Inozyme Pharma Reports Second Quarter 2020 Financial Results and Provides Business Highlights

Upsized IPO in July 2020 raised $128.8 million in gross proceeds

Submitted IND for INZ-701 for the treatment of ENPP1 deficiency; currently on FDA clinical hold pending completion of ongoing GLP toxicology studies

Initiation of INZ-701 Phase 1/2 clinical trials anticipated in early 2021, as previously planned

BOSTON, September 3, 2020 (GLOBE NEWSWIRE) – Inozyme Pharma, Inc. (Nasdaq: INZY), a rare disease biopharmaceutical company developing novel therapeutics for the treatment of diseases of abnormal mineralization impacting the vasculature, soft tissue and skeleton, today reported financial results for the second quarter ended June 30, 2020 and provided recent business highlights.

"We have made substantial progress over the past quarter and in July, including completing our upsized initial public offering in July, acquiring additional ENPP1 deficiency program assets, and submitting our first investigational new drug application (IND) for INZ-701 for the treatment of ENPP1 deficiency," said Axel Bolte, MSc, MBA, co-founder, president and chief executive officer of Inozyme Pharma. "FDA has requested final study reports for our ongoing three-month GLP toxicology studies in mice and non-human primates and has placed our planned Phase 1/2 clinical trial of INZ-701 in adult patients with ENPP1 deficiency in the United States on clinical hold pending submission of the reports. We expect to be able to submit these reports in the fourth quarter of this year and initiate our clinical program, as we had planned, in early 2021."

Recent Business Highlights

- **Completed upsized Initial Public Offering** – In July 2020, Inozyme completed its initial public offering of 8,050,000 shares of common stock at a public offering price of $16.00 per share, including the full exercise of the underwriter's option to purchase additional shares. Gross proceeds from the IPO were $128.8 million and net proceeds from the offering, after deducting underwriting discounts, commissions and offering expenses, were approximately $116.5 million.

- **Submitted an IND for INZ-701 -** On July 30, 2020, Inozyme submitted its first IND for INZ-701 for the treatment of ENPP1 deficiency to the U.S. Food and Drug Administration (FDA). At the end of the 30-day FDA review period, the Company was notified that the IND was placed on clinical hold, pending submission of the final study report for its ongoing three-month toxicology studies in mice and non-human primates (NHPs) being performed in accordance with Good Laboratory Practices (GLP) regulations. Inozyme initiated these GLP toxicology studies prior to submission of the IND and expects to complete the studies and have reports available in the fourth quarter of 2020. The FDA did not request any additional preclinical data to resolve the clinical hold other than the submission of the final study report for the three-month GLP toxicology studies in mice and NHPs. Subject to the submission of the final study reports for the

three-month toxicology studies as recommended by the FDA and the successful resolution of the clinical hold, the Company expects to initiate its Phase 1/2 trials in early 2021 and report initial safety and biomarker data in 2021, as it had planned.

- **Expanded and strengthened Board of Directors with appointment of Doug Treco, Ph.D., as Chairman and Lynne Sullivan, MST as an Independent Director –** In May 2020, Doug Treco joined Inozyme's Board as chairman and Lynne Sullivan joined the Board as an independent director. Dr. Treco co-founded Ra Pharmaceuticals, Inc., where he was chief executive officer and a member of the Board of Directors from its inception until the company was acquired in April 2020.

  Ms. Sullivan is currently the chief financial officer for UNITY Biotechnology, and she previously served as the chief financial officer of Compass Therapeutics and as a senior vice president of finance at Biogen, where she spent 11 years, with global responsibility for Corporate Finance, Financial Planning and Analysis and Corporate Tax.

- **Acquired ENPP1 program assets from Alexion Pharmaceuticals –** In July 2020, Inozyme announced the acquisition of intellectual property and assets from Alexion Pharmaceuticals focusing on ENPP1 gene deficiencies. The acquisition complements Inozyme's ongoing development of INZ-701 and expands the Company's intellectual property portfolio.

- **Initiated disease burden study in ENPP1 deficiency and ABCC6 deficiency –** In May 2020, Inozyme and GACI Global, a patient advocacy organization dedicated to bettering the lives of families affected by Generalized Arterial Calcification of Infancy and/or Autosomal Recessive Hypophosphatemic Rickets Type 2 (GACI/ARHR2), announced the initiation of a study to characterize the burden of disease and understand the systemic progression of disease for the rare genetic diseases of both ENPP1 deficiency and ABCC6 deficiency from the perspective of a patient and/or parent.

- **Expanded physical footprint with move to new office** – To meet the demands of anticipated growth, Inozyme expanded and enhanced its physical office space in August 2020.

Second Quarter 2020 Financial Results

- **Cash Position** – Cash, cash equivalents and short-term investments were $63.9 million as of June 30, 2020, as compared to $40.8 million as of March 31, 2020. Total cash, cash equivalents and short-term investments on June 30, 2020 does not include total net proceeds of approximately $116.5 million from the Company's IPO in July 2020. Based on its current plans, the Company expects that its existing cash, cash equivalents and short-term investments, including the proceeds from its July 2020 IPO, will be sufficient to enable it to fund its operating expenses and capital expenditure requirements into the second half of 2022.

- **Research and Development (R&D) Expenses** – R&D expenses were $7.9 million for the second quarter ended June 30, 2020, compared to $3.5 million for the same period in 2019. The increase was primarily due to higher consulting and professional fees related to the planned filing of an IND for INZ-701, preclinical studies and clinical preparation activities with the Company's CRO, and growth in the number of R&D employees.

- **General and Administrative (G&A) Expenses** – G&A expenses were $1.7 million for the second quarter ended June 30, 2020, compared to $1.1 million for the same period in 2019. The increase was primarily due to the growth in the number of G&A employees, an increase in legal fees related to patents and new contracts, and generally higher fees in areas such as audit, tax and information technology to support the Company's growth.

- **Net Loss** – Net loss was $9.5 million, or $7.57 loss per share, for the second quarter ended June 30, 2020, compared to $4.2 million, or $3.57 loss per share, for the same period in 2019.

About Inozyme Pharma

Inozyme Pharma 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 potentially first-in-class 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 future clinical trials and our research and development programs, the availability of preclinical study and clinical trial data and the period over which we believe that our existing cash and cash equivalents 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 successfully resolve the clinical hold with regard to its

planned Phase 1/2 clinical trial of INZ-701 for ENPP1 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, 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, 2020	December 31, 2019
Cash, cash equivalents and short-term investments	$ 63,867	$ 47,132
Total assets	68,511	47,944
Total liabilities	6,965	3,236
Convertible preferred stock	111,565	77,927
Accumulated deficit	(51,863)	(34,652)
Total stockholders' deficit	(50,019)	(33,219)

Condensed Consolidated Statements of Operations and Comprehensive Loss
(Unaudited)

(in thousands, except share and per share data)

	Three Months Ended June 30,		Six Months Ended June 30,	
	2020	2019	2020	2019
Operating expenses:				
Research and development	$ 7,877	$ 3,489	$ 14,283	$ 7,623
General and administrative	1,671	1,064	3,171	2,094
Total operating expenses	9,548	4,553	17,454	9,717
Loss from operations	(9,548)	(4,553)	(17,454)	(9,717)
Other income (expense):				
Interest income	71	394	242	604
Other income (expense), net	4	(14)	1	(31)
Other income (expense), net	75	380	243	573
Net loss	$ (9,473)	$ (4,173)	$ (17,211)	$ (9,144)
Other comprehensive (loss) income:				
Unrealized (losses) gains on available-for-sale securities	(15)	10	8	12
Total other comprehensive (loss) income	(15)	10	8	12
Comprehensive loss	$ (9,488)	$ (4,163)	$ (17,203)	$ (9,132)
Net loss attributable to common stockholders—basic and diluted	$ (9,473)	$ (4,173)	$ (17,211)	$ (9,144)
Net loss per share attributable to common stockholders—basic and diluted	$ (7.57)	$ (3.57)	$ (14.01)	$ (7.83)
Weighted-average common shares outstanding—basic and diluted	1,251,244	1,170,480	1,228,296	1,167,346

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