Inozyme Pharma Reports Q2 2021 Financial Results and Provides Business Highlights

August 11, 2021

- Received Orphan Drug Designation from the European Medicines Agency for INZ-701 for the treatment of ABCC6 Deficiency –

– Clinical Trial Application in Europe accepted for Phase 1/2 trial of INZ-701 in ABCC6 Deficiency –

– Investigational New Drug application in U.S. accepted for Phase 1/2 trial of INZ-701 in ABCC6 Deficiency –

- Expect to enroll patients in Phase 1/2 clinical trials in ENPP1 Deficiency and ABCC6 Deficiency in Q4 2021 and report preliminary biomarker and safety data in the first half of 2022 –

- Cash, cash equivalents, and investments expected to support continued operations into the fourth quarter of 2022 –

BOSTON, Aug. 11, 2021 (GLOBE NEWSWIRE) -- Inozyme Pharma, Inc. (Nasdaq: INZY), a rare disease biopharmaceutical company developing novel therapeutics for the treatment of abnormal mineralization, today reported financial results for the second quarter ended June 30, 2021, and provided recent business highlights.

"We achieved several important milestones during the second quarter of 2021. We expect to initiate our clinical trials in ENPP1 Deficiency and ABCC6 Deficiency in the fourth quarter of this year and then to report preliminary biomarker and safety data in the first half of 2022," said Axel Bolte, MSc, MBA, Inozyme's co-founder, president, and chief executive officer. "We continue to make progress towards our mission of bringing hope to patients and their families who are living with ENPP1 Deficiency and ABCC6 Deficiency. In addition, we added several talented members to our leadership team who will contribute to this next phase of our growth."

Recent Business Highlights

- **Clinical Trial Application (CTA) in Europe and Investigational New Drug (IND) application in U.S. for ABCC6 Deficiency accepted –** The Company expects to enroll patients in a Phase 1/2 clinical trial in the fourth quarter of 2021 and report preliminary biomarker and safety data in the first half of 2022.
- **Published data from Natural History Study of ENPP1 and ABCC6 Deficiencies –** Peer-reviewed article in the *Journal of Bone and Mineral Research* shows early mortality risk in GACI patients despite attempts to treat with bisphosphonates, high prevalence of rickets almost exclusive to ENPP1 Deficiency, and a spectrum of heterogenous calcification and multiple organ complications with both *ENPP1* and *ABCC6* variants, which suggests an overlapping pathology.
- **Appointed Gayle Gironda as senior vice president, human resources –** Ms. Gironda is a human resources leader with more than 20 years of experience in organizational design, talent recruitment, performance culture, planning and leadership development.
- **Appointed David Thompson, M.A., M.S., Ph.D. as chief development officer –** Dr. Thompson has more than 30 years of experience designing and leading research and development programs focused on bone disorders and phosphate regulation. He previously served as Inozyme's senior vice president and chief scientific officer from 2018 to 2020 and was responsible for scientific research as the Company built its proprietary pipeline of investigational therapies, beginning with INZ-701.

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**
  - **Q4 2021:** Start enrollment for Phase 1/2 clinical trial
  - **Q1 2022:** Initiate prospective natural history study
  - **H1 2022:** Report preliminary safety and biomarker data from Phase 1/2 clinical trial
- **ABCC6 Deficiency**
  - **Q4 2021:** Start enrollment for Phase 1/2 clinical trial
  - **H1 2022:** Report preliminary safety and biomarker data from Phase 1/2 clinical trial

Financial Results for the Quarter Ended Jun 30, 2021

- **Cash Position and Financial Guidance –** Cash, cash equivalents, and investments were $137.5 million as of June 30, 2021. Based on its current plans, the Company expects that its existing cash, cash equivalents, and investments will be sufficient to enable funding of its operating expenses and capital expenditure requirements into the fourth quarter of 2022.

- **Research and Development (R&D) Expenses –** R&D expenses were $8.2 million for the quarter ended June 30, 2021, compared to $7.9 million for the quarter ended June 30, 2020. The increase was primarily due to increased salaries, employee-related costs, and stock-based compensation expense due to the growth in the number of R&D employees. These increases were partially offset by a decrease in preclinical toxicology studies in support of our IND filing for INZ-701, and lower manufacturing costs based on the timing of production runs.
- **General and Administrative (G&A) Expenses –** G&A expenses were $4.4 million for the quarter ended June 30, 2021, compared to $1.7 million for the quarter ended June 30, 2020. The increase was primarily due to the growth in the number of G&A employees, an increase in legal fees related to new contracts and operations as a public company and generally higher fees in areas such as audit, tax, and information technology to support the Company's growth.
- **Net Loss –** Net loss was $12.5 million, or $0.53 loss per share, for the quarter ended June 30, 2021, compared to $9.5 million, or $7.57 loss per share, for the quarter ended June 30, 2020.

About Inozyme Pharma

Inozyme Pharma, Inc. (Nasdaq: INZY), is a 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 to treat the rare genetic diseases of ENPP1 and ABCC6 Deficiencies.

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, the initiation and timing of our natural history study, our research and development programs, the availability of preclinical study and clinical trial data, the timing of our regulatory applications and the period over which we believe that our existing cash, cash equivalents and investments will be sufficient to fund our operating expenses. 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 its 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)

	June 30, 2021	December 31, 2020
Cash, cash equivalents and investments	$ 137,464	$ 159,896
Total assets	148,259	169,363
Total liabilities	9,985	11,260
Additional paid-in-capital	252,920	249,175
Accumulated deficit	(114,666)	(91,076)
Total stockholders' equity	138,274	158,103

Condensed Consolidated Statements of Operations and Comprehensive Loss
(Unaudited)

(in thousands, except share and per share data)

	Three Months Ended June 30,	
	2021	2020

Operating expenses:		
Research and development	$ 8,220	$ 7,877
General and administrative	4,435	1,671
Total operating expenses	12,655	9,548
Loss from operations	(12,655)	(9,548)
Other income (expense):		
Interest income	58	71
Other income (expense)	57	4
Other income (expense), net	115	75
Net loss	$ (12,540)	$ (9,473)
Other comprehensive income (loss):		
Unrealized gains (losses) on available-for-sale securities	6	(15)
Total other comprehensive income (loss)	6	(15)
Comprehensive loss	$ (12,534)	$ (9,488)
Net loss attributable to common stockholders—basic and diluted	$ (12,540)	$ (9,473)
Net loss per share attributable to common stockholders—basic and diluted	$ (0.53)	$ (7.57)
Weighted-average common shares outstanding—basic and diluted	23,490,591	1,251,244

	Six Months Ended June 30,	
	2021	**2020**
Operating expenses:		
Research and development	$ 14,823	$ 14,283
General and administrative	8,804	3,171
Total operating expenses	23,627	17,454
Loss from operations	(23,627)	(17,454)
Other income (expense):		
Interest income	121	242
Other income (expense)	(84)	1
Other income (expense), net	37	243
Net loss	$ (23,590)	$ (17,211)
Other comprehensive income (loss):		
Unrealized gains (losses) on available-for-sale securities	16	8
Total other comprehensive income (loss)	16	8
Comprehensive loss	$ (23,574)	$ (17,203)
Net loss attributable to common stockholders—basic and diluted	$ (23,590)	$ (17,211)
Net loss per share attributable to common stockholders—basic and diluted	$ (1.01)	$ (14.01)
Weighted-average common shares outstanding—basic and diluted	23,460,218	1,228,296

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