Maze Therapeutics Announces Exclusive Worldwide License Agreement with Sanofi for MZE001, an Oral Substrate Reduction Therapy for the Treatment of Pompe Disease

MZE001 was Well-tolerated and Reduced Glycogen Accumulation in Blood Cells and Muscle in Healthy Volunteers

Maze to Receive an Aggregate of $150 Million in Upfront Cash and Equity Investment, with Potential for Approximately $600 Million in Aggregate Development, Regulatory and Commercial Milestones

SOUTH SAN FRANCISCO, CA., May 1, 2023 –Maze Therapeutics, a company translating genetic insights into new precision medicines, today announced the signing of an exclusive worldwide license agreement with Sanofi (Nasdaq: SNY) for Maze's glycogen synthase 1 (GYS1) program, including clinical candidate MZE001, which is currently in development for the treatment of Pompe disease and other potential indications. MZE001, designed and developed by Maze, is an oral GYS1 inhibitor that aims to address Pompe disease by limiting disease-causing glycogen accumulation.

"People with Pompe disease continue to need additional treatment options for this life-threatening condition. Sanofi is a leading global healthcare company with deep experience working with this community and the ideal partner to continue the advancement of MZE001," said Jason Coloma, Ph.D., chief executive officer of Maze. "This agreement provides important validation for the potential of our Compass platform to elucidate novel, genetic insights that inform the discovery and advancement of new medicines. We believe the applicability of these insights is broad, and as we look ahead, Maze will focus on more common disorders, where ultimately, we may best apply our expertise and resources to offer the biggest impact for patients. We are thankful for this partnership and are excited to enter the next phase of Maze's ambitious mission to transform the lives of patients through genetically informed therapies."

"Improving the lives of people with Pompe disease is a key focus for Sanofi, and we believe MZE001 could be an important addition to the patient treatment paradigm," said Karin Knobe, Global Head of Clinical Development Rare Diseases and Rare Blood Disorders, Sanofi. "MZE001 has demonstrated meaningful preclinical proof of concept by inhibiting GYS1, a validated, genetic driver of Pompe disease. We are pleased to enter this agreement with Maze and look forward to continuing the advancement of this program."

Maze has advanced MZE001 through Phase 1 development, and recently reported results of its study in healthy volunteers. In the first-in-human, double-blind, placebo-controlled, single and multiple ascending dose clinical trial, MZE001 was well-tolerated at doses up to 720 mg twice daily. Response to MZE001 was evaluated in patients using a novel biomarker, peripheral blood mononuclear cell (PBMC) glycogen, and demonstrated exposure-dependent reductions in PBMC glycogen across dose levels 10 days after administration, confirming target engagement with GYS1. These results were further confirmed in a muscle biopsy cohort, that showed equivalent reductions in muscle glycogen with MZE001.

Under the terms of the agreement, Maze will receive a $150 million payment consisting of both upfront cash and future equity investment for the rights to further develop and commercialize MZE001, as well as an exclusive license to related GYS1-targeting back-up programs and intellectual property. Maze will be

eligible to receive up to an additional approximately $600 million in potential development, regulatory and sales milestones, as well as meaningful royalties on sales if MZE001 is successfully commercialized.

About Pompe Disease
Pompe disease is a rare, inherited disorder caused by mutations in the gene coding for acid alpha-glucosidase (GAA), which lead to the buildup of glycogen in skeletal muscle, respiratory muscle and cardiac muscle tissues resulting in progressive weakness and respiratory compromise.

About MZE001
MZE001 is an investigational oral glycogen synthase (GYS1) inhibitor that aims to address Pompe disease by limiting disease-causing glycogen buildup. GYS1 is an enzyme responsible for glycogen production. MZE001 is currently being evaluated as a potential oral treatment for patients with Pompe disease, as well as other glycogen storage disorders.

About Maze Therapeutics
Maze Therapeutics is a biopharmaceutical company that is harnessing the power of human genetics to transform the lives of patients, with a focus on genetically informed therapies for common diseases such as chronic kidney disease. Maze applies variant functionalization in tandem with advanced data science methods and a robust suite of research and development capabilities to advance a pipeline of novel precision medicines. Maze has developed the Maze Compass Platform ™, a proprietary, purpose-built platform to understand and integrate the critical step of variant functionalization into each stage of drug development. Utilizing the Maze Compass Platform™, Maze is building a broad portfolio of wholly owned and partnered programs. Maze is based in South San Francisco. For more information, please visit mazetx.com, or follow us on LinkedIn and Twitter.

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