Corporate Presentation

October 2023

Callum
Living with
ENPP1 Deficiency

Legal Disclaimer

This presentation and any statements made orally during this presentation contain estimates and other statistical data made by independent parties and by us relating to market size and other data about our industry. This data involves a number of assumptions and limitations, and you are cautioned not to give undue weight to such data and estimates. In addition, projections, assumptions and estimates of our future performance and the future performance of the markets in which we operate are necessarily subject to a high degree of uncertainty and risk. Neither Inozyme Pharma, Inc. nor its affiliates, advisors or representatives make any representations as to the accuracy or completeness of that data or undertakes to update such data after the date of this presentation.

Forward-Looking Statement Disclaimer

Statements in this presentation about future expectations, plans, and prospects, as well as any other statements regarding matters that are not historical facts, may constitute forward-looking statements that involve substantial risks and uncertainties. These statements include, but are not limited to, statements relating to the initiation and timing of our clinical trials, our research and development programs, the availability of preclinical study and clinical trial data, and the period over which we believe that our existing cash, cash equivalents and short term investments will be sufficient to fund our operating expenses.

The words “anticipate,” “believe,” “continue,” “could,” “estimate,” “expect,” “intend,” “may,” “might,” “plan,” “potential,” “predict,” “project,” “should,” “target,” “will,” “would” and similar expressions are intended to identify forward-looking statements, although not all forward-looking statements contain these identifying words. We may not actually achieve the plans, intentions or expectations disclosed in these forward-looking statements, and you should not place undue reliance on these forward-looking statements. Actual results or events could differ materially from the plans, intentions and expectations disclosed in these forward-looking statements. For a discussion of risks and uncertainties, and other important factors, any of which could cause our actual results to differ from those contained in the forward-looking statements, see the “Risk Factors” section in our most recent Annual Report on Form 10-K filed with the Securities and Exchange Commission, as well as discussions of potential risks, uncertainties and other important factors, in the Company’s most recent filings with the Securities and Exchange Commission.

In addition, the forward-looking statements included in this presentation represent our views as of the date of this presentation. We anticipate that subsequent events and developments will cause our views to change. However, while we may elect to update these forward-looking statements in the future, we specifically disclaim any obligation to do so. These forward-looking statements should not be relied upon as representing our views as of any date subsequent to the date of this presentation.

Inozyme is at the forefront of developing transformative therapies for rare diseases of pathologic mineralization and intimal proliferation

- ✓ **ENPP1 Deficiency and ABCC6 Deficiency are serious diseases with no approved therapies**
  - Sizable patient populations with high mortality/morbidity and substantial patient and caregiver burden
  - >550 ENPP1 patients confirmed, with 210 additional patients identified based on medical record screen
- ✓ **INZ-701 has demonstrated rapid, significant, and sustained increase in PPi levels, and exhibited a favorable safety profile**
  - Finalized ENPP1 Deficiency pediatric pivotal trial design with PPi as primary endpoint in US, supported by trends in appropriate secondary endpoints, and co-primary endpoint (RGI-C of $p<0.2$) in EU for pediatric pivotal trial
  - ENPP1 Deficiency pediatric pivotal trial planned for Oct. 2023 – Topline data expected mid-2025
- ✓ **In a position of financial strength, with several expected upcoming milestones and a pipeline designed for long-term value creation**
  - $209.8M* expected to fund operations into Q4 2025; 61.7M common shares outstanding**
- ✓ **Experienced team with a track record of success in rare disease and a strong focus on execution**

*Proforma cash, cash equivalents & short-term investments as of 6/30/23 (Includes net proceeds of $64.5M from offering which closed in August 2023 and $5.1M of net proceeds from the at-the-market offering in July 2023)
**As of 8/3/23

Dysregulation of mineralization and vascular maintenance can lead to a lifelong risk of high mortality and morbidity in a wide range of diseases

ENPP1 Deficiency

GACI-1
Prevalent Symptoms

~50% mortality by 6 months of age [1]

58% cardiovascular complications [1]

ARHR2
Prevalent Symptoms

70-100% skeletal complications (Rickets /osteomalacia)[1,2]

50-75% hearing loss [1,2]

Musculoskeletal complications (50% treatment for joint pain) [3]

- Genetic Prevalence: **1:64,000** [4]
- Associated with low PPi
- No Approved Therapies

ABCC6 Deficiency

GACI-2
Prevalent Symptoms

~10% mortality by 12 months of age [1]

79% cardiovascular complications [1]

PXE
Prevalent Symptoms

37% visually impaired; 15% blind[5]

Stroke - 15% PXE vs 3% pop [6]
PAD - 50% PXE vs 7% pop [7]

2.0x higher risk for osteoarthritis (shoulder and knee) [8]

- Disease Prevalence: **1:25,000 – 1:50,000** [9]
- Associated with low PPi
- No Approved Therapies

Calciphylaxis

End-stage Kidney Disease

~50% mortality rate 1 year after diagnosis [10]

Severe pain despite use of analgesics [11]

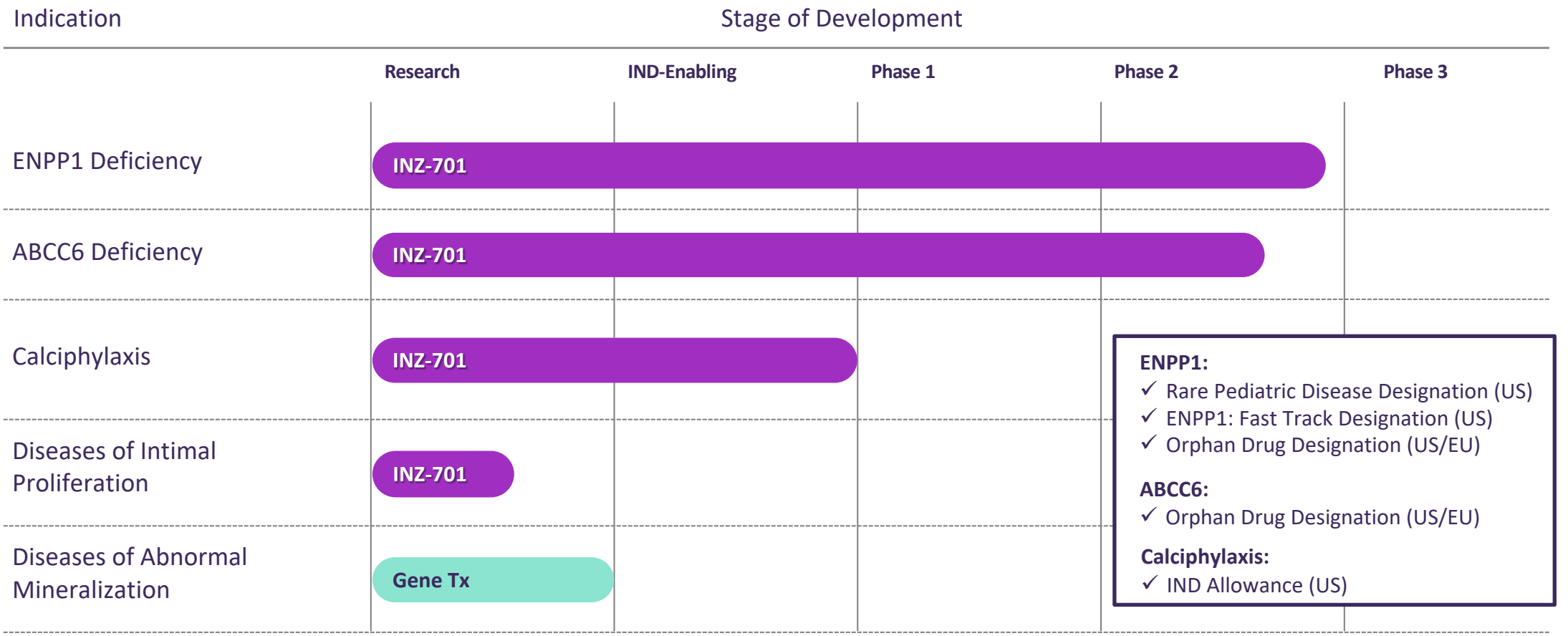
50% bedridden or wheelchair-bound [11]

Wound ulceration requiring surgical debridement [11]

- Disease Incidence: **1-4% of ESKD pop.**[10]
- Associated with low PPi
- No Approved Therapies

Sources: 1. Ferreira et al. JBMR 2021; 2. Ferreira et al. Genet Med, 2020. 3. O'Brien, Khursigara et al. Plos One, 2022. 4. Ferreira et al. Orphanet Journal of Rare Diseases, 2022. 5. Risseeuw et al. Retina 2019; 6 Kauw et al. J Neuro Sci 2017; 7. Verwer et al Eur J Vas Surg. 2023; 8.Gielis et al. J. Clin Med 2020; 9. Kranenburg et al. Eur J Med Gen 2018. 10. Nigwekar. Curr Opim Nephrol Hypetens 2017; 11. Nigwekar et al. NEJM 2018

INZ-701 is a novel enzyme replacement therapy (ERT) with the potential to be an impactful first-to-market therapy in multiple diseases

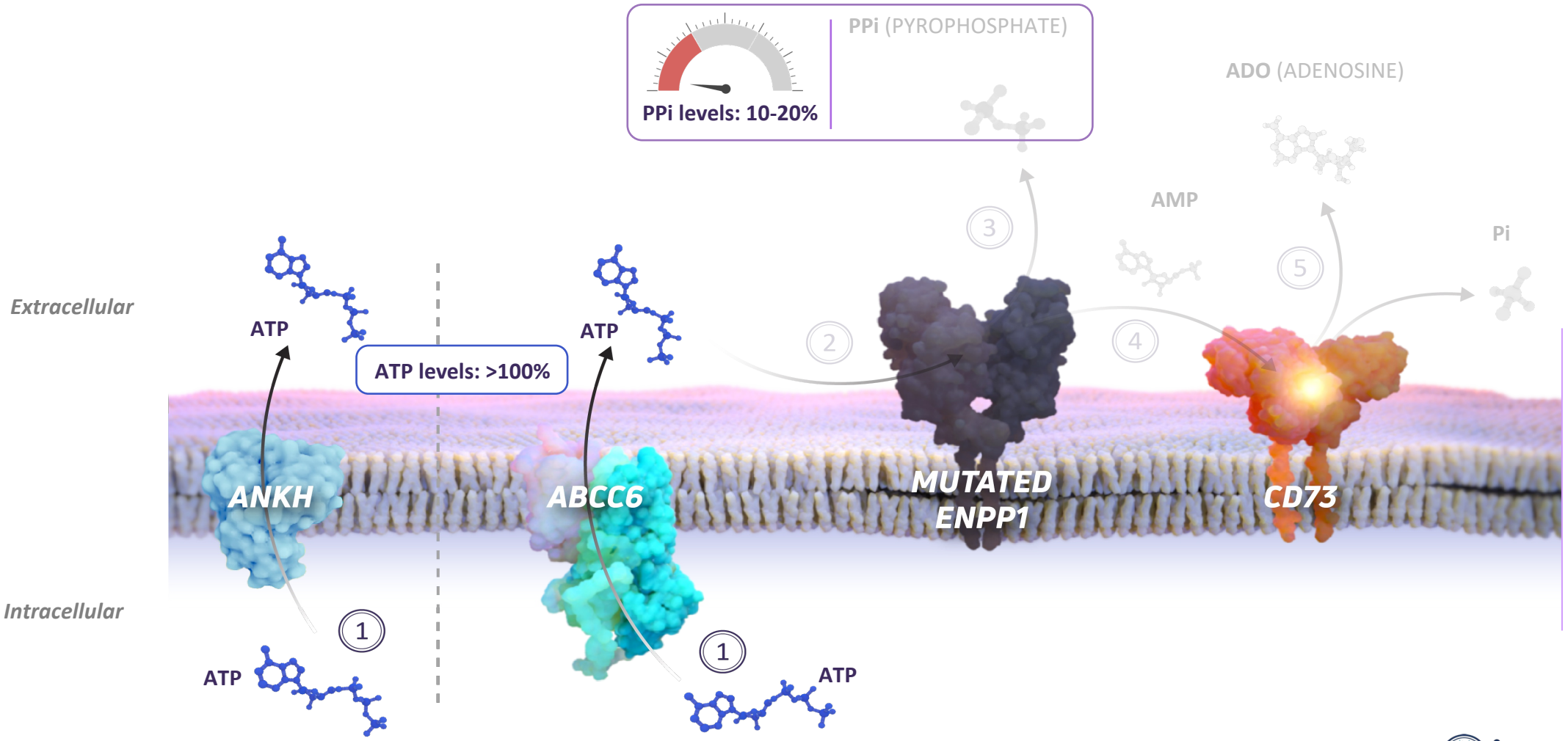


Inozyme retains worldwide, exclusive development and commercial rights to INZ-701

ENPP1 Deficiency

inozyme pharma

Mutations in ENPP1 enzyme result in low PPi and adenosine levels, which leads to progressive pathologic calcification and intimal proliferation



ENPP1 Deficiency is a lifelong, multisystem, rare genetic disease with high mortality and morbidity that evolves throughout a patient's lifetime

GACI/IIAC
0-1 Years (~1-2%)*

50% mortality within 6 months of birth

Severe cardiovascular complications

ARHR2 (Rickets)
1 to <13 years (~25-30%)*

Impaired growth
Orthopedic surgery

Skeletal defects: Rickets

Hearing loss

ARHR2 (Osteomalacia)
13+ Years (~65-70%)*

Bone & joint pathology

Skeletal defects: Osteomalacia

Joint, tendon, and ligament complications

Hearing loss

Genetic Prevalence[1]: 1:64,000

PATIENTS IN US/CANADA	~ 2,800	PATIENTS IN JAPAN	~ 900
PATIENTS IN EUROPE	~ 4,100	PATIENTS IN BRAZIL	~ 1,600

*Estimated percent of total prevalence., 1. Ferreira et al. Orphanet Journal of Rare Diseases, 2022. GACI: Generalized Arterial Calcification of Infancy, IIAC : Idiopathic Infantile Arterial Calcification, ARHR2: Autosomal Recessive Hypophosphatemic Rickets Type 2

Adult ENPP1 Deficiency Phase 1/2 trial

A Phase 1/2, open-label, multiple ascending dose study to evaluate the safety, tolerability, pharmacokinetics, and pharmacodynamics of INZ-701 followed by an open-label long-term extension period in adults with ENPP1 Deficiency

Study Population:
Adults

Eligibility Criteria:

- Age 18-64 years
- Confirmed clinical and genetic diagnosis

9+ patients enrolled

Primary Goals

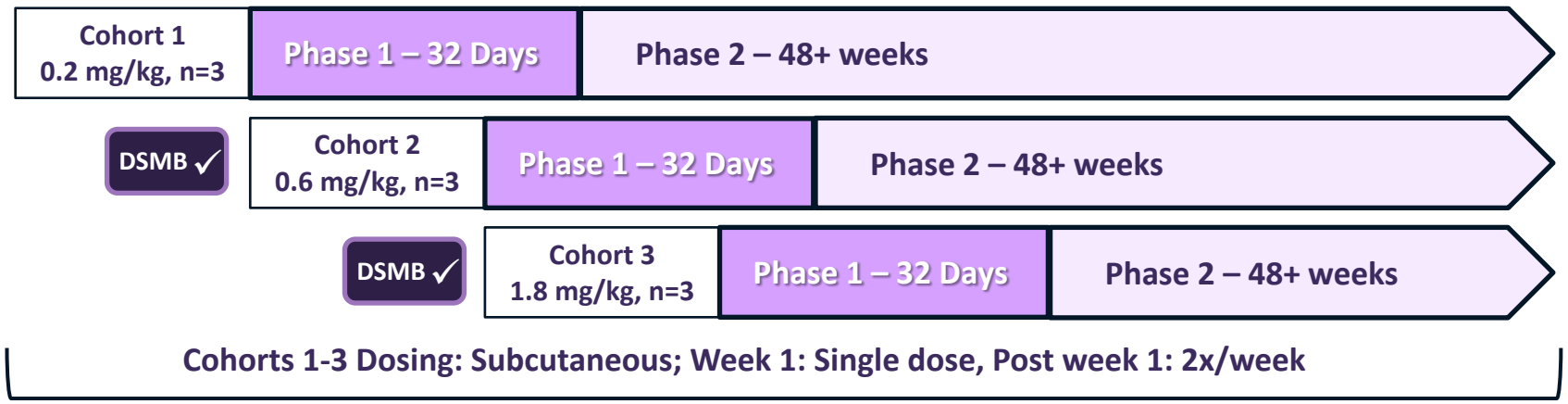
- **Safety** and **tolerability**
- **Immunogenicity**
- **Pharmacokinetic properties**
- **Pharmacodynamics (PPi)**

Secondary Goals

Evaluate potential endpoints for pivotal study

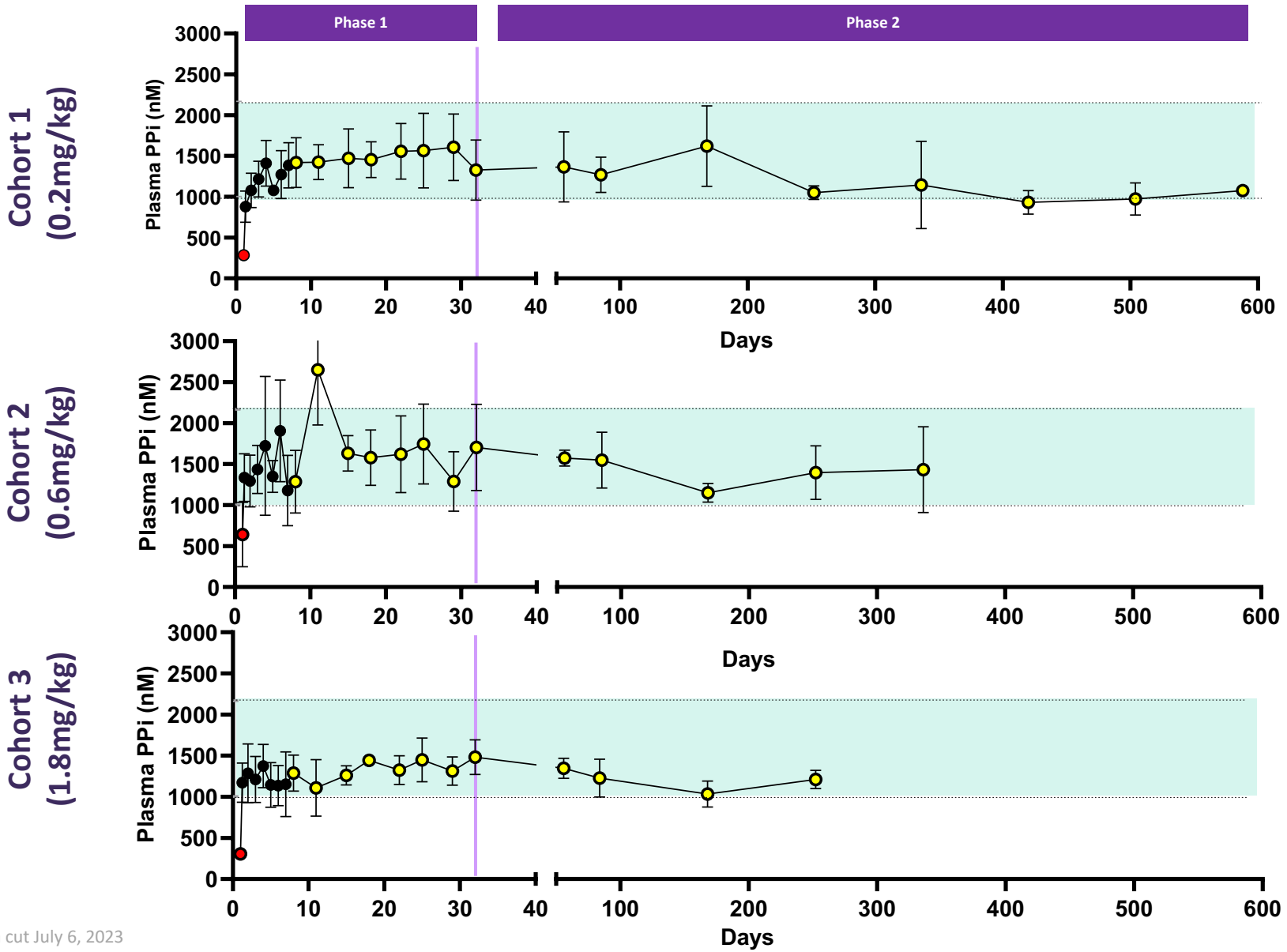
- **Ectopic calcification**, **skeletal**, **vascular** and **physical function**, and **patient reported outcomes**
- Exploratory **biomarkers**

Study Design:



DSMB = Data Safety Monitoring Board. clinicaltrials.gov: NCT04686175

Rapid, significant and sustained increase in PPi observed at all doses



- Rapid increase within 6 hours observed after the 1st dose
- PPi levels reached the healthy volunteer range after the 1st dose

Baseline PPi (pre-dose) + 1st INZ-701 dose
PPi measurement (post-dose)
PPi measurement (pre-dose)
Healthy subject PPi levels; n=10
Data presented as mean ± SEM

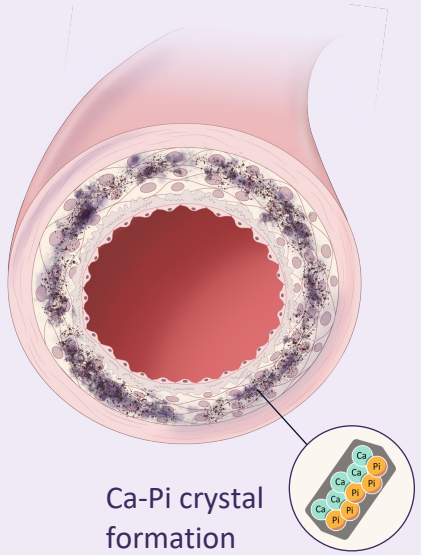
Cohort 1: n=2 post day 84; Cohort 2: n=2 post day 336; Cohort 3: n=2 post day 168

Data cut July 6, 2023

Goal is restoration of proper balance of PPi and Pi to prevent vascular calcification and skeletal abnormalities

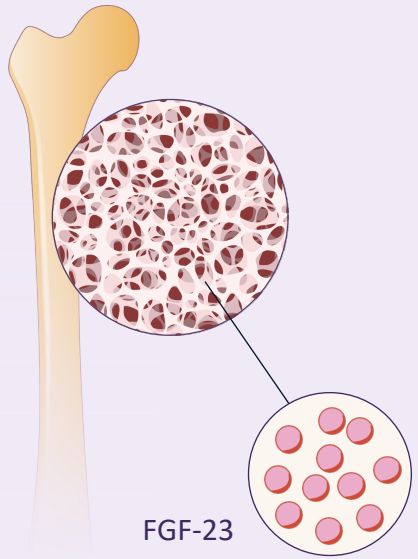
Low PPi

VASCULAR CALCIFICATION



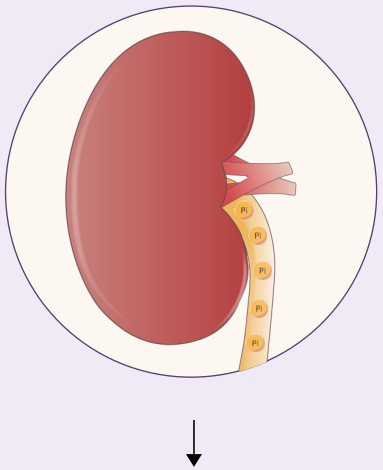
Vascular calcification due to low PPi leads to compensatory FGF-23 increase

INCREASED FGF-23



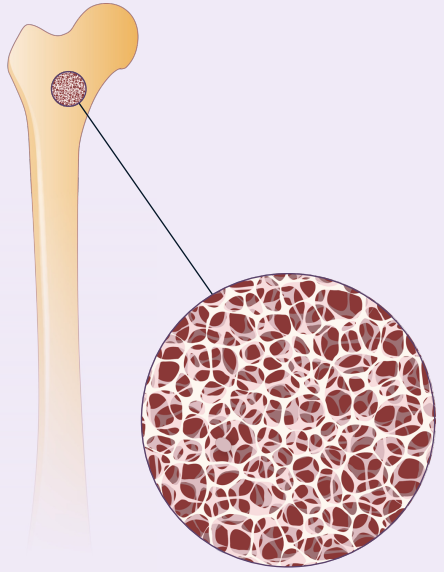
FGF-23, expressed in osteocytes in the bone, is a key regulator of phosphate in the body

RENAL PHOSPHATE WASTING



Increased levels of FGF-23 lead to increased phosphate wasting and hypophosphatemia

RICKETS/ OSTEOMALACIA



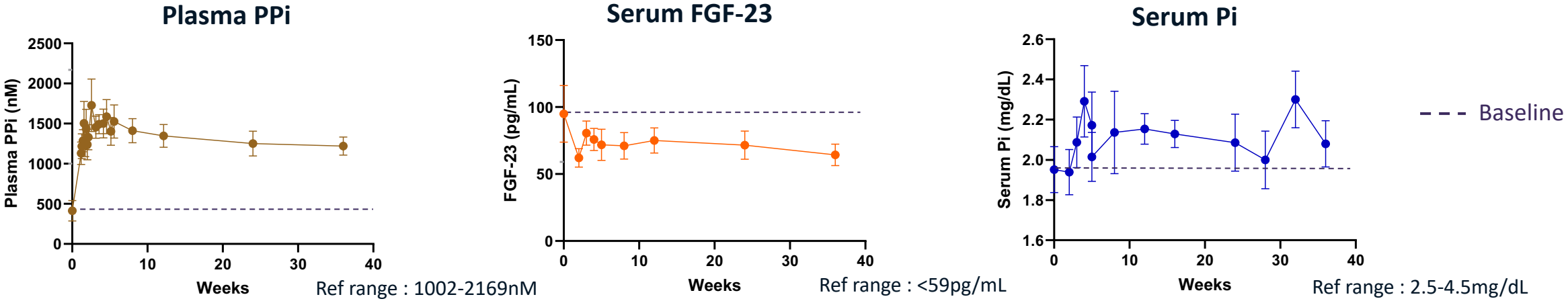
Hypophosphatemia is a central driver of rickets and osteomalacia

Decreasing FGF-23 alone in ENPP1 Deficiency is not sufficient to address the clinical pathology and can exacerbate calcification; therefore, the use of burosumab is contraindicated[1]

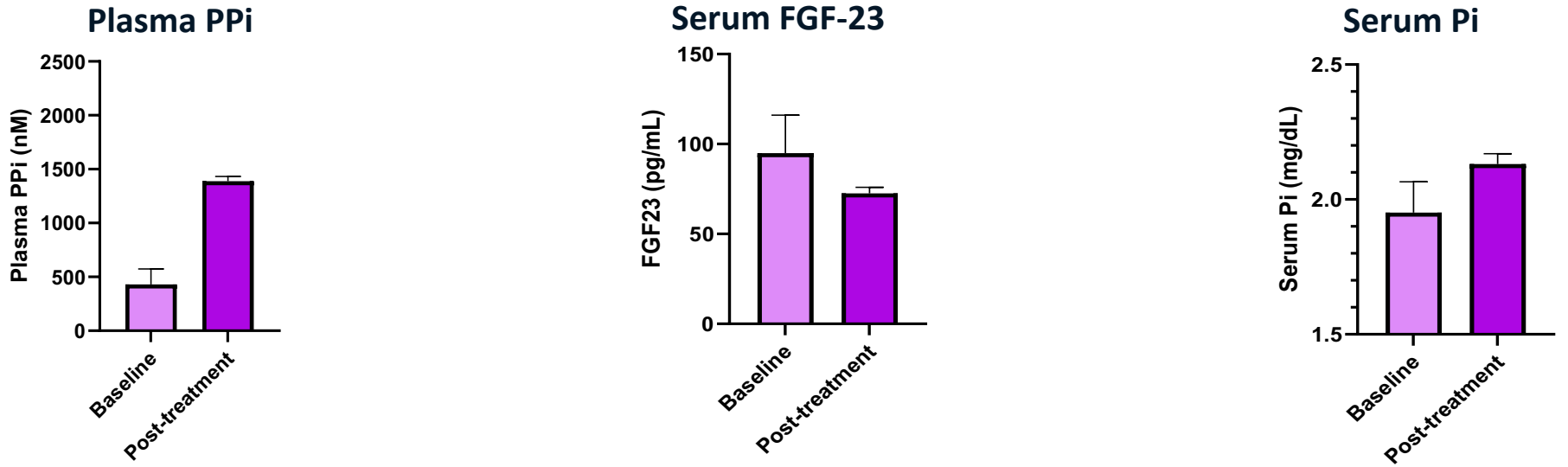
1. Stern et al, Gen in Med 2021

Significant increase in PPi levels were associated with improvement in phosphate and FGF-23 and supports mechanism of action

Pooled Cohorts 1-3: Baseline vs mean Week 2-36 PPi, FGF-23, and Pi levels (±SEM)



Pooled Cohorts 1-3: Mean PPi, FGF-23 and Pi levels (±SEM)



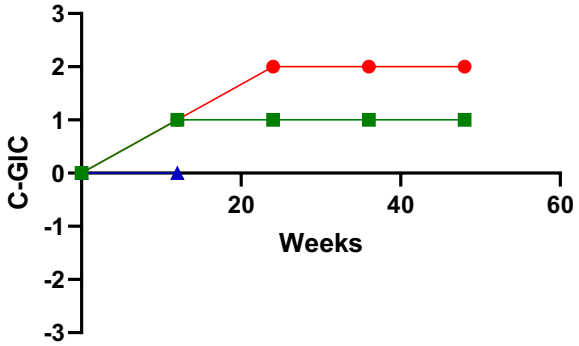
Note: Serum Pi increases observed in absence of phosphate and active vitamin D supplementation

Data cut July 6, 2023; Data presented as mean ± SEM

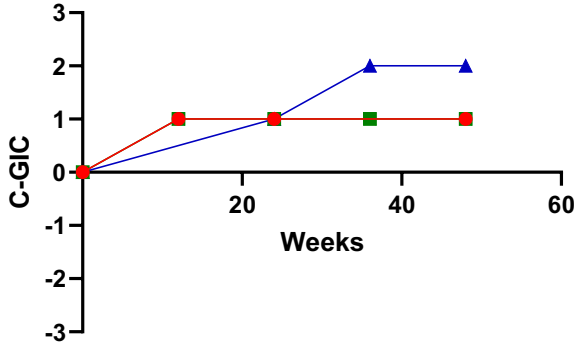
INZ-701 showed concordant improvement in C-GIC and P-GIC in all dose cohorts

Clinician's Global Impression

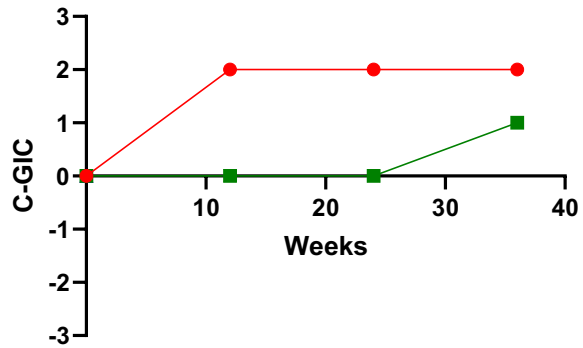
Cohort 1 (0.2mg/kg) (n=3)



Cohort 2 (0.6mg/kg) (n=3)

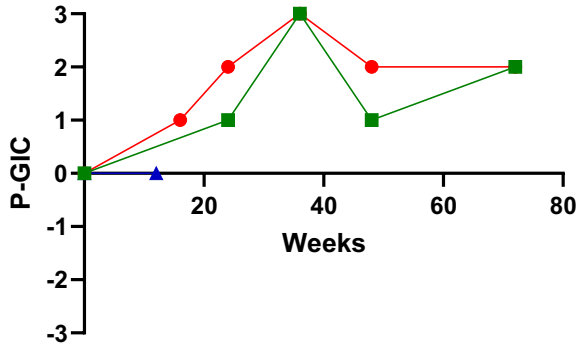


Cohort 3 (1.8mg/kg) (n=2) *

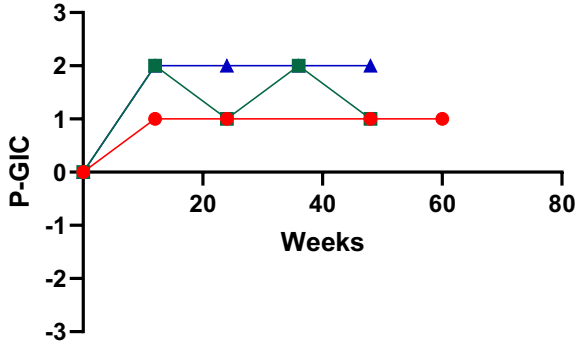


Patient's Global Impression

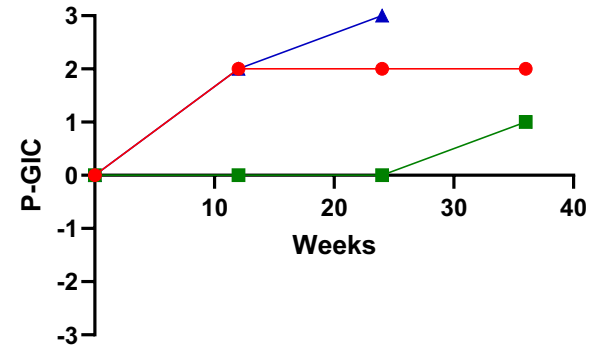
Cohort 1 (0.2mg/kg) (n=3)

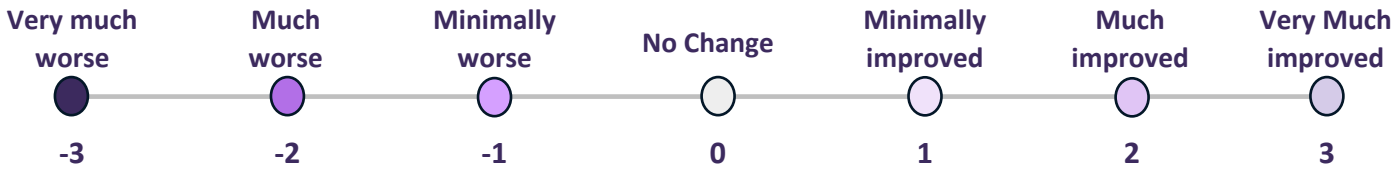


Cohort 2 (0.6mg/kg) (n=3)



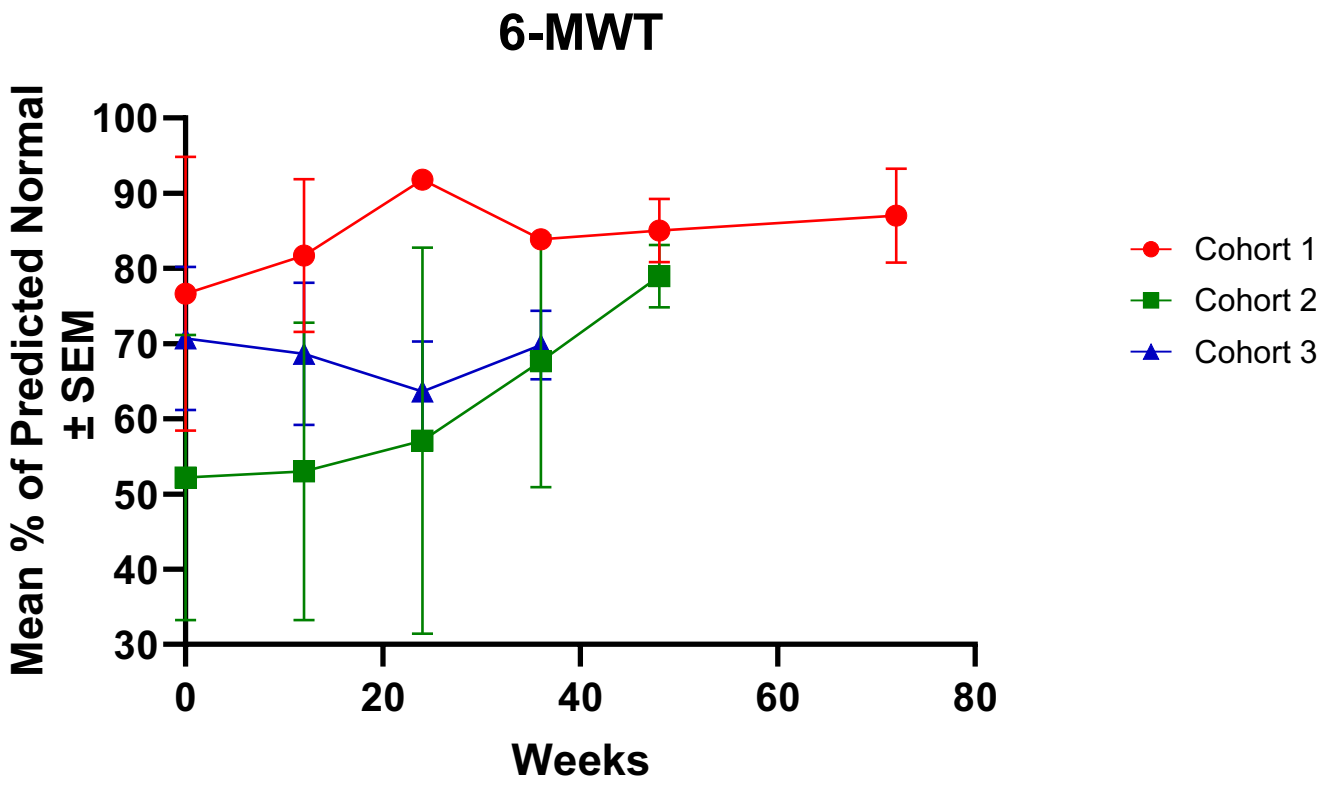
Cohort 3 (1.8mg/kg) (n=3)





Colors represent individual patients in respective cohorts, *Assessment not performed by 1 clinician

INZ-701 showed trend for improvement in 6-minute walk test (6-MWT)

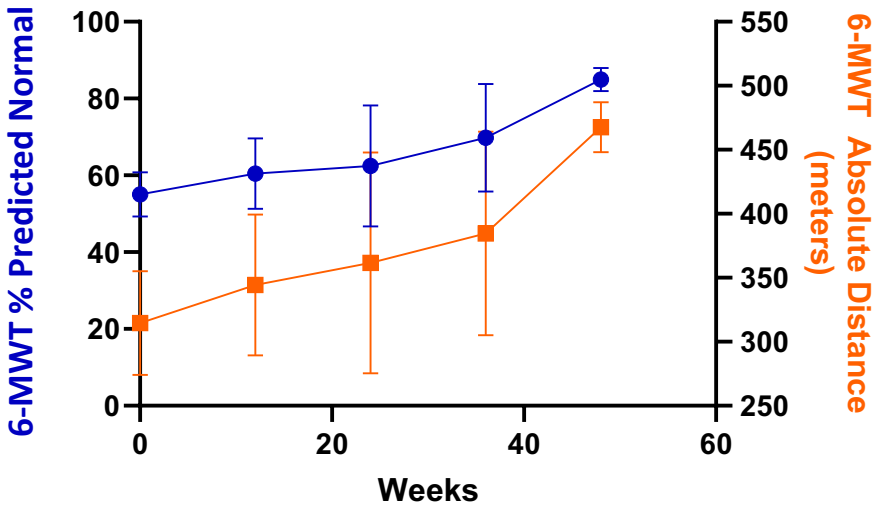


Percent predicted normal adjusts for subject age, gender, height and weight

Data cut July 6, 2023

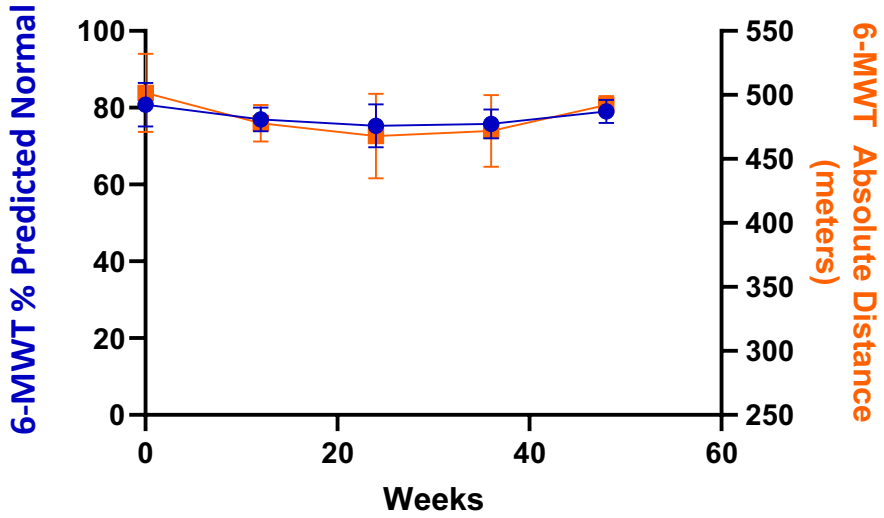
Subgroup analysis: 6-minute walk test results

Patients with <70% predicted of healthy 6-MWT at baseline (n=5)



Greater improvement observed in patients with poor baseline 6-MWT

Patients with >70% predicted of healthy 6-MWT at baseline (n=4)



Stable 6-MWT scores observed in patients with higher baseline values

Data presented as ± SEM; Data cut July 6, 2023

Adult ENPP1 Deficiency Phase 1/2 trial: Primary goals met

Safety and immunogenicity

- ✓ Well-tolerated, **no serious adverse events** related to study drug
- ✓ Support for first studies in infants (ongoing) and children (pending)
- ✓ Low, sometimes transient levels of non-neutralizing anti-drug antibodies

Pharmacokinetics

- ✓ 126-hour **half-life supports once-weekly dosing**
- ✓ Informs and validates PK model

Pharmacodynamics

- ✓ Significant elevation of plasma pyrophosphate (PPi), maintained for over 18 months
- ✓ Changes in **key biomarkers** (i.e., FGF-23 and phosphate (Pi)) **support clinical hypothesis**
- ✓ Rapid increase in PPi at 1 week correlated with decreases in FGF-23 levels (p= 0.0371)
- ✓ Dose ranging data support adult dose of 1.8 mg/kg/week

Identify clinically meaningful outcome measures to inform design of future study in adults

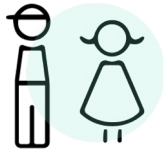
- ✓ Functional **improvements can be measured by 6-minute walk test and patient-reported outcomes**; strongest improvements seen in patients with greatest impairment at baseline
- ✓ Identified areas of bone pathology (low BMC/BMD) may represent locations for radiographic scoring for improvements
- ✓ Subset analyses reveal patient populations most likely to benefit from INZ-701 treatment in future adult studies

BMD = Bone mineral density; BMC = Bone mineral content

ENERGY-3: Pivotal trial in pediatric patients with ENPP1 Deficiency (ARHR2)

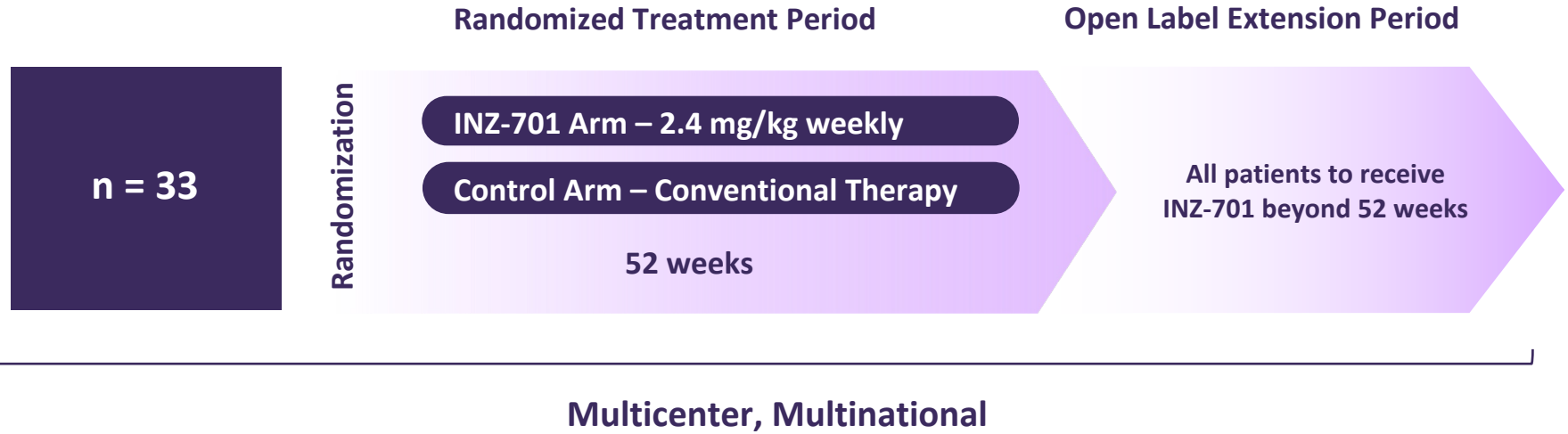
Initiation planned for October 2023

Population: *Pediatric*



- Confirmed genetic diagnosis
- Radiographic evidence of skeletal abnormalities
- ≥1 year and <13 years
- Low plasma PPi

Design: Randomized (2:1), Open Label



Endpoints

US

- **Primary**: Change in plasma PPi from baseline over time
- **Secondary**: Trends in RGI-C score, RSS, Growth Z-score; PK

EU

- **Co-Primary**:
  - Change in plasma PPi from baseline over time
  - RGI-C score (with $p<0.2$)
- **Secondary**: RSS, Growth Z-score; PK

RGI-C: Radiographic Global Impression of Change, RSS: Rickets Severity Score, PK: Pharmacodynamic, ARHR2: Autosomal Recessive Hypophosphatemic Rickets Type 2

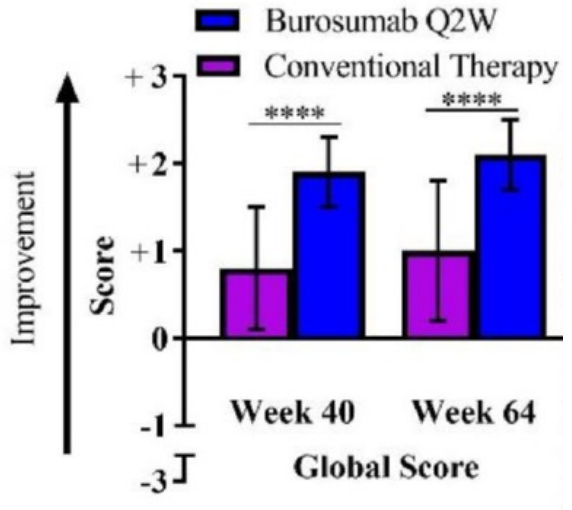
RGI-C (Radiographic Global Impression of Change): An accepted quantitative score for rickets/skeletal abnormalities

- RGI-C has supported the approval of therapies for other genetic forms of rickets

Whyte et al, J Bone Min Res, 33:868, 2018

- Each form of rickets has disease-specific radiographic features
  - We are currently developing an RGI-C specific for ENPP1 Deficiency
  - Scoring system based on extensive natural history x-ray database

Burosumab treatment in XLH resulted in RGI-C of +1.9 vs +0.8 for conventional therapy at week 40



****p<0.0001

Imel et al, Lancet, 393: 2416, 2019

RGI-C: Radiographic Global Impression of Change, XLH: X-Linked Hypophosphatemia

ENERGY-1 Trial: Phase 1b in infants with ENPP1 Deficiency (GACI)

Initiated in Q2 2023

Population: *Infant*

- 1-12 months
- Confirmed genetic diagnosis

Design: Single arm, Open Label

Dosing: Subcutaneous; Range from 0.2 mg/kg once weekly to 0.6 mg/kg twice weekly;
Intra-and interpatient dose escalation based on data review committee recommendation

Multicenter, Multinational

Endpoints:

Primary

- **Safety** and **tolerability** of INZ-701

Additional

- Plasma **PPi**
- **Survival**, **growth**, **development**, **functional performance**, **cardiac function**, **biomarkers** related to **bone** and **mineral metabolism**, healthcare utilization

clinicaltrials.gov: NCT05734196
GACI: Generalized Arterial Calcification of Infancy

Planned Path to Global Approval of INZ-701 in ENPP1 Deficiency

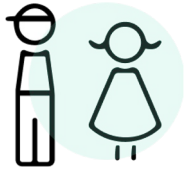
ENERGY-1: Infant (1-12 mos.)
Phase 1b
Single arm (n=8)

Endpoints

Safety** and **tolerability

ENERGY-2: Infant (0-12 mos.)
Pivotal
Single arm per agreed PIP** (n=12)

***PPi + survival** as co-primary*



ENERGY-3: Pediatric (≥1-<13 yrs.)
Pivotal
Randomized – 2:1 (n=33)

***PPi** as sole primary* (US) and co-primary with **RGI-C** (EU)*

ENERGY-4: Adolescent and Adult (13+)
Pivotal
Randomized – 2:1 (n=30)***

***PPi** as sole primary* (US) and co-primary with **BMC/BMD** expected (EU)*

*Supported by trends in appropriate secondary endpoints
**Plan to conduct this trial ex-U.S.. Discussions are ongoing with FDA regarding design of this trial in the U.S.
***Subject to regulatory discussions and appropriate financial resources

Basis for Planned Marketing Applications

1st BLA/MAA

- Adult Phase 1/2 full data
- ENERGY-3 full data
- ENERGY-1 available data
- ENERGY-2 available data
  - Natural history control group; patients matched on covariates associated with mortality

Additional filings

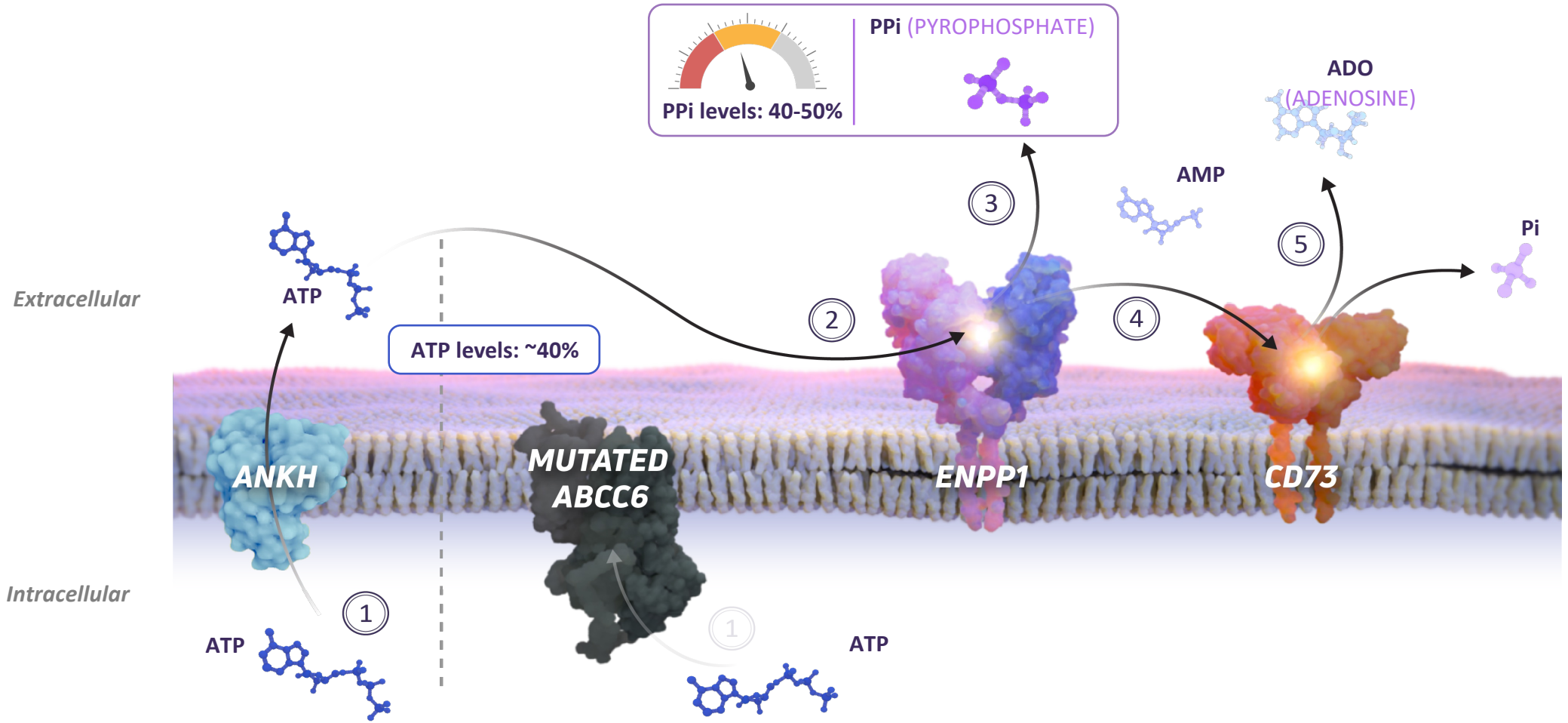
- ENERGY-4 full data (Supplemental BLA/MAA)
- Japan, Brazil, Middle East

RGI-C: Radiographic Global Impression of Change, BMC/BMD: Bone mineral content/density, BLA: Biologics license application, MAA: Marketing authorisation application

ABCC6 Deficiency

inozyme pharma

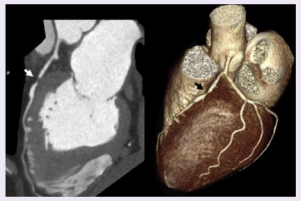
Mutations in the ABCC6 gene leads to reduced ATP levels, resulting in low levels of PPi and adenosine



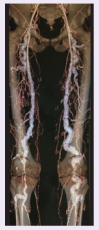
Adult ABCC6 Deficiency (PXE) is a multisystem disease associated with a broad range of organ complications and high degree of patient burden

Disease Prevalence: 1:25,000[5] - 1:50,000[6]

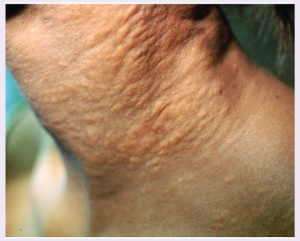
Neointimal proliferation
Karam et al. J Cardio
Comp Tomo 2015



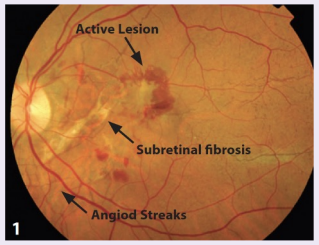
Vascular calcification
Mowafy et al. Vascular
& Endovascular Review 2019



Skin lesions
Borst et al. Trends in Biochemical
Sciences, February 2019



Retinal abnormalities
Zaria et al. eye news
OCTOBER/NOVEMBER 2015



Complications of Adult ABCC6 Deficiency (PXE) [1-4]

Onset in 20-30s

Prevalence by 70 years

- Calcium deposits in mid-dermis
- Skin papules, and progressive in-elasticity

>90%

- Calcification of retina
- Visual impairment

37%

- Arterial calcification
- Vascular complications*

~30%

* Early-onset of Peripheral Arterial Disease (PAD) and Intermittent Claudication (IC), and ischemic stroke/transient ischemic attack (TIA)

Sources: 1. Risseeuw et al. Retina, 2019; 2. Leftheriotis et al. J Vasc Surg, 2011. 3. Vanakker et al. Hum Mutat, 2008; 4. Van den Berg et al. Cerebrovasc Dis, 2000., 5. Internal, Unpublished Data; 6. Ferreira et al. Genet Med, 2021; PXE, pseudoxanthoma elasticum

Adult ABCC6 Deficiency (PXE) Phase 1/2 trial

A Phase 1/2, open-label, multiple ascending dose study to evaluate the safety, tolerability, pharmacokinetics, and pharmacodynamics of INZ-701 followed by an open-label long-term extension period in adults with ABCC6 Deficiency

Study Population:
Adults

Eligibility Criteria:

- Age 18-69 years
- Confirmed clinical and genetic diagnosis

10 patients enrolled

Primary Goals

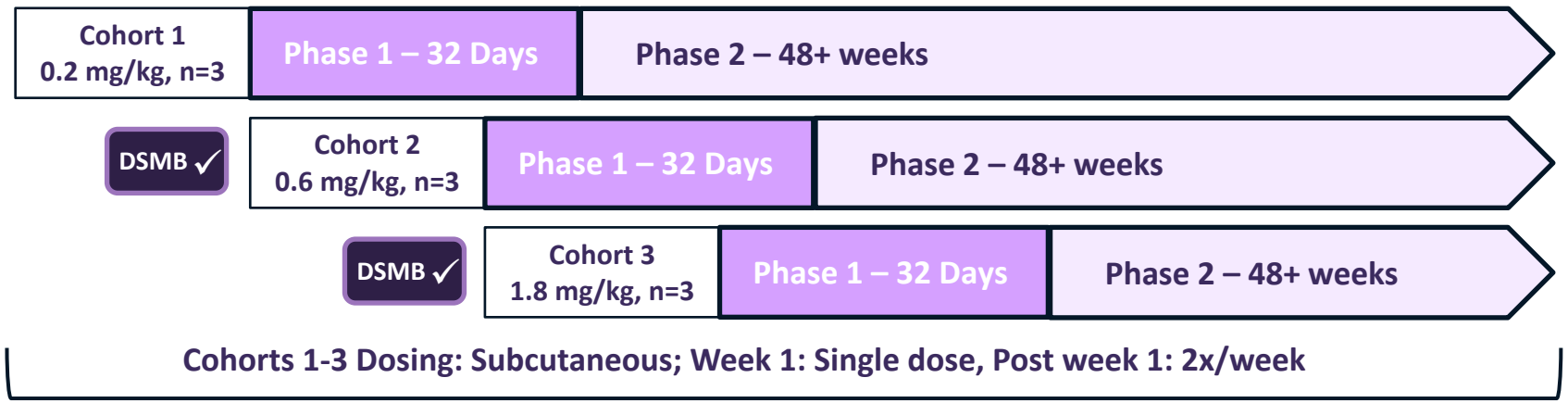
- **Safety** and **tolerability**
- **Immunogenicity**
- **Pharmacokinetic properties**
- **Pharmacodynamics (PPi)**

Secondary Goals

Evaluate potential endpoints for pivotal study

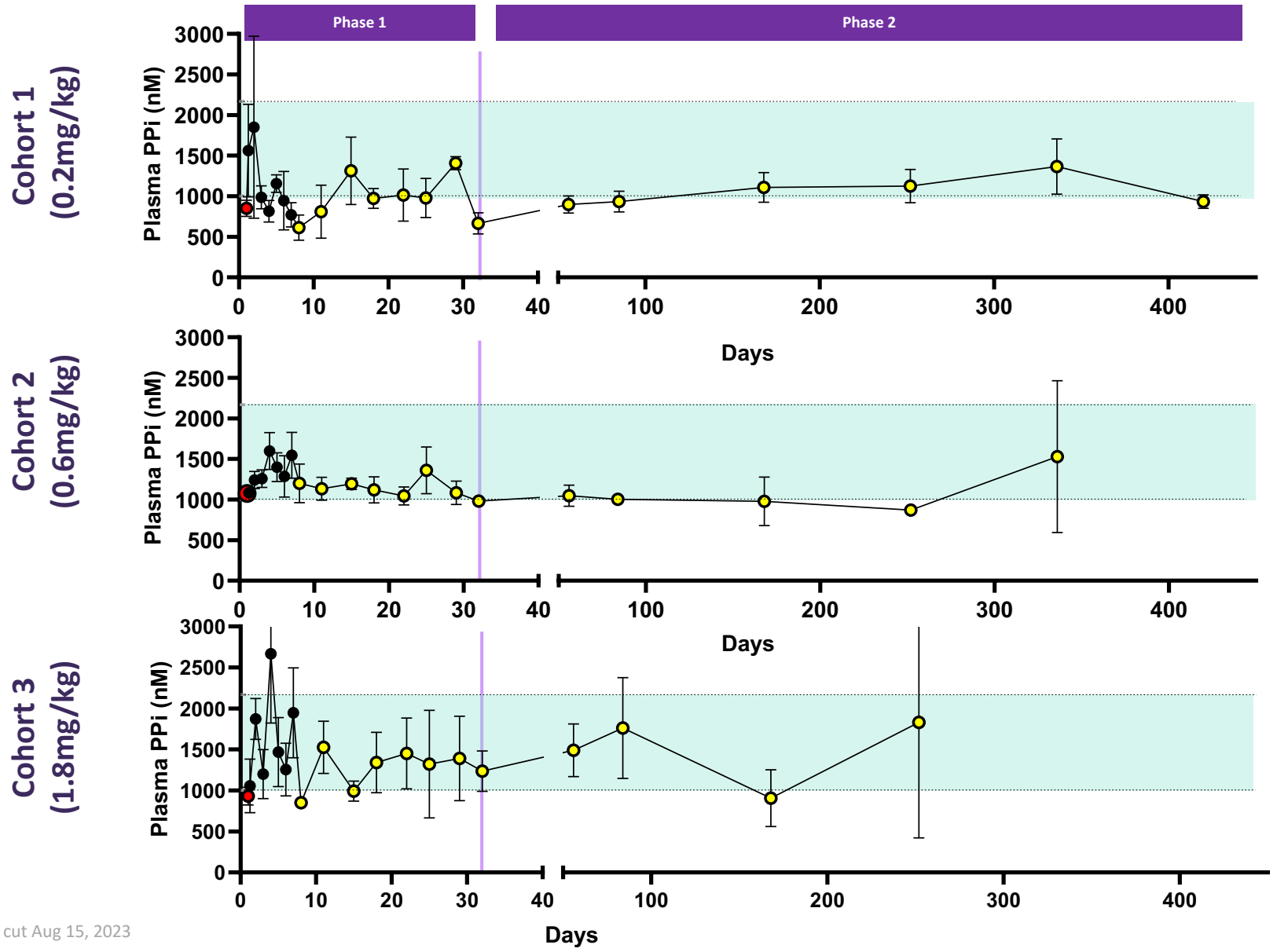
- **Ophthalmologic disease**, **ectopic calcification**, **cardiovascular disease, physical function** and **PROs**
- Exploratory **biomarkers**

Study Design:



DSMB = Data Safety Monitoring Board. clinicaltrials.gov: NCT04686175

Rapid and sustained increase in PPi observed at 1.8 mg/kg dose



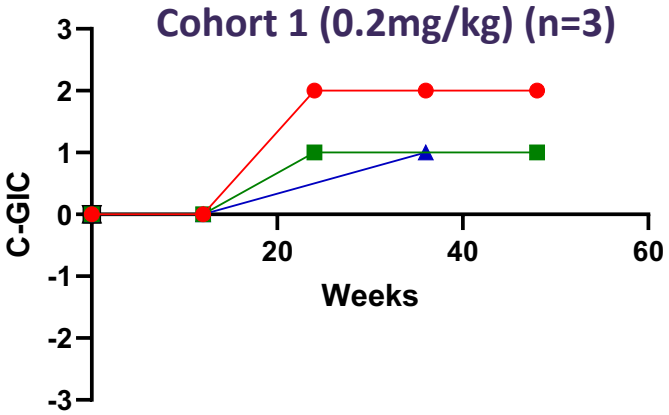
- Rapid increase observed after the 1st dose
- PPi levels reached the healthy volunteer range after the 1st dose

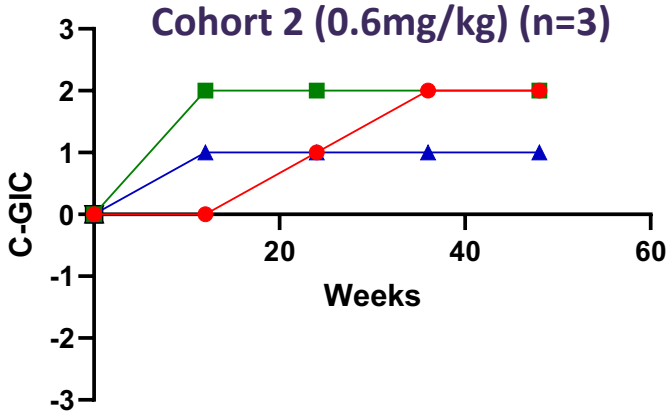
Baseline PPi (pre-dose) + 1st INZ-701 dose
PPi measurement (post-dose)
PPi measurement (pre-dose)
Healthy subject PPi levels; n=10
Data presented as mean ± SEM

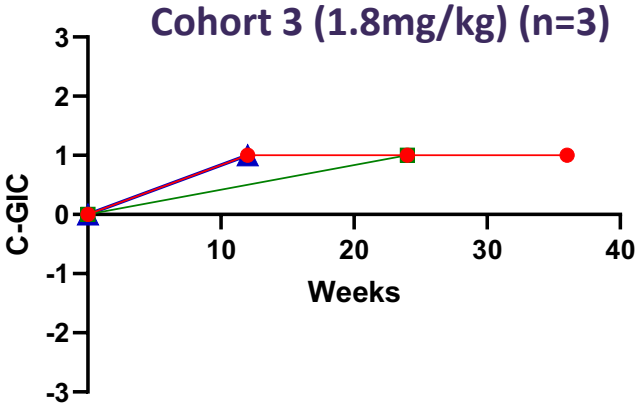
Data cut Aug 15, 2023

Majority of timepoints showed improvement in C-GIC and P-GIC in all dose cohorts

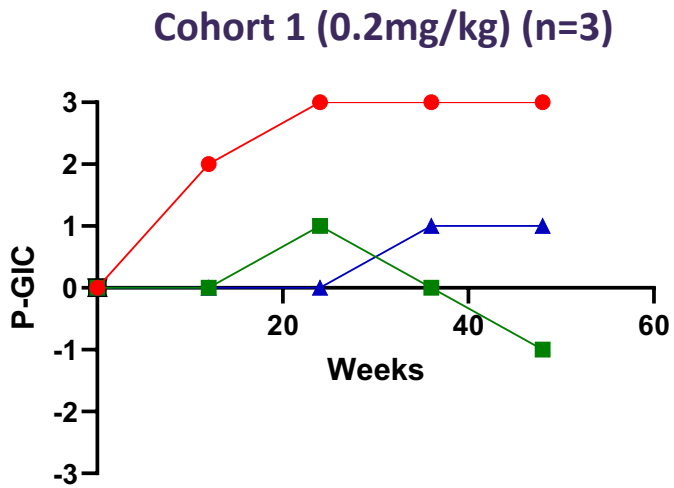
Clinician's Global Impression

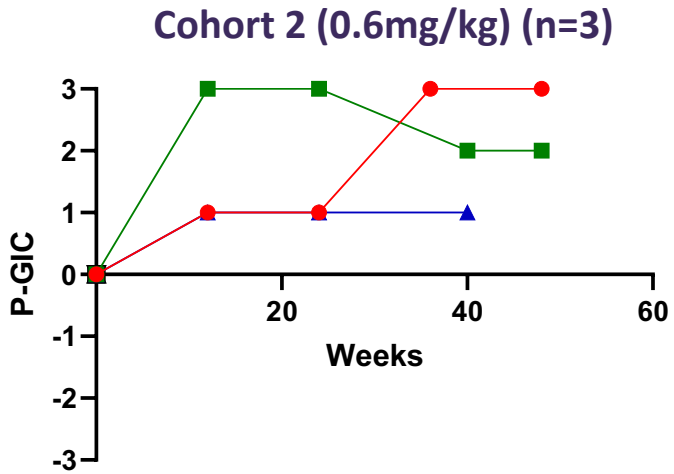


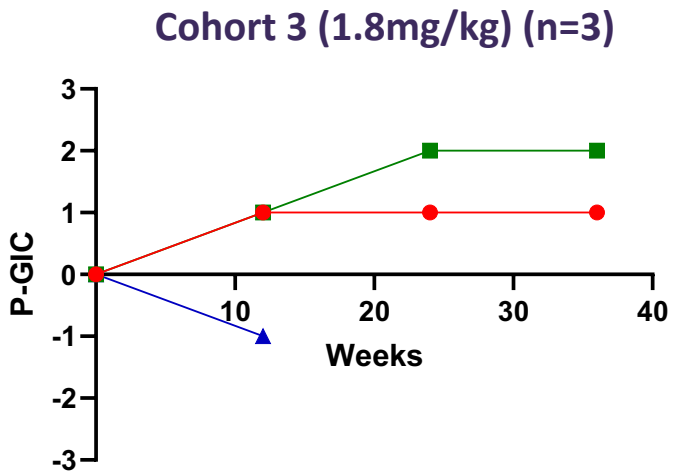


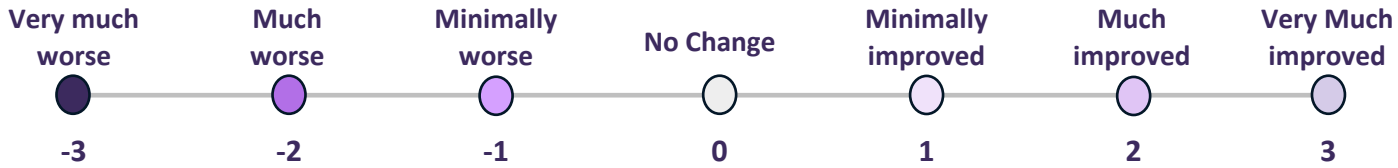


Patient's Global Impression









Colors represent individual patients in respective cohorts

Phase 1/2 trial of INZ-701 in patients with ABCC6 Deficiency

Safety

- ✓ INZ-701 was generally well-tolerated, and exhibited a favorable safety profile
- ✓ ADA titers generally low, with no evidence of neutralizing ADA

Pharmacokinetics

- ✓ Consistent PK observed in all patients as measured by immunoassay and enzymatic activity

Pharmacodynamics

- ✓ Rapid increase in PPi in all patients to levels comparable to those observed in healthy subjects
- ✓ Most sustained increase observed at highest dose level

Identify clinically meaningful outcome measures to inform design of future study in adults

- ✓ Global impression of change (GIC): improvement noted in 9/9 (C-GIC) and 7/9 (P-GIC)
- ✓ Concordance between C-GIC and P-GIC

Planned roadmap for clinical development of INZ-701 in ABCC6 Deficiency

	Pharmacodynamic Marker	Intermediate Endpoints	Clinical Outcome Measures
Infants	**PPi**	▪ Cardiovascular function ▪ Ectopic calcification	▪ Survival ▪ Developmental milestones/ Physical growth ▪ Days in hospital/ ICU/ on ventilator
Adults	**PPi**	▪ Vascular calcification ▪ Cardiac calcification ▪ Cardiovascular function ▪ Retinal abnormalities	▪ Cardiovascular events ▪ Peripheral arterial disease ▪ Vision function

Specific pivotal trial design and endpoints pending regulatory discussions and approval

Building a Rare Disease Franchise

inozyme pharma

Potential to deliver approved therapy for ENPP1 Deficiency and capitalize on significant commercial opportunity

Sizeable patient population with high unmet need

Connected, collaborative, and motivated patient community

Aligned KOLs/HCPs at well known global centers of excellence

Inozyme Readiness

- 10,000+ patients expected in major addressable markets – North America, Europe, Japan, Brazil, and Middle East
- Advancing understanding of the genetic prevalence – Estimated 1:64,000[1] pregnancies worldwide

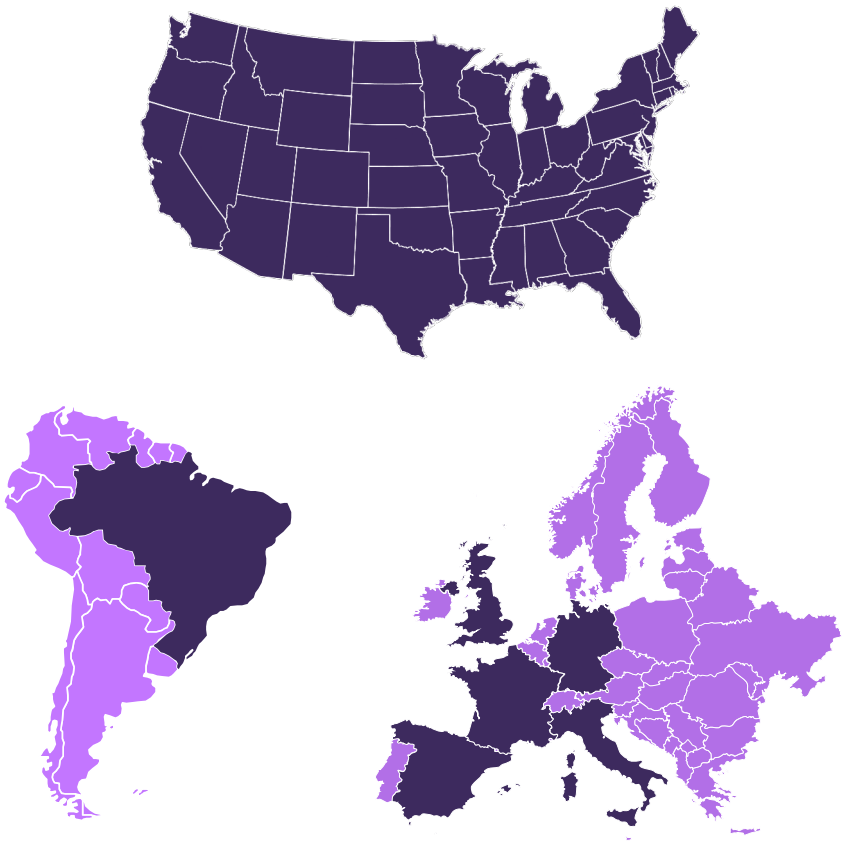
- Building strong relationships with patient and caregiver community
- Supporting programs and research partnerships to further characterize disease burden

- Executing data generation strategy to drive scientific leadership
- Broadening reach to increase awareness of HCPs and improve time to diagnosis

Source: 1. Ferreira et al. Orphanet Journal of Rare Diseases, 2022.

Ongoing efforts to increase disease awareness, educate patient and medical communities, and improve access to genetic testing

Growing Our Global Footprint



Currently evaluating Inozyme presence in Japan and Middle East

Expanding HCP Audience

Infant and Pediatric ENPP1

- Fetal and pediatric cardiology
- Neonatology
- Pediatric endocrinology
- Maternal-fetal medicine
- Genetics

Adult ENPP1

- Endocrinology
- Nephrology
- Genetics
- Bone specialists

Increasing Congress Attendance

NeoHeart – Cardiovascular Management of the Neonate
Society for Maternal-Fetal Medicine
PES Pediatric Endocrine Society
ASHG American Society of Human Genetics
ASBMR The American Society for Bone and Mineral Research
Human Growth Foundation
ECTS European Calcified Tissue Society
8th World Congress of Pediatric Cardiology and Cardiac Surgery

Partnering to Remove Barriers to Diagnosis

ipm.ai PART OF REAL CHEMISTRY
BeginNGS NEWBORN GENOMIC SEQUENCING to end the diagnostic odyssey
GENOMENON®
PREVENTION GENETICS
INVITAE
BlueDil int'l
GACI Global circulating hope
CENTOGENE
Rady Children's

Identifying ENPP1 patients to support market potential – strong progress in patient identification to date

Genetic Prevalence[8]: 1:64,000

750+ Global patients identified with **confirmed, known, or suspected** ENPP1 Deficiency

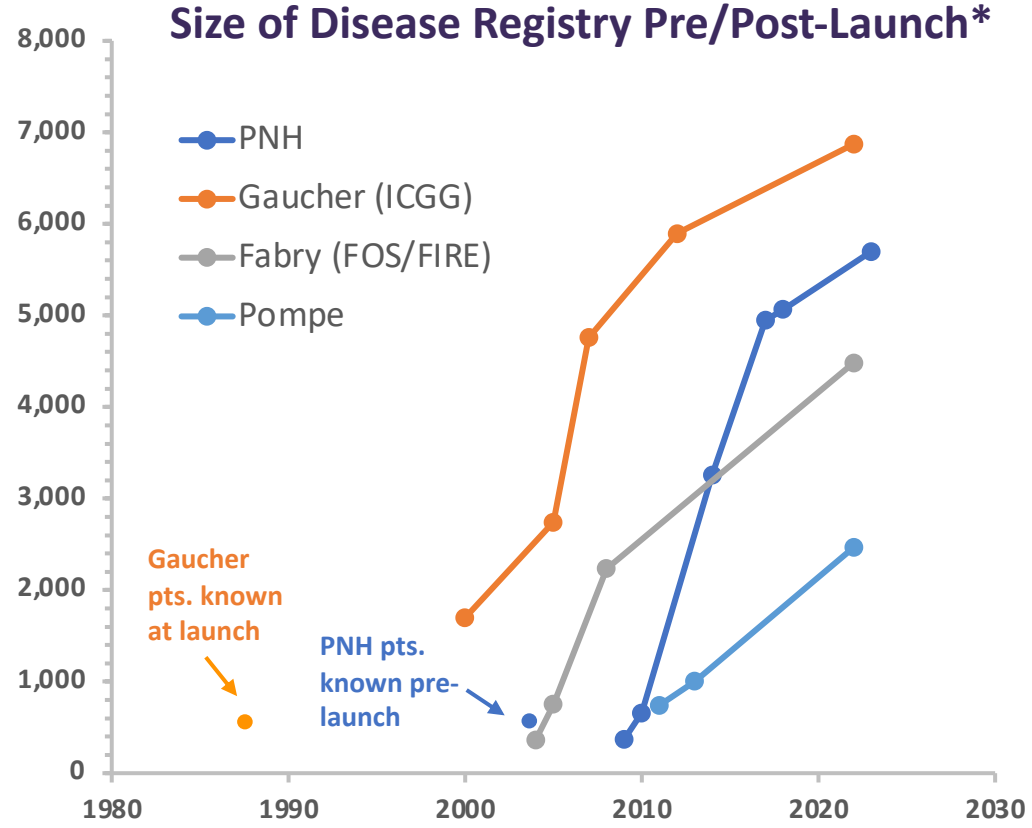
200+
Suspected Patients
Identified using AI model of large US claims/EMR database

300+
Known Patients*
Clinical diagnosis only

250+
Confirmed Patients
Clinical and genetic diagnoses

Internal data as of 5/31/23

Size of Disease Registry Pre/Post-Launch*



	Incidence Est.*	2022 Sales Est.*
Gaucher	1:57,00-142,000[1]	$1.1B[5]
PNH	1:62,500[2]	>$2.8B[6]
Fabry	1:37,000-117,000 (males)[3]	$1.4B[7]
Pompe	1:23,232[4]	~0.9B[8]

Sources: 1. Gaucher Institute: https://gaucher-institute.com/burden-of-disease/epidemiology-of-gaucher-disease, 2. Hillmen (2008), 3. Beck et al. (2022), 4. Park (2022), 5, Vpriv (Takeda) + Cerezyme (Sanofi); 2022 filings, 6. Astra Zeneca full-year 2022 result; Assumes 50% of total Soliris/Ultomiris sales. 7. Est. (not broken out by AZ), Replagal (Takeda) + Fabrazyme (Sanofi); 2022 filings, Est. based on 1H2022 sales, **Based on Inozyme estimates from publicly available sources.* 8. Ferreira et al. Orphanet Journal of Rare Diseases, 2022; 1:64,000 pregnancies

Anticipated milestones provide robust news flow

Milestone	2023	2024	2025
ENPP1 Deficiency			
Initiate ENERGY-1 – Phase 1b Trial in Infants	✓		
Interim Data - Adult Phase 1/2 Trial – Cohorts 1-3	✓		
Initiate ENERGY-3 – Pivotal Trial Pediatric Patients	Oct. 23		
Topline Data – Adult Phase 1/2 Trial*		Q1 24	
Initiate ENERGY-2 – Pivotal Trial in Infants – Ex. U.S.		Q2 24	
Interim Data – ENERGY-1 Trial		2H 24	
Topline Data – ENERGY-3 Trial			Mid-Year
ABCC6 Deficiency			
Interim Data - Adult Phase 1/2 Trial	✓		
Topline Data – Adult Phase 1/2 Trial*		Q1 24	
Initiate Phase 3 Trial**		Q4 24	

*48-week data from Cohorts 1-3, **Pending regulatory discussions and appropriate financial resources

Inozyme is at the forefront of developing transformative therapies for rare diseases of pathologic mineralization and intimal proliferation

- ✓ **ENPP1 Deficiency and ABCC6 Deficiency are serious diseases with no approved therapies**
  - Sizable patient populations with high mortality/morbidity and substantial patient and caregiver burden
  - >550 ENPP1 patients confirmed, with 210 additional patients identified based on medical record screen
- ✓ **INZ-701 has demonstrated rapid, significant, and sustained increase in PPi levels, and exhibited a favorable safety profile**
  - Finalized ENPP1 Deficiency pediatric pivotal trial design with PPi as primary endpoint in US, supported by trends in appropriate secondary endpoints, and co-primary endpoint (RGI-C of $p<0.2$) in EU for pediatric pivotal trial
  - ENPP1 Deficiency pediatric pivotal trial planned for Oct. 2023 – Topline data expected mid-2025
- ✓ **In a position of financial strength, with several expected upcoming milestones and a pipeline designed for long-term value creation**
  - $209.8M* expected to fund operations into Q4 2025; 61.7M common shares outstanding**
- ✓ **Experienced team with a track record of success in rare disease and a strong focus on execution**

*Proforma cash, cash equivalents & short-term investments as of 6/30/23 (Includes net proceeds of $64.5M from offering which closed in August 2023 and $5.1M of net proceeds from the at-the-market offering in July 2023)
**As of 8/3/23

Thank you


Nora
Living with
ENPP1 Deficiency
Callum
Living with
ENPP1 Deficiency