# Inozyme Pharma Reports First Quarter 2022 Financial Results and Provides Business Highlights

*- Recently reported positive preliminary biomarker and safety data from ongoing Phase 1/2 clinical trial of INZ-701 in ENPP1 Deficiency -*

*- Preliminary biomarker and safety data from ongoing Phase 1/2 trial of INZ-701 in ABCC6 Deficiency on track for the second quarter of 2022 -*

*- Cash, cash equivalents and investments as of quarter end, together with proceeds from April 2022 offering, extends runway into the fourth quarter of 2023 -*

BOSTON, May 10, 2022 (GLOBE NEWSWIRE) -- Inozyme Pharma, Inc. (Nasdaq: INZY), a clinical-stage rare disease biopharmaceutical company developing novel therapeutics for the treatment of abnormal mineralization, today reported financial results for the first quarter ended March 31, 2022 and provided recent business highlights.

"Inozyme is off to an exceptionally strong start for the year. We now have preliminary clinical data in hand supporting INZ-701's potential for therapeutic benefit in patients with ENPP1 Deficiency. We expect topline data for all three cohorts from the ongoing Phase 1/2 trial in the second half of the year," said Axel Bolte, MSc, MBA, Inozyme's co-founder, president, and chief executive officer. "In addition, dosing is now underway in our Phase 1/2 trial in patients with ABCC6 Deficiency, and we remain on track to report preliminary biomarker and safety data later this quarter. We believe our recent follow-on financing positions us well to execute on our mission of delivering therapeutic options to patients with diseases of abnormal mineralization."

**Recent Updates**

- **Phase 1/2 Trial of INZ-701 in Adults with ENPP1 Deficiency.** In April 2022, the Company reported positive preliminary biomarker and safety data from its ongoing Phase 1/2 trial of INZ-701 in patients with ENPP1 Deficiency. Rapid, significant, and sustained increases in plasma pyrophosphate (PPi) levels were observed in all three patients in the trial's lowest dose cohort (0.2 mg/kg), and the range of peak PPi levels observed during the 32-day dose evaluation period across the three patients was comparable to those levels observed in the Company's study of healthy subjects. INZ-701 was generally well tolerated and exhibited a favorable initial safety profile. As recently announced, the second dose escalation cohort (0.6 mg/kg) is now fully enrolled, and dosing is underway. Inozyme plans to report topline data from the ongoing trial in the second half of 2022.
- **Phase 1/2 Trial of INZ-701 in Adults with ABCC6 Deficiency.** Dosing is underway in the Company's Phase 1/2 trial of INZ-701 in patients with ABCC6 Deficiency. The Company is on track to report preliminary biomarker and safety data in the second quarter of 2022.
- **Data Presented at Recent ECTS Congress and ESC FCVB 2022 Meeting.** Multiple presentations were featured at the European Calcified Tissue Society (ECTS) Congress held May 7-10, 2022 in Helsinki and the European Society of Cardiology (ESC) Frontiers in CardioVascular Biomedicine (FCVB) meeting held April 29-May 1, 2022 in Budapest. Copies of the presentations from the ECTS Congress, entitled, "Calciphylaxis is associated with pyrophosphate deficiency in dialysis patients: exploratory study" and "Evaluating the effect of food, fasting and exercise on inorganic pyrophosphate (PPi) levels in healthy subjects," and from the ESC FCVB meeting, entitled, "INZ-701, a recombinant ENPP1-Fc protein, effectively treats and prevents neointimal proliferation in WT and ENPP1 deficient mice," will be available in the Papers & Publications section of the

Inozyme website.

- **ENPP1 Deficiency and ABCC6 Deficiency Natural History Studies.** Patient enrollment is underway in a longitudinal, retrospective natural history study in ENPP1 Deficiency and ABCC6 Deficiency. The study is designed to test and validate findings from the Company's cross-sectional retrospective natural history study. Inozyme expects to commence prospective natural history studies of both populations in the second quarter of 2022.
- **Closed $73 Million Underwritten Offering Priced At-the-Market.** In April 2022, the Company closed an underwritten offering of 16,276,987 shares of its common stock at a price of $3.69 per share and, to certain investors in lieu of common stock, pre-funded warrants to purchase 3,523,013 shares of common stock at a price of $3.6899 per pre-funded warrant. Net proceeds from the offering were approximately $68.3 million, after deducting underwriting discounts and commissions and other offering expenses. Purchasers in the offering included a select group of healthcare focused institutional investors, including new and existing investors.
- **Strengthened Management Team with Two Executive Appointments**. The Company recently announced the appointment of Sanjay Subramanian, MBA, as senior vice president and chief financial officer, and Soojin Kim, Ph.D., as senior vice president and chief technical operations officer.

**First Quarter 2022 Financial Results**

- **Cash Position and Financial Guidance -** Cash, cash equivalents, and investments were $97.8 million as of March 31, 2022. Based on its current plans, the Company expects that its existing cash, cash equivalents, and investment, together with the net proceeds of $68.3 million from its April 2022 offering, will be sufficient to fund its operations into the fourth quarter of 2023.
- **Research and Development (R&D) Expenses** - R&D expenses were $11.8 million for the quarter ended March 31, 2022, compared to $6.6 million for the prior-year period. This increase was primarily due to increased clinical trial costs, increased manufacturing costs, and increased employee-related costs and fees for outsourced services to support the growth of the business.

- **General and Administrative (G&A) Expenses** - G&A expenses were $5.0 million for the quarter ended March 31, 2022, compared to $4.4 million for the prior-year period. The increase was primarily due to an increase in the number of general and administrative employees, consulting expenses, and expenses to support our operations as a public company.

- **Net Loss** - Net loss was $16.9 million, or $0.71 loss per share, for the quarter ended March 31, 2022, compared to $11.1 million, or $0.47 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 neointimal 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 experience rickets, a condition also known as autosomal-recessive hypophosphatemic rickets type 2 (ARHR2), while adults experience osteomalacia (softened bones), and they can exhibit a range of signs and symptoms that include hearing loss, arterial calcification, and cardiac and/or neurological involvement. 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 we believe affects more than 67,000 individuals worldwide. Infants with ABCC6 Deficiency are diagnosed with GACI type 2, a vascular condition that resembles GACI type 1, the acute infantile form of ENPP1 Deficiency. In older patients, 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 cardiovascular system. There are no approved therapies for ABCC6 Deficiency.

**About INZ-701**

INZ-701 is a clinical-stage enzyme replacement therapy in development for the treatment of mineralization disorders of the circulatory system, bones, and kidneys. In preclinical studies, the experimental therapy has shown potential to generate PPi and to restore it to appropriate physiological levels, thereby preventing calcification in the vasculature and kidneys, while at the same time normalizing bone mineralization. Inozyme is developing INZ-701 for certain rare, life-threatening, and devastating genetic disorders such as ENPP1 Deficiency and ABCC6 Deficiency in which PPi levels are below the normal physiological levels. INZ-701 is currently in Phase 1/2 clinical trials for the treatment of ENPP1 Deficiency and ABCC6 Deficiency.

**About Inozyme Pharma**

Inozyme Pharma, Inc. (Nasdaq: INZY) is a clinical-stage rare disease biopharmaceutical company developing novel therapeutics for the treatment of diseases of abnormal mineralization impacting the vasculature, soft tissue, and skeleton. Through our in-depth understanding of the biological pathways involved in mineralization, we are pursuing the development of therapeutics to address the underlying causes of these debilitating diseases. It is well established that two genes, ENPP1 and ABCC6, play key roles in a critical mineralization pathway and that defects in these genes lead to abnormal mineralization. We are initially focused on developing a novel therapy, INZ-701, to treat the rare genetic diseases of ENPP1 and ABCC6 Deficiencies. INZ-701 is currently in Phase 1/2 clinical trials for the treatment of ENPP1 Deficiency and ABCC6 Deficiency.

Inozyme Pharma was founded in 2017 by Joseph Schlessinger, Ph.D., Demetrios Braddock, M.D., Ph.D., and Axel Bolte, MSc, MBA, with technology developed by Dr. Braddock and licensed from Yale University. For more information, please visit www.inozyme.com.

**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 of our clinical trials and other studies, trial design, the availability of data from clinical trials and other studies, the potential benefits of INZ-701 and the sufficiency of the Company's cash resources. 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 Phase 1/2 clinical trials of INZ-701 for ENPP1 Deficiency and ABCC6 Deficiency; obtain and maintain necessary approvals from the FDA 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; 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; 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, 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 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)**

**(in thousands)**

| | March 31, 2022 | December 31, 2021 |
|---|---|---|
| Cash, cash equivalents, and investments | $ 97,773 | $ 111,801 |
| Total assets | $ 108,931 | $ 123,541 |
| Total liabilities | $ 14,702 | $ 14,273 |
| Additional paid-in-capital | $ 258,940 | $ 256,948 |
| Accumulated deficit | $ (164,584) | $ (147,700) |
| Total stockholders' equity | $ 94,229 | $ 109,268 |

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

**(in thousands, except share and per share data)**

| | Three Months Ended March 31, | |
|---|---|---|
| | 2022 | 2021 |
| **Operating expenses:** | | |
| Research and development | $11,814 | $6,603 |
| General and administrative | 5,025 | 4,369 |
| Total operating expenses | 16,839 | 10,972 |
| Loss from operations | (16,839) | (10,972) |
| Other income (expense): | - | - |
| Interest income | 60 | 63 |
| Other expenses | (105) | (141) |
| Other expenses, net | (45) | (78) |
| **Net loss** | $(16,884) | $(11,050) |
| Other comprehensive (loss) income: | | |
| Unrealized (losses) gains on available-for-sale securities | (132) | 10 |
| Foreign currency translation adjustment | (15) | - |
| Total other comprehensive (loss) income | (147) | 10 |
| **Comprehensive loss** | $(17,031) | $(11,040) |
| Net loss attributable to common stockholders-basic and diluted | $(16,884) | $(11,050) |
| Net loss per share attributable to common stockholders-basic and diluted | $(0.71) | $(0.47) |
| Weighted-average common shares outstanding-basic and diluted | 23,686,351 | 23,429,507 |

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