue endpoints and a functional endpoint.
- The drug-drug interaction study was completed.

## Events after the end of the period

- The first long-term toxicological study (six months) was completed.
- The validation study of fatigue as an endpoint was completed.

## 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 long-term toxicological studies ✓
- Initiate registrational Phase 2/3 study.

## DISEASE AREA

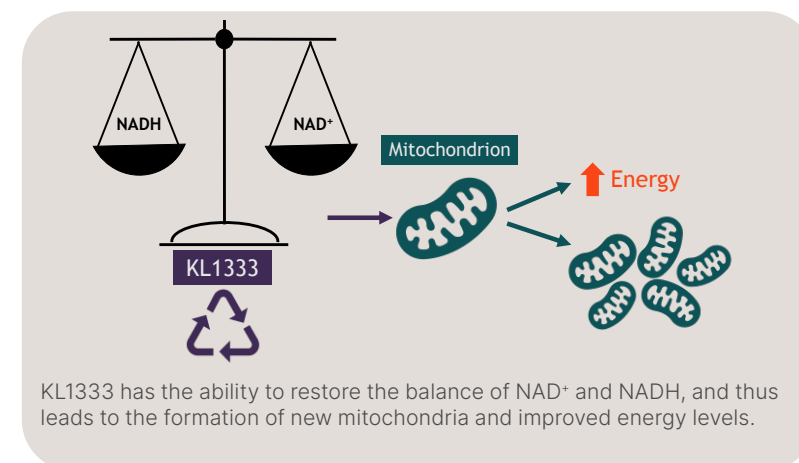
KL1333 is being developed towards a treatment for a subset of adult primary mitochondrial disease patients suffering from multiple debilitating symptoms, including chronic fatigue and myopathy. Diagnoses can include MELAS-MIDD and KSS-CPEO spectrum disorders as well as MERRF syndrome.

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 2024. The number of patients in the target group for treatment with KL1333 is approximately 40,000<sup>1)</sup> in Europe and the US. At typical orphan drug pricing, this translates into a blockbuster opportunity.

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



# NV354

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

### Finalizing pharmacology studies Preparing for healthy volunteer studies



#### Events in the first quarter

- During the quarter, the preclinical safety studies were completed and evaluation is ongoing. In parallel, the preclinical pharmacology studies continue and the company is preparing for the clinical program.

#### Objectives for 2021

- Complete preclinical pharmacology and safety studies
- Produce NV354 clinical trial material for clinical studies.
- Complete regulatory documentation to support clinical entrance.

#### 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.

#### 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.<sup>1)</sup>

#### 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.

#### Mitochondrion with Leigh syndrome



↓ Energy

NV354



↑ Energy

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

<sup>1</sup> Gorman et al., Prevalence of Nuclear and Mitochondrial DNA Mutations Related to Adult Mitochondrial Disease, 2015

## Non-core asset

**The company is seeking a strategic partner for the continued development of NeuroSTAT. It has initiated preliminary discussions with the TRACK-TBI network on a potential collaboration for a Phase 2 traumatic brain injury study with NeuroSTAT under the Precision Medicine project<sup>1) 2)</sup> funded by the U.S. Department of Defense.**

### ■ NEUROSTAT – FOR TREATMENT OF 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.

#### 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<sup>1) 2)</sup> for a Phase 2 study on traumatic brain injury with NeuroSTAT. The study, if authorized by US Department of Defence (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 second quarter of 2021 was SEK 18,000 (97,000) and relates to sales to Oroboros. Other operating revenues for the second quarter were SEK 0,000 (34,000), last year pertained to compensation for sick pay. During the first six months of 2021 the consolidated turnover was KSEK 18,000 (105,000). Other operating revenues for the first six months amounted SEK 0,000 (34,000).

## Results of operations

The operating loss for the second quarter was SEK 30,311,000 (20,305,000) and for the first six months the operating loss amounted SEK -51,762,000 (36,832,000). The net loss before tax for the second quarter amounted to KSEK 30,314,000 (20,312,000). For the first six months the loss before tax was -51,770,000 (-36,849,000).

The operating loss was affected by other external expenses, which for the first six months were SEK 39,694,000 (27,846,000). Expenses related to development projects, as a part of external expenses, have affected the result with SEK 33,030,000 (17,591,000) whereof SEK 28,736,000 (13,048,000) relates to project in clinical phase. Personnel expenses during the first six months amounts to SEK 10,449,000 (7,783,000) including notice period and severance pay to former CEO of SEK 2,881,000. Other operating expenses amount to, SEK 270,000 (78,000) and pertain to exchange-rate losses.

(SEK 000)	Note	1 Apr, 2021 30 Jun, 2021	1 Apr, 2020 30 Jun, 2020	1 Jan, 2021 30 Jun, 2021	1 Jan, 2020 30 Jun, 2020	1 Jan, 2020 31 Dec, 2020
Net sales		18	97	18	105	216
Other operating income		0	34	0	34	1,648
		<b>18</b>	<b>130</b>	<b>18</b>	<b>138</b>	<b>1,864</b>
<b>Operating expenses</b>						
Other external expenses		-24,154	-15,889	-39,694	-27,846	-46,072
Personnel cost		-5,453	-4,233	-10,499	-7,783	-13,305
Depreciation and write-down of tangible and intangible assets		-659	-636	-1,317	-1,263	-2,558
Other operating expenses		-63	324	-270	-78	-
		<b>-30,329</b>	<b>-20,435</b>	<b>-51,780</b>	<b>-36,971</b>	<b>-61,935</b>
<b>Operating income</b>		<b>-30,311</b>	<b>-20,305</b>	<b>-51,762</b>	<b>-36,832</b>	<b>-60,071</b>
<b>Profit/loss from financial items</b>						
Result from other securities and receivables related to non current assets		-	-	-	-	107
Financial income		-	-	-	-	-
Financial costs		-3	-8	-8	-17	-30
		<b>-3</b>	<b>-8</b>	<b>-8</b>	<b>-17</b>	<b>77</b>
<b>Profit/loss before tax</b>		<b>-30,314</b>	<b>-20,312</b>	<b>-51,770</b>	<b>-36,849</b>	<b>-59,994</b>
Income tax	2	-	-	-	-	-
<b>Profit/loss for the period</b>		<b>-30,314</b>	<b>-20,312</b>	<b>-51,770</b>	<b>-36,849</b>	<b>-59,994</b>
<b>Other comprehensive income</b>						
<i>Items that may be reclassified to profit or loss</i>						
Translation differences on foreign subsidiaries		-4	-3	-3	-	-3
<b>Total comprehensive income for the period</b>		<b>-30,318</b>	<b>-20,315</b>	<b>-51,772</b>	<b>-36,850</b>	<b>-59,997</b>
<b>Loss for the period attributable to:</b>						
Parent company shareholders		-30,314	-20,312	-51,769	-36,848	-59,989
Non-controlling interests		-1	-	-1	-1	-5
		<b>-30,314</b>	<b>-20,312</b>	<b>-51,770</b>	<b>-36,849</b>	<b>-59,994</b>
<b>Total comprehensive income for the period</b>						
Parent company shareholders		-30,317	-20,314	-51,771	-36,849	-59,992
Non-controlling interests		-1	-	-1	-1	-5
		<b>-30,318</b>	<b>-20,315</b>	<b>-51,772</b>	<b>-36,850</b>	<b>-59,997</b>
Earnings per share before and after dilution(SEK) based on average number of shares		-0.08	-0.14	-0.13	-0.18	-0.24

## Consolidated Statement of Financial Position

### Financial position

The equity/assets ratio was 90 (90) percent as of 30 June 2021, and equity was SEK 164,504,000 (145,044,000). The equity includes funds from the in April and May completed two tranches directed share issue, which provided the company with SEK 75,900,000 after deduction of issue costs of SEK 4,100,000. Cash and cash equivalents amounted to SEK 94,146,000 (69,109,000) as of 30 June 2021, an increase of SEK 32,503,000 from the beginning of the year. Total assets as of 30 June 2021 were SEK 183,040,000 (160,395,000).

### 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	30 Jun, 2021	30 Jun, 2020	31 Dec, 2020
<b>ASSETS</b>				
<b>Non-current assets</b>				
<b>Intangible assets</b>	1			
Development costs		51,706	51,706	51,706
Patents		20,268	21,465	20,971
Other Intangible assets		1,277	1,411	1,344
		<b>73,251</b>	<b>74,583</b>	<b>74,021</b>
<b>Tangible assets</b>				
Equipment		82	68	41
Rigth of use asset leases		172	515	343
		<b>254</b>	<b>583</b>	<b>384</b>
<b>Financial assets</b>				
Other long-term securities		13,101	13,101	13,101
		<b>13,101</b>	<b>13,101</b>	<b>13,101</b>
<b>Total non-current assets</b>		<b>86,606</b>	<b>88,267</b>	<b>87,506</b>
<b>Current assets</b>				
Other receivables		1,325	1,366	928
Prepaid expenses and accrued income		963	1,653	586
Cash and cash equivalents		94,146	69,109	61,643
		<b>96,434</b>	<b>72,128</b>	<b>63,157</b>
<b>TOTAL ASSETS</b>		<b>183,040</b>	<b>160,395</b>	<b>150,663</b>

# Consolidated Statement of Financial Position

(SEK 000)	Note	30 Jun, 2021	30 Jun, 2020	31 Dec, 2020
<b>EQUITY AND LIABILITIES</b>				
<b>Equity attributable to the shareholders of the parent company</b>				
Share capital		20,150	13,484	14,817
Additional paid in capital		730,592	642,893	660,025
Translation reserve		617	621	616
Retained earnings		-586,868	-511,958	-535,096
<b>Total equity attributable to the shareholders of the parent</b>		<b>164,491</b>	<b>145,039</b>	<b>140,363</b>
Non-controlling interests		13	5	0
<b>Total equity</b>		<b>164,504</b>	<b>145,044</b>	<b>140,363</b>
<b>Long-term liabilities</b>				
Other longterm liabilities		-	361	92
		-	<b>361</b>	<b>92</b>
<b>Short-term liabilities</b>				
Accounts payable		8,856	9,258	4,201
Other liabilities		415	535	675
Accrued expenses and deferred income		9,265	5,197	5,333
		<b>18,536</b>	<b>14,991</b>	<b>10,209</b>
<b>Total liabilities</b>		<b>18,536</b>	<b>15,712</b>	<b>10,392</b>
<b>TOTAL EQUITY AND LIABILITIES</b>		<b>183,040</b>	<b>160,395</b>	<b>150,663</b>

# Consolidated Statement of Changes in Equity

(SEK 000)	Equity attributable to the shareholders of the parent company					Non-controlling interests	Total equity
	Share-capital	Additional paid in capital	Translation reserve	Retained earnings	Total		
<b>Opening balance, 1 January 2021</b>	<b>14,817</b>	<b>660,025</b>	<b>616</b>	<b>-535,095</b>	<b>140,362</b>	<b>-0</b>	<b>140,362</b>
<b>Comprehensive profit/loss for the period</b>							
Profit/loss for the period	-	-	-	-51,769	-51,769	-1	-51,770
Other comprehensive income							
Translation differences	-	-	1	-4	-3	-	-3
<b>Other comprehensive profit/loss for the period, net after tax</b>	<b>-</b>	<b>-</b>	<b>1</b>	<b>-4</b>	<b>-3</b>	<b>-</b>	<b>-3</b>
<b>Total comprehensive profit/loss</b>	<b>-</b>	<b>-</b>	<b>1</b>	<b>-51,773</b>	<b>-51,771</b>	<b>-1</b>	<b>-51,772</b>
<b>Transactions with shareholders</b>							
Rights Issue*	5,333	70,567	-	-	75,900	-	75,900
Shareholder contribution	-	-	-	-	-	14	14
<b>Total transactions with shareholders</b>	<b>5,333</b>	<b>70,567</b>	<b>-</b>	<b>-</b>	<b>75,900</b>	<b>14</b>	<b>75,914</b>
<b>Closing balance, 30 June 2021</b>	<b>20,150</b>	<b>730,592</b>	<b>617</b>	<b>-586,869</b>	<b>164,491</b>	<b>13</b>	<b>164,504</b>
<b>Opening balance, 1 January 2020</b>	<b>9,298</b>	<b>592,980</b>	<b>619</b>	<b>-475,107</b>	<b>127,791</b>	<b>5</b>	<b>127,795</b>
<b>Comprehensive profit/loss for the period</b>							
Profit/loss for the period	-	-	-	-36,848	-36,848	-1	-36,849
Other comprehensive income							
Translation differences	-	-	2	-3	-	-	-
<b>Other comprehensive profit/loss for the period, net after tax</b>	<b>-</b>	<b>-</b>	<b>2</b>	<b>-3</b>	<b>-</b>	<b>-</b>	<b>-</b>
<b>Total comprehensive profit/loss</b>	<b>-</b>	<b>-</b>	<b>2</b>	<b>-36,851</b>	<b>-36,849</b>	<b>-1</b>	<b>-36,850</b>
<b>Transactions with shareholders</b>							
Rights Issue*	4,186	49,912	-	-	54,098	-	54,098
<b>Total transactions with shareholders</b>	<b>4,186</b>	<b>49,912</b>	<b>-</b>	<b>-</b>	<b>54,098</b>	<b>-</b>	<b>54,098</b>
<b>Closing balance, 30 June 2020</b>	<b>13,484</b>	<b>642,892</b>	<b>621</b>	<b>-511,958</b>	<b>145,039</b>	<b>5</b>	<b>145,044</b>
<b>Opening balance, 1 January 2020</b>	<b>9,298</b>	<b>592,980</b>	<b>619</b>	<b>-475,107</b>	<b>127,791</b>	<b>5</b>	<b>127,795</b>
<b>Comprehensive profit/loss for the period</b>	<b>-</b>	<b>-</b>	<b>-</b>	<b>-</b>	<b>-</b>	<b>-</b>	<b>-</b>
Profit/loss for the period	-	-	-	-59,989	-59,989	-5	-59,994
Other comprehensive income	-	-	-	-	-	-	-
Translation differences	-	-	-3	-	-3	-	-3
<b>Other comprehensive profit/loss for the period, net after tax</b>	<b>-</b>	<b>-</b>	<b>-3</b>	<b>-</b>	<b>-3</b>	<b>-</b>	<b>-3</b>
<b>Total comprehensive profit/loss</b>	<b>-</b>	<b>-</b>	<b>-3</b>	<b>-59,989</b>	<b>-59,992</b>	<b>-5</b>	<b>-59,997</b>
<b>Transactions with shareholders</b>	<b>-</b>	<b>-</b>	<b>-</b>	<b>-</b>	<b>-</b>	<b>-</b>	<b>-</b>
Rights Issue	5,519	67,045	-	-	72,564	-	72,564
<b>Total transactions with shareholders</b>	<b>5,519</b>	<b>67,045</b>	<b>-</b>	<b>-</b>	<b>72,564</b>	<b>-</b>	<b>72,564</b>
<b>Closing balance, 31 December 2020</b>	<b>14,817</b>	<b>660,025</b>	<b>616</b>	<b>-535,095</b>	<b>140,362</b>	<b>-0</b>	<b>140,362</b>

\*Total equity includes funds from the April 6, 2021 and May 4th completed directed share issue with SEK 75,900,000 after expenses SEK 4,100,000.

## Consolidated Statement of Cash Flows

### Cash flow and investments

Operating cash flow for the second quarter was SEK -29,448,000 ( -14,183,000). For the first six months the operating cash flow amounted SEK -42,990,000 (-42,329,000). The cash flow effect related to investments in intangibles equals SEK -269,000 (-805,000) for the first six months. Cash flow for the second quarter equals SEK 46,166,000 (39,546,000). Cashflow for the first six months equals SEK 32,499,000 (10,790,000).

(SEK 000)	1 Apr, 2021 30 Jun, 2021	1 Apr, 2020 30 Jun, 2020	1 Jan, 2021 30 Jun, 2021	1 Jan, 2020 30 Jun, 2020	1 Jan, 2020 31 Dec, 2020
<b>Cash flow from operating activities</b>					
<b>Operating income</b>	-30,311	-20,304	-51,762	-36,832	-60,071
<b>Adjustments for non-cash items:</b>					
Depreciation	659	636	1,317	1,263	2,558
Currency differences on intercompany items	-7	-	-7	-	-
Result from other securities and receivables related to non current assets	-	-	-	-	107
Interest received	-	-	-	-	-
Interest paid	-3	-8	-8	-17	-30
<b>Net cash from operating activities before changes in working capital</b>	<b>-29,663</b>	<b>-19,677</b>	<b>-50,460</b>	<b>-35,586</b>	<b>-57,436</b>
<b>Changes in working capital</b>					
Increase/decrease of other current assets	-156	-1,188	-773	-1,418	86
Increase/decrease of other short-term liabilities	371	6,681	8,243	-5,325	-10,208
<b>Changes in working capital</b>	<b>215</b>	<b>5,494</b>	<b>7,471</b>	<b>-6,743</b>	<b>-10,122</b>
<b>Cash flow from operating activities</b>	<b>-29,448</b>	<b>-14,183</b>	<b>-42,990</b>	<b>-42,329</b>	<b>-67,558</b>
<b>Investing activities</b>					
Acquisition of intangible assets	-235	-280	-269	-805	-1,407
Acquisition of tangible assets	-65	-	-65	-	-
Increase in other financial assets	-	-	-	-	-
<b>Cash flow from investing activities</b>	<b>-300</b>	<b>-280</b>	<b>-334</b>	<b>-805</b>	<b>-1,407</b>
<b>Financing activities</b>					
Shareholder contribution subsidiary	14	-	14	-	-
New share issue	75,900	54,098	75,900	54,098	72,564
Amoritzation lease	-	-90	-92	-174	-269
<b>Cash flow from financing activities</b>	<b>75,914</b>	<b>54,008</b>	<b>75,822</b>	<b>53,924</b>	<b>72,295</b>
Cash flow for the period	46,166	39,546	32,499	10,790	3,330
Cash and cash equivalents at the beginning of the period	47,976	29,568	61,643	58,319	58,319
Effect of exchange rate changes on cash	4	-5	5	0	-6
<b>Cash and cash equivalents at end of period</b>	<b>94,146</b>	<b>69,109</b>	<b>94,146</b>	<b>69,109</b>	<b>61,643</b>

## Parent Company

## Income Statement

## Parental company

Company earnings after tax for the first quarter amounts to SEK -29,082,000 (-20,343,000). Earnings after tax for the first six months amount to SEK -50,538,000 (-36,876,000). Most of the Group's operations are conducted within the parent company. Accordingly, no further specific information regarding the parent company is presented.

## Parent Company

## Statement of Comprehensive Income

(SEK 000)		1 Apr, 2021	1 Apr, 2020	1 Jan, 2021	1 Jan, 2020	1 Jan, 2020
	Note	30 Jun, 2021	30 Jun, 2020	30 Jun, 2021	30 Jun, 2020	31 Dec, 2020
Net sales		18	97	18	105	216
Other operating income		0	-	0	-	1,648
		<b>18</b>	<b>97</b>	<b>18</b>	<b>105</b>	<b>1,864</b>
<b>Operating expenses</b>						
Other external expenses		-24,185	-15,980	-39,815	-28,028	-46,411
Personnel cost		-4,279	-4,233	-9,326	-7,783	-13,305
Depreciation and write-down of tangible and intangible assets		-573	-550	-1,145	-1,091	-2,215
Other operating expenses		-63	324	-270	-78	-
		<b>-29,100</b>	<b>-20,439</b>	<b>-50,556</b>	<b>-36,980</b>	<b>-61,931</b>
<b>Operating income</b>		<b>-29,082</b>	<b>-20,343</b>	<b>-50,538</b>	<b>-36,876</b>	<b>-60,067</b>
<b>Profit/loss from financial items</b>						
Result from other securities and receivables related to non current assets		-	-	-	-	107
Interest expenses and other similar loss items		-	-	-	-	-1
		<b>-</b>	<b>-</b>	<b>-</b>	<b>-</b>	<b>106</b>
<b>Profit/loss before tax</b>		<b>-29,082</b>	<b>-20,343</b>	<b>-50,538</b>	<b>-36,876</b>	<b>-59,961</b>
Income tax	2	-	-	-	-	-
<b>Profit/loss for the period</b>		<b>-29,082</b>	<b>-20,343</b>	<b>-50,538</b>	<b>-36,876</b>	<b>-59,961</b>

(SEK 000)		1 Apr, 2021	1 Apr, 2020	1 Jan, 2021	1 Jan, 2020	1 Jan, 2020
	Note	30 Jun, 2021	30 Jun, 2020	30 Jun, 2021	30 Jun, 2020	31 Dec, 2020
Profit/loss for the period		-29,082	-20,343	-50,538	-36,876	-59,961
Other comprehensive income		-	-	-	-	-
<b>Total comprehensive profit/loss for the period</b>		<b>-29,082</b>	<b>-20,343</b>	<b>-50,538</b>	<b>-36,876</b>	<b>-59,961</b>

# Parent Company

## Balance Sheet

(SEK 000)	Note	30 Jun, 2021	30 Jun, 2020	31 Dec, 2020
<b>ASSETS</b>				
<b>Non-current assets</b>				
<i>Intangible assets</i>	1			
Development costs		51,706	51,706	51,706
Patents		20,268	21,465	20,971
Other intangible assets		1,277	1,411	1,344
		<b>73,251</b>	<b>74,584</b>	<b>74,021</b>
<b>Tangible assets</b>				
Equipment		82	68	41
		<b>82</b>	<b>68</b>	<b>41</b>
<b>Financial assets</b>				
Other long-term placement		13,101	13,101	13,101
Shares in subsidiaries	3	24,558	23,625	23,625
		<b>37,659</b>	<b>36,726</b>	<b>36,726</b>
<b>Total non-current assets</b>		<b>110,991</b>	<b>111,377</b>	<b>110,788</b>
<b>Current assets</b>				
<i>Short term receivables</i>				
Receivables from group companies		498	-	-
Other receivables		1,305	1,363	926
Prepaid expenses and accrued income		962	1,653	585
		<b>2,765</b>	<b>3,015</b>	<b>1,511</b>
Cash and bank balances		93,884	69,083	<b>61,634</b>
<b>Total current assets</b>		<b>96,650</b>	<b>72,098</b>	<b>63,145</b>
<b>TOTAL ASSETS</b>		<b>207,642</b>	<b>183,475</b>	<b>173,933</b>

# Parent Company

## Balance Sheet

(SEK 000)	Note	30 Jun, 2021	30 Jun, 2020	31 Dec, 2020
<b>EQUITY AND LIABILITIES</b>				
<b>Equity</b>				
<i>Restricted equity</i>				
Share capital		20,150	13,484	14,817
Statutory reserve		1,856	1,856	1,856
Development expenditure reserve		12,873	14,070	13,576
		<b>34,879</b>	<b>29,411</b>	<b>30,249</b>
<b>Unrestricted equity</b>				
Share premium reserve		137,611	152,978	67,045
Retained earnings		67,418	23,115	126,676
Profit/loss for the period		-50,538	-36,842	-59,961
		<b>154,492</b>	<b>139,251</b>	<b>133,759</b>
<b>Total equity</b>		<b>189,371</b>	<b>168,662</b>	<b>164,009</b>
<b>Short-term liabilities</b>				
Accounts payable		8,848	9,258	4,201
Other liabilities		233	362	406
Accrued expenses and deferred income		9,190	5,194	5,317
		<b>18,271</b>	<b>14,814</b>	<b>9,924</b>
<b>TOTAL EQUITY AND LIABILITIES</b>		<b>207,642</b>	<b>183,475</b>	<b>173,933</b>

# Notes

## Note 1 — Intangible assets

(SEK 000)	Development costs	Patents	Other	Total
<b>ACCUMULATED COST</b>				
Opening balance 1 Jan. 2021	51,706	33,771	2,864	88,341
Additions	-	351	-	351
Closing balance 30 Jun. 2021	51,706	34,122	2,864	88,692
<b>ACCUMULATED DEPRECIATION</b>				
Opening balance 1 Jan. 2021	-	-12,800	-1,519	-14,319
Depreciation for the period	-	-1,054	-67	-1,121
Closing balance 30 Jun. 2021	-	-13,854	-1,586	-15,440
Residual value 30 Jun. 2021	51,706	20,268	1,278	73,252
<b>(SEK 000)</b>				
<b>ACCUMULATED COST</b>				
Opening balance 1 Jan. 2020	51,706	32,279	2,864	86,849
Additions	-	1,492	-	1,492
Closing balance 31 Dec. 2020	51,706	33,771	2,864	88,341
<b>ACCUMULATED DEPRECIATION</b>				
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	51,706	20,971	1,345	74,022

## Note 2 – Tax

The group's total loss carry-forward amount to SEK 643,842,000 as of 30 June 2021 (581,413,000). The parent company's total loss carry-forwards amounts to SEK 670,953,000 as of 30 June 2021 (555,581,000). 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 wholly owned american subsidiary Abliva Inc., registered in March 2021 and the newly formed Swedish subsidiary Abliva Incentive AB, holding option program for the CEO.

## Other disclosures

### 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.

(SEK 000)	1 Jan. 2021 30 Jun. 2021	1 Jan. 2020 31 Dec. 2020
Eskil Elmér, CSO	-	6
Magnus Hansson, CMO	-	4
<b>Total</b>	<b>-</b>	<b>10</b>

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. Apart from remuneration to senior executives no transactions with related parties have occurred.

### 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 6 (4) are women.

### 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

Q3 Report January-September 2021	November 19, 2021
Year-End Report 2021	February 22, 2022

The interim reports and the Annual Year Report are available at: [www.abliva.com](http://www.abliva.com)

## 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. The Board of Directors works continuously to secure the business operation's need for financing.

### 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 profession-

als to better meet the influx of COVID-19 patients. There is a risk that the start of the upcoming phase II / III study, which is expected to begin in the second half of 2021, will be delayed. Abliva's preparations in the form of preclinical safety studies during 2021 to apply for permission to initiate clinical trials for the drug candidate NV354 for Leigh's syndrome are currently not considered to be affected to a greater extent by the COVID-19 pandemic.

### Capitalized Development Costs

The Board of Swedish Accounting Supervision has examined the Company's interim report as of September 30, 2020 and the Annual report for 2020 regarding the accounting of capitalized development costs. The matter has been forwarded to the Swedish Financial Supervisory Authority (SFSA) on 12 July 2021. SFSA has informed the Company that it intends to return to the Company regarding continued handling.

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.

The group and parent company have 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 financial 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, 19 August, 2021

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

**David Beijker**  
Board member

**Roger Franklin**  
Board member

**Denise Goode**  
Board member

**Jan Törnell**  
Board member

**Ellen Donnelly**  
Chief Executive Officer



David Laskow-Pooley



David Beijker



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. CEST on 19 August, 2021.

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 stroke-like 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<sup>+</sup>/NADH.** A coenzyme involved in metabolism. NAD<sup>+</sup> 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<sup>+</sup> regulator, is in clinical development and has been granted orphan drug designation in Europe and the US. NV354, an energy replacement (succinate) therapy, is in preclinical development. 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).

## Abliva AB (publ)

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