Inozyme Pharma Reports Full Year 2021 Financial Results and Provides Business Highlights

March 15, 2022

– Phase 1/2 trial of INZ-701 in adult patients with ENPP1 Deficiency underway –

– Expect to enroll first patient in Phase 1/2 trial of INZ-701 for ABCC6 Deficiency in the second quarter of 2022 –

– Preliminary safety and biomarker data from Phase 1/2 trials in ENPP1 Deficiency and ABCC6 Deficiency on track for the first half of 2022 –

BOSTON, March 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 full year ended December 31, 2021, and provided recent business highlights.

“2021 was a transformational year for Inozyme, as we continued to progress our INZ-701 program and graduated to a clinical-stage company. We initiated our first clinical trial for patients with ENPP1 Deficiency and are screening patients for our clinical trial in ABCC6 Deficiency, and have continued to deepen our understanding of the pathophysiology of these rare and devastating mineralization disorders,” said Axel Bolte, MSc, MBA, Inozyme’s co-founder, president, and chief executive officer. “We firmly believe that INZ-701 could be a meaningful intervention in patients with ENPP1 Deficiency and ABCC6 Deficiency, and we are committed to realizing the full potential of this promising therapy.”

Recent Updates

- **Phase 1/2 Trial of INZ-701 in Adults with ENPP1 Deficiency.** Patient dosing is underway in the Company’s Phase 1/2 trial of INZ-701 in patients with ENPP1 Deficiency. The Company remains on track to report preliminary biomarker and safety data in the first half of 2022.
- **Phase 1/2 Trial of INZ-701 in Adults with ABCC6 Deficiency.** Patient screening is underway in the Company’s Phase 1/2 trial of INZ-701 in patients with ABCC6 Deficiency. The Company expects to enroll its first patient in the Phase 1/2 trial in the second quarter of 2022. The Company remains on track to report preliminary biomarker and safety data in the first half of 2022.
- **First Patient Enrolled in Longitudinal Retrospective Natural History Study in ENPP1 Deficiency and ABCC6 Deficiency.** This study is designed to test and validate findings from the Company’s Cross-sectional Retrospective Natural History Study.
- **Strengthened Management Team with Two Executive Appointments**. The Company recently announced the appointment of Sanjay S. Subramanian, MBA, as chief financial officer, effective as of March 21, 2022, and Soojin Kim, Ph.D., as senior vice president and chief technical operations officer.
- **Presented Data on ENPP1 Program.** At the American Society for Bone and Mineral Research (ASBMR) 2021 Annual Meeting in October 2021, the Company presented data from its ENPP1 Deficiency Natural History Study and gene therapy program supporting the urgent need for novel interventions for ENPP1 Deficiency.

Upcoming Anticipated Milestones

The Company also announced the following anticipated milestones for the INZ-701 clinical development program, subject to COVID-19-related restrictions:

- **ENPP1 Deficiency**
  - H1 2022: Report preliminary safety and biomarker data from Phase 1/2 clinical trial
  - Q2 2022: Initiate prospective natural history study
- **ABCC6 Deficiency**
  - Q2 2022: First patient dosing in Phase 1/2 clinical trial
  - H1 2022: Report preliminary safety and biomarker data from Phase 1/2 clinical trial
  - Q2 2022: Initiate prospective natural history study

Financial Result for the Year Ended December 31, 2021

- **Cash Position and Financial Guidance –** Cash, cash equivalents, and investments were $111.8 million as of December 31, 2021. Based on its current plans, the Company expects that its existing cash, cash equivalents, and investment will be sufficient to fund its operating expenses and capital expenditures into the first quarter of 2023.
- **Research and Development (R&D) Expenses** – R&D expenses were $37.7 million for the year ended December 31,

2021, compared to $46.5 million for the year ended December 31, 2020. This decrease was primarily due to the non-recurring, non-cash purchase of in-process research and development intellectual property assets from Alexion Pharmaceuticals, Inc. in exchange for the Company's stock in in July 2020, as well as decreases in research and development expenses such as toxicology studies and decreases in manufacturing operations based on the timing of production runs, partially offset by increases in clinical trials costs and increased employee-related costs to support the growth of the business.

- **General and Administrative (G&A) Expenses** – G&A expenses were $18.9 million for the year ended December 31, 2021, compared to $10.5 million for the year ended December 31, 2020. The increase was primarily due to the growth in the number of G&A employees, an increase in legal fees and generally higher fees in areas such as audit, tax, and information technology to support the Company's growth and operations as a public company.

- **Net Loss** – Net loss was $56.6 million, or $2.40 loss per share, for the year ended December 31, 2021, compared to $56.4 million, or $5.11 loss per share, for the year ended December 31, 2020.

About ENPP1 Deficiency

ENPP1 Deficiency is a progressive condition that manifests as a spectrum of diseases. Those who present in utero or 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 45% to 50% of infants with ENPP1 Deficiency die within 6 months of birth. Children and adults with ENPP1 Deficiency typically experience rickets and osteomalacia (softened bones), also termed autosomal-recessive hypophosphatemic rickets type 2 (ARHR2), and can exhibit a range of signs and symptoms that can include hearing loss, arterial calcification, cardiac, and 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. The condition is characterized by pathological mineralization in blood vessels and soft tissues throughout the body that can drive devastating medical problems. Some infants with ABCC6 Deficiency are diagnosed with generalized arterial calcification of infancy (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), a rare, inherited disorder in which individuals develop calcification of soft connective tissues, including in the eyes, cardiovascular system, and skin. There are no approved therapies for 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 a Phase 1/2 clinical trial for ENPP1 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 initiation and timing of our clinical trials, trial design, the availability of clinical trial data and the potential benefits of INZ-701. 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 initiate and conduct its ongoing and planned 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)

	December 31, 2021	December 31, 2020
Cash, cash equivalents and investments	$ 111,801	$ 159,896
Total assets	123,541	169,363
Total liabilities	14,273	11,260
Additional paid-in-capital	256,948	249,175
Accumulated deficit	(147,700)	(91,076)
Total stockholders' equity	109,268	158,103

Condensed Consolidated Statements of Operations and Comprehensive Loss
(Unaudited)

(in thousands, except share and per share data)

	Year Ended December 31,	
	2021	**2020**
Operating expenses:		
Research and development	$ 37,720	$ 46,493
General and administrative	18,926	10,548
Total operating expenses	56,646	57,041
Loss from operations	(56,646)	(57,041)
Other income:		
Interest income	211	370
Other income (expense), net	(189)	247
Other income, net	22	617
Net loss	$ (56,624)	$ (56,424)
Other comprehensive (loss) income:		
Unrealized losses on available-for-sale securities	(4)	(3)
Foreign currency translation adjustment	20	—
Total other comprehensive income (loss)	16	(3)
Comprehensive loss	$ (56,608)	$ (56,427)
Net loss attributable to common stockholders—basic and diluted	$ (56,624)	$ (56,424)
Net loss per share attributable to common stockholders—basic and diluted	$ (2.40)	$ (5.11)
Weighted-average common shares outstanding—basic and diluted	23,558,306	11,036,500

Contacts

Investors:
Inozyme Pharma
Stefan Riley, Director of Investor Relations
stefan.riley@inozyme.com

Media:
SmithSolve
Matt Pera
(973) 886-9150
matt.pera@smithsolve.com