Inozyme Pharma Reports Second Quarter 2022 Financial Results and Provides Business Updates

August 15, 2022

- Recently reported positive preliminary biomarker and safety data from ongoing Phase 1/2 clinical trials of INZ-701 in ENPP1 Deficiency and ABCC6 Deficiency –

- Appointed Kurt Gunter, M.D. as senior vice president and chief medical officer -

- Secured flexible debt facility for up to $70 million -

- Cash, cash equivalents and investments as of quarter end, together with first tranche of debt facility, funds cash flow requirements into the second quarter of 2024 –

BOSTON, Aug. 15, 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 second quarter ended June 30, 2022 and provided recent business highlights.

"I am pleased with the increasing momentum in our clinical development and the strength of our balance sheet to deliver important data milestones across our clinical trials in patients with ENPP1 Deficiency and ABCC6 Deficiency," said Axel Bolte, MSc, MBA, Inozyme's co-founder, president and chief executive officer. "We recently reported positive preliminary data in our Phase 1/2 trial of INZ-701 in patients with ABCC6 Deficiency, providing the first evidence that INZ-701 increased PPi levels in subjects with functional ENPP1 enzyme, which we believe potentially opens the possibility to expand to additional indications that are characterized by low levels of PPi. We look forward to presenting data from the higher dose cohorts in the Phase 1/2 trial of INZ-701 in ABCC6 Deficiency."

Recent Clinical Trial Updates

- **Phase 1/2 Clinical Trial of INZ-701 in Adults with ABCC6 Deficiency (pseudoxanthoma elasticum or PXE).** In July 2022, the Company reported positive preliminary biomarker, safety and pharmacokinetic data from its ongoing Phase 1/2 trial of INZ-701 in patients with ABCC6 Deficiency. Dosing is underway in the second dose escalation cohort (0.6 mg/kg). The Company plans to report topline data from the ongoing trial in the first quarter of 2023.

- **Phase 1/2 Clinical Trial of INZ-701 in Adults with ENPP1 Deficiency.** The 32-day dose evaluation period is complete for the second dose escalation cohort (0.6 mg/kg), and following review of the preliminary data, an independent Data Safety Monitoring Board (DSMB) recommended the trial continue as planned to the highest dose cohort (1.8 mg/kg). The Company plans to report topline data from the ongoing trial in the fourth quarter of 2022. The Company is also actively engaged in designing and planning a clinical trial of INZ-701 in infants and adolescents with ENPP1 Deficiency.

- **Prospective Natural History Study.** Patient enrollment is underway in a prospective natural history study in ENPP1 Deficiency and ABCC6 Deficiency. The study is designed to test and validate findings from the Company's previously published cross-sectional retrospective natural history study. Patient enrollment is also underway in a longitudinal, retrospective natural history study in ENPP1 Deficiency and ABCC6 Deficiency.

- **Planned Clinical Trial of INZ-701 in Calciphylaxis.** In May 2022, the Company presented data at the European Calcified Tissue Society Congress (ECTS) which showed that morbidity and mortality in patients with calciphylaxis were associated with low levels of PPi. Based on these data, the Company plans to finalize the regulatory pathway in the fourth quarter of 2022 to initiate a clinical trial of INZ-701 in calciphylaxis.

Recent Corporate Updates

- **Scientific Publications.** Peer-reviewed article on Burden of Illness study in *PLOS ONE* showed that individuals with ENPP1 Deficiency or infant onset ABCC6 Deficiency experience lifelong morbidity causing substantial physical and emotional burden to patients and caregivers. Peer-reviewed article on preclinical data in *Experimental Dermatology* showed that INZ-701 increased PPi and prevented skin calcification and therefore might provide therapeutic benefit in ABCC6 Deficiency.

- **Collaboration with Rady Children's Institute for Genomic Medicine (RCIGM).** Inozyme is a founding member of the Public-Private BeginNGS™ Consortium established by RCIGM to advance and evaluate a novel newborn screening technology to facilitate the diagnosis of genetic diseases. The collaboration focuses on a diagnostic and precision medicine guidance tool called BeginNGS™, which incorporates rapid Whole Genome Sequencing (rWGS®) to currently screen newborns for approximately 400 genetic diseases, including generalized arterial calcification of infancy (GACI), the infant

form of ENPP1 Deficiency. RCIGM is conducting a pilot evaluation that aims to supplement existing newborn screening protocols at birthing hospitals in the United States, with the ultimate goal for BeginNGS to test ~1,000 disorders and sequence 3.7 million newborns annually.

- **Flexible Debt Facility for up to $70 million.** In July 2022, the Company entered into a debt financing facility for up to $70 million with K2 HealthVentures (K2HV). Under the terms of the agreement, Inozyme drew down $5 million from the facility upon execution, and has an option to draw down an additional $20 million from the first tranche through March 31, 2023. Subsequently, an additional $20 million will be available in two tranches, subject to the achievement of certain time-based, clinical and regulatory milestones, and the final $25 million will be available to be drawn down through August 31, 2025, subject to use of proceeds limitations and lender approval. The Company is under no obligation to draw funds in the future.

- **Appointment of Kurt Gunter, M.D., as senior vice president and chief medical officer.** Dr. Gunter, who most recently served as chief medical officer, cell therapy, and head of regulatory affairs at Athenex, brings to Inozyme over 30 years of expertise in regulatory affairs and clinical development.

Second Quarter 2022 Financial Results

- **Cash Position and Financial Guidance –** Cash, cash equivalents, and investments were $151.5 million as of June 30, 2022. Based on its current plans, the Company expects that its cash, cash equivalents, and investments as of June 30, 2022, together with the $25 million available under the first tranche of its debt facility, will enable the Company to fund its cash flow requirements into the second quarter of 2024.

- **Research and Development (R&D) Expenses** – R&D expenses were $10.0 million for the quarter ended June 30, 2022, compared to $8.2 million for the prior-year period. This increase was primarily due to increased clinical trial costs and fees for outsourced services to support the growth of the business.

- **General and Administrative (G&A) Expenses** – G&A expenses were $5.4 million for the quarter ended June 30, 2022, compared to $4.4 million for the prior-year period. The increase was primarily due to an increase in personnel costs.

- **Net Loss** – Net loss was $15.3 million, or $0.38 loss per share, for the quarter ended June 30, 2022, compared to $12.5 million, or $0.53 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 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 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 ongoing and planned clinical trials and other studies, the availability of data from clinical trials, the potential benefits of INZ-701, the impact of the debt facility on the Company's balance sheet 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 and Quarterly Report on Form 10-Q 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)

	June 30, 2022	December 31, 2021
Cash, cash equivalents and investments	$ 151,480	$ 111,801
Total assets	$ 161,989	$ 123,541
Total liabilities	$ 12,813	$ 14,273
Additional paid-in-capital	$ 329,414	$ 256,948
Accumulated deficit	$ (179,845)	$ (147,700)
Total stockholders' equity	$ 149,176	$ 109,268

Condensed Consolidated Statements of Operations and Comprehensive Loss
(Unaudited)

(in thousands, except share and per share data)

	Three Months Ended June 30,	
	2022	2021
Operating expenses:		
Research and development	$ 10,007	$ 8,220
General and administrative	5,384	4,435
Total operating expenses	15,391	12,655
Loss from operations	(15,391)	(12,655)
Other income (expense):		
Interest income	321	58
Other (expenses) income	(191)	57
Other income, net	130	115
Net loss	$ (15,261)	$ (12,540)
Other comprehensive (loss) income:		
Unrealized (losses) gains on available-for-sale securities	(225)	6

Foreign currency translation adjustment	(43)	—
Total other comprehensive (loss) income	(268)	6
Comprehensive loss	$ (15,529)	$ (12,534)
Net loss attributable to common stockholders—basic and diluted	$ (15,261)	$ (12,540)
Net loss per share attributable to common stockholders—basic and diluted	$ (0.38)	$ (0.53)
Weighted-average common shares and pre-funded warrants outstanding—basic and diluted	39,703,550	23,490,591

	Six Months Ended June 30,	
	2022	**2021**
Operating expenses:		
Research and development	$ 21,821	$ 14,823
General and administrative	10,409	8,804
Total operating expenses	32,230	23,627
Loss from operations	(32,230)	(23,627)
Other income (expense):		
Interest income	381	121
Other expense	(296)	(84)
Other income, net	85	37
Net loss	$ (32,145)	$ (23,590)
Other comprehensive (loss) income:		
Unrealized (losses) gains on available-for-sale securities	(357)	16
Foreign currency translation adjustment	(58)	—
Total other comprehensive (loss) income	(415)	16
Comprehensive loss	$ (32,560)	$ (23,574)
Net loss attributable to common stockholders—basic and diluted	$ (32,145)	$ (23,590)
Net loss per share attributable to common stockholders—basic and diluted	$ (1.01)	$ (1.01)
Weighted-average common shares and pre-funded warrants outstanding—basic and diluted	31,739,197	23,460,218

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