Inozyme Pharma Raises $67 Million in Series A2 Financing to Develop Therapies for Rare Mineralization Disorders

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– Financing to Advance Lead Development Program, INZ-701, through Clinical Proof of Concept in Two Indications –

– Company Expands Pipeline with New Program for a Rare Mineralization Disorder –

Boston, Mass., April 10, 2019 – Inozyme Pharma, Inc., a biotechnology company developing novel medicines to treat rare, debilitating, and life-threatening mineralization disorders, today announced that the Company raised $67 million in a Series A2 financing led by Pivotal bioVenture Partners and Sofinnova Investments. Cross-over investors, including RA Capital Management, Cowen Healthcare Investments, and Rock Springs Capital, as well as the Company's previous investors, Longitude Capital, NEA, Novo Holdings, and Sanofi Ventures, also joined this round.

Launched in 2017, Inozyme has to date raised $116 million in venture capital. The Company will use the proceeds from the Series A2 financing to advance its lead enzyme replacement therapy, INZ-701, into clinical development for severe disorders of calcification associated with deficiencies in the enzyme ENPP1, including Generalized Arterial Calcification of Infancy (GACI) and Autosomal Recessive Hypophosphatemic Rickets Type 2 (ARHR2).
Inozyme also announced that it is expanding the INZ-701 development program by adding ABCC6 Deficiency as a second indication. Similar to mutated *ENPP1*, mutations in the *ABCC6* gene are linked to calcification disorders resulting from low pyrophosphate (PPi) and can lead to a condition known as pseudoxanthoma elasticum (PXE). Furthermore, the Company has broadened its pipeline by adding a second research program targeting an undisclosed rare bone disease.

"Since our inception, we have advanced our lead candidate INZ-701 through preclinical development. Our goal is to develop first-in-class therapies for patients with rare mineralization disorders," said Axel Bolte, chief executive officer and co-founder of Inozyme Pharma. "With the addition of a second indication for INZ-701 and a second rare bone disease program, we are expanding our footprint in these underserved disorders. This latest round of financing enables us to broaden our development pipeline and to bring INZ-701 through clinical proof of concept. We are excited to evolve our investor base by adding five highly regarded new investors who are committed to helping us achieve our goals, and we appreciate the continued support of our existing investors as Inozyme continues to grow. We will continue to evaluate additional assets that are highly innovative and have the potential to make a meaningful impact for patients with mineralization disorders."

The Company has already demonstrated preclinical proof of concept for INZ-701 in GACI and ARHR2. Both of these disorders result from mutations in the *ENPP1* gene, leading to ENPP1 Deficiency. The Company is currently completing the IND-enabling studies for INZ-701 and expects to initiate clinical trials in 2020.

"Pivotal bioVenture Partners is delighted to join the Inozyme Pharma team in its quest to develop therapeutic solutions for patients with rare, debilitating, and life-threatening mineralization disorders," commented Rob Hopfner, RPh, Ph.D., MBA, managing partner of Pivotal bioVenture Partners. "Inozyme has assembled a world class team of drug hunters and drug developers to advance first-in-class therapies for rare orphan diseases. We have been highly impressed by their solid science, well-defined regulatory strategy, and deep understanding of the biology of rare mineralization disorders."

Inozyme also announced the addition of Dr. Hopfner to its Board of Directors, along with Sarah Bhagat, Ph.D., principal at Sofinnova Investments.

About INZ-701

INZ-701 is an enzyme replacement therapy in development for the treatment of mineralization disorders of the circulatory system, bones, and kidneys. In preclinical studies, the experimental therapy has shown potential to generate plasma pyrophosphate (PPi) and to restore it to appropriate physiological levels, thereby preventing calcification in the vasculature and kidneys, while at the same time normalizing bone mineralization. Inozyme is developing INZ-701 for certain rare, life-threatening, and devastating genetic disorders such as ENPP1 Deficiency (GACI and ARHR2) and ABCC6 Deficiency (PXE) in which PPi levels are below the normal physiological levels. For more information about INZ-701, please visit: http://www.inozyme.com/our-science/.

About Inozyme Pharma, Inc.

Inozyme Pharma is a biotechnology company committed to developing novel medicines for the treatment of rare mineralization disorders. Inozyme Pharma was founded in 2017 by Joseph Schlessinger Ph.D., Demetrios Braddock, M.D., Ph.D., and Axel Bolte, MSc., MBA. The Company licensed technology from Yale University, developed in the laboratory of Dr.
Braddock.

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