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

- Topline data from Phase 1/2 clinical trial of INZ-701 in ENPP1 Deficiency on track for the fourth quarter of 2022 -

- Topline data from Phase 1/2 clinical trial of INZ-701 in ABCC6 Deficiency on track for the first quarter of 2023 -

- Liquidity and capital resources as of quarter end funds cash flow requirements into the second quarter of 2024 -

BOSTON, Nov. 10, 2022 (GLOBE NEWSWIRE) -- Inozyme Pharma, Inc. (Nasdaq: INZY), 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 third quarter ended September 30, 2022 and provided recent business highlights.

"Progress continues with our ongoing Phase 1/2 clinical trials of INZ-701 in patients with ENPP1 Deficiency and ABCC6 Deficiency, with dosing now underway in the third dose cohort of both trials. We recently introduced self-administration of INZ-701 in the Phase 2 extension portion of the ongoing ENPP1 Deficiency trial, improving patient convenience. In parallel, we remain focused on initiating a clinical trial in pediatric patients with ENPP1 Deficiency," said Axel Bolte, MSc, MBA, Inozyme's co-founder, president and chief executive officer. "Our partnerships with leading disease experts and research organizations continue to yield new insights into ENPP1 Deficiency. Recent data published in a peer-reviewed journal reported a threefold increase in characterized disease-causing *ENPP1* variants and identified symptomatic patients with heterozygous mutations which emphasizes the urgent need for therapeutic options in this severe disease."

Recent Clinical Trial Updates

- **Phase 1/2 Clinical Trial of INZ-701 in Adults with ENPP1 Deficiency.** Dosing is underway in the third dose escalation cohort (1.8 mg/kg), and the Company is on track to report topline data from the ongoing trial in the fourth quarter of 2022. The Company recently announced the first self-administration of INZ-701 in the Phase 2 extension portion of this trial. The Company is also actively engaged in designing and planning a clinical trial of INZ-701 in pediatric patients with ENPP1 Deficiency.
- **Phase 1/2 Clinical Trial of INZ-701 in Adults with ABCC6 Deficiency (pseudoxanthoma elasticum or PXE).** Dosing is underway in the third dose escalation cohort (1.8 mg/kg). The Company is on track to report topline data from the ongoing trial in the first quarter of 2023.
- **Natural History Studies in ENPP1 Deficiency and ABCC6 Deficiency.** Patient enrollment continues in the Company's prospective natural history study and longitudinal retrospective natural history study in ENPP1 Deficiency and ABCC6 Deficiency. These studies are designed to test and validate findings from Inozyme's previously published cross-sectional retrospective natural history study.

Additional Recent Updates

- **ENPP1 Variant Database Publication.** Peer-reviewed article in *Human Mutation* titled "ENPP1 Deficiency: A clinical update on the relevance of individual variants using a locus-specific database" reported 3-fold increase in pathogenic/likely pathogenic *ENPP1* variants. Analysis also identified severe phenotypes in patients with monoallelic heterozygous *ENPP1* variants.
- **Data Presented at ASBMR 2022 Annual Meeting.** Two preclinical presentations supporting INZ-701's potential application in indications marked by pathologic mineralization were featured at the American Society for Bone and

Mineral Research (ASBMR) 2022 Annual Meeting in September. Copies of the presentations titled, "Treatment with ENPP1-Fc reduces vascular calcifications in a CKD rat model" and "ENPP1-Fc expressing AAV vector prevents ectopic tissue calcification and restores bone parameters in ENPP1 deficient mice", are available in the Investor Relations section of the Inozyme website.

- **Data Presented at IVBM 2022 Annual Meeting.** Preclinical data supporting INZ-701's potential to treat intimal proliferation was featured at the International Vascular Biology Meeting (IVBM) 2022 Annual Meeting in October. A copy of the presentation titled, "ENPP1-Fc inhibits proliferation of pathological synthetic phenotype vascular smooth muscle cells (VSMCs) in the presence of ATP: the role of ecto-5'-nucleotidase CD73", is available in the Investor Relations section of the Inozyme website.

Third Quarter 2022 Financial Results

- **Cash Position and Financial Guidance -** Cash, cash equivalents, and investments were $141.5 million as of September 30, 2022. Based on its current plans, the Company expects that its cash, cash equivalents, and investments together with the remainder of the first tranche of its debt facility as of September 30, 2022 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 $12.2 million for the quarter ended September 30, 2022, compared to $9.3 million for the prior-year period. This increase was primarily due to the progression of the clinical trials of INZ-701 for ENPP1 Deficiency and ABCC6 Deficiency, manufacturing operations, and consultants to support the ongoing trials.

- **General and Administrative (G&A) Expenses** - G&A expenses were $4.7 million for the quarter ended September 30, 2022, compared to $4.9 million for the prior-year period. The decrease was primarily due to a decrease in professional services and other administrative expenses partially offset by an increase in personnel costs.

- **Net Loss** - Net loss was $16.4 million, or $0.38 loss per share, for the quarter ended September 30, 2022, compared to $14.3 million, or $0.60 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 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, which can drive morbidity and mortality in devastating genetic disorders such as ENPP1 Deficiency and ABCC6 Deficiency. 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 impacting the vasculature, soft tissue, and skeleton. We are developing INZ-701, a potential first-in-class enzyme therapy, to address pathologic mineralization and intimal proliferation which can drive morbidity and mortality in these severe diseases. INZ-701 is currently in Phase 1/2 clinical trials for the treatment of ENPP1 Deficiency and ABCC6 Deficiency.

For more information, please visit www.inozyme.com and follow us on LinkedIn, 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 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)

	September 30, 2022	December 31, 2021
Cash, cash equivalents and investments	$141,452	$111,801
Total assets	$154,021	$123,541
Total liabilities	$19,241	$14,273
Additional paid-in-capital	$331,470	$256,948
Accumulated deficit	$(196,217)	$(147,700)
Total stockholders' equity	$134,780	$109,268

Condensed Consolidated Statements of Operations and Comprehensive Loss
(Unaudited)

(in thousands, except share and per share data)

	Three Months Ended September 30,	
	2022	2021
Operating expenses:		
Research and development	$12,191	$9,346
General and administrative	4,721	4,916
Total operating expenses	16,912	14,262
Loss from operations	(16,912)	(14,262)
Other income (expense):		
Interest income	737	47
Other expenses	(197)	(65)
Other income, net	540	(18)
Net loss	$(16,372)	$(14,280)
Other comprehensive (loss) income:		
Unrealized gains (losses) on available-for-sale securities	(60)	(6)
Foreign currency translation adjustment	(20)	(9)
Total other comprehensive (loss) income	(80)	(15)
Comprehensive loss	$(16,452)	$(14,295)
Net loss attributable to common stockholders-basic and diluted	$(16,372)	$(14,280)
Net loss per share attributable to common stockholders-basic and diluted	$(0.38)	$(0.60)
Weighted-average common shares outstanding-basic and diluted	43,657,718	23,643,494

	Nine Months Ended September 30,	

	2022	2021
Operating expenses:		
Research and development	$34,012	$24,169
General and administrative	15,130	13,720
Total operating expenses	49,142	37,889
Loss from operations	(49,142)	(37,889)
Other income (expense):		
Interest income	1,118	168
Other expenses	(493)	(149)
Other income, net	625	19
Net loss	$(48,517)	$(37,870)
Other comprehensive (loss) income:		
Unrealized gains (losses) on available-for-sale securities	(417)	10
Foreign currency translation adjustment	(78)	(9)
Total other comprehensive (loss) income	(495)	1
Comprehensive loss	$(49,012)	$(37,869)
Net loss attributable to common stockholders-basic and diluted	$(48,517)	$(37,870)
Net loss per share attributable to common stockholders-basic and diluted	$(1.36)	$(1.61)
Weighted-average common shares outstanding-basic and diluted	35,755,695	23,521,981

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