



## Interim Report

January - March 2022



The ability to raise this round given the current market situation is a strong indication of support for the company vision and strategy, and the development plans for KL1333 and NV354.

Ellen Donnelly, CEO

Delivering mitochondrial health

# First quarter summary

Financing enables start of the registrational FALCON study with KL1333

NV354 data is being assembled for a clinical trial application



## Important events first quarter (Jan - Mar 2022)

- 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.

## Important events after the reporting period

- On May 31, Abliva's Board of Directors announced funding of approximately SEK 200 million that will provide the company with the capital required to pursue the Phase 2/3 study with KL1333 to a key interim analysis, as well as prepare NV354 for clinical phase. For more information see pages 10-11.
- Annual General Meeting was held on May 20, 2022.

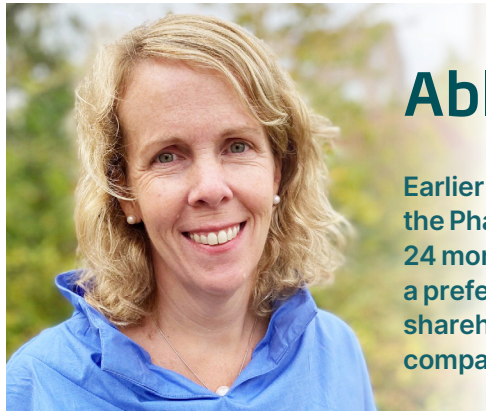
## Financial information

### January-March 2022\*

- Net revenues: SEK 0,000 (0,000)
- Other operating income: SEK 0,000 (0,000)
- Loss before tax: SEK 22,028,000 (21,456,000)
- Loss per share before dilution: SEK 0.05 (0.07)
- Diluted loss per share: SEK 0.05 (0.07)

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





# Abliva Announces SEK 200 Million Financing Round

Earlier today Abliva announced a SEK 200 million financing that will provide the company with the capital necessary to run the Phase 2/3 study with KL1333 to a key interim analysis, progress NV354 to be clinic-ready, and provide the company with 24 months of cash runway. Pending final approval by the Board of directors this evening, the financing round will include a preferential rights issue of SEK 50 million and a directed share issue of SEK 150 million (comprised of existing and new shareholders). The ability to raise this round given the current market situation is a strong indication of support for the company vision and strategy, and the development plans for KL1333 and NV354.

## Details on the Financing

The announcement today of the SEK 200 million financing round is truly a landmark event for the company. Not only have we completed an unprecedented raise in a very challenging market environment (ECM transactions for European biotechs are down 95% versus last year), but we have also built a syndicate in the directed share issue of high-quality investors who have the financial power to support the company for the longer term. Equally important, we also gave our current shareholders the opportunity to participate in this round with a preferential rights issue. Finally, in a separate transaction (not as part of the DSI or RI), lead investor Hadean Ventures will convert the convertible loan from January 2022 into shares.

## Use of Proceeds

The majority of the proceeds will be used to initiate the registration, Phase 2/3 clinical trial evaluating KL1333 for the treatment of adults suffering from primary mitochondrial disease. The start of this global study is expected in the second half of 2022 with our first patient dosed in early 2023. The financing procured will allow us to enroll the first 40 patients in the study and progress to a meaningful interim analysis that will provide important safety and efficacy information (through a conditional power analysis) to allow us to move into the second stage

of the seamless platform design study (provided the study is not futile), with either continuation at the planned study size or expansion up to a pre-defined cap. After discussion with future potential investors and strategic partners we are confident that the interim analysis will provide the information necessary to attract additional interest in the company.

*“The ability to raise this round given the current market situation is a strong indication of support for the company vision and strategy, and the development plans for KL1333 and NV354”*

The rest of the proceeds will be used to progress NV354 and support the company. NV354 will continue towards the clinic with the manufacturing of clinical trial material and the submission of the appropriate regulatory documentation to support a Phase 1 start. The company will continue to be run in lean manner with limited overhead. The financing will provide the company with twenty-four months of runway through mid-2024.

## Acknowledgements

A historic raise such as this could not have been possible without the help of Abliva family, friends and shareholders. First, I'd like to thank the shareholders, who patiently waited as we

worked to raise this money. We appreciate your continued confidence as we strove to do the **right** deal, not just **a** deal.

To our lead investor, Hadean Ventures, a 'thank you' seems insufficient at this point. We are truly fortunate to have an investor in Abliva who clearly shares our passion for building a global company and a portfolio of therapeutics focused on primary mitochondrial diseases.

To our new investors, welcome. We look forward to growing our relationship as we dose patients across the world with KL1333.

And to our banks, Kempen and Penser. Thank you for your professionalism, confidence and perseverance throughout this process.

And to the physicians and patients who have been waiting for news of study start. Let's do this!

Onward with thanks and humility-

**Ellen Donnelly**  
CEO



**Primary mitochondrial diseases** are metabolic diseases that affect the cells' ability to convert energy. The diseases can manifest very differently depending on the organs impacted and the number of dysfunctional mitochondria in that organ. Historically viewed as clinical syndromes, our knowledge about the various mutations underlying mitochondrial diseases has increased, improving our ability to identify and treat these patients. 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.

PROJECT	DISEASE	DISCOVERY	PRECLINICAL	PHASE 1	PHASE 2/3	MARKET
KL1333*	PMD (mtDNA disorders)					
NV354	PMD (Leigh syndrome)					
Early programs	PMD					

\*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 biotech 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 biotech 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

Mitochondria function as the powerhouses of our cells and are crucial for the cells' energy metabolism. Mitochondrial diseases 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<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 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<sup>1</sup>.

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 mitochondrial disease Leigh Syndrome initially with potential to expand to other indications that have a dysfunctional complex I in the electron transport chain.

Further, 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<sup>2,3</sup>.

In addition, we have sought scientific 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) Gorman et al., Prevalence of Nuclear and Mitochondrial DNA Mutations Related to Adult Mitochondrial Disease, 2015

2) Jayasundara et al. Orphanet J of Rare Dis. Estimating the clinical cost of drug development for orphan versus non-orphan drugs. 2019. 3) EvaluatePharma, Orphan Drug Report 2019.

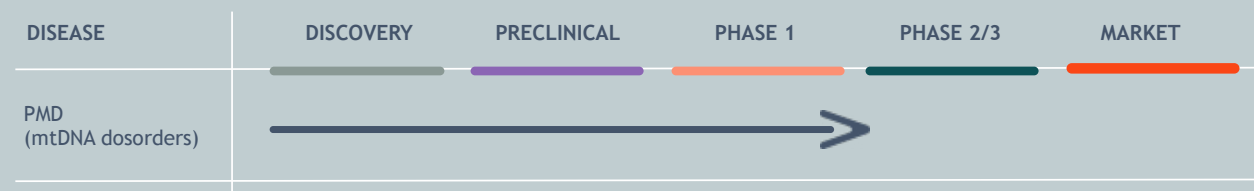
# KL1333 Innovative therapy heading to registrational Phase 2/3 study

Financing enables start of the registrational Phase 2/3 study in 2022

Fully financed to interim analysis

Phase 1a/b study: Positive safety results and signs of efficacy

Orphan drug designation in both the United States and Europe



## Events in the first quarter

- During the quarter, Abliva continued with preparations for the start of the registrational FALCON study, including through interactions with regulatory authorities and clinical sites in the countries involved, as well as work to finalize the clinical protocol.

## Events after the reporting period

- The company received approval also from the UK regulatory agencies to start the Phase 2/3 study.

## Objectives for 2022

- Regulatory approvals in select countries.
- Initiate the start of the FALCON study.

## DISEASE AREA

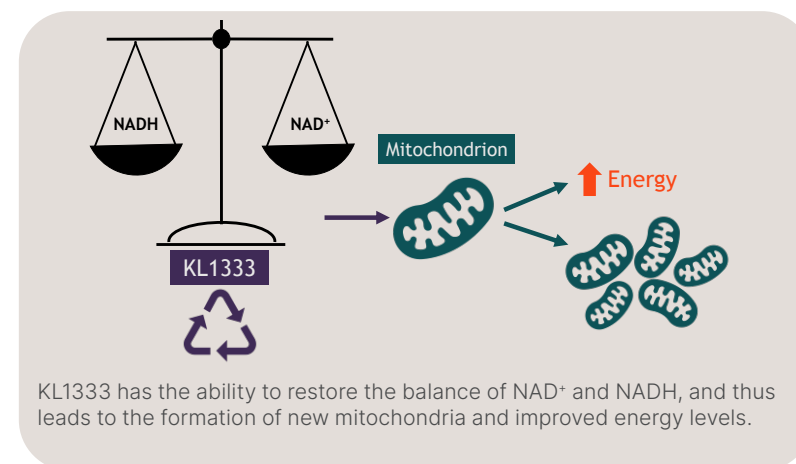
KL1333 is being developed as a treatment for a subset of adult primary mitochondrial disease patients suffering from multiple debilitating symptoms, including mitochondrial fatigue and myopathy. Diagnoses can include MELAS-MIDD and KSS-CPEO spectrum disorders as well as MERRF syndrome.

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 2026. 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.

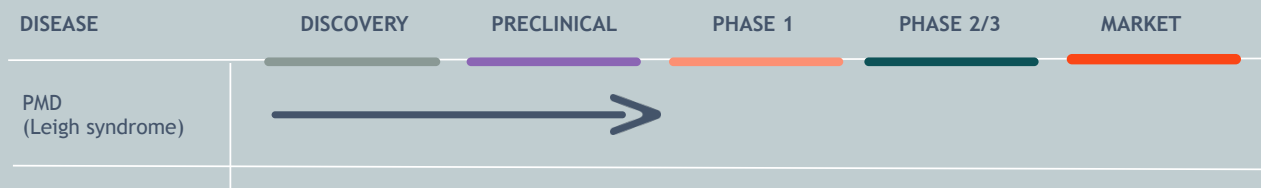
1) Gorman et al., Prevalence of Nuclear and Mitochondrial DNA Mutations Related to Adult Mitochondrial Disease, 2015



# NV354

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

Positive feedback from UK MHRA scientific advice meeting  
Preparation of the program for clinical studies 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 2022

- Produce NV354 clinical trial material for clinical Phase 1 study.
- 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: NeuroSTAT - For treatment of Traumatic Brain Injury

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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 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.

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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 first quarter of 2022 was SEK 0,000 (0,000). Other operating revenues for the first quarter were SEK 0,000 (0,000).

## Results of operations

The operating loss for the first quarter was SEK 21,550,000 (21,451,000). The net loss before tax for the first quarter amounted to SEK 22,028,000 (21,456,000).

The operating loss was affected by other external expenses, which for the full were SEK 17,350,000 (15,540,000). During the first quarter, expenses related to development projects, as a part of external expenses, have affected the result with SEK 15,702,000 (12,318,000) whereof SEK 15,264,000 (11,328,000) relates to project in clinical phase. Personnel expenses during the first quarter amount to SEK 3,448,000 (5,047,000). Other operating expenses amount to, SEK 145,000 (207,000) and pertains to exchange-rate losses.

(SEK 000)	Note	1 Jan, 2022 31 Mar, 2022	1 Jan, 2021 31 Mar, 2021	1 Jan, 2021 31 Dec, 2021
Net sales		-	-	151
Other operating income		-	-	-
		-	-	1,151
<b>Operating expenses</b>				
Other external expenses		-17,350	-15,540	-103,695
Personnel cost		-3,448	-5,047	-16,844
Depreciation and write-down of tangible and intangible assets		-608	-658	-2,764
Other operating expenses		-145	-207	-330
		-21,550	-21,451	-123,633
<b>Operating income</b>		-21,550	-21,451	-123,482
<b>Profit/loss from financial items</b>				
Result from other securities and receivables related to non current assets		-	-	-
Financial costs		-478	-5	-12
		-478	-5	-12
<b>Profit/loss before tax</b>		-22,028	-21,456	-123,494
Income tax	2	-	-	-4
<b>Profit/loss for the period</b>		-22,028	-21,456	-123,498
<b>Other comprehensive income</b>				
<i>Items that may be reclassified to profit or loss</i>				
Translation differences on foreign subsidiaries		23	1	71
<b>Total comprehensive income for the period</b>		-22,005	-21,455	-123,427
<b>Loss for the period attributable to:</b>				
Parent company shareholders		-22,028	-21,456	-123,492
Non-controlling interests		-1	-	-6
		-22,028	-21,456	-123,498
<b>Total comprehensive income for the period</b>				
Parent company shareholders		-22,004	-21,455	-123,420
Non-controlling interests		-1	-	-7
		-22,005	-21,455	-123,427
Earnings per share before and after dilution(SEK) based on average number of shares		-0.05	-0.07	-0.33
Average number of shares before and after dilution		403,006,798	296,340,132	370,168,023

## Consolidated Statement of Financial Position

### Financial position

The equity/assets ratio was 33 (79) percent as of 31 March 2022, and equity was SEK 20,114,000 (41,528,000) compared to beginning of the year. The company's long-term liabilities relates to a convertible loan from Hadean Ventures of SEK 26,000,000. The Convertible Issue consists of 260 convertibles at a nominal value of SEK 100,000 per convertible. The subscription price shall be equal to the nominal value of the Convertibles with a deduction of a set-up fee of 3.95 percent. The Convertibles carries an annual interest rate of 10 percent of their nominal value. The term of the Convertibles is 12 months with a maturity date on December 20, 2022, to the extent conversion has not taken place before such date. The conversion price will either be (i) same price as for other investors in a potential capital raise, or (ii) 10-day VWAP prior to conversion request date, net amount of SEK 24,223 thousand after set-up fees and overheads. Book value corresponds to the payment less the set-up fee of 3.95% and the brokerage fee of SEK 750,000. Hadean Ventures has declared that they intend to Current liabilities amounted to SEK 17,348,000 (18,140,000) as of March 31, 2022. Cash and cash equivalents amounted to SEK 23,880,000 (47,976,000) as of 31 March 2022, an increase of SEK 1,541,000 from the beginning of the year. Total assets as of March 31, 2022 were SEK 61,686,000 (85,342,000).

Abliva's Board of Directors intends to resolve, on May 31, 2022, based on the authorization granted at the Annual General Meeting on May 20, 2022, on an issue of shares with preferential rights for the Company's shareholders of approximately SEK 50 million and on a directed issue of approximately SEK 150 million. The shares in both issues will be issued at an issue discount of a maximum of 10 percent compared to the closing price on Tuesday, May 31, 2022. Further, on May 31, 2022, Hadean Ventures declared that they intend to convert the convertible loan into shares.

(SEK 000)	Note	31 Mar, 2022	31 Mar, 2021	31 Dec, 2021
<b>ASSETS</b>				
<b>Non-current assets</b>				
<b>Intangible assets</b>	1			
Development costs		-	-	-
Patents		20,083	20,536	20,293
Other Intangible assets		1,176	1,311	1,210
		<b>21,259</b>	<b>21,847</b>	<b>21,503</b>
<b>Tangible assets</b>				
Equipment		49	30	60
Righ of use asset leases		-	258	-
		<b>49</b>	<b>287</b>	<b>60</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>34,409</b>	<b>35,236</b>	<b>34,664</b>
<b>Current assets</b>				
Other receivables		1,328	1,071	912
Prepaid expenses and accrued income		2,070	1,059	1,003
Cash and cash equivalents		23,880	47,976	22,339
		<b>27,278</b>	<b>50,106</b>	<b>24,254</b>
<b>TOTAL ASSETS</b>		<b>61,686</b>	<b>85,342</b>	<b>58,918</b>

## Consolidated Statement of Financial Position

### 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 Mar, 2022	31 Mar, 2021	31 Dec, 2021
<b>EQUITY AND LIABILITIES</b>				
<b>Equity attributable to the shareholders of the parent company</b>				
Share capital		20,150	14,817	20,150
Additional paid in capital		730,560	660,025	730,560
Translation reserve		711	617	688
Retained earnings*		-731,316	-608,257	-709,879
<b>Total equity attributable to the shareholders of the parent</b>		<b>20,106</b>	<b>67,202</b>	<b>41,519</b>
Non-controlling interests		8	-	9
<b>Total equity</b>		<b>20,114</b>	<b>67,202</b>	<b>41,528</b>
<b>Long-term liabilities</b>				
Other longterm liabilities		24,223	-	-
		<b>24,223</b>	<b>-</b>	<b>-</b>
<b>Short-term liabilities</b>				
Accounts payable		9,289	9,049	9,616
Other liabilities		1,393	640	277
Accrued expenses and deferred income		6,667	8,451	7,497
		<b>17,348</b>	<b>18,140</b>	<b>17,390</b>
<b>Total liabilities</b>		<b>41,571</b>	<b>18,140</b>	<b>17,390</b>
<b>TOTAL EQUITY AND LIABILITIES</b>		<b>61,686</b>	<b>85,342</b>	<b>58,918</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>-586,802</b>	<b>88,656</b>	<b>-0</b>	<b>88,656</b>
Comprehensive profit/loss for the period	-	-	-	-	-	-	-
Profit/loss for the period	-	-	-	-21,455	-21,455	-0	-21,456
Other comprehensive income	-	-	-	-	-	-	-
Translation differences	-	-	1	-	1	-	1
<b>Other comprehensive profit/loss for the period, net after tax</b>	<b>-</b>	<b>-</b>	<b>1</b>	<b>-</b>	<b>1</b>	<b>-</b>	<b>1</b>
<b>Total comprehensive profit/loss</b>	<b>-</b>	<b>-</b>	<b>1</b>	<b>-21,455</b>	<b>-21,454</b>	<b>-</b>	<b>-21,455</b>
Transactions with shareholders	-	-	-	-	-	-	-
<b>Total transactions with shareholders</b>	<b>-</b>	<b>-</b>	<b>-</b>	<b>-</b>	<b>-</b>	<b>-</b>	<b>-</b>
<b>Closing balance, 31 March 2021</b>	<b>14,817</b>	<b>660,025</b>	<b>617</b>	<b>-608,257</b>	<b>67,202</b>	<b>0</b>	<b>67,202</b>
<b>Opening balance, 1 January 2021</b>	<b>14,817</b>	<b>660,025</b>	<b>616</b>	<b>-586,802</b>	<b>88,656</b>	<b>0</b>	<b>88,656</b>
Comprehensive profit/loss for the period	-	-	-	-123,492	-123,492	-6	-123,498
Profit/loss for the period	-	-	-	-123,492	-123,492	-6	-123,498
Other comprehensive income	-	-	-	-	-	-	-
Translation differences	-	-	72	-	72	-1	71
<b>Other comprehensive profit/loss for the period, net after tax</b>	<b>-</b>	<b>-</b>	<b>72</b>	<b>-</b>	<b>72</b>	<b>-1</b>	<b>71</b>
<b>Total comprehensive profit/loss</b>	<b>-</b>	<b>-</b>	<b>72</b>	<b>-123,492</b>	<b>-123,420</b>	<b>-7</b>	<b>-123,427</b>
<b>Transactions with shareholders</b>							
Rights Issue*	5,333	70,534	-	-	75,868	-	75,868
Share-based payment	-	-	-	415	415	-	415
Shareholder contribution	-	-	-	-	-	16	16
<b>Total transactions with shareholders</b>	<b>5,333</b>	<b>70,534</b>	<b>-</b>	<b>415</b>	<b>76,283</b>	<b>16</b>	<b>76,299</b>
<b>Closing balance, 31 December 2021</b>	<b>20,150</b>	<b>730,560</b>	<b>688</b>	<b>-709,879</b>	<b>41,519</b>	<b>9</b>	<b>41,528</b>
<b>Opening balance, 1 January 2022</b>	<b>20,150</b>	<b>730,560</b>	<b>688</b>	<b>-709,879</b>	<b>41,519</b>	<b>9</b>	<b>41,528</b>
<b>Comprehensive profit/loss for the period</b>							
Profit/loss for the period	-	-	-	-22,028	-22,028	-1	-22,028
Other comprehensive income	-	-	-	-	-	-	-
Translation differences	-	-	23	-	23	-	23
<b>Other comprehensive profit/loss for the period, net after tax</b>	<b>-</b>	<b>-</b>	<b>23</b>	<b>-</b>	<b>23</b>	<b>-</b>	<b>23</b>
<b>Total comprehensive profit/loss</b>	<b>-</b>	<b>-</b>	<b>23</b>	<b>-22,028</b>	<b>-22,005</b>	<b>-1</b>	<b>-22,005</b>
<b>Transactions with shareholders</b>							
Share-based payment	-	-	-	591	591	-	591
<b>Total transactions with shareholders</b>	<b>-</b>	<b>-</b>	<b>-</b>	<b>591</b>	<b>591</b>	<b>-</b>	<b>591</b>
<b>Closing balance, 31 March 2022</b>	<b>20,150</b>	<b>730,560</b>	<b>711</b>	<b>-731,316</b>	<b>20,106</b>	<b>8</b>	<b>20,114</b>



## Consolidated Statement of Cash Flows

### Cash flow and investments

Operating cash flow for the first quarter was SEK -22,718,000 (-13,542,000). The cash flow effect related to investments in intangibles equals SEK -13,000 (-34,000) for the first quarter. Cash flow for the first quarter equals SEK 1,519,000 (13,668,000).

(SEK 000)	1 Jan, 2022 31 Mar, 2022	1 Jan, 2021 31 Mar, 2021	1 Jan, 2021 31 Dec, 2021
<b>Cash flow from operating activities</b>			
Operating income	-21,550	-21,451	-123,482
<b>Adjustments for non-cash items:</b>			
Depreciation	608	-	2,660
Currency differences on intercompany items	14	658	-7
Impaired Value	-	-	104
Share-based payments	591	-	415
Interest paid	-	-5	-12
Paid taxes			
<b>Net cash from operating activities before changes in working capital</b>	<b>-20,337</b>	<b>-20,797</b>	<b>-120,326</b>
<b>Changes in working capital</b>			
Increase/decrease of other current assets	-1,483	-617	-400
Increase/decrease of other short-term liabilities	-898	7,873	6,651
<b>Changes in working capital</b>	<b>-2,381</b>	<b>7,256</b>	<b>6,251</b>
<b>Cash flow from operating activities</b>	<b>-22,718</b>	<b>-13,542</b>	<b>-114,075</b>
<b>Investing activities</b>			
Acquisition of intangible assets	13	-34	-1,024
Acquisition of tangible assets	-0	-	-65
<b>Cash flow from investing activities</b>	<b>13</b>	<b>-34</b>	<b>-1,089</b>
<b>Financing activities</b>			
Shareholder contribution subsidiary	-	-	-
New share issue	-	-	75,868
Amortization lease	-	-92	-92
Increase/decrease of long-term liabilities	24,223	-	-
<b>Cash flow from financing activities</b>	<b>24,223</b>	<b>-92</b>	<b>75,792</b>
Cash flow for the period	1,519	-13,667	-39,372
Cash and cash equivalents at the beginning of the period	22,339	61,643	61,643
Effect of exchange rate changes on cash	22	1	68
<b>Cash and cash equivalents at end of period</b>	<b>23,880</b>	<b>47,976</b>	<b>22,339</b>

## Parent Company

## Income Statement

**Parental company**

Company earnings after tax for the first quarter amounts to SEK -21,434,000 (-21,456,000). Accordingly, no further specific information regarding the parent company is presented.

(SEK 000)		1 Jan, 2022	1 Jan, 2021	1 Jan, 2021
	Note	31 Mar, 2022	31 Mar, 2021	31 Dec, 2021
Net sales		-	-	151
Other operating income		-	-	-
		-	-	151
<b>Operating expenses</b>				
Other external expenses		-18,218	-15,630	-107,521
Personnel cost		-1,987	-5,047	-12,952
Depreciation and write-down of tangible and intangible assets		-608	-572	-2,420
Other operating expenses		-145	-207	-330
		-20,957	-21,456	-123,223
<b>Operating income</b>		-20,956	-21,456	-123,072
<b>Profit/loss from financial items</b>				
Interest expenses and other similar loss items		-478	-	-
		-478	-	-
<b>Profit/loss before tax</b>		-21,434	-21,456	-123,072
Income tax	2	-	-	-
<b>Profit/loss for the period</b>		-21,434	-21,456	-123,072

## Parent Company

## Statement of Comprehensive Income

(SEK 000)		1 Jan, 2022	1 Jan, 2021	1 Jan, 2021
	Note	31 Mar, 2022	31 Mar, 2021	31 Dec, 2021
Profit/loss for the period		-21,434	-21,456	-123,072
Other comprehensive income		-	-	-
<b>Total comprehensive profit/loss for the period</b>		-21,434	-21,456	-123,072

# Parent Company

## Balance Sheet

(SEK 000)	Note	31 Mar, 2022	31 Mar, 2021	31 Dec, 2021
<b>ASSETS</b>				
<b>Non-current assets</b>				
<i>Intangible assets</i>	1			
Development costs		-	-	-
Patents		20,083	20,536	20,293
Other intangible assets		1,176	1,311	1,210
		<b>21,259</b>	<b>21,847</b>	<b>21,503</b>
<b>Tangible assets</b>				
Equipment		49	30	60
		<b>49</b>	<b>30</b>	<b>60</b>
<b>Financial assets</b>				
Other long-term placement		13,100	13,101	24,557
Shares in subsidiaries	3	24,558	23,625	13,101
		<b>37,658</b>	<b>36,726</b>	<b>37,658</b>
<b>Total non-current assets</b>		<b>58,966</b>	<b>58,602</b>	<b>59,221</b>
<b>Current assets</b>				
<i>Short term receivables</i>				
Other receivables		1,306	1,068	890
Prepaid expenses and accrued income		2,070	1,059	1,003
		3,376	2,127	1,893
Cash and bank balances		23,607	47,966	21,696
<b>Total current assets</b>		<b>26,984</b>	<b>50,093</b>	<b>23,589</b>
<b>TOTAL ASSETS</b>		<b>85,950</b>	<b>108,696</b>	<b>82,810</b>

# Parent Company

## Balance Sheet

(SEK 000)	Note	31 Mar, 2022	31 Mar, 2021	31 Dec, 2021
<b>EQUITY AND LIABILITIES</b>				
<b>Equity</b>				
<i>Restricted equity</i>				
Share capital		20,150	14,817	20,150
Statutory reserve		1,856	1,856	1,856
Development expenditure reserve		2,402	3,386	2,613
		<b>24,409</b>	<b>20,060</b>	<b>24,619</b>
<b>Unrestricted equity</b>				
Share premium reserve		70,534	67,045	70,534
Retained earnings*		-29,845	25,198	93,017
Profit/loss for the period		-21,434	-21,456	-123,072
		<b>19,255</b>	<b>70,787</b>	<b>40,479</b>
<b>Total equity</b>		<b>43,664</b>	<b>90,847</b>	<b>65,098</b>
<b>Long-term liabilities</b>				
Other longterm liabilities		24,223	-	-
		24,223	-	-
<b>Short-term liabilities</b>				
Accounts payable		9,268	9,049	9,616
Liabilities subsidiary		1,783	-	1,253
Other liabilities		1,389	368	273
Accrued expenses and deferred income		5,623	8,432	6,570
		<b>18,063</b>	<b>17,849</b>	<b>17,712</b>
<b>TOTAL EQUITY AND LIABILITIES</b>		<b>85,950</b>	<b>108,696</b>	<b>82,810</b>



# Notes

## Note 1 — Intangible assets

(SEK 000)	Patents	Other	Total
<b>ACCUMULATED COST</b>			
Opening balance 1 Jan. 2022	35,180	2,864	38,044
Additions	352	-	352
Closing balance 31 Mar. 2022	35,532	2,864	38,396
<b>ACCUMULATED DEPRECIATION</b>			
Opening balance 1 Jan. 2022	-14,887	-1,654	-16,541
Depreciation for the period	-562	-34	-596
Closing balance 31 Mar. 2022	-15,449	-1,688	-17,137
Residual value 31 Mar. 2022	20,083	1,176	21,259

(SEK 000)	Patents	Other	Total
<b>ACCUMULATED COST</b>			
Opening balance 1 Jan. 2020	33,771	2,864	36,635
Additions	1,562	-	1,562
Impaired value	-153	-	-153
Closing balance 31 Dec. 2020	35,180	2,864	38,044
<b>ACCUMULATED DEPRECIATION</b>			
Opening balance 1 Jan. 2020	-12,800	-1,519	-14,319
Depreciation for the period	-2,136	-135	-2,271
Impaired value	49	-	49
Closing balance 31 Dec. 2020	-14,887	-1,654	-16,541
Residual value 31 Dec. 2020	20,293	1,210	21,503

## Note 2 – Tax

The group's total loss carry-forwards amounts to SEK 767,871,000 as of 31 March 2022 (638,330,000). The parent company's total loss carry-forwards amounts to SEK 741,933,000 as of 31 March 2022 (612,472,000). 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., Boston and the 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- 31 Mar. 2021	1 Jan.2021- 31 Dec. 2021
Eskil Elmér, CSO	-	5
Magnus Hansson, CMO	-	3
<b>Total</b>	<b>-</b>	<b>8</b>

No 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 March 2022 was 8 (6), of which 5 (5) are women.

### Important events during the first quarter (Jan-Mar 2022)

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 ore. 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

Q2 Report January-June 2022	August 19, 2022
Q3 Report January-September 2022	November 22, 2022
Year-End Report 2022	February 24, 2023

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.

### 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 to interim analysis. If the company does not succeed in funding the KL1333 program after the interim analysis, the program may be delayed.

### 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 late 2022, with dosing in the first patient in early 2023, will be further delayed due to Covid-19. The company's second drug candidate NV354, which is being prepared for clinical development, also risks being delayed due to the Covid-19 pandemic.

### Impact of the Ukraine war

The Ukraine war has created unrest and insecurity in the world. The business impact is difficult to predict, but the ongoing war has caused unfavorable market conditions and may affect the ability to raise capital.

Abliva is not involved in any disputes.

For more detail of risks and uncertainty factors, refer to the Statutory Administration Report in the 2021 Annual Report 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.

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 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 2021 on pages 43-58.

## 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, May 31, 2022

**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 9:00 p.m. CEST on May 31, 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 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 discovers and 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 has prioritized two projects. KL1333, a powerful regulator of the essential co-enzymes NAD<sup>+</sup> and NADH, is entering late-stage development. NV354, an energy replacement therapy, has completed preclinical development. Abliva is based in Lund, Sweden.

## What is primary mitochondrial disease?

Primary mitochondrial diseases are metabolic diseases that affect the cells' ability to convert energy. The diseases can manifest very differently depending on the organs impacted and the number of dysfunctional mitochondria in that organ. Historically viewed as clinical syndromes, our knowledge about the various mutations underlying mitochondrial diseases has increased, improving our ability to identify and treat these patients. It is estimated that 125 persons per million have a primary mitochondrial disease.

Abliva's discovery projects focus on gaining a deeper understanding of the mechanisms underlying primary mitochondrial diseases in order to enable us to design new molecules and develop the 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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