THERANEXUS

Innovative biopharmaceutical company specialized in the treatment of rare central nervous system disorders

INVESTOR PRESENTATION

June 2023

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HEC Challenge + Program

Thierry LAMBERT

Chief Financial Officer

Business Administration at the University of Birmingham

MBA (Master of Business Administration) from INSEAD

ICAEW Certified Public Accountant (Institute of Chartered Accountants in England and Wales)

Theranexus, a company with a Phase 3 ready clinical asset

...developing drug candidates for rare neurological disorders

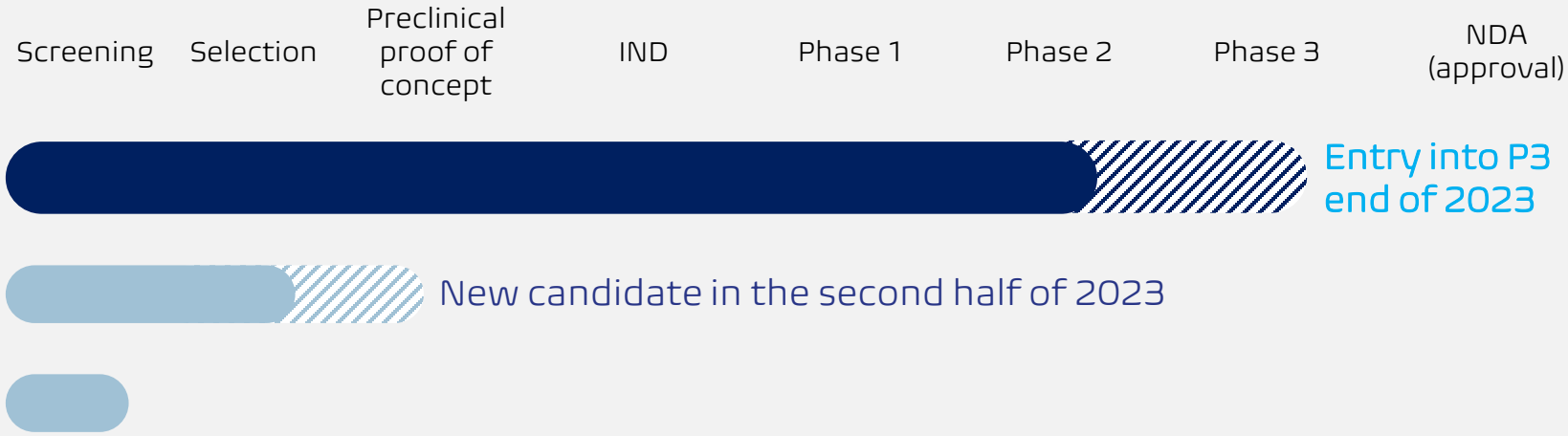
Batten-1

BEYOND BATTEN DISEASE FOUNDATION

Juvenile Batten disease (CLN3)
P1-2 fully recruited and in progress

Platform dedicated to lysosomal diseases

Platform dedicated to diseases with neuronal hyperexcitability



The R&D platform is supported by

3 scientific collaborations:

Neurolead	INSERM	AlstroSight
Platform for the identification of drug candidates based on pathological models of rare neurological diseases	Identification of innovative drug candidates (ASO)[1] in rare neurological diseases	Innovative numerical methods for the search of new drug candidates to treat rare neurological diseases

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[1]ASO = Antisense oligonucleotide

Batten-1, a particularly attractive clinical-stage asset

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A particularly promising first clinical stage asset in juvenile Batten disease (CLN3)

A structuring partnership

- BBDF: the leading patient foundation in this field

BEYOND BATTEN DISEASE FOUNDATION

Recent discovery of the disease mechanism

- Central role of GSL accumulation[1] in neuronal death[2 3]

Use of a registered molecule with a clinically validated mechanism of action

- Proven ability to cross BBB[4] and inhibit GSL formation in patients with CNS lysosomal diseases

Batten-1: an exclusive liquid formulation

- Best suited to patients' needs
- Several levels of protection

Dense short and medium-term newsflow

- Phase 3 US/Europe launch end 2023
- P1/2 Results end 2023 and 2024
- Interim results Phase 3 from 2025

First-to-market in a high-potential market

- Only drug in clinical development for CLN3
- Final results Phase 3 end 2026
- Registration S1-2027
- Estimated peak sales of >$500 million

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[1]Glycosphingolipids
[2]Work carried out by Emyr Llyod Evans' laboratory at Cardiff University (funded by the Beyond Batten Disease foundation).
[3] Glycosphingolipid reduction with miglustat as a therapeutic strategy for CLN3 and otherneuronal ceroid lipofuscinoses, Dr Emyr Lloyd-Evans, Cardiff University
4BBB=Blood-brain barrier

CLN3: the mechanism of the disease has been discovered thanks to recent academic work financed by BBDF

700 to 1 000 patients in the U.S.[1]

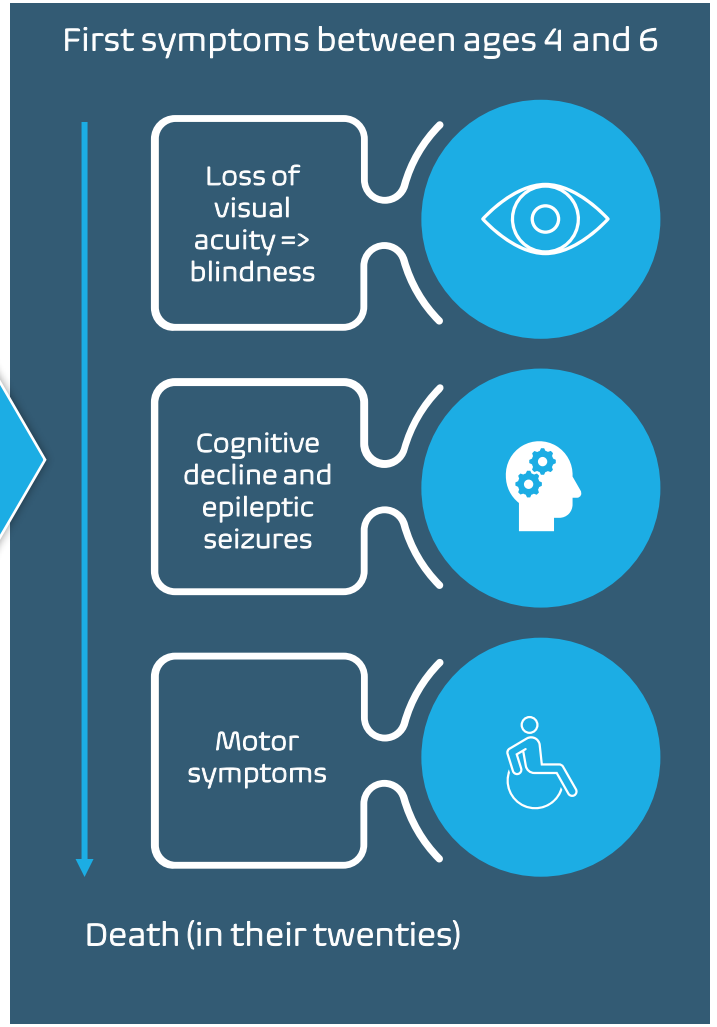
800 to 1 000 patients in the EU[2]

No treatment registered

Autosomal recessive transmission, founder effect localized in Nordic countries

Scientific rationale that led to Batten-1: how to reduce glycosphingolipid accumulation?

Mutation in the CLN3 protein → Toxic accumulation of glycosphingolipids (GSL) in the brain[3] → Neuronal death leading to disabling neurological symptoms[3]

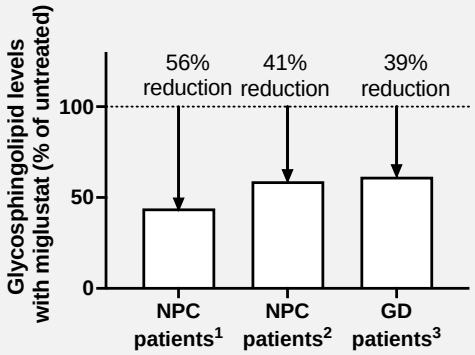


[1]Based on health insurance data (Decision Resources Group)
[2]Orphanet data
[3]Work carried out by Emyr Lloyd Evans' laboratory at Cardiff University (funded by the Beyond Batten Disease foundation) and confirmatory work carried out by Theranexus

Batten-1 (miglustat) targets glycosphingolipid synthesis, whose accumulation is toxic to neurons

GSL reduction by miglustat already validated in humans for other indications…

Significantly reduced GSL levels (clinical demonstration in similar indications)

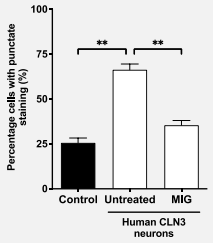


[1] Lachmann et al, 2004
[2] Fan et al, 2013
[3] Zavesca labeling package, EMA, 2005

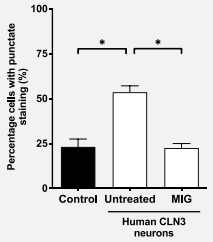
..an activity also validated on preclinical models in CLN3…

GSL accumulation strongly reduced in CLN3

Ganglioside GM1



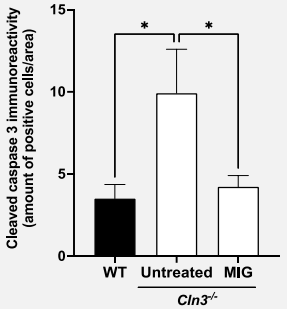
Globoside Gb4



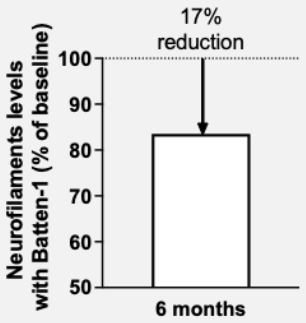
Human CLN3 cell models[1]

…leading to a reduction of cell death in the brain

Apoptosis (preclinical data)[2]



Reduction in NfL, biomarker of neuronal death (P1/2 clinical data)[3]



Batten-1 : a well-known mechanism of action already validated in humans

[1]1 Glycosphingolipid reduction with miglustat as a therapeutic strategy for CLN3 and otherneuronal ceroid lipofuscinoses, Dr Emyr Lloyd-Evans, Cardiff University, Work carried out by Emyr Lloyd Evans' laboratory at Cardiff University (funded by the Beyond Batten Disease foundation), and confirmatory work carried out by Theranexus.
[2]Work performed at Marco Sardiello's lab (Baylor College of Medicine, US) funded by BBDF
[3]Preliminary results after 6 months of treatment in Phase 1/2 of Batten-1

Encouraging preliminary Phase 1/2 clinical results

A tolerance profile already validated in P1/2; encouraging first signs of efficiency

- **6 patients** with juvenile Batten disease, **aged 17 and more**
- Study conducted by Batten disease reference centers in the United States
- Good safety profile and pharmacokinetics in line with expectations after 9 weeks

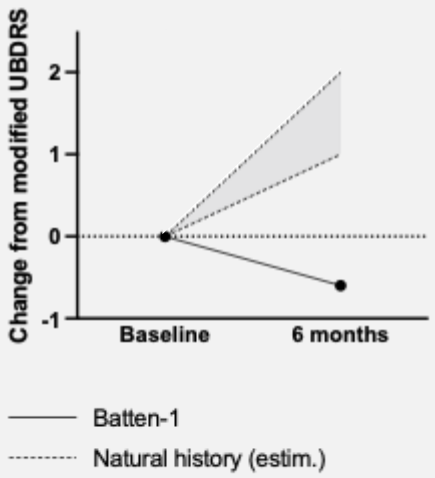
Preliminary results at 6 months

- **Decrease in neurofilament light chain (NFL)** in the blood of patients dosed
- **Stabilization of motor symptoms** compared to deterioration expected according to the natural progression of the disease

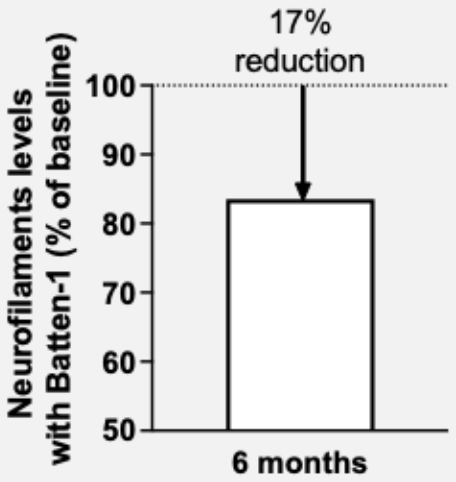
MAIN CLINICAL RESULTS

EXPECTED LATE 2023 AND 2024

Stabilization of motor symptoms



Reduction in NfL, biomarker of neuronal death
(P1/2 clinical data)



Preliminary results suggesting an effect of Batten-1 on neuronal death and first signs of clinical efficacy

[1] LogMAR (or Log MAR) is a unit for quantifying visual acuity, enabling statistical calculations such as mean, standard deviation, etc. to be made.

Batten-1: Phase 3 study design validated by the FDA and EMA

FDA and EMA approvals obtained for design and efficacy endpoints of pivotal Phase 3 study

- **A single Phase 3 study** would enable registration of the Batten-1 candidate in Batten disease
- Study conducted in parallel at several centers in the **United States and Europe**

- **Randomized**, double-blind **study** evaluating the efficacy of Batten-1 over 2 years of treatment, versus placebo

Target population: approximately 60 patients aged 4 to 16 years old

Additional cohort of 9 open-label patients generating intermediate results every 6 month

Primary endpoint:
Visual acuity

Evaluated by :

- A quantitative measurement scale in LogMar[1]
- A qualitative scale for the most severely affected patients who cannot be assessed quantitatively

Secondary endpoints :

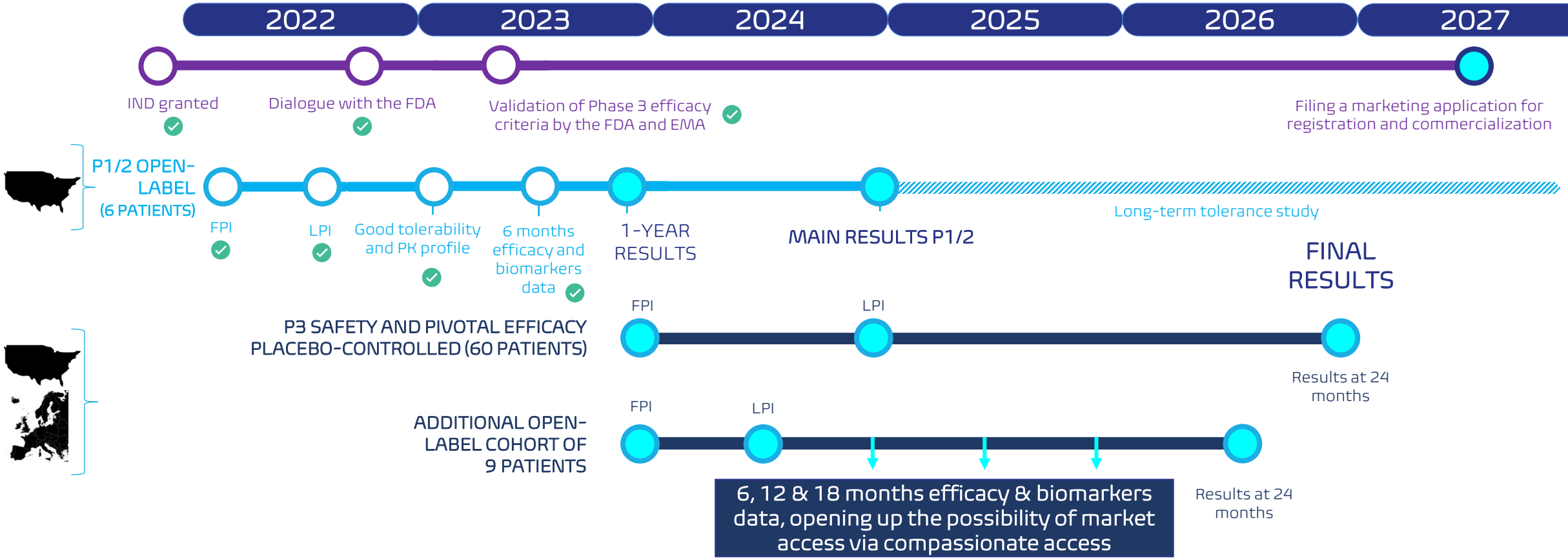
Cognitive functions

Motor functions

A P3 design validated by the FDA and EMA, offering a steady newsflow until Batten-1 registration

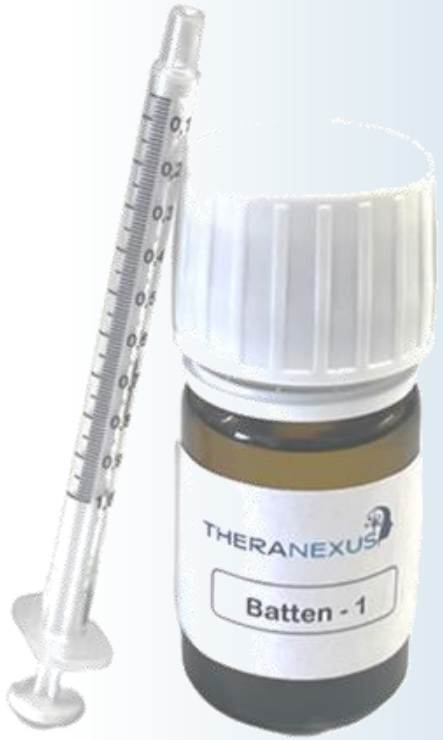
[1]LogMAR (or Log MAR) is a unit for quantifying visual acuity, enabling statistical calculations such as mean, standard deviation, etc. to be made.

Development plan with Phase 3 results by the end of 2026, followed by marketing authorization



Final results by the end of 2026, followed by marketing approval and commercialization (in particular in the US and Europe)

Batten-1: development of a proprietary oral solution for CLN3 patients



A drinkable solution that makes treatment easier to take and modulate

- Drinkable solution :
  - Suitable for pediatric patients and patients presenting swallowing difficulties.
  - High concentration, enabling treatment of all patients, regardless of age or weight.
- Graduated pipette
  - Fine dose modulation, aiming for the best possible tolerance by gradually increasing the dose at the start of treatment.

Protected by :

-1-

Method-of-use and formulation patents filed in main markets (2021 and 2022)

-2-

Orphan Drug Designations granted by the FDA (USA) and EMA (EU)

Batten-1: global sales peak at over $500 million a year

Market estimates obtained by an independent study commissioned by Theranexus, demonstrating a strong market opportunity

Territories	Currently diagnosed patients / estimated prevalence[4]	Sales based on diagnosed patients (usd millions)[1]	Sales based on estimated patients (usd millions)[1]
United States	750[2] / 1,200	480	765
EU5 (Germany, France, Italy, Spain, United Kingdom)	340[3] / 500	50	75
Nordic countries	120[3] / 180	15	20
MARKET OPPORTUNITY		$545 million	$860 million

1: Price estimates based on interviews with payers and KOLs (source: Justin Stindt Consulting report).
2: Estimated population diagnosed based on insurance claims (source: DRG - Clarivate analytical report for Theranexus).
3: Estimated population diagnosed based on data from hospital discharge reports (RW Health report for Theranexus).
4: Estimates based on literature analysis (DRG - Clarivate analytical report for Theranexus; RW Health report for Theranexus)

A favorable pricing environment for rare diseases

Product (company)	Target population (7MM)[1]	US annual price[2]
Zavesca (J&J)	1,950	$653k
Brineura (Biomarin)	550	$795k
Procysbi (Horizon)	1,250	$1,074k
Xenpozyme (Sanofi)	~100	$1,949k

[1] 7 Major Markets
[2]Drug price per patient in the United States ; source GlobalData: Drug Pricing Intelligence (POLI) Database

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Financial information

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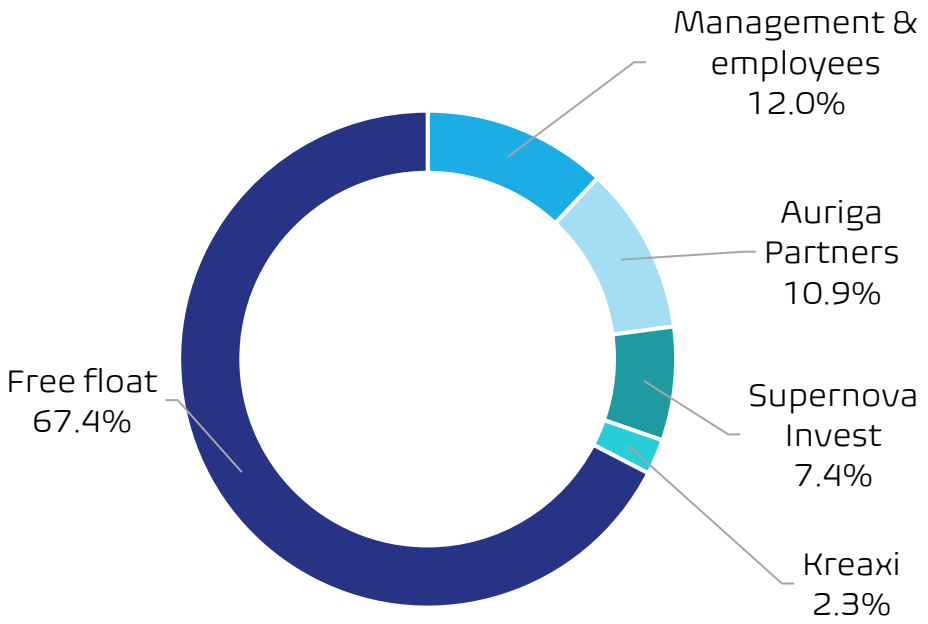
Listing and shareholders

Financial data

- ISIN: FR0013286259 - Mnemo: ALTHX
- Market: Euronext Growth
- Share price at June 8, 2023: 2.04 €
- Market capitalization: 10.9 M€
- Coverage: ODDO BHF, Portzamparc
- Liquidity contract: Portzamparc

Capital structure

Number of shares: 5,346,213



Simplified income statement

In thousands of euros (French accounting standards)	2022	2021
Operating income	**1,246**	**25**
Other purchases and external charges	3,967	5,591
Salaries and social charges	3,165	2,689
Depreciation and amortization	332	1,204
Other expenses	39	53
Operating results	**(6,257)**	**(9,512)**
Financial result	(158)	(396)
Extraordinary expenses	(771)	-
Income tax	619	1,758
Net income	**(6,567)**	**(8,150)**

Cash and cash equivalents at March 31st 2023: €7,300 K

Theranexus: a strong opportunity to create value for investors

Innovative biopharmaceutical company with many strenghts

Batten-1:

- A clinical asset approaching Phase 3, with a mechanism validated in humans
- Encouraging Phase 1/ 2 results, providing early evidence of clinical efficacy
- A first-to-market drug candidate with a market opportunity of over $500 million

An R&D platform dedicated to rare neurological diseases:

- Supported by top-level scientific collaborations
- With a new ASO candidate in development in the second half of 2023

A dynamic Batten-1 newsflow over the next 3 years

S2 2023 and S2 2024
Main results Phase 1/2 at one and two years of treatment

2024 & 2025
Open cohort efficacy & biomarker data at 6, 12, 18 & 24 months

S2 2026
Phase 3 results

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THANK YOU FOR YOUR ATTENTION!

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