OxThera

Corporate Presentation – Pareto Securities Healthcare Conference

September 2020

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OxThera – Snapshot

- **Developing novel treatment options for people with primary hyperoxaluria (PH)**
  - Ultra-rare kidney disease; significant morbidity and mortality; currently no approved therapies

- **Oxabact® – oral bi-modal enteric biotherapy of human gut bacteria *Oxalobacter formigenes***
  - Highly encouraging 2-year clinical data in dialysis patients underpins mechanism of action
  - Secured Orphan Drug Designation in both US and EU
  - Secured Rare Pediatric Disease Designation in US

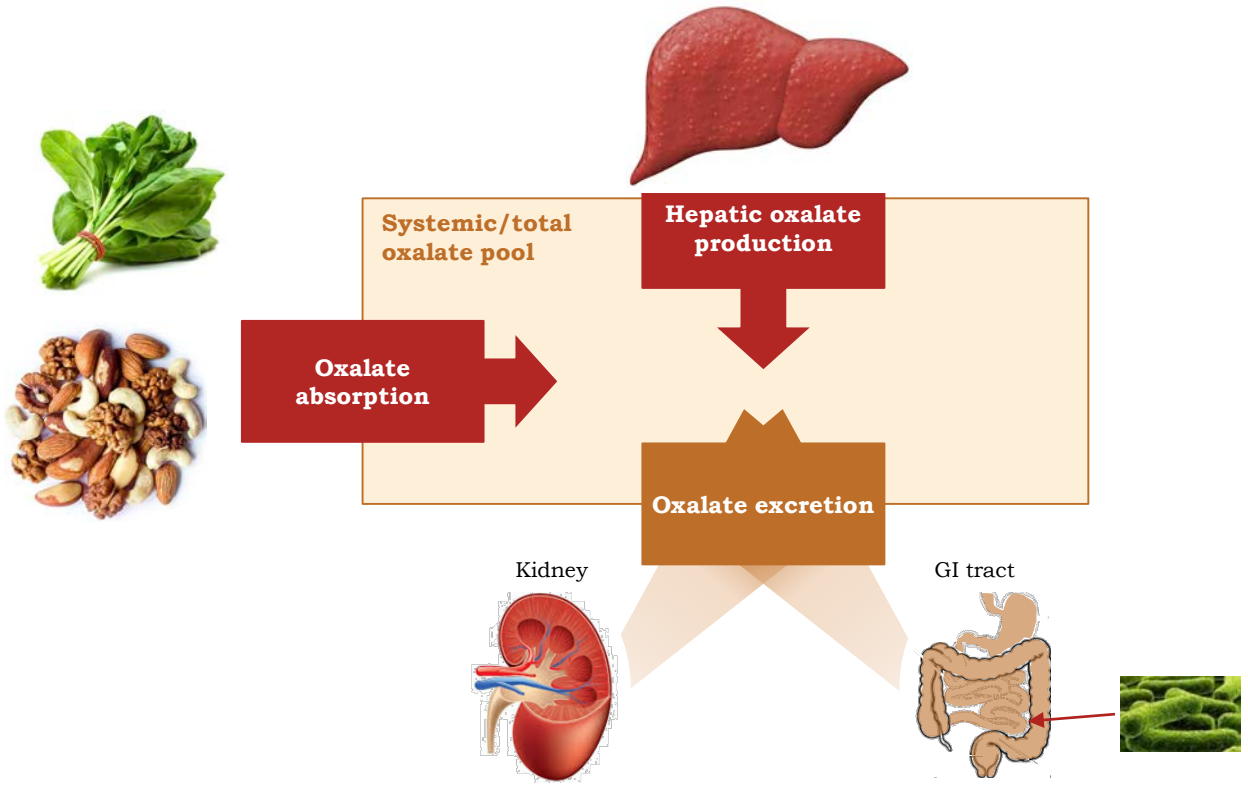
- **Phase III *ePHex* trial under way, key data due mid 2021**
  - Fully enrolled
  - Plan to pursue accelerated approval path to market based on POx as surrogate marker

- **Attractive commercial opportunity with potential addressable market of $1B+ in PH**
  - Broad applicability across all PH sub-types; consolidated centers of excellence

- **Headquartered in Sweden and backed by leading international life science investors**
  - LSP, Kurma Partners, Industrifonden, Sunstone and Ysios

Sources of Oxalate

External versus internal sources of oxalate



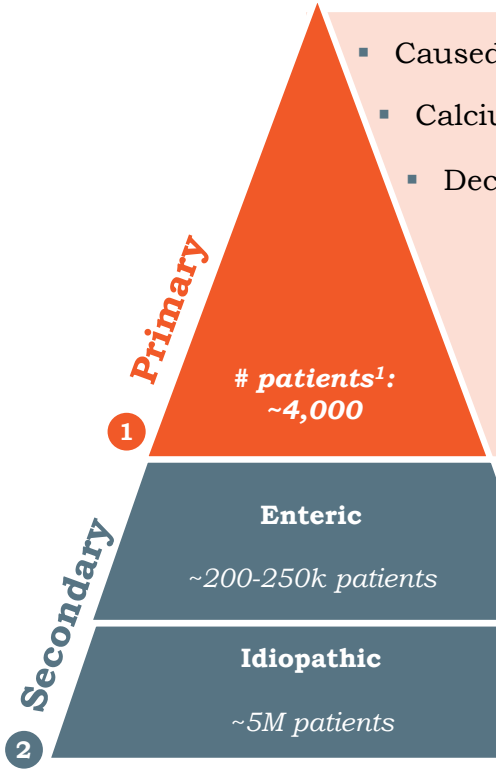
Comments

- Oxalate (oxalic acid, $C_2H_2O_4$) is an organic compound and is used in the body as a chelating compound for many cations, due to its negative charge
- It occurs naturally in many foods (spinach, nuts, chocolate)
- It is also produced in the liver of mammals as an end-product of metabolism
- It *cannot* be metabolized by mammals and is mainly excreted by renal filtration into urine, to a lesser extent secreted into the GI tract

- O. formigenes is an anaerobic, non-pathogenic gut bacteria with the ability to metabolize oxalate

Animals/humans colonized with *Oxalobacter formigenes* have a lower risk for kidney stones, nephrocalcinosis and renal failure

Primary Hyperoxaluria – A Rare and Severe Type of Hyperoxaluria



- Caused by aberrant liver enzymes leading to **over-production** and **accumulation** of oxalate
- Calcium oxalate crystals form in body fluids and soft tissues incl. kidney, heart, bone, eye
- Decline in kidney function, systemic oxalosis and kidney failure (median ESRD age ~25 yrs)

Type 1	**Type 2**	**Type 3**
- Deficiency of AGT[3]	- Deficiency of GRHPR[3]	- Deficiency of HOGA[3]
- Most prevalent type of PH	- Chronic stones	- Chronic recurring stones
- Severe disease course	- Kidney failure risk	- Low risk kidney failure
- High risk for ESRD	- Slow progression to ESRD	- Slow progression to ESRD
- ~2,400 patients[2]	- ~300 patients[2]	- ~300 patients[2]

- Related to a chronic, underlying GI disorder (e.g. gastric bypass surgery, cystic fibrosis, short bowel syndrome etc.) and overabsorption of oxalate

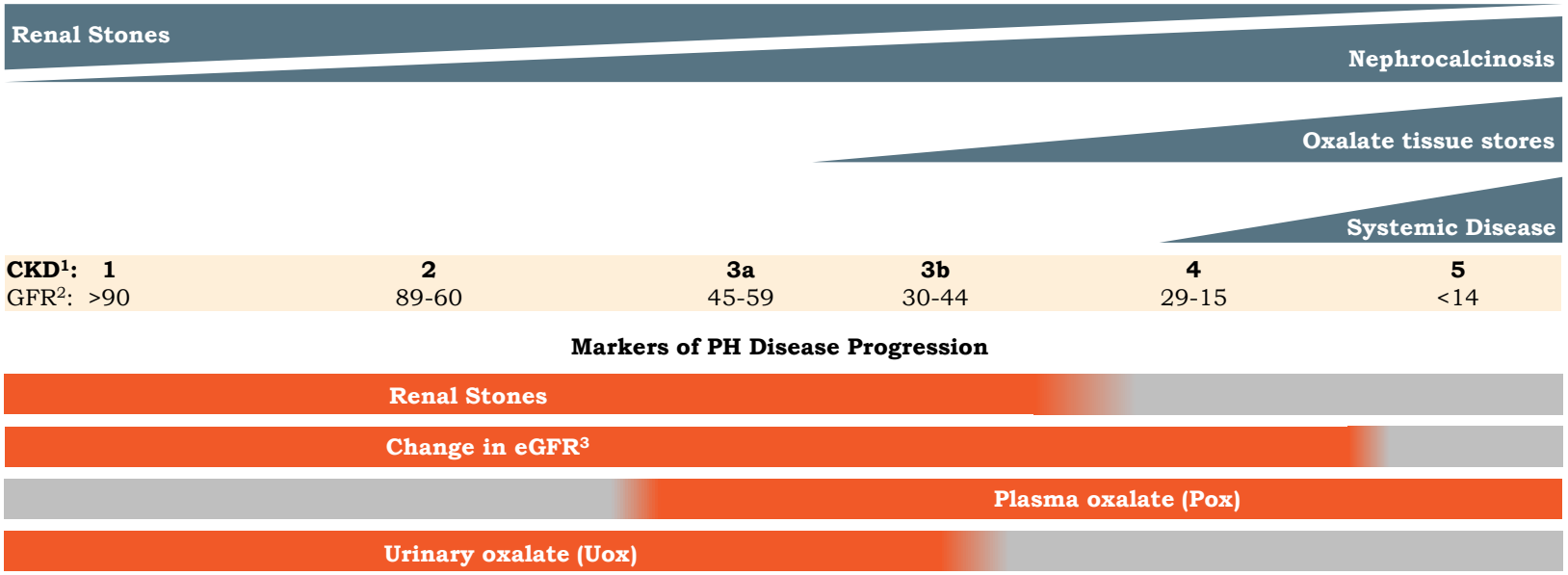
- Unknown underlying cause

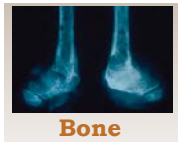
Despite low prevalence, Primary Hyperoxaluria represents a high value opportunity due to unmet need and paediatric patient pool

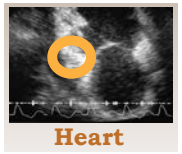
Source: Management estimates, van der Hoeven et al. 2012, Allena Pharmaceuticals
1: Prevalence of ~5 per million in the US, Canada and Western Europe
2: Amount of total Primary Hyperoxaluria prevalence
3: AGT = alanine-glyoxalate and serine-pyrovate aminotransferase; encoding gene: AGXT, GRHPR = glyoxylate and hydroxypyruvate reductase; encoding gene: GRHPR, HOGA: 4-hydroxy-2-oxogluterate aldolase; encoding gene: HOGA1

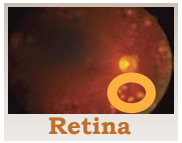
Primary Hyperoxaluria – Multi-Organ Complications with Severe Morbidity

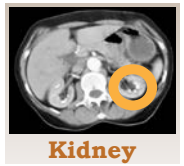
Markedly elevated oxalate has devastating consequences









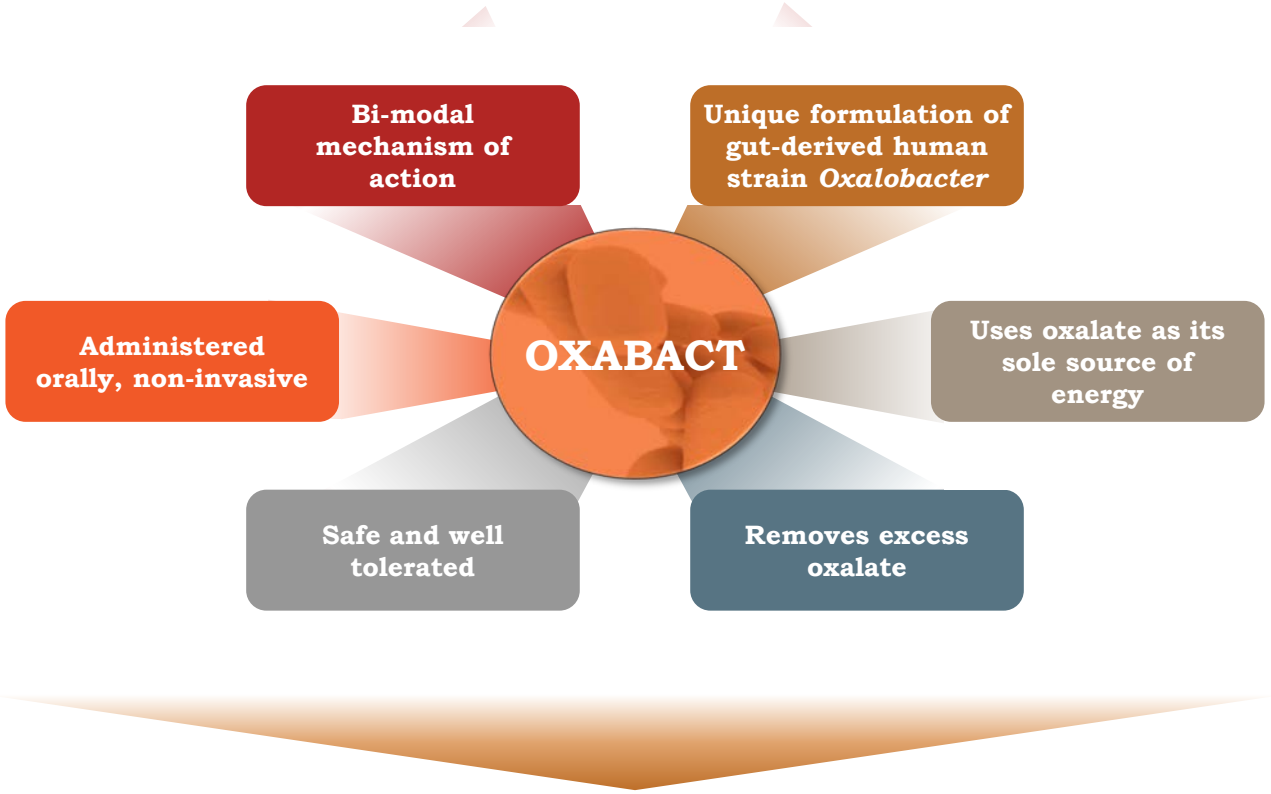


Premature death
PH1 – median age 30 yrs, if not transplanted

Early Onset ESRD
PH1 - Median age 24-35 yrs

1: CKD = chronic kidney disease
2: GFR = glomerular filtration rate (measure of kidney function)
3: eGFR = *estimated* glomerular filtration rate

Oxabact has potential in all PH types

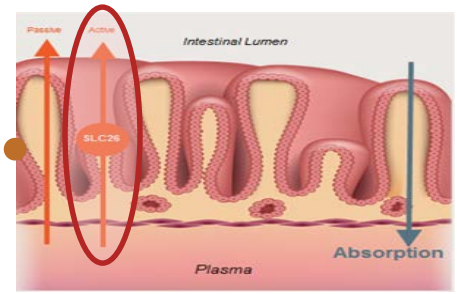


✓ **Orphan Drug Designation** ✓ **Broad patent protection**

Unique Mechanism of Action Reduces Systemic Oxalate

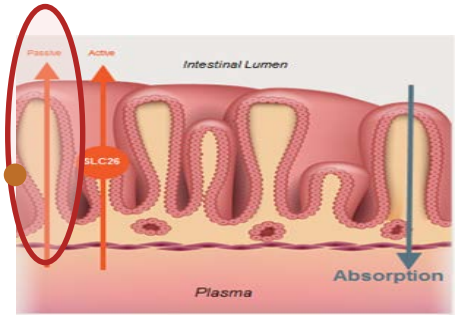
OXABACT

A **Enhances *active secretion* of oxalate from plasma to intestinal lumen**



- Emits secretagogues which enhance active transport of endogenous oxalate from blood to the intestine via SLC26[1]
- Oxalate is excreted through intestines

B **Enhances *passive transport* of oxalate from plasma to intestinal lumen**



- Metabolizes oxalate (its sole source of energy) at rates of >100 mmol oxalate/day/capsule
  - PH patients produce up to 7mmol/day of oxalate
- Elimination of oxalate from gut further increases passive excretion from plasma to the intestines

A **B**

Decreased formation of new CaOx crystals

Reduced systemic oxalate load

Dissolution of already present CaOx crystals

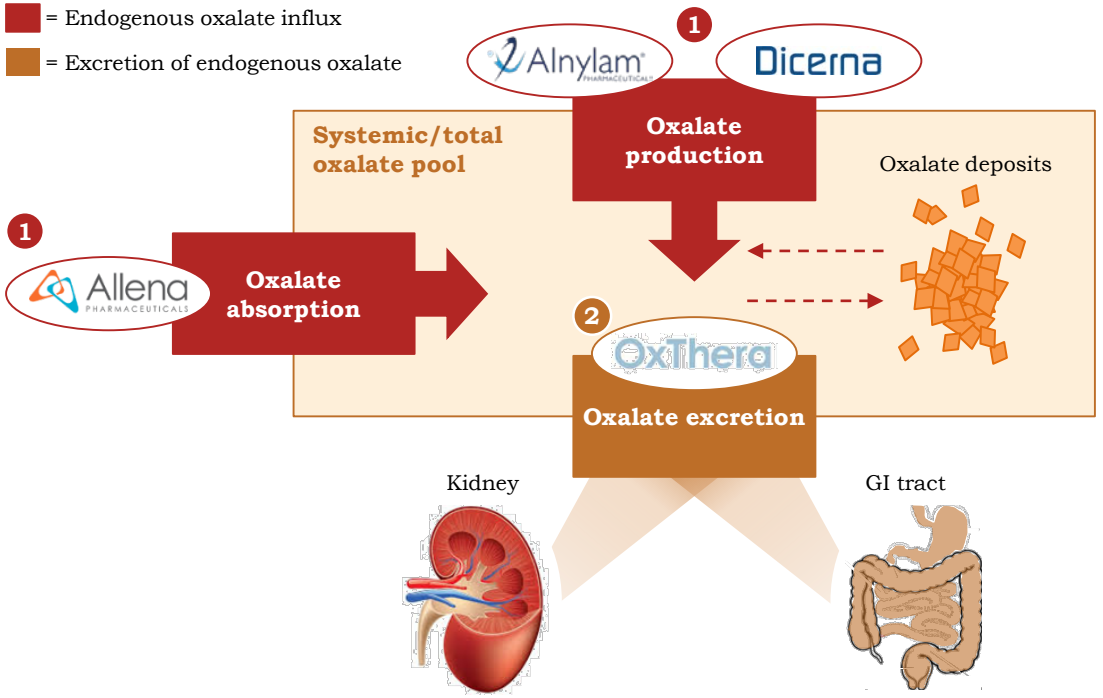
Stabilization/ improvement of kidney/cardiac function

Source: Hatch et al., Urolithiasis, 2016. Arvans et al., JASN, 2016
1: SLC26 = multifunctional anion exchangers

As Crystals are a Key Disease Driver, Oxabact's Unique MoA has Advantages to Other Therapeutic Approaches

Oxabact acts to excrete oxalate from plasma, preventing crystallization and dissolving existing crystals – other compounds aim to reduce the endogenous influx of oxalate

Oxabact focus vs. other therapeutic approaches



Comments

1. Other approaches aim to reduce internal *production* of oxalate or reduce *absorption* of dietary oxalate

2. Oxabact doesn't affect endogenous oxalate influx but instead seeks to increase the *excretion* of the endogenous oxalate deposits in the body

   - Clinical benefit of siRNA in patients with crystal deposits is to be proven
   - Oxabact acts on crystals deposits and has demonstrated preliminary evidence for crystal dissolution

Oxabact has an important role in symptomatic patients and has applicability across all forms of PH

Oxabact Represents a Potential Paradigm Change

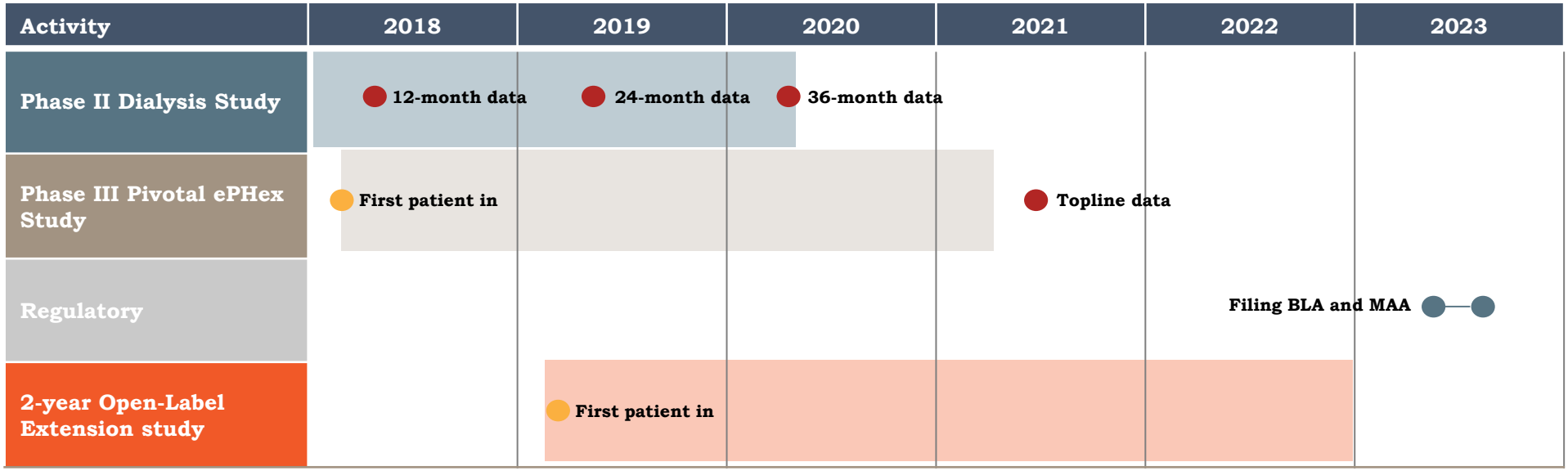
No approved therapeutic treatment options; most patients poorly managed with current standard of care; high morbidity and mortality with transplant

Disease progression:	**Maintained renal function**	**Declining renal function**	**End-stage renal disease**		**Post-transplantation**
	Pre-Dialysis patients		**On dialysis**	**Within 1 year post transplantation**	**Cured post transplantation**
Share of patients:	**~50-55%**		**~15%**	**~15-20%**	**~15%**
Current standard of care:	▪ High Fluid intake ▪ Potassium citrate ▪ Pyridoxine		▪ Typically 6 days per week haemodialysis treatment ▪ May use additional peritoneal dialysis	▪ Combined or staged liver/kidney transplant most common for PH1	▪ Intensive dialysis often performed for 1-2 weeks following
Associated treatment cost:	**Up to USD ~30k per intervention[2]**		**USD ~90k/yr[3]**	**USD ~1m[4]** →	
Potential Oxabact role:	***Chronic treatment aimed at maintaining renal function and delaying/preventing renal failure***		***Reduce / stabilize oxalate levels and dialysis frequency; potential improvement post-transplantation outcome***	***Management of Uox levels to protect from oxalate damage to new renal graft***	

Source: United States Renal Data System, bioStrategies Group, Duke Medicine, Milliman Research Report 2017
1: Pyridoxine (Vitamin B6) is a co-factor for AGT
2: Costs are not significant unless surgical intervention is required for kidney stones. Average cost for such interventions (lithotripsy) are USD ~15k per kidney stone, reaching as high as USD ~30k in most severe cases
3: Annual cost of dialysis in the US 3 times per week
4: 5 year cost of kidney and liver transplantation of USD ~1m

Oxabact – Clinical Development Strategy and Timeline

Completed recruitment Pivotal Phase III ePHex study in Q2 2020 with topline data by mid 2021

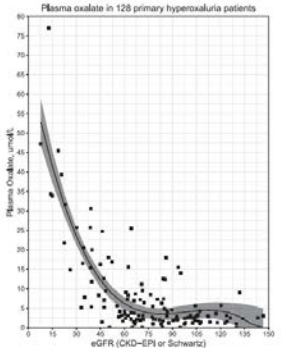


Phase III Pivotal ePHex Study – Hypothesis Foundation

Epidemiological data and completed clinical trials provided OxThera with unique knowledge base essential for designing the current development program for Oxabact

Higher Pox levels associated with lower eGFR

- Pox levels correlate with disease progression (i.e. renal function decline) in both epidemiologic[1] and previous Oxabact studies[2]



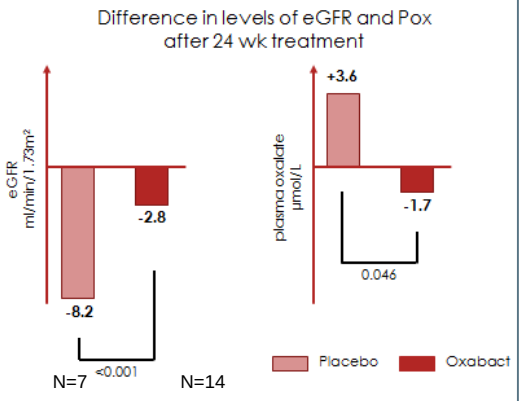
Relationship between plasma oxalate and eGFR in 128 patients with PH

OxThera Study	N (Active/Placebo)	Correlation (r)	P-value
OC3-DB-01	19/23	-0.52	0.0007
OC3-DB-02	21/15	-0.38	0.026
OC5-DB-01	14/13	-0.54	0.049

Completed placebo-controlled Oxabact® studies

Oxabact treatment has the ability to reduce Pox and slow decline in kidney function

- Lower decline in eGFR observed in Oxabact patients vs. placebo
- Greater reduction / stabilization of Pox levels observed in Oxabact patients vs. placebo[2]

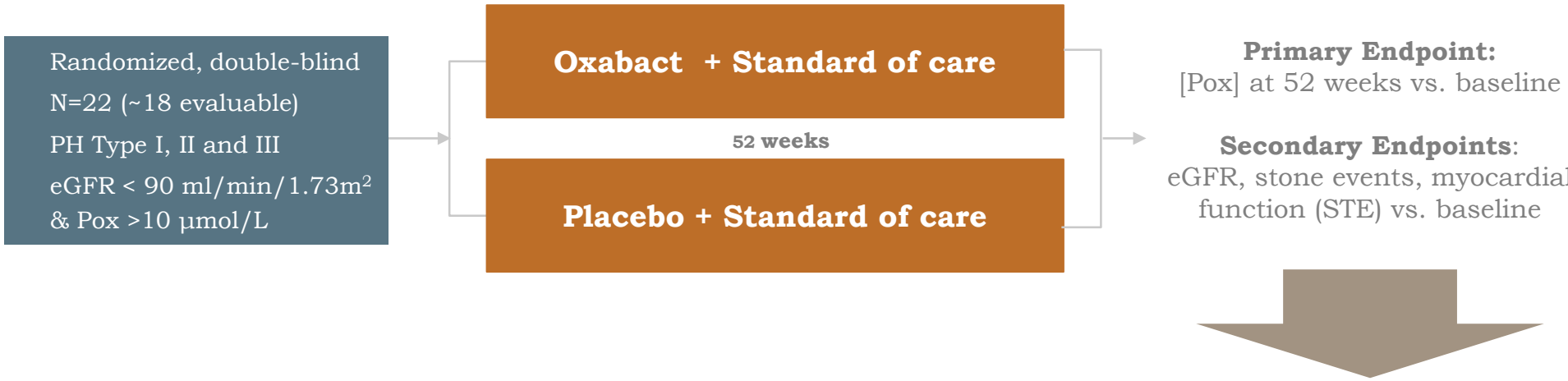


Oxabact well positioned to enter Phase III

- ✓ Demonstrated reductions in Pox levels and evidence of reduced disease progression
- ✓ Data support unique mechanism of action
- ✓ Provides solid base for designing the pivotal study
- ✓ Renal function supported by FDA as an approvable endpoint

1: Milliner D. et al., 2020
2: OxThera data on file

Phase III Pivotal ePHex Trial – Design and Status

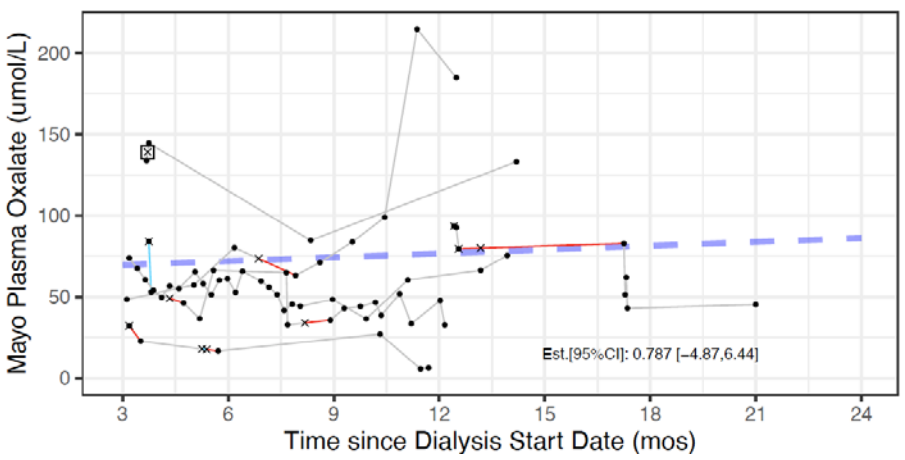


- **Currently 25 patients randomized – minimum threshold met**
- **No further patients required**
- **11 patients rolled over to Open Label Extension**

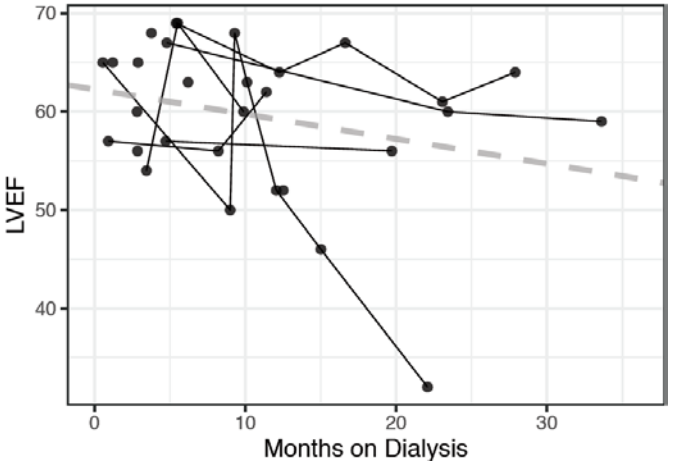
In Late-Stage Patients with ESRD, Pox Increases and Cardiac Function Worsens Despite Intensification of Dialysis Regimen

Plasma Oxalate: Mayo Clinic Natural History Cohort

Patients followed for at least 1 year



Cardiac Function: Mayo Clinic Natural History Cohort



- Consensus in literature in that ***Pox increases with time on dialysis*** (in PH and non-PH ESRD patients)[1]
- No method of dialysis is ideal, however, ***intensive daily dialysis*** is recommended[2]
- In Mayo cohort, dialysis frequency was increased from 4.3 to 5.2 days/week on average in the first year – still, ***Pox could not be controlled***

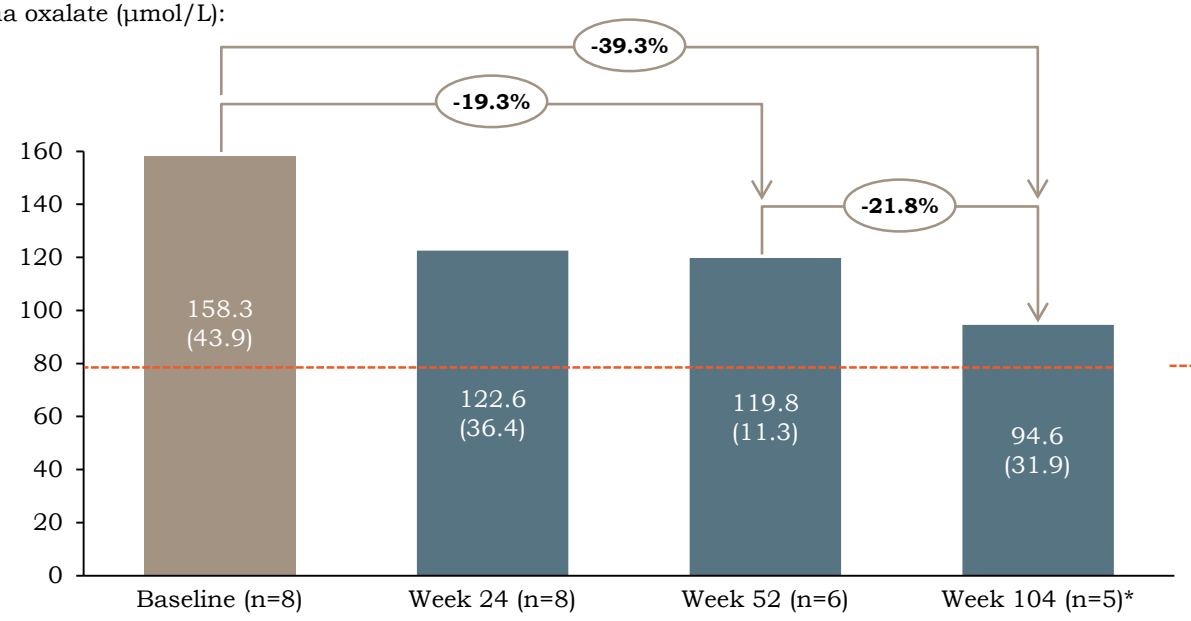
1: Costello et al. 1991, Hoppe et al. 1996, Bunchman et al. 1994
2: Cochat et al. 2012

With Oxabact Treatment, Significant Reduction in Pox in the Absence of Dialysis Intensification

Highly encouraging 2-year data indicates that Oxabact facilitates the dissolution of oxalate crystals, thus validating the bi-modal MoA

Total Pox over time

Mean (SD) values of plasma oxalate (µmol/L):



Note: Three patients discontinued early: non-compliance with study procedures/study drug (2) and liver transplantation (1)

***p<0.0001**
p-value based on mixed model of repeated measures including all values over time from baseline to week 104

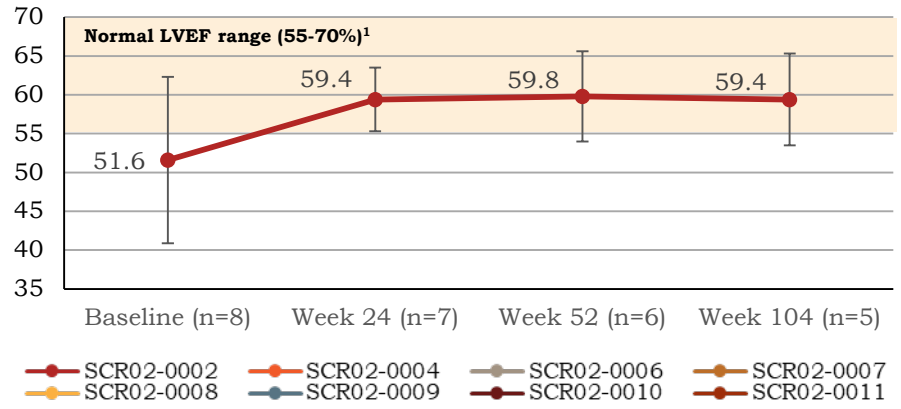
The study design (i.e. no change to dialysis regimen) ensured that observed reduction in Pox is attributable to Oxabact treatment.

Dialysis Study – Positive Impact on Cardiac Function

2-year data demonstrates Oxabact therapy has the potential to improve and stabilize cardiac function in PH patients on dialysis

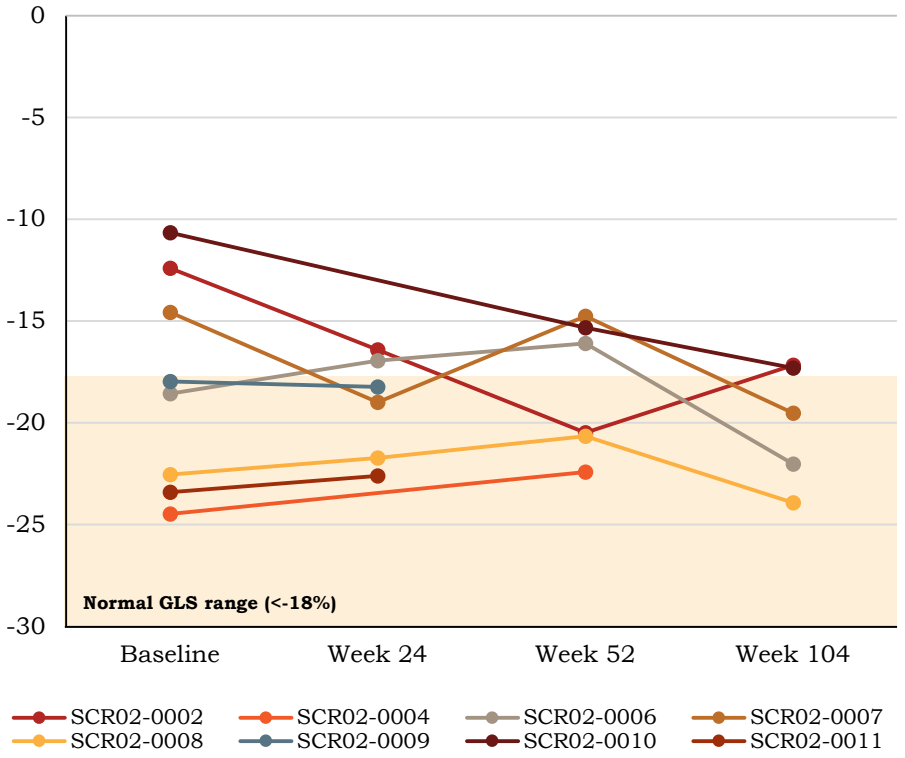
Left Ventricular Ejection Fraction (LVEF)

Mean (SD) LVEF (%) over time:



Speckle Tracking Echocardiography

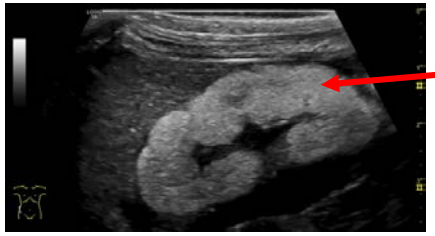
GLS (%):



1: Source: Cleveland Clinic

Oxabact Treatment in a Child with Infantile Oxalosis: Reduction in Pox and Prevention of Worsening of Systemic Oxalosis

Kidney Ultrasound



Ultrasound and retinography:

Severe nephrocalcinosis

and

macular retinal deposits

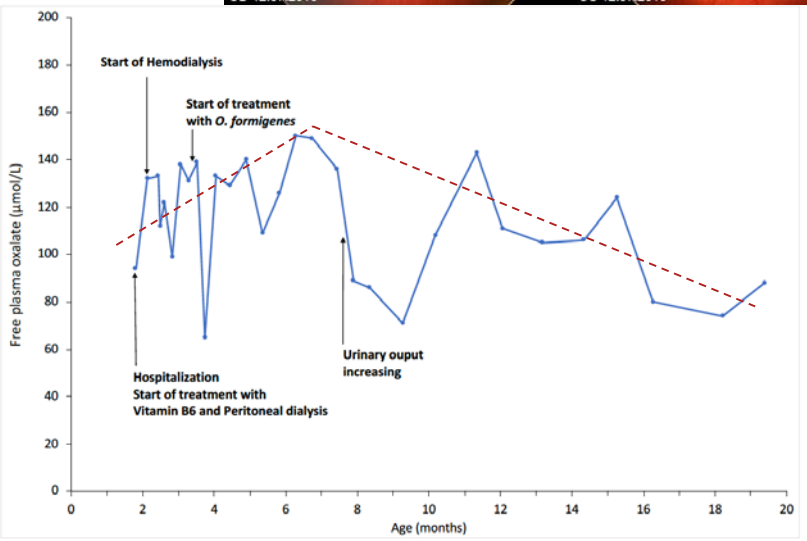
Retinography

- 2 mo. old girl with negative family h/o PH
- Hospitalized for failure to thrive
- Diagnosed with ESRD, no urine production
- Pox 94 µmol/L
- Named patient use request for Oxabact received Dec. 2017, treatment until end of October 2019 (21 months)

- Dialysis regimen:
  - 3 x per week HD
  - Daily home PD

- Other medications: Vit B6, ESA, growth hormone

"Halting of disease progression" described by treating physician

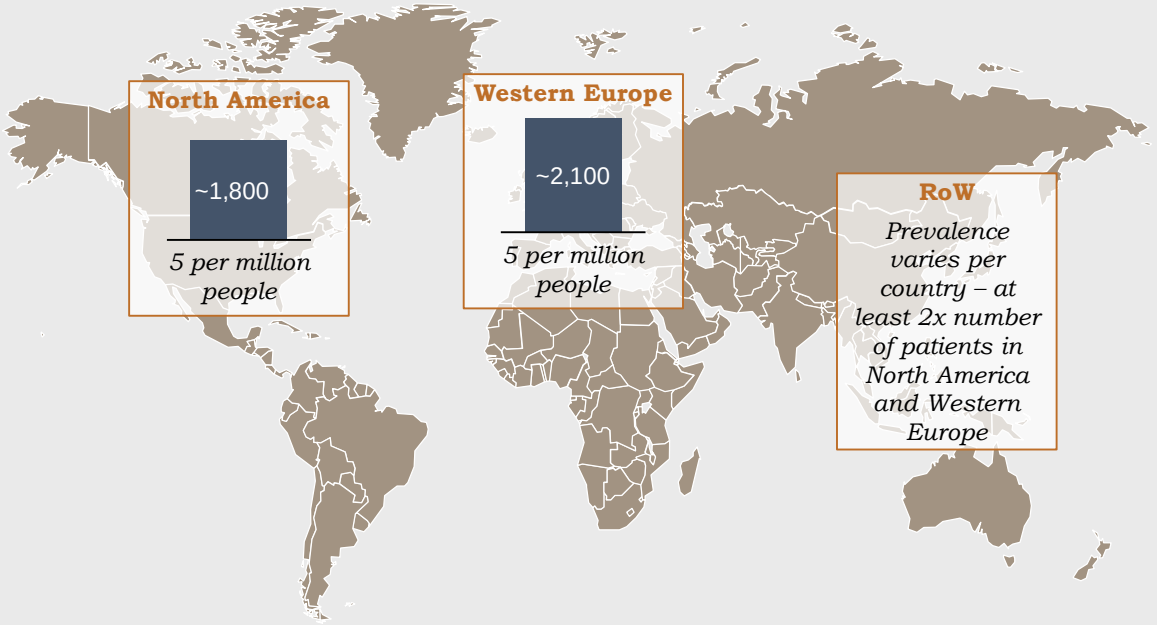


Successful combined liver/kidney transplantation October 2019 with normal renal function and normal Pox since tx.

Case report published in *Pediatric Nephrology Pape et al 2020*

Primary Hyperoxaluria – $1 Bn+ Market Potential despite Low Prevalence

Prevalence of Primary Hyperoxaluria



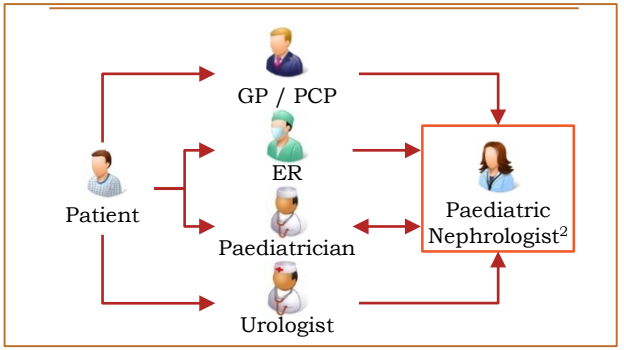
- ✓ ***Total number of patients in key regions Western Europe[1] and North America[1] ~4,000***
- ✓ ***Primary Hyperoxaluria is not constrained by ethnic or regional differences***
- ✓ ***More prevalent in countries where consanguineous marriages are common (some North African and Middle Eastern nations)***

As treatment options surface, patient reporting typically increases and reported incidence figures rise

Market potential

- Assuming ~4,000 PH patients in Western Europe and North America
- Pricing in line with other ultra-orphan drugs
- **With conservative pricing assumptions, Primary Hyperoxaluria alone has a total market potential of at least $1 Bn**

Few provider touchpoints



- ✓ ***Patients typically diagnosed by a Paediatric Nephrologist***
- ✓ ***Limited number of treatment centers allow for more straightforward commercialization***

1: North America includes United States and Canada, and Western Europe includes the western EU countries
2: Nephrologists typically diagnose adult patients
3: Adult form of disease typically not diagnosed before later in life

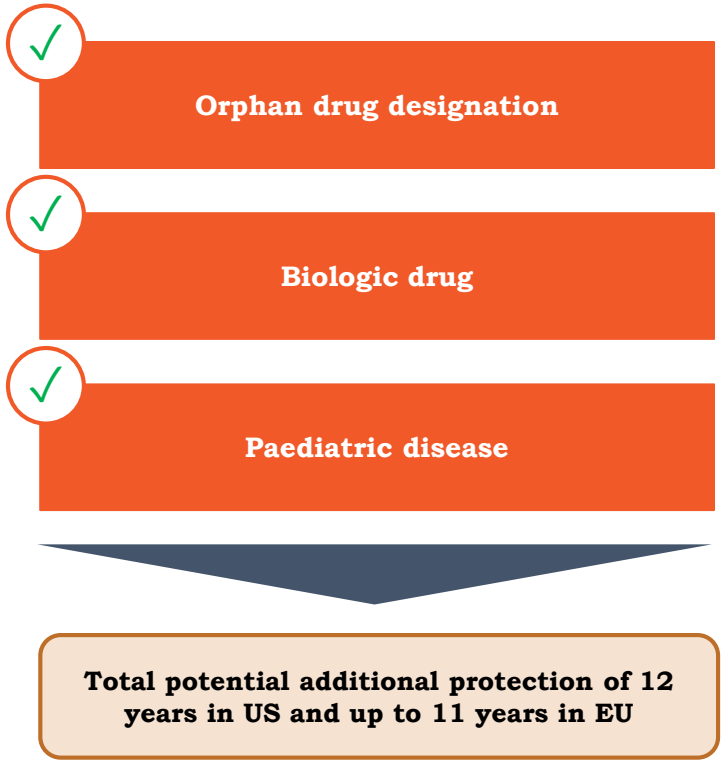
Oxabact Product Protection

Broad patent portfolio with 100+ patents granted in major markets – additional exclusivity secured through orphan drug designation, biologic product and paediatric disease focus

Patent protection and intellectual property

- Multiple patents covering bacterial product, pharmaceutical formulations comprising the product and medical uses/methods thereof
- Patents directed to the treatment of Hyperoxaluria and other oxalate-related conditions
- Freedom-to-operate for current product performed in major markets
- Strong know-how and expertise within relevant CMC processes
- Royalty obligation of up to 1% of net sales during the patent lifetime in India (until 2020) and US (until 2025) and then up to 0.5% for 10 years post patent expiry

Additional market exclusivity



Anticipated Near-Term News Flow

Multiple near-term value creating milestones, leading up to Top Line Data in Mid 2021

H1: Secured RPDD in US

Q2: Complete dialysis study

Q2: Complete enrolment Phase III ePHex study

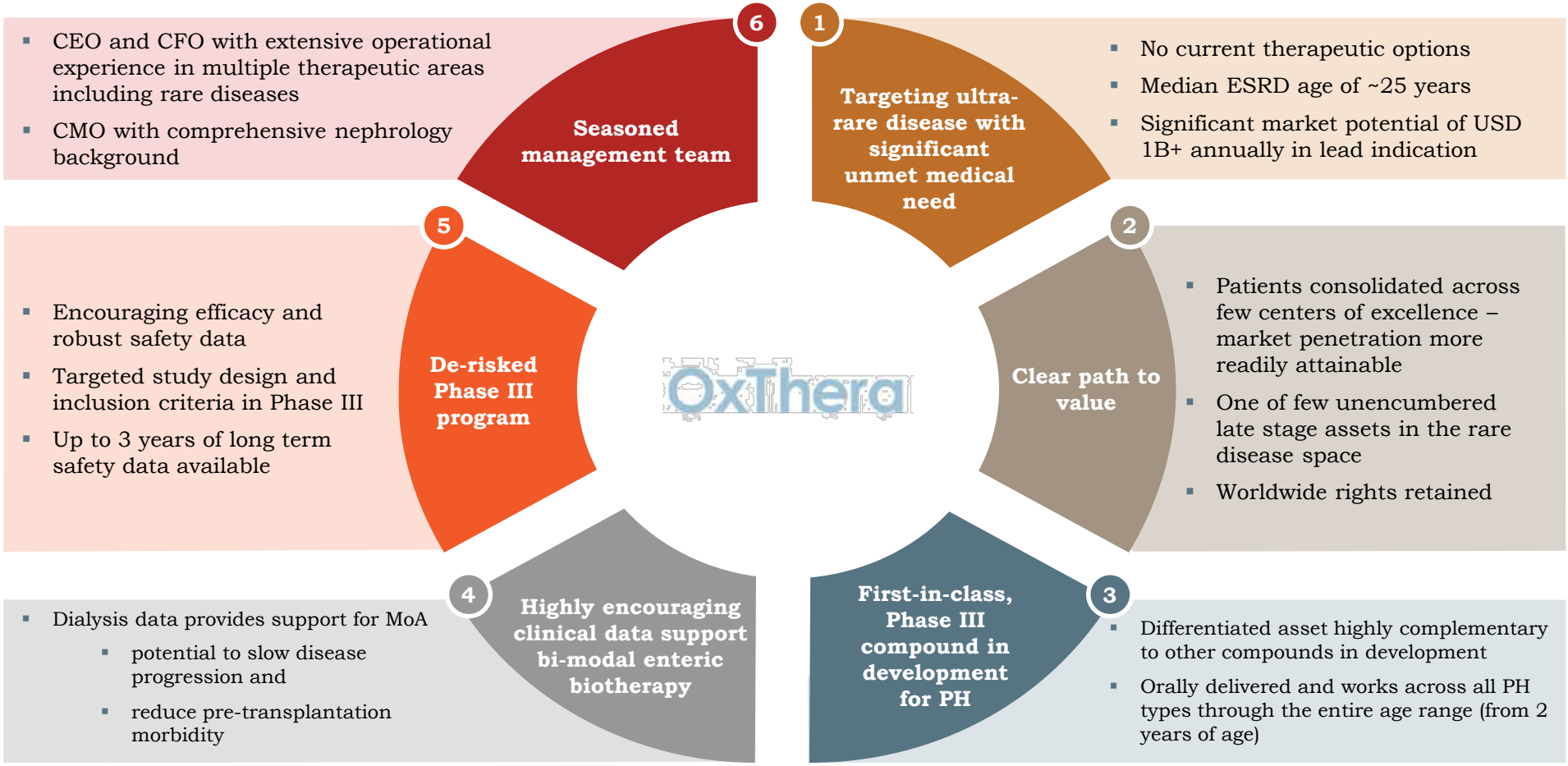
Q3: Publication 24 mos data dialysis Study

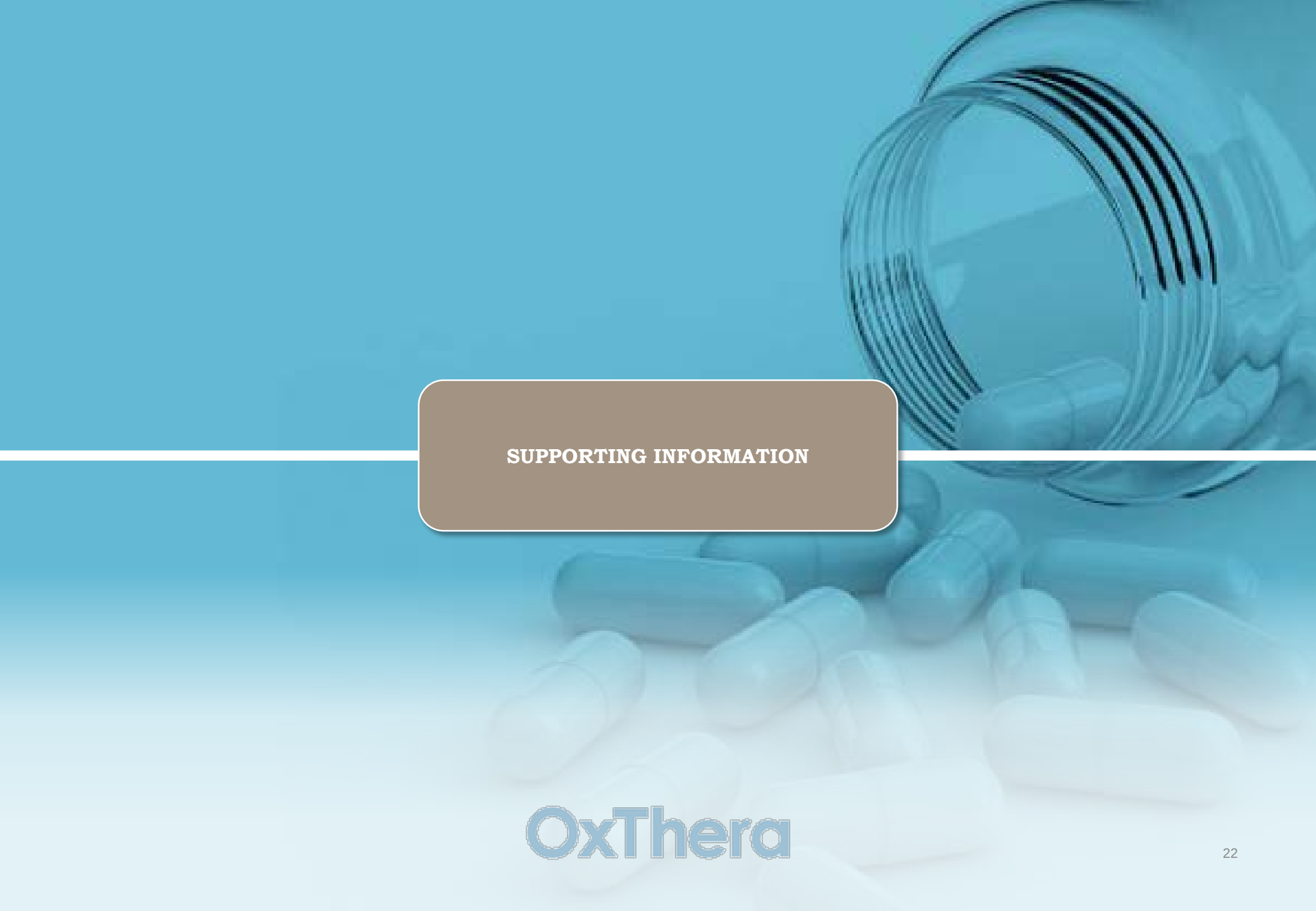
Q2: LPLV Phase III ePHex

Mid Year: Topline results

H2: 12-mo interim analysis ePHex OLE study

OxThera - Key Highlights





SUPPORTING INFORMATION

OxThera

Safety Profile

Through extensive clinical testing in 100+ patients with treatment durations up to 36 months, Oxabact has been safe and well tolerated

Comments

- To date, Oxabact has been administered to more than 110 subjects for time periods of 4 weeks to 36 months
- Administration of *Oxalobacter* at various doses as a frozen cell paste, capsule or buffered powder for suspension has been well tolerated with no treatment related serious adverse events (SAEs) reported
- In all studies, the treatment was well tolerated and the majority of adverse events (AEs) were reported as mild to moderate and unlikely related to the treatment
- No discernible differences in adverse events between treatment and placebo arms were observed
- Most frequently reported AEs in completed studies were GI disorders (e.g. abdominal pain, nausea, vomiting, diarrhea)[1]
- No drug related suspected unexpected serious adverse reactions (SUSARs) reported

Key takeaways

✓ **No treatment related SAEs**

✓ **Most reported AEs were mild to moderate and not related to treatment**

✓ **No difference in adverse events between Oxabact and placebo**

1: Others include infections and infestations, nervous system disorders, and respiratory, thoracic and mediastinal disorders

Renowned Biotech Investor Base

Comments

- OxThera is supported by a strong group of international life science investors
  - Largest owner is institutional investor LSP
- OxThera has successfully completed four financing rounds since 2005, with the most recent round in November 2016, raising a total of EUR 32m in 3 tranches
  - Enabled the company to initiate pivotal study
- The company currently has an employee incentive program with 39.8m employee stock options outstanding allocated to management and board
  - The program is currently under review

Select investors

Supported by leading specialist investors

LSP
Kurma Partners
YSIOS CAPITAL
Industrifonden
Sunstone LIFE SCIENCE VENTURES

Overview of shareholders

#	Shareholder	# of shares	% ownership
1	LSP V Coöperatieve	51,850,995	22.98%
2	Kurma Partners	29,685,078	13.16%
3	Idinvest Partners	29,540,037	13.09%
4	Ysios BioFund II Innvierte	25,925,497	11.49%
5	Sunstone Life Science Ventures Fund III	25,925,497	11.49%
6	Stiftelsen Industrifonden	25,018,021	11.09%
7	Flerie Invest AB	15,555,298	6.89%
8	Brohuvudet AB	15,117,314	6.70%
9	Mayo Clinic	4,538,553	2.01%
10	Khalid Islam	1,279,228	0.57%
11	Malmsten Invest AB	873,440	0.39%
12	Q-Med AB	290,782	0.13%
13	Mayo Foundation for Medical Education and Research	39,000	0.02%
	Sum	**225,638,740**	**100.00%**

OxThera Management – An Introduction

MATTHEW GANTZ,
Chief Executive Officer

- CEO of OxThera since 2017
- Former PathoGenesis, Chiron and BTG executive
- Built and led several successful high growth businesses in the US and Europe across the specialty pharma and medical device space in multiple therapeutic areas
- Gained approval and successfully launched two rare disease drugs, TOBI and Voraxaze, in Europe and US respectively
- Board Member of SOBI, a publically listed Swedish Rare Disease Company

MATS-OLOF WALLIN,
Chief Financial Officer

- CFO of OxThera since 2020
- Previously CFO of several listed Swedish life science companies (Karo Pharma, Sobi and Biotage)
- Various senior executive positions in the Pharmacia Group

BASTIAN DEHMEL,
Chief Medical Officer

- Medical Doctor, CMO of OxThera since 2017
- Previously with Amgen where he led global development efforts for Amgen's Calcimimetic franchise
- Extensive experience in the Nephrology space as medical lead for regulatory approvals of etelcalcetide and cinacalcet

ELISABETH LINDNER,
Strategic Consultant

- Joined OxThera in 2012 and COO between 2017 and 2020
- Held senior executive roles including CEO of Diamyd, Head of Process R&D at OctaPharma, and Head of Development at Metcon Medicin
- Board Director at Cobra Biologics

Strong Board of Directors

GEORGES GEMAYEL,
Chairman

- Chairman for OxThera since October 2013
- Chairman of Enterome Bioscience, Orphazyme and Dynacure, and Board Member of Momenta Pharmaceuticals and Supernus Pharmaceuticals
- Previously Executive Chairman of Syndexa Pharmaceuticals and FoldRx Pharmaceuticals
- Also served as CEO of Altus Pharmaceuticals and EVP of Genzyme Corporation

MARTIJN KLEIJWEGT,
Board member

- Founded Life Sciences partners in 1998 and is currently Managing Partner and co-owner of the firm
- Director of a large number of companies, amongst others Qiagen, Crucell, Movetis, Prosensa Holding and Kiadis Pharma
- One of Europe's most experienced healthcare investors, with over 30 years of hands-on finance and investment experience

RÉMI DROLLER,
Board member

- Managing Partner at Kurma Partners
- Serves on the Board of Directors of AM Pharma, Orphazyme, Dynacure, Pharvaris and ImCheck
- Previously partner at Idinvest Partners where he developed investment activity in the life sciences

STEN VERLAND,
Board member

- Senior Partner of Sunstone Life Science Ventures and one of the co-founders of Sunstone
- Serves on the Board of Directors of Orphazyme, Vaximm, Anergis, Minervax and the Danish Venture Capital Association
- Serial entrepreneur and business angel and has co-founded ten start-ups, primarily in the biotech and CRO fields

MAARTEN DE CHATEAU,
Board member

- CEO of Sixera Pharma and Buzzard Pharmaceuticals
- Chairman of Atrogi and Board Member of AddBio, Cavis Technologies and Gesynta Pharma
- Co-founder and previously CEO of Cormorant Pharmaceuticals developing an antibody approach in immuno-oncology, acquired by Bristol-Myers Squibb
- Previously Medical Director at Swedish Orphan Biovitrum, within medical/clinical development

JONAS BRAMBECK,
Board member

- Investment Manager at Industrifonden responsible for Life Sciences area
- Serves on the Board of Directors of Oncopeptides and deputy member of the Board of Directors of Avidicare Holding and Airsonett Holding
- ~20 years experience from venture capital within life sciences, also holding a PhD in organic chemistry

ALAIN MUNOZ,
Board member

- Chairman of the Strategic Board of Hybrigenics, Board Member of Valneva and Zealand Pharma and adviser to KURMA Biofund
- MD in cardiology and anesthesiology with over 25 years experience in the pharmaceutical industry
- In-depth knowledge of the EU, US and Japanese regulatory process and markets
- Previously a senior executive in Sanofi Group and Fournier Laboratories as well as member of the scientific committee of the French Drug Agency

LANCE BERMAN,
Board member

- Venture Partner at Ysios
- Over 20 years of clinical development, medical affairs and corporate leadership experience in both the big pharma and small biotech start-up spaces
- Previously CMO and SVP at Relypsa, CMO at CPEX Pharmaceuticals and several roles at Pfizer, Schering-Plough Corporation and Janssen Pharmaceuticals

ANDERS FINK VADSHOLT,
Board member

- CFO of Orphazyme
- Over 20 years of corporate finance, business development and corporate leadership experience from both the financial and biotech industry
- Previously roles CEO at Topotarget, Partner at Bankinvest Biomedical Venture and other roles at 7TM Pharma and Carnegie Bank