# Inozyme Pharma Reports Second Quarter 2023 Financial Results and Provides Business Highlights

August 8, 2023

*– Interim data readouts from ongoing Phase 1/2 trials of INZ-701 in adults with ENPP1 Deficiency and ABCC6 Deficiency expected in September 2023 –*

*– Plasma pyrophosphate (PPi) to serve as primary endpoint in the U.S. and co-primary endpoint in the EU for ENERGY-3 pediatric pivotal trial for ENPP1 Deficiency; initiation expected in October 2023 –*

*– Cash, cash equivalents, and short-term investments as of June 30, 2023, together with proceeds from July 2023 offering, expected to fund cash flow requirements into the fourth quarter of 2025 –*

BOSTON, Aug. 08, 2023 (GLOBE NEWSWIRE) -- Inozyme Pharma, Inc. (Nasdaq: INZY) ("Inozyme" or "Company"), a clinical-stage rare disease biopharmaceutical company developing novel therapeutics for the treatment of pathologic mineralization and intimal proliferation, today reported financial results for the second quarter ended June 30, 2023, and provided business highlights.

"We continue to make tremendous progress advancing our clinical programs of INZ-701 in patients with ENPP1 Deficiency and ABCC6 Deficiency, with interim data from both programs expected in September 2023," said Douglas A. Treco, Ph.D., chief executive officer and chairman of Inozyme's board of directors. "Notably, following regulatory discussions with the FDA and EMA, we have outlined a clear path forward toward supporting potential regulatory filings in ENPP1 Deficiency."

**Recent Clinical Highlights**

- **Global Development Strategy Update for INZ-701 in Patients with ENPP1 Deficiency.** In July 2023, Inozyme announced that based on guidance from the U.S. Food and Drug Administration (FDA) and the Paediatric Committee (PDCO) of the European Medicines Agency (EMA), change in plasma PPi will serve as the primary endpoint in the U.S., supported by consistent trends in appropriate secondary endpoints, and a co-primary endpoint in the EU for the ENERGY-3 pivotal trial in pediatric patients.

  Based on regulatory feedback from the FDA and EMA, positive data from the ongoing and planned clinical trials of INZ-701 in patients with ENPP1 Deficiency, including comprehensive data demonstrating clinical impact of plasma PPi, could provide the basis for the Company's submission of marketing applications in both the U.S. and EU. These data will include final results from the ongoing Phase 1/2 trial in adults, available results from the ongoing ENERGY-1 Phase 1b and planned ENERGY-2 pivotal trial in infants, and final results from the planned ENERGY-3 pivotal trial in pediatric patients.

  If these marketing applications are approved, the Company expects to commercially launch INZ-701 for infant and pediatric patients as early as the second half of 2026. Pending regulatory discussions and appropriate financial resources, the Company also plans to conduct the ENERGY-4 pivotal trial in adolescents and adults with ENPP1 Deficiency. Data from the planned ENERGY-4 trial may provide the basis for supplemental marketing applications.

- **ENERGY-1 Phase 1b Trial of INZ-701 in Infants with ENPP1 Deficiency.** Dosing is now underway in the Phase 1b ENERGY-1 trial designed to primarily assess the safety, tolerability, pharmacokinetic, and pharmacodynamic profile of INZ-701 in infants with ENPP1 Deficiency.
- **Medical Conference Presentations.** In the second quarter, Yves Sabbagh, Ph.D., Inozyme's senior vice president and chief scientific officer, and Kurt Gunter, M.D., Inozyme's senior vice president and chief medical officer, presented previously reported interim data from the ongoing Phase 1/2 clinical trial of INZ-701 in patients with ENPP1 Deficiency at the European Calcified Tissue Society Congress and European Congress of Endocrinology, respectively.

**Recent Corporate Highlights**

- **Closed Underwritten Offering.** In August 2023, the Company closed an underwritten public offering of 14,375,000 shares of its common stock at a price of $4.80 per share, for net proceeds of approximately $64.5 million from the offering, after deducting underwriting discounts and commissions and estimated offering expenses.

**Anticipated Milestones**

- **ENPP1 Deficiency**
  - Interim data from Cohorts 1-3 in the Phase 2 portion of the ongoing Phase 1/2 trial in adults – Sep. 2023
  - Initiation of the ENERGY-3 pivotal trial in pediatric patients – Oct. 2023
  - Topline data from Cohorts 1-3 in the ongoing Phase 1/2 trial in adults – Q1 2024
  - Initiation of the ENERGY-2 pivotal trial in infants, ex-U.S. – Q2 2024

- Interim data from the ENERGY-1 Phase 1b trial in infants – 2H 2024
- Topline data from the ENERGY-3 pivotal trial in pediatric patients – Mid-2025

- **ABCC6 Deficiency**

  - Interim data from the Phase 2 portion of the ongoing Phase 1/2 trial in adults – Sep. 2023
  - Topline data from the Phase 2 portion of the ongoing Phase 1/2 trial in adults – Q1 2024
  - Initiation of Phase 2 clinical trial of INZ-701 in adult patients with ABCC6 Deficiency, subject to regulatory review and sufficient funding – Q4 2024

**Second Quarter 2023 Financial Results**

- **Cash Position and Financial Guidance.** Cash, cash equivalents, and short-term investments were $140.2 million as of June 30, 2023. Based on its current plans, the Company now anticipates its cash, cash equivalents, and short-term investments as of June 30, 2023, together with approximately $64.5 million in proceeds from the July 2023 public offering, will enable the Company to fund its cash flow requirements into Q4 2025.
- **Research and Development (R&D) Expenses.** R&D expenses were $11.7 million for the quarter ended June 30, 2023, compared to $10.0 million for the prior-year period. This increase was primarily due to increased chemistry, manufacturing, and controls expenses, clinical development costs, and personnel-related expenses to support our clinical trials.
- **General and Administrative (G&A) Expenses.** G&A expenses were $4.7 million for the quarter ended June 30, 2023, compared to $5.4 million for the prior-year period. The decrease was primarily related to a decrease in stock-based compensation expense and cost-saving initiatives.
- **Net Loss.** Net loss was $15.6 million, or $0.35 loss per share, for the quarter ended June 30, 2023, compared to $15.3 million, or $0.38 loss per share, for the prior-year period.

**About ENPP1 Deficiency**

ENPP1 Deficiency is a progressive condition that manifests as a spectrum of diseases. Individuals who present in utero or in infancy are typically diagnosed with generalized arterial calcification of infancy (GACI), which is characterized by extensive vascular calcification and intimal proliferation (overgrowth of smooth muscle cells inside blood vessels), resulting in myocardial infarction, stroke, or cardiac or multiorgan failure. Approximately 50% of infants with ENPP1 Deficiency die within six months of birth. Children with ENPP1 Deficiency typically develop rickets, a condition diagnosed as autosomal-recessive hypophosphatemic rickets type 2 (ARHR2), while adolescents and adults can develop osteomalacia (softened bones). ARHR2 and osteomalacia lead to pain and mobility issues. Patients can also exhibit signs and symptoms of hearing loss, arterial and joint calcification, and cardiovascular complications. There are no approved therapies for ENPP1 Deficiency.

**About ABCC6 Deficiency**

ABCC6 Deficiency is a rare, severe, inherited disorder caused by mutations in the ABCC6 gene, leading to low levels of PPi. PPi is essential for preventing harmful soft tissue calcification and regulating bone mineralization. ABCC6 Deficiency is a systemic and progressively debilitating condition, which affects more than 67,000 individuals worldwide. Infants with ABCC6 Deficiency are diagnosed with generalized arterial calcification of infancy (GACI) type 2, a condition that resembles GACI type 1, the infant form of ENPP1 Deficiency. In older individuals, ABCC6 Deficiency presents as pseudoxanthoma elasticum (PXE), which is characterized by pathological mineralization in blood vessels and soft tissues clinically affecting the skin, eyes, and vascular system. There are no approved therapies for ABCC6 Deficiency.

**About INZ-701**

INZ-701, a recombinant Fc fusion protein, is an ENPP1 enzyme replacement therapy in development for the treatment of rare disorders of the vasculature, soft tissue, and skeleton. In preclinical studies, the experimental therapy has shown potential to prevent pathologic mineralization and intimal proliferation (the overgrowth of smooth muscle cells inside blood vessels), which can drive morbidity and mortality in devastating genetic disorders such as ENPP1 Deficiency and ABCC6 Deficiency. INZ-701 is currently in clinical trials for the treatment of ENPP1 Deficiency and ABCC6 Deficiency.

**About Inozyme Pharma**

Inozyme Pharma, Inc. is a clinical-stage rare disease biopharmaceutical company developing novel therapeutics for the treatment of diseases impacting the vasculature, soft tissue and skeleton. Inozyme is developing INZ-701, an enzyme replacement therapy, to address pathologic mineralization and intimal proliferation which can drive morbidity and mortality in these severe diseases. INZ-701 is currently in clinical trials for the treatment of ENPP1 Deficiency and ABCC6 Deficiency.

For more information, please visit **www.inozyme.com** and follow us on **LinkedIn**, **X (formerly Twitter)** , and **Facebook**.

**Cautionary Note Regarding Forward-Looking Statements**

Statements in this press release about future expectations, plans, and prospects, as well as any other statements regarding matters that are not historical facts, may constitute "forward-looking statements" within the meaning of The Private Securities Litigation Reform Act of 1995. These statements include, but are not limited to, statements relating to the timing and design of our clinical trials, the potential benefits of INZ-701, the timing and contents of our planned global development strategy, the availability and timing of clinical trial data, planned regulatory filings and the basis for such filings, the timing of the planned commercial launch of INZ-701, if approved, and the period over which we believe that our existing cash, cash equivalents, and short-term investments will be sufficient to fund our cash flow requirements. The words "anticipate," "believe," "continue," "could," "estimate," "expect," "intend," "may," "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. Any forward-looking statements are

based on management's current expectations of future events and are subject to a number of risks and uncertainties that could cause actual results to differ materially and adversely from those set forth in, or implied by, such forward-looking statements. These risks and uncertainties include, but are not limited to, risks associated with the Company's ability to conduct its ongoing clinical trials of INZ-701 for ENPP1 Deficiency and ABCC6 Deficiency; enroll patients in ongoing and planned trials; obtain and maintain necessary approvals from the FDA, EMA, and other regulatory authorities; continue to advance its product candidates in preclinical studies and clinical trials; replicate in later clinical trials positive results found in preclinical studies and early-stage clinical trials of its product candidates; obtain clinically meaningful results with respect to novel endpoints; advance the development of its product candidates under the timelines it anticipates in planned and future clinical trials; obtain, maintain, and protect intellectual property rights related to its product candidates; manage expenses; comply with the covenants under its outstanding loan agreement; and raise the substantial additional capital needed to achieve its business objectives. For a discussion of other risks and uncertainties, and other important factors, any of which could cause the Company's actual results to differ from those contained in the forward-looking statements, see the "Risk Factors" section in the Company's most recent Annual Report on Form 10-K filed with the Securities and Exchange Commission (SEC), as well as discussions of potential risks, uncertainties, and other important factors, in the Company's most recent filings with the SEC. In addition, the forward-looking statements included in this press release represent the Company's views as of the date hereof and should not be relied upon as representing the Company's views as of any date subsequent to the date hereof. The Company anticipates that subsequent events and developments will cause the Company's views to change. However, while the Company may elect to update these forward-looking statements at some point in the future, the Company specifically disclaims any obligation to do so.

**Condensed Consolidated Balance Sheet Data**
**(Unaudited)**

| | June 30, 2023 | December 31, 2022 |
|---|---|---|
| Cash, cash equivalents, and short-term investments | $ 140,247 | $ 127,866 |
| Total assets | $ 150,929 | $ 139,195 |
| Total liabilities | $ 45,336 | $ 20,801 |
| Additional paid-in-capital | $ 353,285 | $ 333,356 |
| Accumulated deficit | $ (247,748) | $ (214,761) |
| Total stockholders' equity | $ 105,593 | $ 118,394 |

**Condensed Consolidated Statements of Operations and Comprehensive Loss**
**(Unaudited)**

| | Three Months Ended June 30, | | Six Months Ended June 30, | |
|---|---|---|---|---|
| | 2023 | 2022 | 2023 | 2022 |
| **Operating expenses:** | | | | |
| Research and development | $ 11,666 | $ 10,007 | $ 23,523 | $ 21,821 |
| General and administrative | 4,728 | 5,384 | 11,240 | 10,409 |
| Total operating expenses | 16,394 | 15,391 | 34,763 | 32,230 |
| Loss from operations | (16,394) | (15,391) | (34,763) | (32,230) |
| Other income (expense): | | | | |
| Interest income, net | 839 | 321 | 1,838 | 381 |
| Other expense, net | (28) | (191) | (62) | (296) |
| Other income, net | 811 | 130 | 1,776 | 85 |
| **Net loss** | $ (15,583) | $ (15,261) | $ (32,987) | $ (32,145) |
| Other comprehensive income (loss): | | | | |
| Unrealized gains (losses) on available-for-sale securities | 76 | (225) | 226 | (357) |
| Foreign currency translation adjustment | 11 | (43) | 30 | (58) |
| Total other comprehensive income (loss) | 87 | (268) | 256 | (415) |
| **Comprehensive loss** | $ (15,496) | $ (15,529) | $ (32,731) | $ (32,560) |
| Net loss attributable to common stockholders—basic and diluted | $ (15,583) | $ (15,261) | $ (32,987) | $ (32,145) |
| Net loss per share attributable to common stockholders—basic and diluted | $ (0.35) | $ (0.38) | $ (0.74) | $ (1.01) |
| Weighted-average common shares and pre-funded warrants outstanding—basic and diluted | 44,860,279 | 39,703,550 | 44,293,577 | 31,739,197 |

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