Corporate Presentation

May 2024

Callum
Living with
ENPP1 Deficiency

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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.

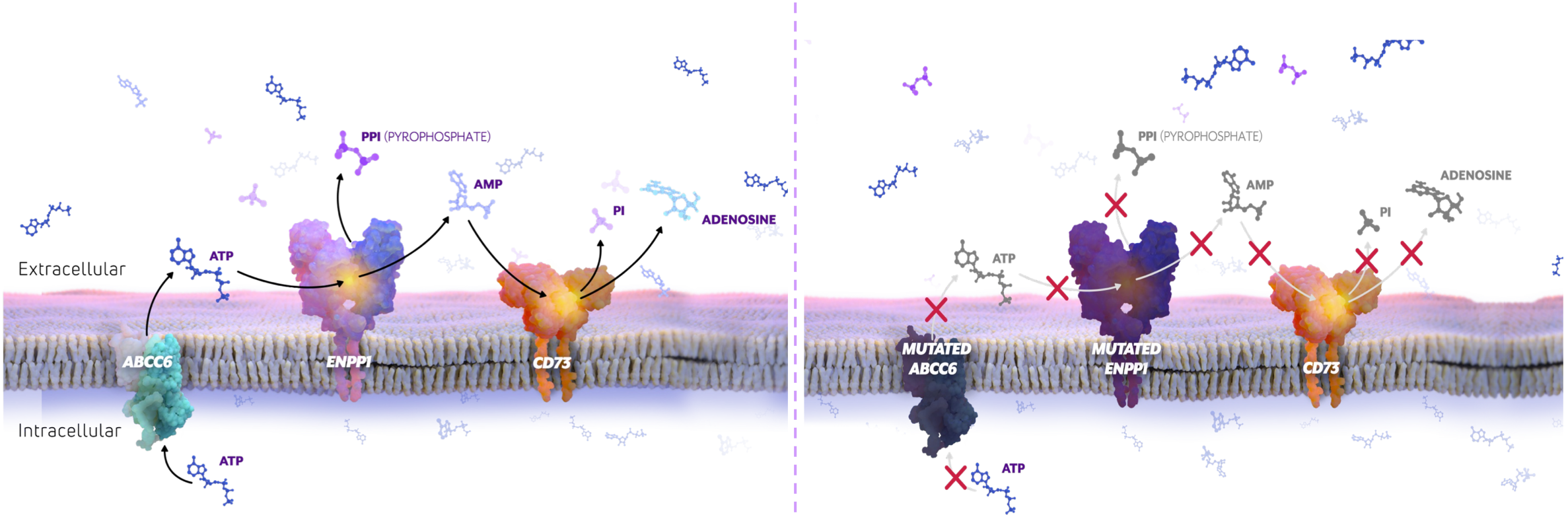
The words “anticipate,” “believe,” “continue,” “could,” “estimate,” “expect,” “intend,” “may,” “might,” “plan,” “potential,” “predict,” “project,” “should,” “target,” “will,” “would” and similar expressions are intended to identify forward-looking statements, although not all forward-looking statements 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 pyrophosphate (PPi) deficiency

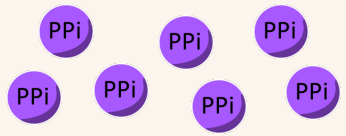
- ✓ **ENPP1 Deficiency, ABCC6 Deficiency and calciphylaxis are serious diseases with no approved therapies**
- ✓ **INZ-701 has demonstrated rapid, significant, and sustained increase in PPi levels, and exhibited a favorable safety profile across multiple clinical trials**
- ✓ **Currently in pivotal trial for ENPP1 Deficiency; Completed Phase 2 trial for ABCC6 Deficiency**
- ✓ **Experienced team with a track record of success in rare disease and a strong focus on execution**
- ✓ **In a position of financial strength, with several expected upcoming milestones and a pipeline designed for long-term value creation**
  - $166.2M expected to fund operations into Q4 2025 as of 3/31/24
  - 61.85M common shares outstanding as of 5/2/24

The PPi-Adenosine Pathway - ABCC6, ENPP1, and CD73 deliver extracellular ATP to generate PPi and adenosine

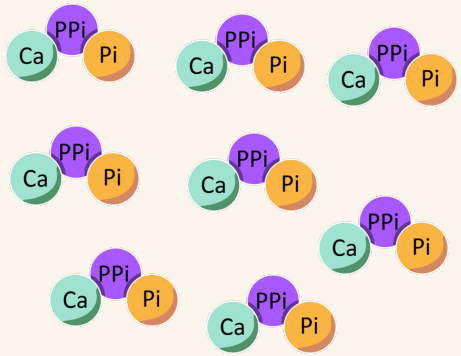


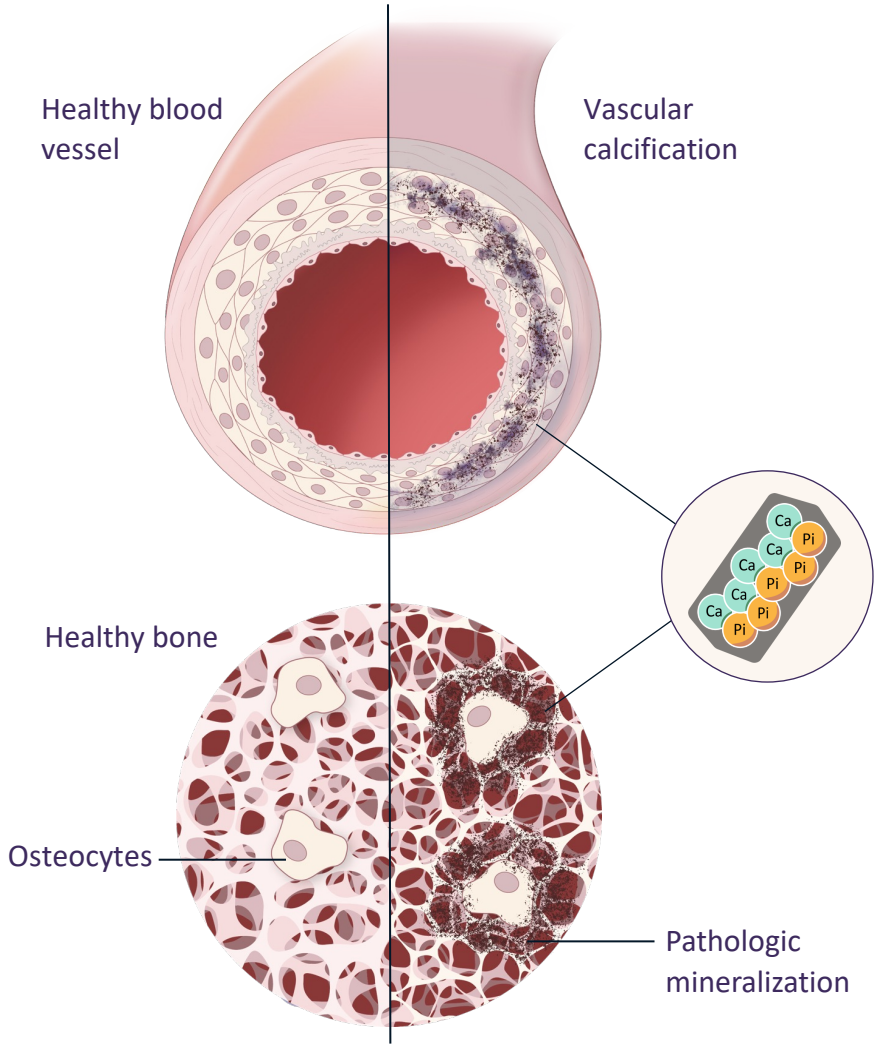
PPi is a master regulator of mineralization

Normal levels of PPi



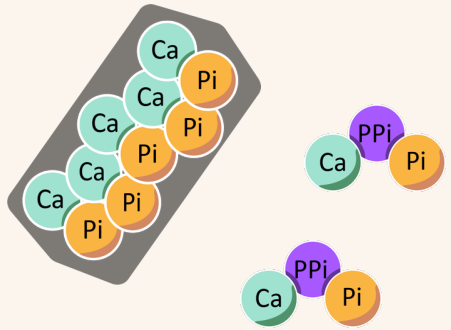
PPi prevents and inhibits Ca-Pi crystal formation





Low levels of PPi

Low PPi leads to Ca-Pi crystal formation



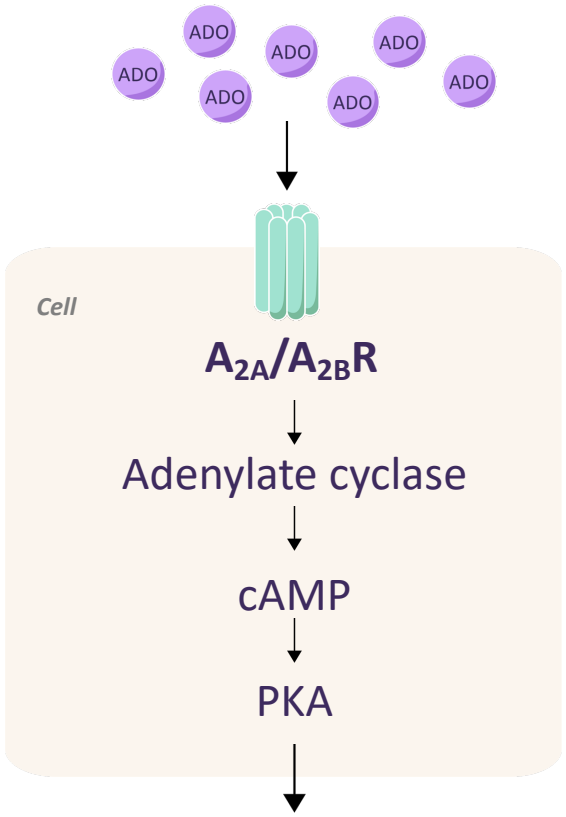
Inorganic pyrophosphate (PPi) PPi **Calcium** Ca **Phosphate** Pi

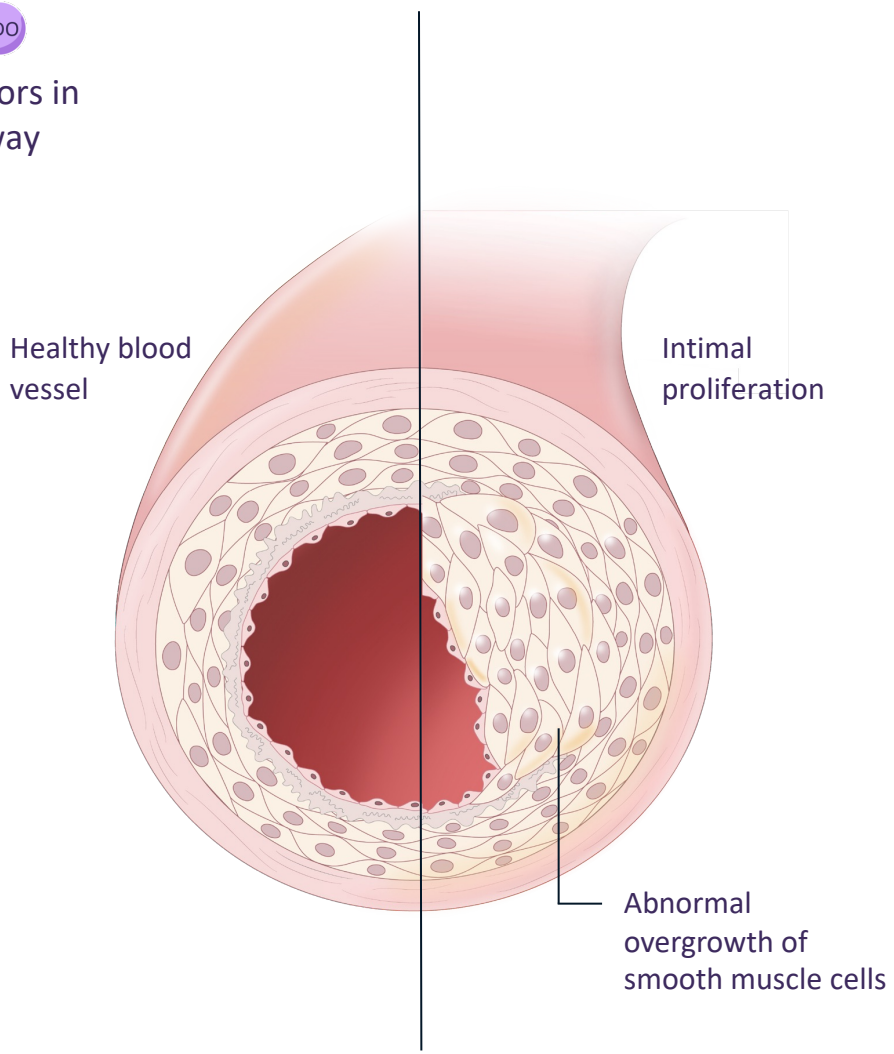
Adenosine is a potent inhibitor of intimal proliferation

Normal levels of adenosine (ADO)

ADO

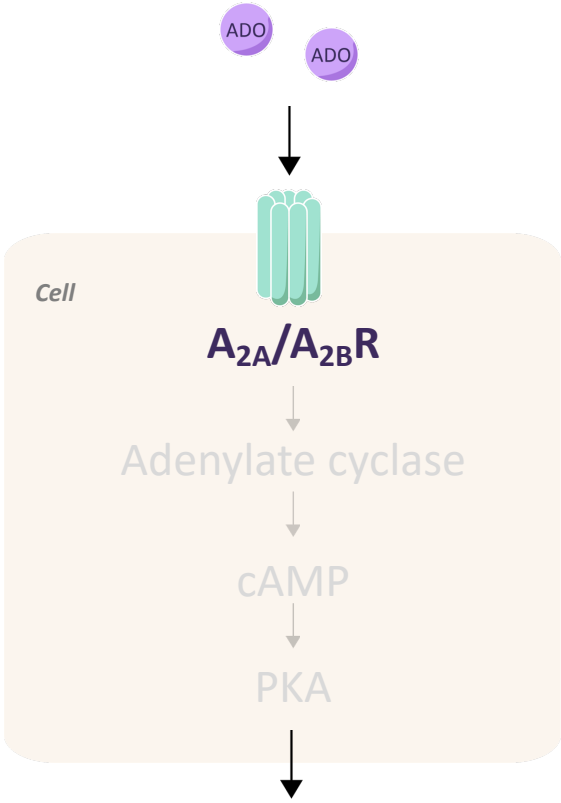
Adenosine directly binds adenosine receptors in the smooth muscle cell proliferation pathway





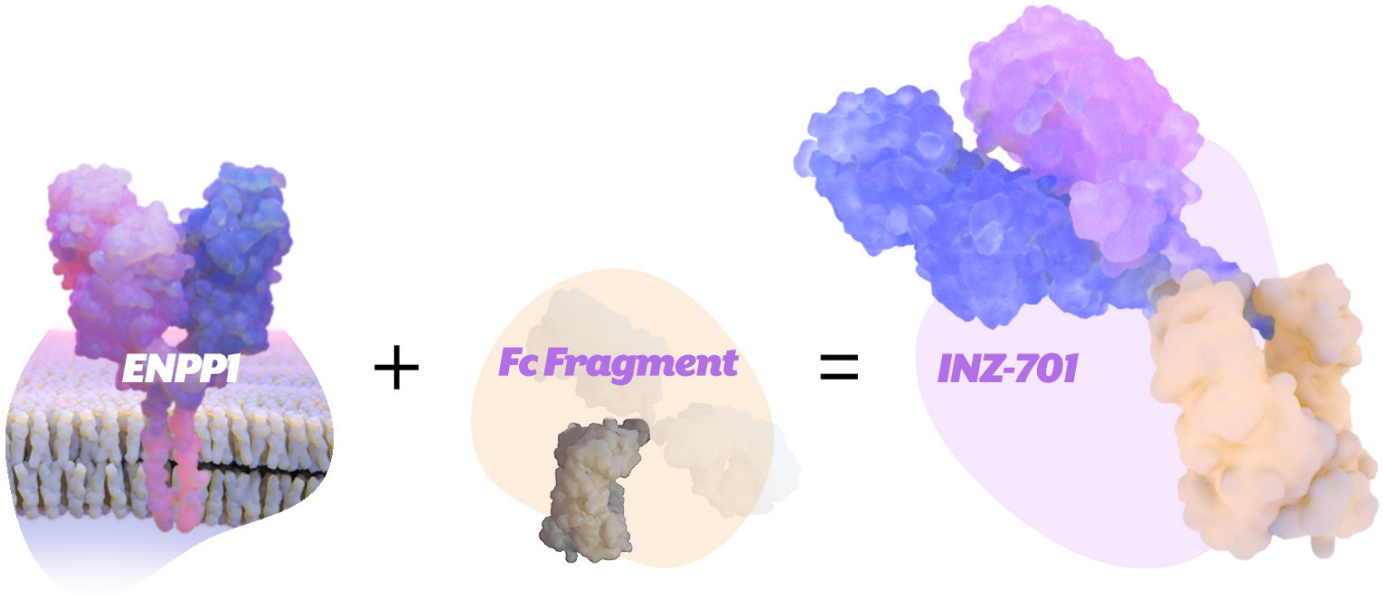
Low levels of adenosine

Low adenosine leads to narrowing and obstruction of blood vessels



INZ-701 is designed to increase PPi and adenosine

Designed for systemic availability versus native membrane-bound ENPP1 enzyme



Construct

Recombinant Fc fusion protein with soluble extracellular domain designed to improve pharmacokinetic (PK) properties

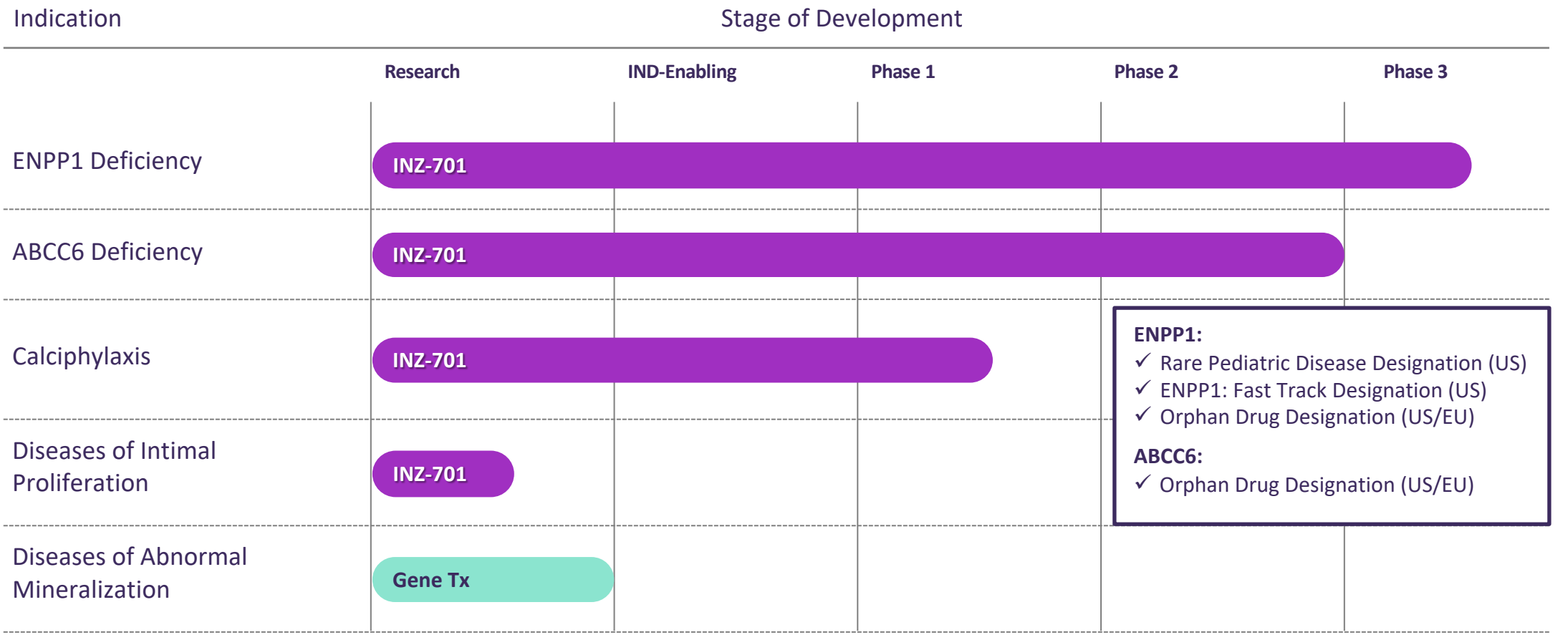
Delivery

Subcutaneous for systemic bioavailability

Enzymatic properties

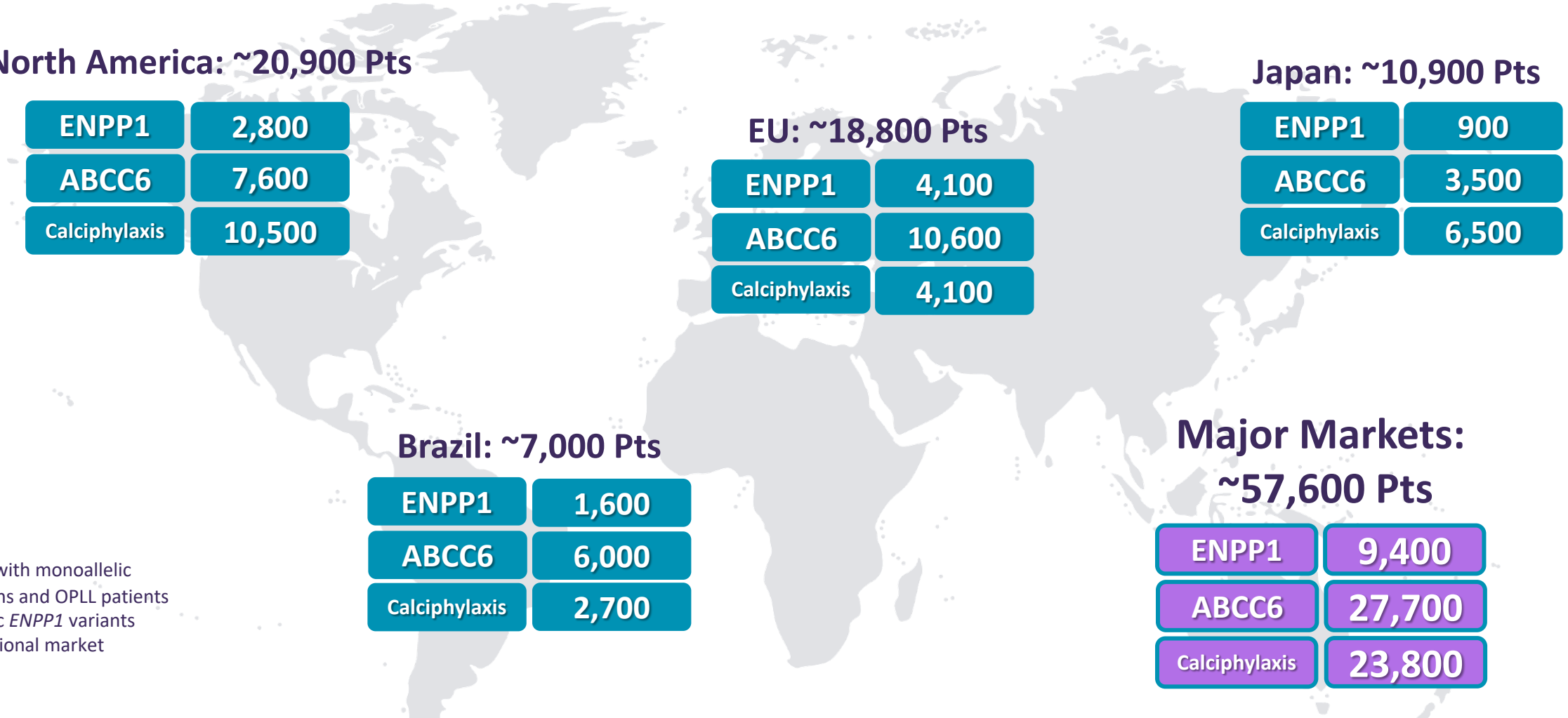
High catalytic efficiency (Kcat/Km)

INZ-701 increased PPi levels in clinical trials - Potential to be an impactful first-to-market therapy in multiple diseases



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

Significant opportunity for INZ-701 across major markets with potential for further geographic and targeted patient expansion



North America: ~20,900 Pts

ENPP1	2,800
ABCC6	7,600
Calciphylaxis	10,500

EU: ~18,800 Pts

ENPP1	4,100
ABCC6	10,600
Calciphylaxis	4,100

Japan: ~10,900 Pts

ENPP1	900
ABCC6	3,500
Calciphylaxis	6,500

Brazil: ~7,000 Pts

ENPP1	1,600
ABCC6	6,000
Calciphylaxis	2,700

Major Markets: ~57,600 Pts

ENPP1	9,400
ABCC6	27,700
Calciphylaxis	23,800

Note: Patients with monoallelic *ENPP1* mutations and OPLL patients with pathogenic *ENPP1* variants represent additional market opportunities

Sources: Company estimates. Ferreira et al. Genet Med, 2021. Ferreira et al. Orphanet Journal of Rare Diseases, 2022. Nigwekar SU, et al. J Gen Intern Med. 2014; Nigwekar SU, et al. J Am Soc Nephrol. 2016. Chinnadurai, R., Huckle, A., Hegarty, J. et al. Calciphylaxis in end-stage kidney disease: outcome data from the United Kingdom Calciphylaxis Study. J Nephrol 34, 1537–1545 (2021). https://doi.org/10.1007/s40620-020-00908-9 USRDS Annual Data Report 2021. https://adr.usrds.org/2021/end-stage-renal-disease/1-incidence-prevalence-patient-characteristics-and-treatment-modalities. Supplemented ERA-EDTA Registry data evaluated the frequency of dialysis, kidney transplantation, and comprehensive conservative management for patients with kidney failure in Europe - Kidney International (kidney-international.org) Prevalence of calciphylaxis estimated to be 2% of hemodialysis patients; North America (excl Mexico); EU = EU5 + UK.

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ENPP1 Deficiency

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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

Biallelic 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

Note: Estimates do not include symptomatic patients with monoallelic mutations

*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

Completed Phase 1/2 trial of INZ-701 in adults with ENPP1 Deficiency successfully met all study objectives

Safety

- ✓ **Favorable safety profile** was maintained
- ✓ Low/moderate, sometimes transient, ADA titers

PK/PD

- ✓ **PK data** from cohort 4 **support once-weekly dosing**
- ✓ **PPi remained elevated** with long-term treatment

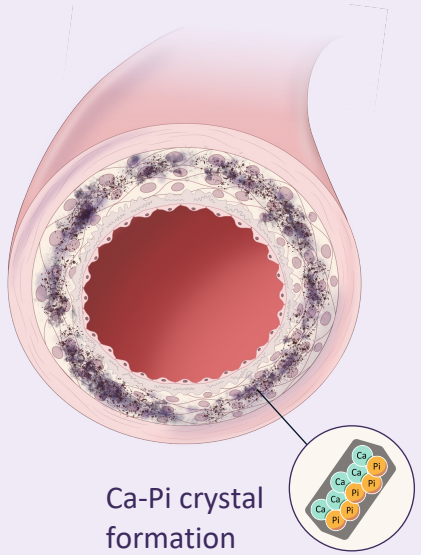
Clinical

- ✓ Favorable response on **clinical outcomes** (PROs and 6MWT) was **maintained**
- ✓ Bone biomarker response consistent with restoring proper bone mineralization

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

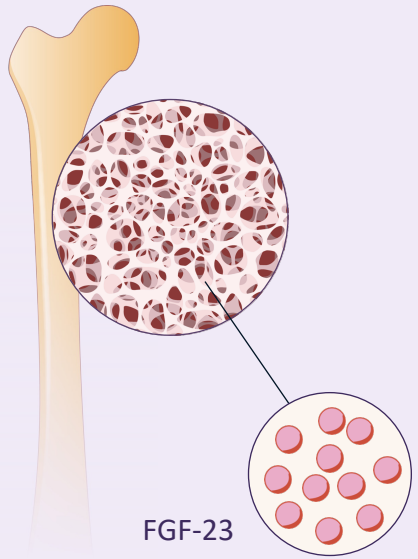
Low PPi

VASCULAR CALCIFICATION



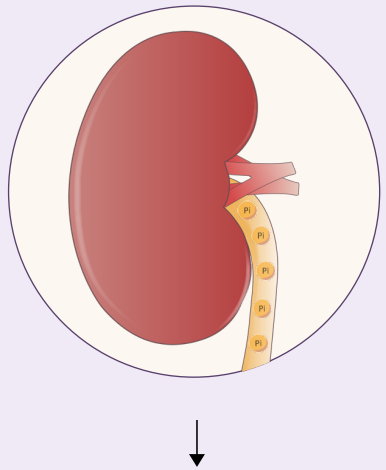
Vascular calcification and abnormal calcification surrounding bone cells leads to elevated FGF-23 levels

INCREASED FGF-23



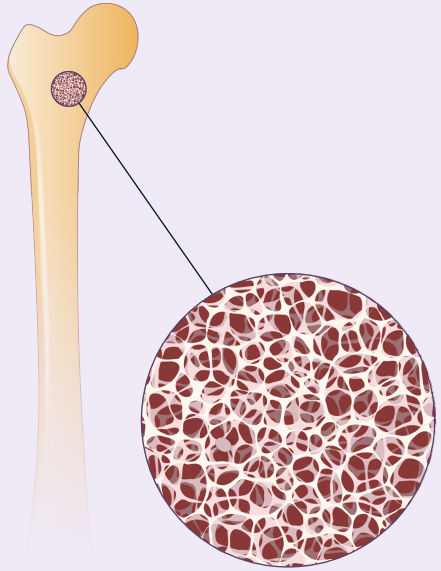
FGF-23, expressed in osteocytes in the bone, is a key regulator of phosphate in the body

RENAL PHOSPHATE WASTING



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

RICKETS/ OSTEOMALACIA



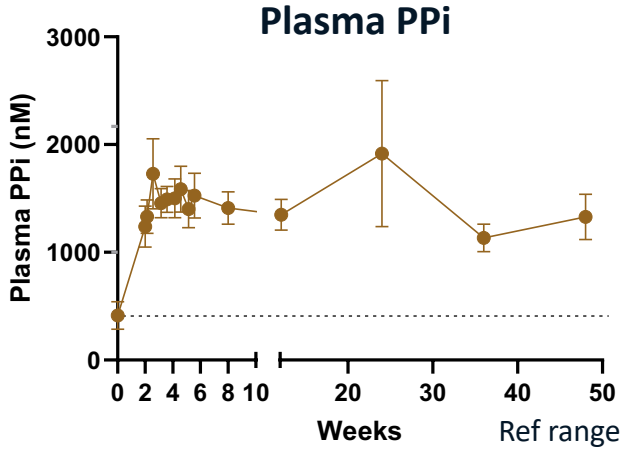
Hypophosphatemia is a central driver of rickets and osteomalacia

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]

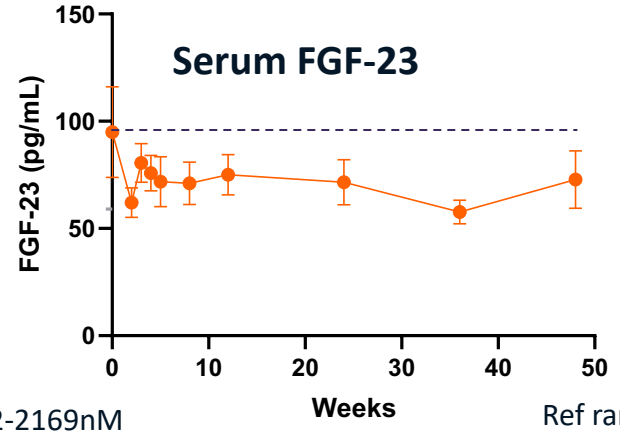
1. Stern et al, Gen in Med 2021

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

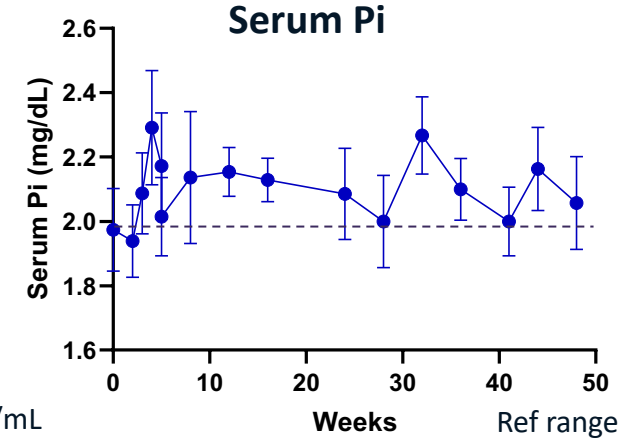
Pooled Cohorts 1-3: Baseline vs mean Week 2-48 PPi, FGF-23, and Pi levels (±SEM)



Ref range : 1002-2169nM



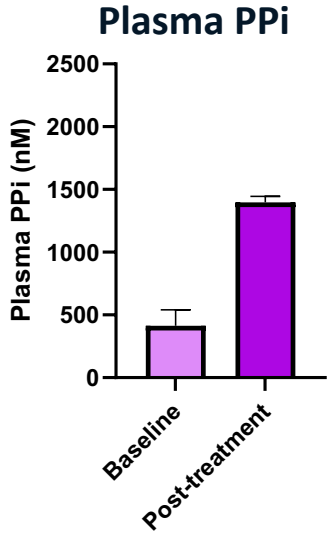
Ref range : <59pg/mL

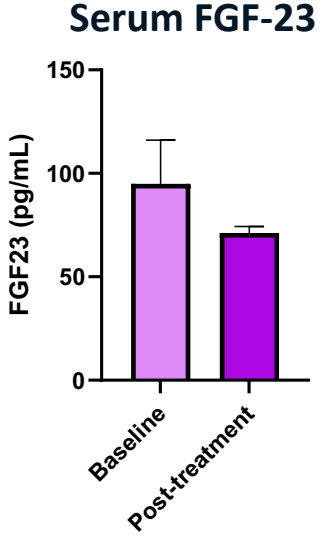


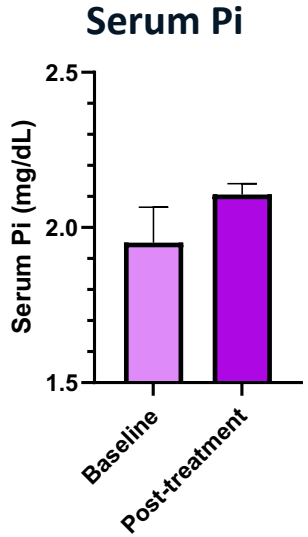
Baseline

Ref range : 2.5-4.5mg/dL

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







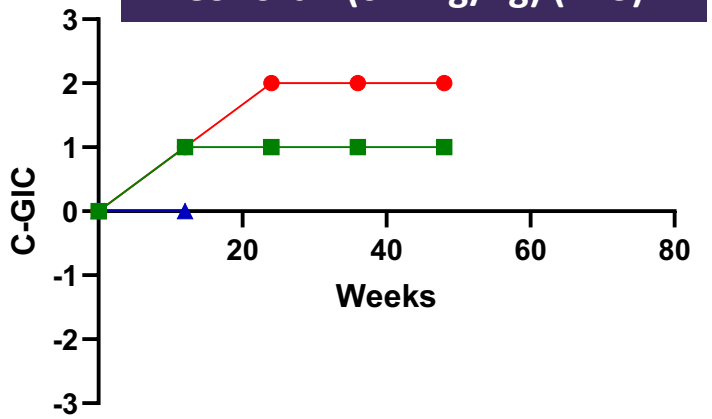
Note: Serum Pi increases observed in absence of phosphate and active vitamin D supplementation

Data cut Jan 25 2024 ; Data presented as mean ± SEM

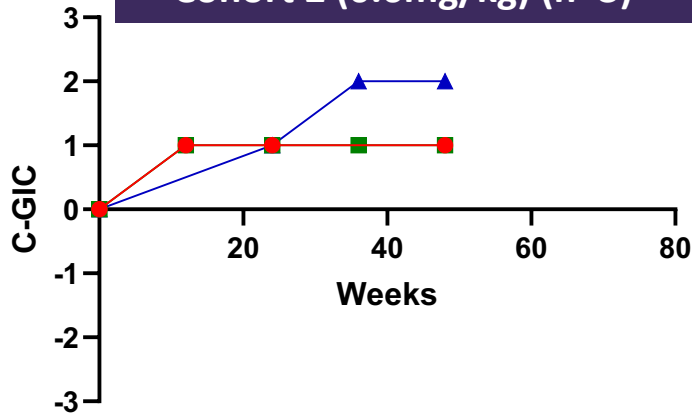
Global Impression of Change Scale: Concordant improvement in C-GIC and P-GIC in all three dose cohorts

Clinician's Global Impression

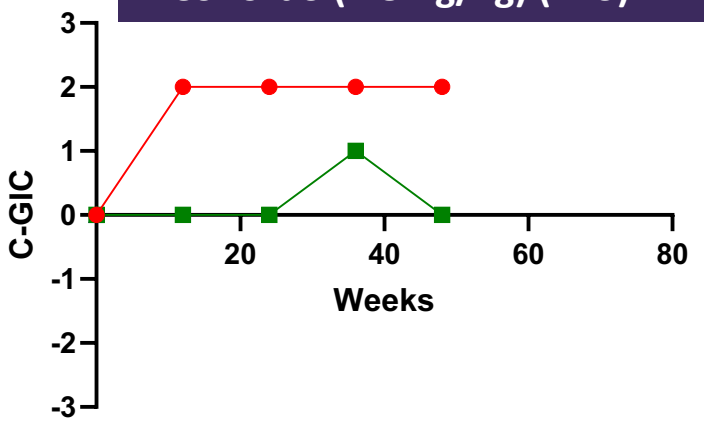
Cohort 1 (0.2mg/kg) (n=3)



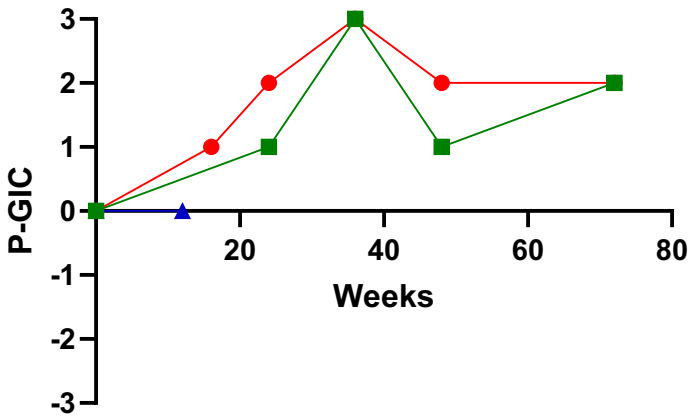
Cohort 2 (0.6mg/kg) (n=3)

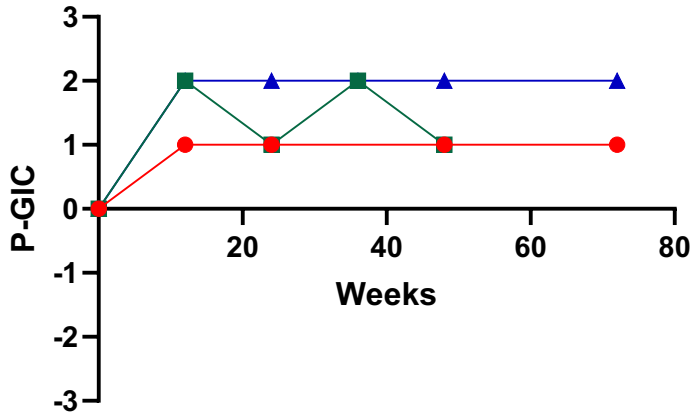


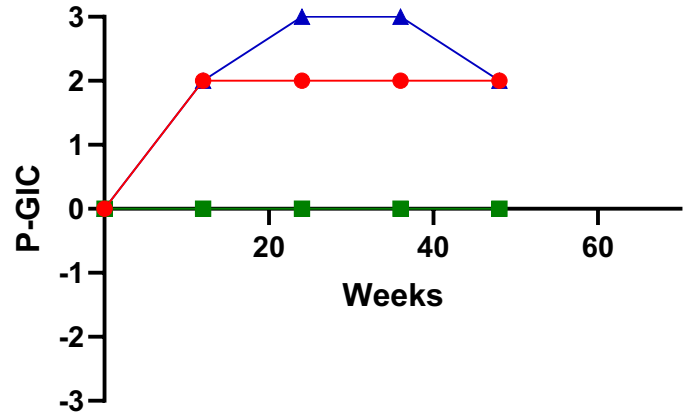
Cohort 3 (1.8mg/kg) (n=3)*

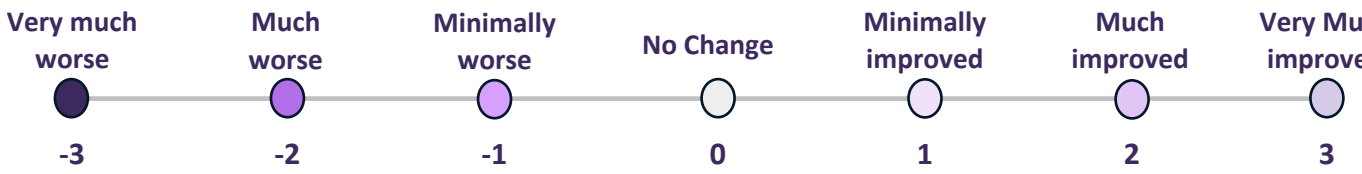


Patient's Global Impression







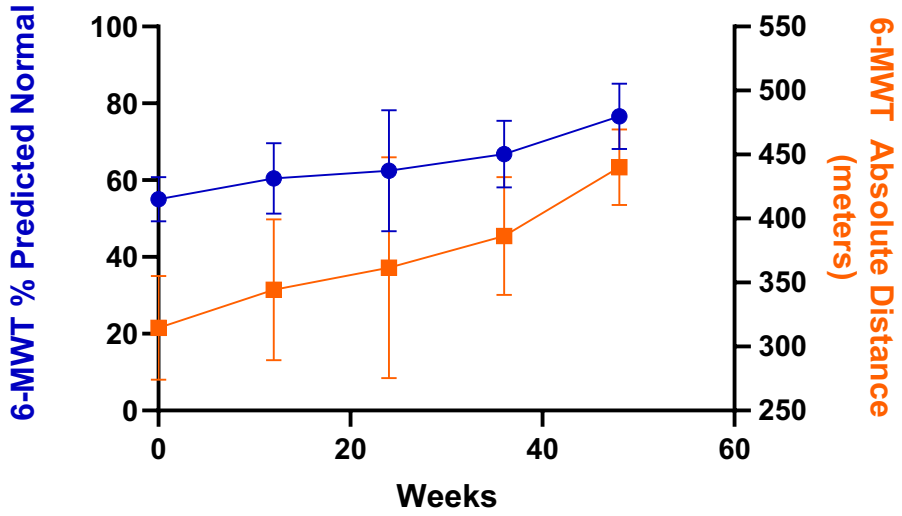


* n=2 for C-GIC

Data cut 25 Jan 24
Colors represent individual patients in respective cohorts

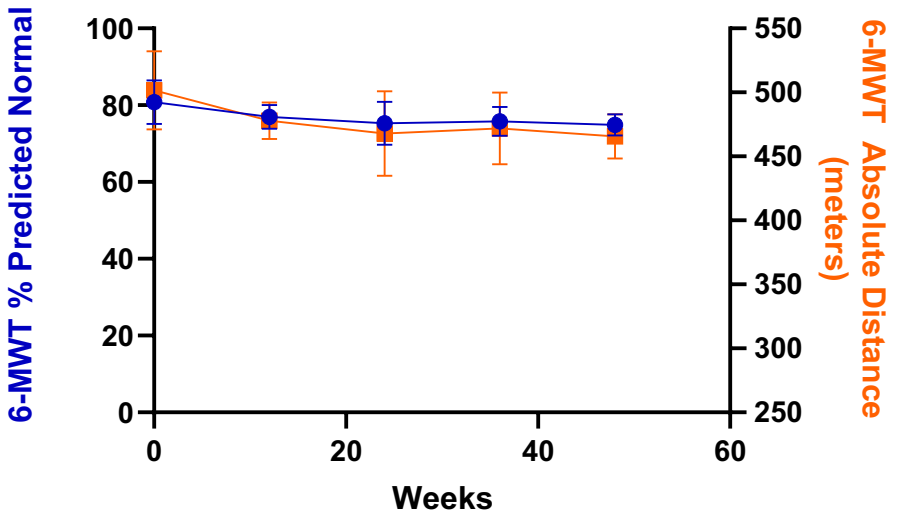
INZ-701 showed trend for improvement in 6-minute walk test (6-MWT)

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 cut Jan 25, 2024; Data presented as ± SEM

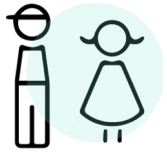
ENPP1 Deficiency: Planned Path to Global Approval

Pivotal Trial in Pediatric ENPP1 Deficiency Ongoing

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

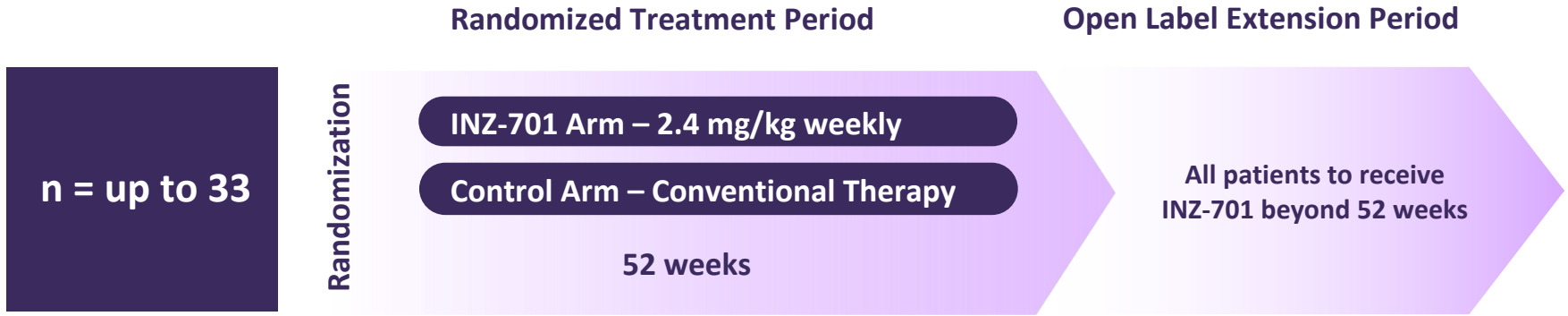
Patient recruitment underway – Topline data expected mid-2025

Population: *Pediatric*



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

Design: Randomized (2:1), Open Label



Multicenter, Multinational

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: Pharmacokinetic, ARHR2: Autosomal Recessive Hypophosphatemic Rickets Type 2, ClinicalTrials.gov: NCT06046820

Planned path to global approval of INZ-701 in ENPP1 Deficiency

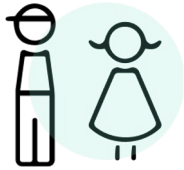
ENERGY-1: Infant (0-12 mos.)
Phase 1b
Single arm (n=8)

Endpoints

Safety *and* ***tolerability*** *as primary;* ***PPi*** *and* ***survival*** *as secondary*

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

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ABCC6 Deficiency is a multisystem, rare genetic disease: High morbidity and a continuum of effects across age groups

GACI-2
0-1 Years

~10% mortality within 12 months of birth [1]

Severe cardiovascular complications and pulmonary hypertension

Pediatric
1 to <18 years

Multisystem vasculopathy and strokes [2]

Progressive cardiovascular calcification/stenosis of major arteries

Cerebrovascular calcification -including stroke

Initial retinal calcification

PXE
18+ Years

Blindness, cardiovascular disease and mobility impairment [3-7]

Progressive arterial calcification

Increased incidence of stroke and dementia

Retinal calcification – Angioid streaks, atrophy

Progressive calcification and fragmentation of elastic fibers

Genetic Prevalence: 1:25,000 - 1:50,000 [8-9]

Sources: *1. Ferreira et al. JBMR 2021; 2. Grossi et al, Eur J Med Genet, 63 2020; 3. Shimada et al. Int.J.Mol.Sci. 2021; 4.Risseeuw et al. Retina, 2019; 5. Leftheriotis et al. J Vasc Surg, 2011; 6. Vanakker et al. Hum Mutat. 2008; 7. Van den Berg et al. Cerebrovasc Dis, 2000; 8. Internal, Unpublished Data; 9. Ferreira et al. Genet Med, 2021*

Completed Phase 1/2 trial of INZ-701 in adults with ABCC6 Deficiency successfully met all study objectives

Safety

- ✓ INZ-701 demonstrated a **favorable safety profile**
- ✓ No serious or severe adverse events
- ✓ Low/moderate, sometimes transient, ADA titers

PK/PD

- ✓ **Rapid** and **sustained increase in PPi** observed in highest dose cohort (1.8 mg/kg)

Clinical

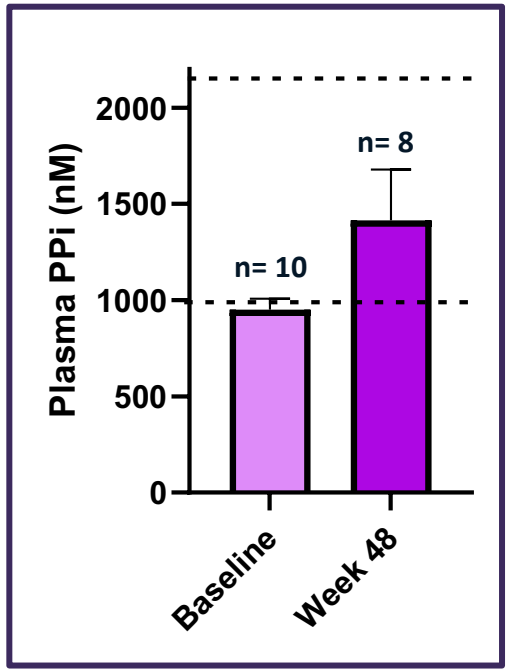
- ✓ **Positive changes** in multiple affected organ systems (cerebrovasculature and choroidal layer of eye) support **improvements in vascular health**
- ✓ Improvement in visual function (VFQ-25) and multiple PROs observed

VFQ-25: Visual Function Questionnaire; PRO: Patient Reported Outcomes; ADA: Anti-drug antibodies

INZ-701 showed benefit across multiple domains relevant for future pivotal trial

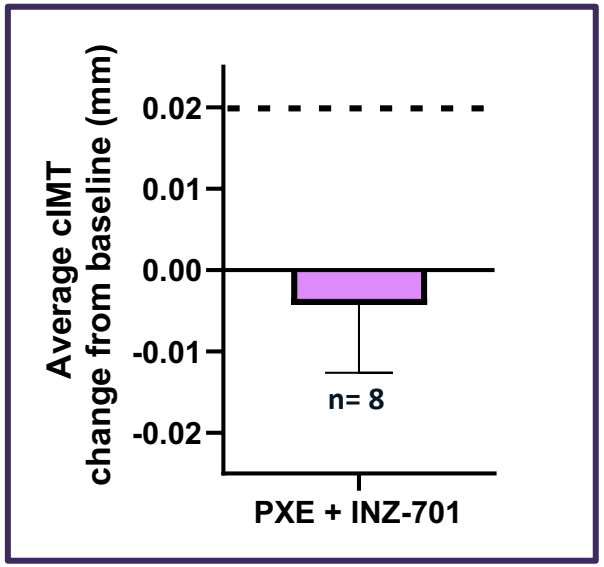
Combined cohort 1-3 data comparing baseline to week 48

PPi increased



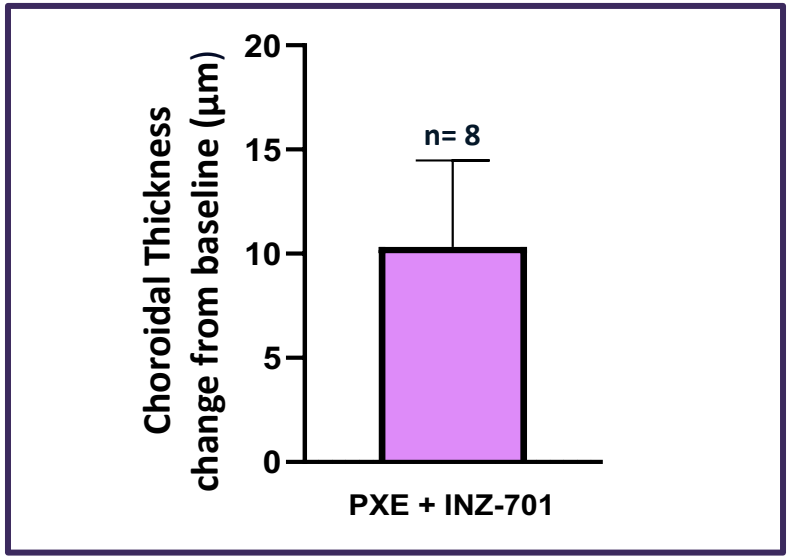
Normal range

Carotid artery intima-media thickness decreased (cIMT)



Mean annual cIMT change in TEMP study[1]

Choroidal thickness increased

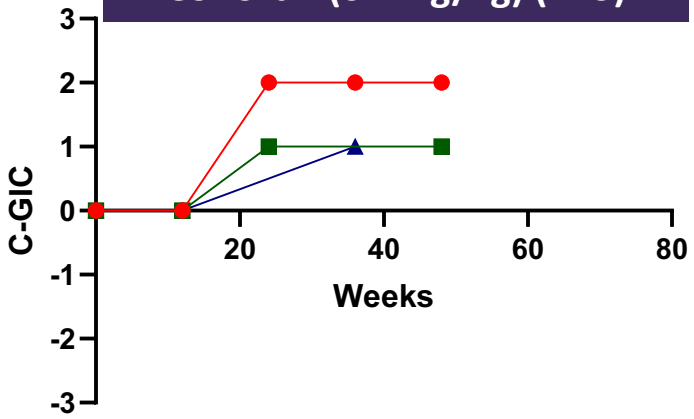


Data cut – 10 Jan 2024; Data presented as mean ± SEM **Source:** 1. *Kranenburg et al, JACC, 2018*

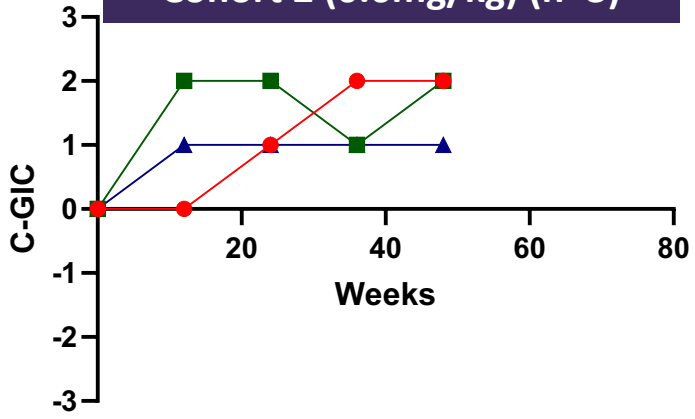
Global Impression of Change Scale: Concordant improvement in C-GIC and P-GIC in all three dose cohorts

Clinician's Global Impression

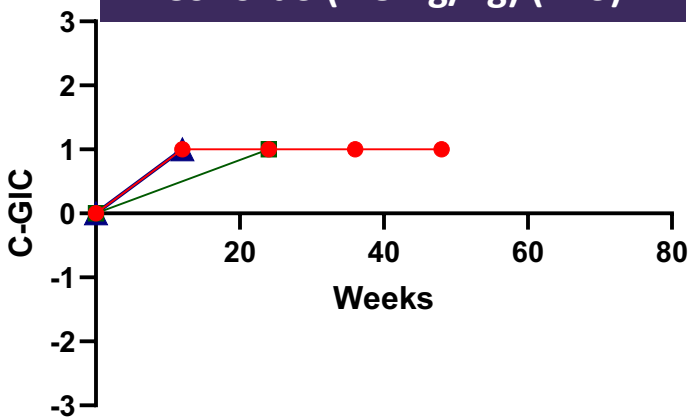
Cohort 1 (0.2mg/kg) (n=3)



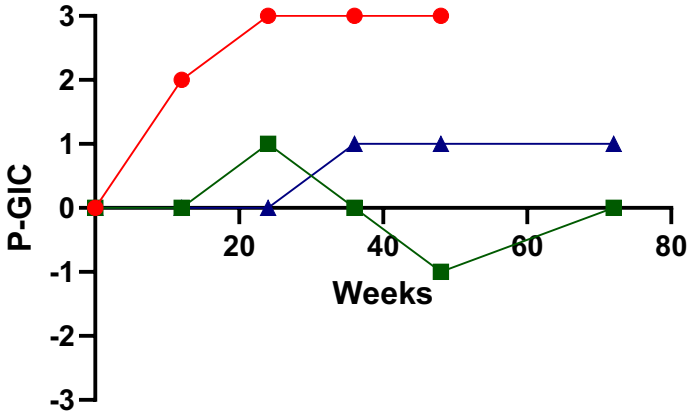
Cohort 2 (0.6mg/kg) (n=3)

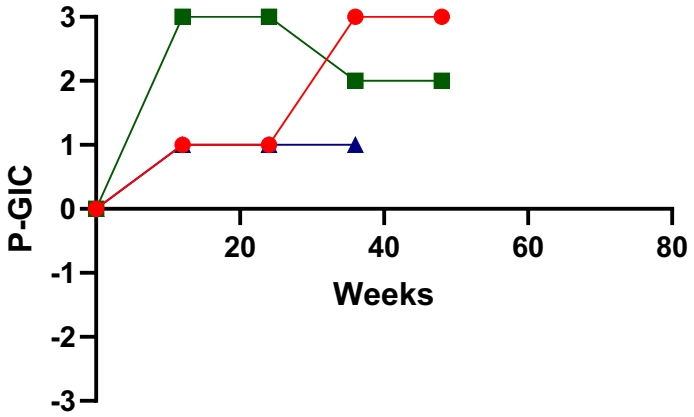


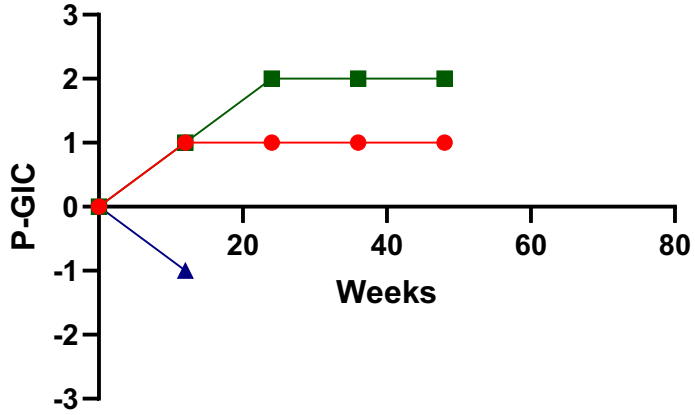
Cohort 3 (1.8mg/kg) (n=3)

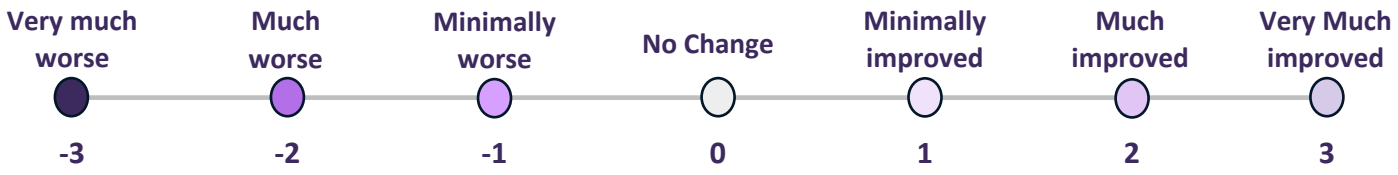


Patient's Global Impression









Data cut – 10 Jan 2024
Colors represent individual patients in respective cohorts

ABCC6 Deficiency Development Plan

Focused on pediatric population with ABCC6 Deficiency

Unmet Need

- ✓ Retrospective natural history study (early-onset) and interventional study (adults) identified **risk of stroke** and **retinal disease** as consistent presentation in ABCC6 Deficiency

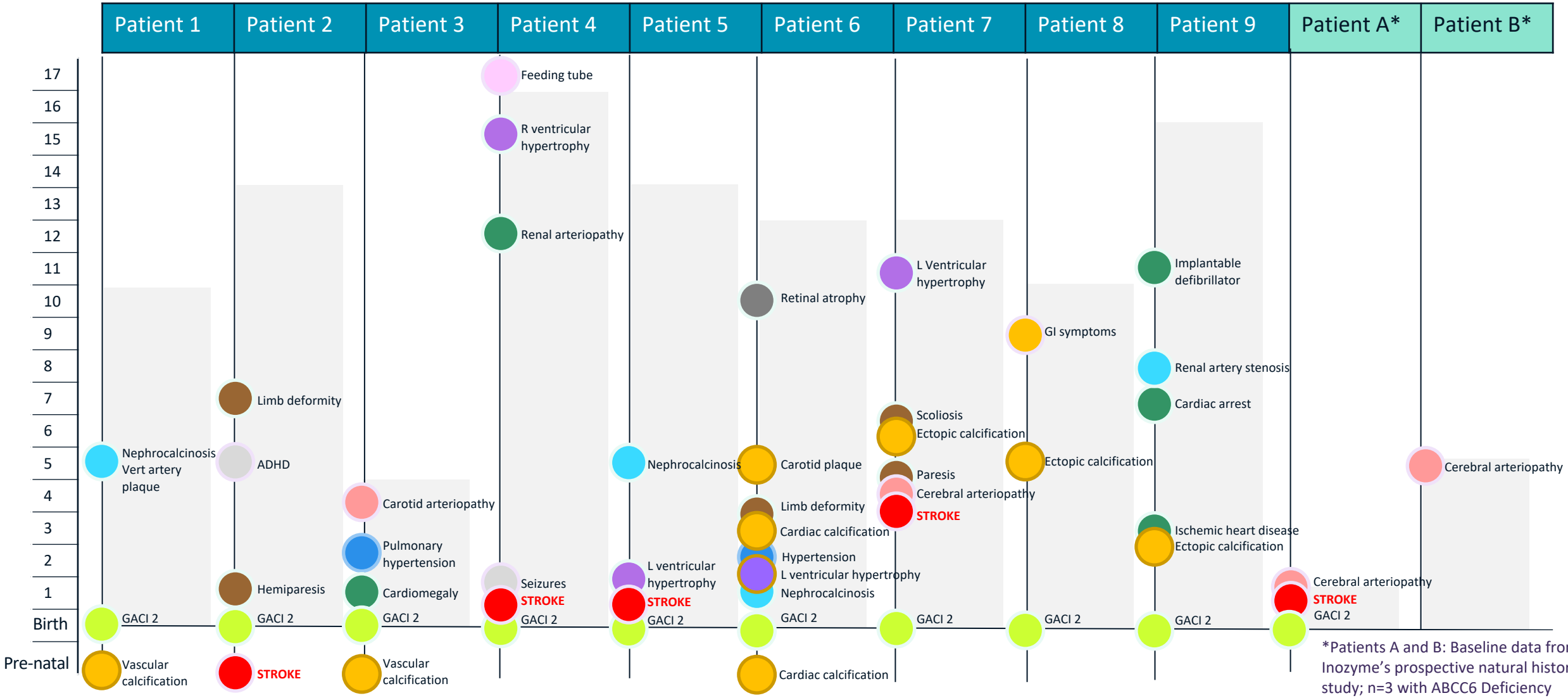
Market

- ✓ Market research identified **substantial pediatric population** that represents the most important unmet need in ABCC6 Deficiency

Regulatory

- ✓ Pivotal trial design planning in progress
- ✓ Plan to seek **accelerated approval** based on imaging metric predictive of ischemic stroke

Retrospective Natural History Study: ABCC6 Deficiency patients had a heavy disease burden early in life



*Patients A and B: Baseline data from Inozyme's prospective natural history study; n=3 with ABCC6 Deficiency

Data cut – 10 Jan 2024

Shaded area indicates age at time of informed consent

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

Ongoing Study

ENERGY-1: Infant (0-12 mos.)
Phase 1b
Single arm

- ***Safety*** *and* ***tolerability*** *as primary*
- ***PPi*** *and* ***survival*** *as secondary*

Future Studies

Pediatric (≥1-<18 yrs.)*
Pivotal
Randomized, controlled

- *Potential accelerated approval based on endpoints predictive of clinical benefit over 12–18-month randomized period*
- *Monitor for* ***cerebrovascular, cardiovascular and ophthalmic*** *outcomes against untreated control population over 2-4 years to support full approval*

Adult – PXE (18+)*
Pivotal
Randomized, controlled

- *Composite endpoint comprised of* ***retinal measurements, peripheral arterial disease outcomes*** *and* ***PPi***

Completed Study

Adult – PXE (18+)
Phase 1/2
Single arm – MAD

- *Generally safe and well tolerated*
- *Consistently elevated PPi at highest dose*
- *Signals of clinical activity on vascular and ophthalmic for retinal endpoints*

*Subject to regulatory discussions and appropriate financial resources

Basis for Potential Accelerated Approval (US) /Conditional Approval (EU)

1st BLA/MAA

- Adult Phase 1/2 full data
- ENERGY-1 available data
- Pediatric Pivotal trial data

Additional filings

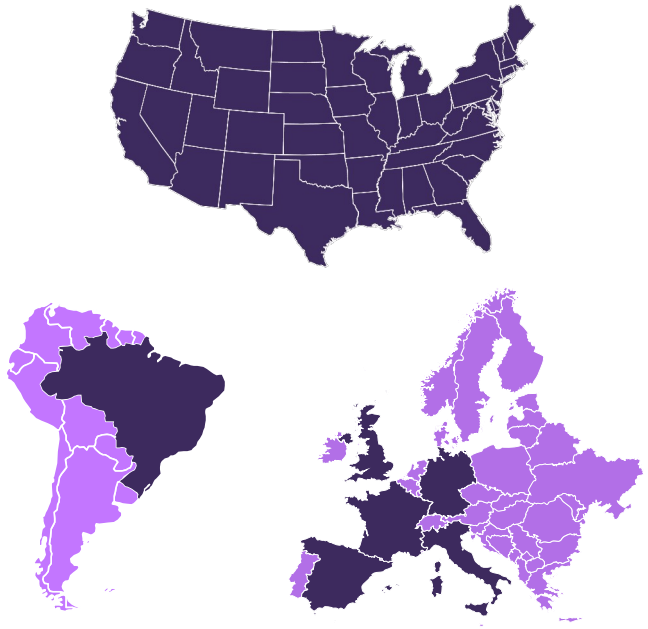
- Adult (18+) study (Supplemental BLA/MAA)
- Japan, Brazil, Middle East

Building a Rare Disease Franchise

inozyme pharma

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

Newborn Screening

US – Rady Children's Hospital Network
UK – Genomics England
Efforts ongoing to add to other panels across the globe

Expanding HCP Audience

Infant and Pediatric ENPP1/ABCC6

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

Adult ENPP1/ABCC6

- 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

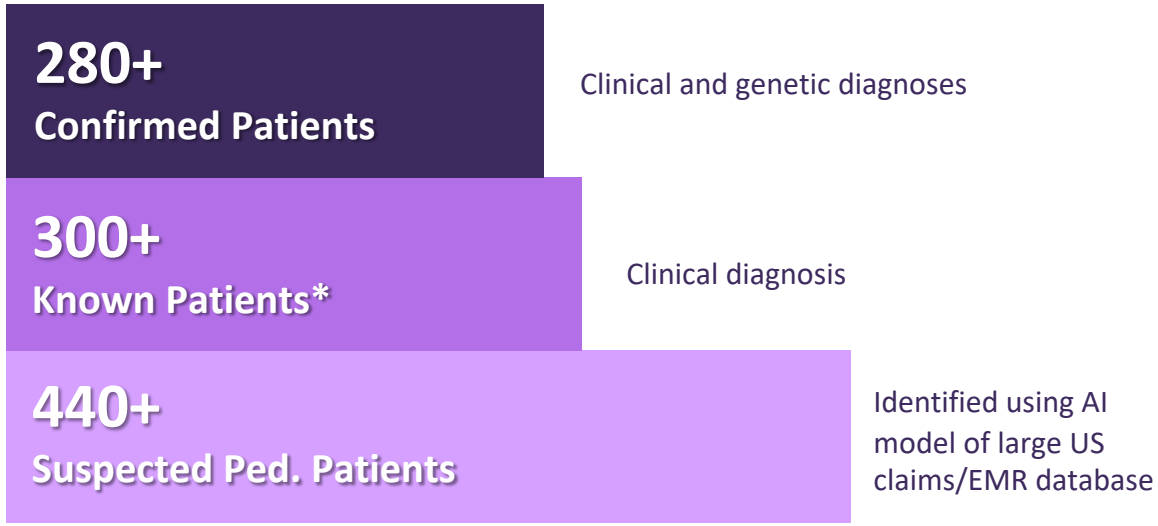
Partnering to Remove Barriers to Diagnosis

GACI Global – circulating hope
BeginNGS – Newborn Genomic Sequencing to end the diagnostic odyssey
Genomenon
ipm.ai – Part of Real Chemistry
Invitae
BlueDil int'l
Prevention Genetics
Genomics England
CentoGene
Rady Children's

Identifying ENPP1 patients to support market potential – strong progress to date and expanding efforts into patients with monoallelic mutations

Biallelic Genetic Prevalence[1]: 1:64,000

1000+ Global patients identified with **confirmed, known, or suspected** ENPP1 Deficiency



Internal data as of 1/4/24; number of confirmed patients expected to increase with patient/physician education, initiation of clinical trials, and progress towards potential regulatory approval

* Phenotypic findings of disease only

- Identified ~82 confirmed symptomatic monoallelic ENPP1 patients
  - Identified patients range in age from 0 to 70+ years of age
- Majority of patients identified through Skeletal Disorders or Hypophosphatemia gene panels
  - Suggests monoallelic patients can have clinical symptomatology similar to those with biallelic ENPP1 Deficiency
- Conducting observational study to characterize clinical features of adults with monoallelic *ENPP1* mutations

Sources: 1. Ferreira et al. Orphanet Journal of Rare Diseases, 2022; 1:64,000 pregnancies

~1,300 likely U.S. pediatric patients with ABCC6 Deficiency were identified, representing ~70% of estimated genetic prevalence

Pediatric ABCC6 Deficiency: U.S. Patient estimates

Ischemic Stroke
940 patients

- *Ischemic stroke between ages 1-18*
- *Genetic panel ordered between ages 1 and <18 **OR** mild neurological symptoms occurred prior to stroke*
- *PXE or a phosphorous disorder diagnosis code in all history*
- *Exclusion of differential diagnoses*

Angioid Streaks
264 patients

- *Angioid streaks between ages 1 and <18*
- *Exclusion of differential diagnoses and eye injuries*

Retinal Imaging/OCT
60 patients

- *Optical coherence tomography (OCT) between ages 1 and <18*
- *Genetic panel ordered **AND** mild neurological symptoms occurred between ages 1 and <18*
- *PXE or a phosphorous disorder diagnosis code in all history*
- *Exclusion of differential diagnoses*

Cardiovascular Anomaly
24 patients

- *Cardiovascular anomaly **AND** arterial calcification between ages 1-and <18*
- *PXE or a phosphorous disorder diagnosis code in all history*
- *Exclusion of differential diagnoses*

Identified 1,288 likely U.S. pediatric patients with ABCC6 Deficiency

Sources: *IPM Longitudinal claims database An all-history (2009 – present) lookback period was utilized for clinical profiling Patients with 2+ claims for any one differential diagnosis (sickle cell disorders, Ehlers-Danlos syndrome, Paget's disease) were excluded. Based on 65% medical claims capture rate. No projection factor was used for claims coverage.*

Calciphylaxis

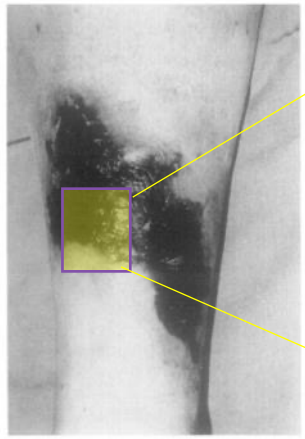
Calciphylaxis: A severe complication of ESKD with high mortality and morbidity

Calciphylaxis Incidence: 3.5 : 1,000 ESKD Patients[7]

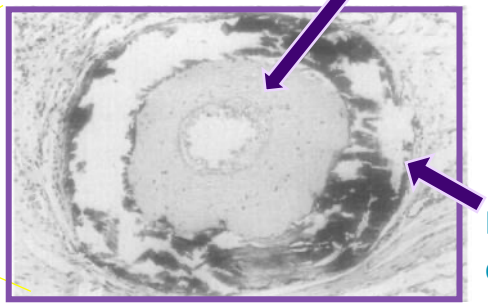
Major Markets Estimate: 5,000 patients/year

Primarily affects end stage kidney disease (ESKD) patients[2]

Vascular calcification-mediated disease



(Hafner et al, JAAD, 1995)

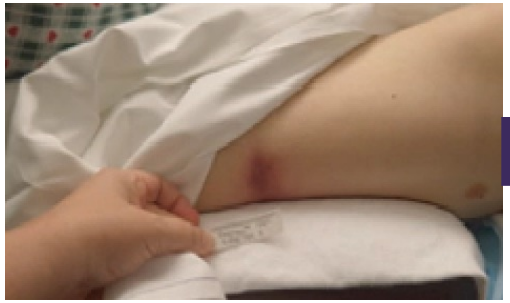


(Hafner et al, JAAD, 1995)

Microvascular occlusion of skin arterioles caused by medial calcification, intimal proliferation, and thrombosis; Low PPi

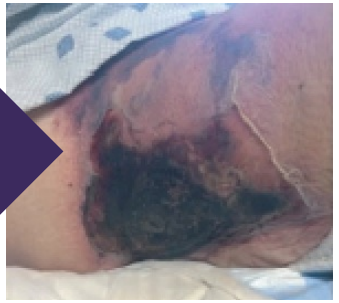
Significant morbidity and mortality

Initial skin lesions typically present as extremely painful plaques and nodules, and progress to necrotic ulcers



Ghosh T, et al. Int J Dermatol. 2017

2 months



Ghosh T, et al. Int J Dermatol. 2017

>70% require hospitalization for severe ulcerations[4]

~50% of patients are bedridden or wheelchair-bound[4]

~50% mortality

1 year after diagnosis[7]

- Median survival time: 2.6 months[4]
- Sepsis most common cause of death[4-6]

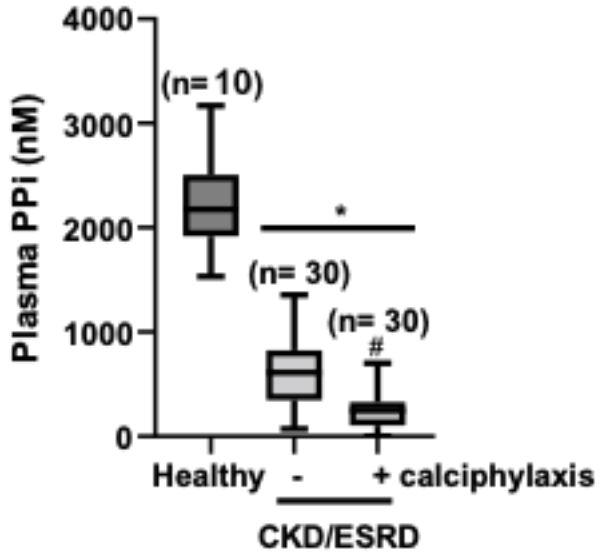
No approved therapy

Sources: 1. Nigwekar SU, et al. J Gen Intern Med. 2014; 2. Nigwekar SU, et al. J Am Soc Nephrol. 2016; 3. USRDS Annual Data Report 2021. https://adr.usrds.org/2021/end-stage-renal-disease/1-incidence-prevalence-patient-characteristics-and-treatment-modalities ;4. Weenig RH, et al. J Am Acad Dermatol. 2007; 5. Nigwekar SU, et al. Clin J Am Soc Nephrol. 2008; 6. Bazari H, et al. N Engl J Med. 2007; 7. Nigwekar et al. NEJM 2018.

Calciphylaxis is associated with PPi deficiency

Arteriolar calcification largely develops due to imbalance between calcification inhibitors and promoters[1-3]

PPi levels lower in calciphylaxis



PPi levels predicted 6-month mortality among patients with calciphylaxis

Data presented as median ± interquartile range

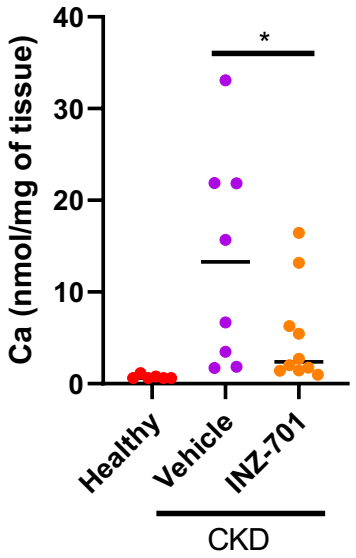
- ESKD patients had significantly lower PPi levels compared to healthy subjects[4]
- Calciphylaxis patients had significantly lower plasma PPi levels when compared with non-calciphylaxis ESKD patients[4]
- Published data showed correlation between PPi levels and severity of calcification

Sources: 1. Kramann R, et al. Nephrol Dial Transplant. 2013; 2. Moe SM, et al . Pediatr Nephrol. 2003; 3. Shroff R, et al. J Am Soc Nephrol. 2013; 4. Internal, Unpublished Data, ECTS 2022 Nigwekar et al; ESKD = End stage kidney disease, CKD = Chronic kidney disease

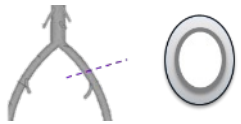
INZ-701 reduced vascular calcification in a CKD model

INZ-701 prevented extensive calcification in iliac artery

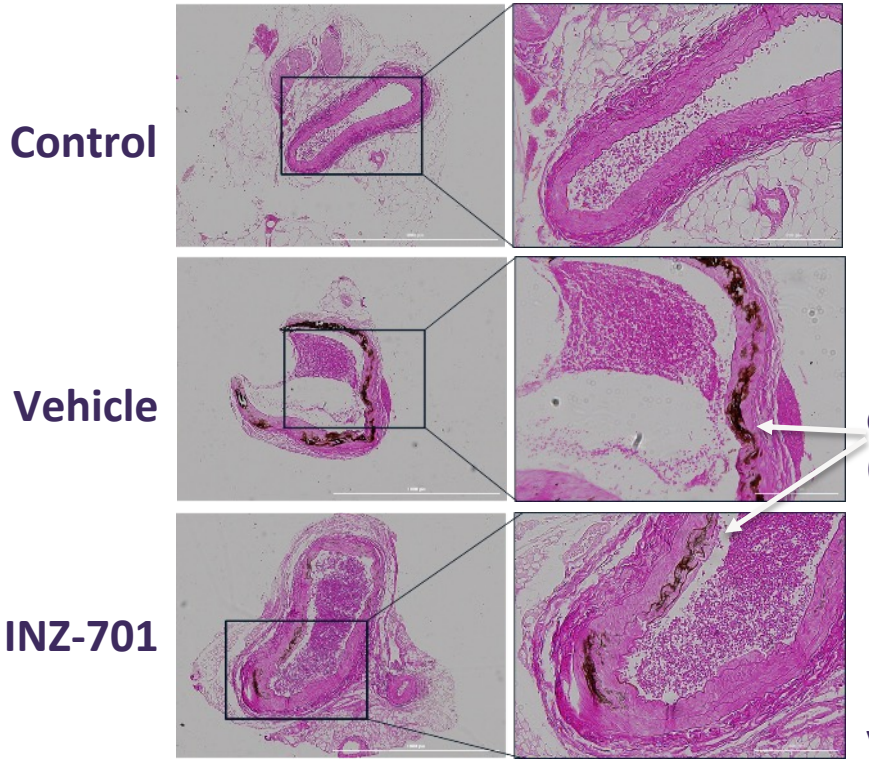
Right Iliac Artery



* p=0.0337 vs Vehicle
one tailed t-test



Left Iliac Artery



- CKD rats dosed with vehicle showed extensive, often circumferential, medial calcification that extended over multiple levels
- In contrast, CKD rats dosed with INZ-701, had a significant reduction in calcification
- Similar prevention of calcification observed in ascending aorta

Chronic kidney disease (CKD) was induced in Sprague Dawley (SD) rats with an adenine diet. 100ng/kg calcitriol SC was also given to the rats; INZ-701 = 10mg/kg, SC QOD for 9 weeks initiated with adenine diet. Sources: Internal, Unpublished Data

SEAPORT-1: Phase 1 trial in patients with end-stage kidney disease (ESKD) receiving hemodialysis

Interim data expected in Q4 2024

Study Population:
Adults

Eligibility Criteria:

- ≥18 to <70 years
- ESKD and receiving hemodialysis
- Undergoing 3 treatments of HD per week
- Low plasma PPi

Design: Single arm, Open Label

n = up to 10

INZ-701 – 1.8 mg/kg Weekly
coinciding with dialysis days

30 days treatment, weekly dosing

Up to 3 US sites

Primary Goals

- Change from baseline in **plasma PPi** concentration
- **Safety**

Secondary Goals

- **Pharmacokinetic (PK)** and **pharmacodynamic (PD)** parameters

Clinicaltrials.gov: NCT06302439

Anticipated milestones provide robust news flow

Milestone	2024	2025
ENPP1 Deficiency		
Topline Data – Adult Phase 1/2 Trial	✓	
Initiate – ENERGY-2 Pivotal Trial in Infants – Ex. U.S.	**2H 24**	
Interim Data – ENERGY-1 Phase 1 Infant Trial	**2H 24**	
Topline Data – ENERGY-3 Pivotal Pediatric Trial		**Mid-Year**
ABCC6 Deficiency		
Topline Data – Adult Phase 1/2 Trial	✓	
Initiate Pivotal Trial*		**Q1 25**
Calciphylaxis		
Initiate SEAPORT-1 - Phase 1 Trial**	✓	
Interim Data - SEAPORT-1 Phase 1 Trial	**Q4 24**	

*Pending regulatory discussions and appropriate financial resources, **Phase 1 trial in patients with end-stage kidney disease (ESKD) receiving hemodialysis

Inozyme is at the forefront of developing transformative therapies for rare diseases of pyrophosphate (PPi) deficiency

- ✓ **ENPP1 Deficiency, ABCC6 Deficiency and calciphylaxis are serious diseases with no approved therapies**
- ✓ **INZ-701 has demonstrated rapid, significant, and sustained increase in PPi levels, and exhibited a favorable safety profile across multiple clinical trials**
- ✓ **Currently in pivotal trial for ENPP1 Deficiency; Completed Phase 2 trial for ABCC6 Deficiency**
- ✓ **Experienced team with a track record of success in rare disease and a strong focus on execution**
- ✓ **In a position of financial strength, with several expected upcoming milestones and a pipeline designed for long-term value creation**
  - $166.2M expected to fund operations into Q4 2025 as of 3/31/24
  - 61.85M common shares outstanding as of 5/2/24

Thank you


Nora
Living with
ENPP1 Deficiency
Callum
Living with
ENPP1 Deficiency