Inozyme Pharma Announces Authorization to Proceed in U.S. and U.K. with Phase 1/2 Clinical Trial of INZ-701 for the Treatment of ENPP1 Deficiency

January 4, 2021

– U.S. Food and Drug Administration cleared Investigational New Drug Application –

– United Kingdom Medicines and Healthcare Products Regulatory Agency authorized Clinical Trial Application –

– Program addressing rare mineralization disorders expected to enroll first subject in H1'21 and provide preliminary safety and biomarker data in H2'21 –

BOSTON, Jan. 04, 2021 (GLOBE NEWSWIRE) -- **Inozyme Pharma, Inc. (Nasdaq: INZY),** a clinical-stage biopharmaceutical company developing novel therapeutics for the treatment of rare diseases of abnormal mineralization impacting the vasculature, soft tissue and skeleton, today announced that the U.S. Food and Drug Administration (FDA) has cleared the Company's Investigational New Drug (IND) application and that the United Kingdom Medicines and Healthcare Products Regulatory Agency (MHRA) has authorized its Clinical Trial Application (CTA) for a Phase 1/2 clinical trial evaluating INZ-701 in adults with ENPP1 deficiency. The Company expects to enroll the first subject in the first half of 2021 and provide preliminary safety and biomarker data in the second half of 2021.

"With these important regulatory clearances for our first-in-human clinical trial for INZ-701 in subjects with ENPP1 deficiency, we have transitioned from a research-stage to a clinical-stage company. This is a significant milestone in our mission to develop therapeutic breakthroughs in diseases of abnormal mineralization," said Axel Bolte, MSc, MBA, co-founder, president and chief executive officer of Inozyme Pharma. "We are pleased to begin 2021 by ramping up study start up activities and look forward to dosing subjects in the first half of the year."

About the INZ701-101 Phase 1/2 Clinical Trial of INZ-701 in Adults with ENPP1 Deficiency

The Phase 1/2 clinical trial is a multi-center, open-label, first-in-human, multiple ascending dose study in adults with ENPP1 deficiency. The trial is expected to enroll nine adult subjects across three dose cohorts with three subjects per cohort. Subjects will participate in a pre-dosing screening period followed by a four-week treatment period in which subjects will receive INZ-701 subcutaneously twice weekly. The Phase 1/2 clinical trial will primarily investigate the safety and tolerability of INZ-701 and characterize its pharmacokinetic and pharmacodynamic profile, including plasma pyrophosphate (PPi) and other biomarker levels, to establish a recommended dosing regimen for further clinical development. Exploratory objectives include obtaining baseline measurements of calcification, patient reported outcomes and quality of life.

Additional details can be found:

https://clinicaltrials.gov/ct2/show/NCT04686175

About INZ-701

INZ-701 is a soluble, recombinant protein containing the extracellular domain of native human ENPP1 fused to the Fc domain of the immunoglobulin IgG1 that is designed to correct a defect in the mineralization pathway caused by ENPP1 and ABCC6 deficiencies. In preclinical studies conducted in ENPP1-deficient and ABCC6-deficient mouse models, dosing with INZ-701 resulted in normalized levels of PPi and reduced tissue calcification. The FDA has granted orphan drug, rare pediatric disease, and fast track designations to INZ-701 for the treatment of ENPP1 deficiency. The European Medicines Agency has also granted orphan drug designation to INZ-701 for the treatment of ENPP1 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. 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 ENPP1 and ABCC6 deficiencies. ENPP1 and ABCC6 deficiencies are chronic, systemic, and progressive diseases occurring over the course of a patient's lifetime, starting as early as fetal development and spanning into adulthood. ENPP1 and ABCC6 deficiencies are estimated to occur in approximately one in 200,000 and one in 50,000 births, respectively.

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, our research and development programs, the availability of preclinical study and clinical trial data, and the timing of our regulatory applications. 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 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.

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