PRESSRELEASE

SHIONOGI

Shionogi & Co., Ltd. and Maze Therapeutics, Inc. Announce Exclusive Worldwide License Agreement for MZE001, a Novel Therapeutic Candidate for the Treatment of Pompe Disease

- Agreement Adds to Shionogi's Growing Pipeline in Rare Disease
- MZE001 has the Potential to be the First Oral Therapy for Pompe Disease

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OSAKA, Japan, and South San Francisco, California, May 10, 2024 – Shionogi & Co., Ltd. (Head Office: Osaka, Japan; Chief Executive Officer: Isao Teshirogi, Ph.D.; hereafter "Shionogi") and Maze Therapeutics, Inc. (Head Office: South San Francisco, California, USA; Chief Executive Officer: Jason Coloma, Ph.D.; hereafter "Maze") announced the companies have completed an exclusive worldwide license agreement for the rights to MZE001, an investigational oral glycogen synthase 1 (GYS1) inhibitor that aims to address Pompe disease by limiting disease-causing glycogen buildup.

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

Under the terms of the agreement, Shionogi has acquired exclusive worldwide rights for MZE001 as well as related programs and intellectual property. Shionogi will pay an upfront fee of $150 million, and Maze will be eligible for milestone payments based on development, regulatory and commercial achievements plus tiered royalties based upon future net sales. Shionogi and Maze have passed the required 30-day waiting period outlined in United States Hart-Scott-Rodino (HSR) Act, and the transaction is completed.

"This agreement is a strong strategic fit for Shionogi. It will help meaningfully advance our commitment to develop innovative medicines that address unmet medical needs and complement Shionogi's rapidly expanding pipeline in the focus areas designated in our Medium-Term Business Plan STS2030 Revision," said Isao Teshirogi, Ph.D., CEO of Shionogi. "The science behind MZE001 is differentiated and promising, and we look forward to developing the compound as both monotherapy and add-on therapy to enzyme replacement therapies."

MZE001 is a small molecule and specific inhibitor of GYS1, an enzyme involved in glycogen synthesis. It reduces the glycogen concentration in muscles by inhibiting this enzyme, and the results from the Phase 1 study of MZE001 suggest that it has the potential to be the first oral therapy for the treatment of Pompe disease[ii]. MZE001 has the potential to be used both as a monotherapy option and as an add-on therapy with enzyme replacement, the current standard of care, to enhance the treatment of patients with Pompe disease.

"Shionogi is committed to advancing and commercializing MZE001 because they understand the potential this therapy has for patients and the unmet medical needs it could address," said Jason Coloma,

Ph.D., CEO of Maze. “Shionogi has a track record of developing and delivering innovative medicines to patients worldwide, and we’re confident they are the right partner to continue to advance MZE001 through clinical trials so that it may reach patients with this life-threatening condition as soon as possible.”

In 2022, the U.S. Food and Drug Administration (FDA) granted Orphan Drug Designation to MZE001.[iii] The FDA grants Orphan Drug designation to prevent, diagnose or treat a rare disease or condition. Under the FDA's Orphan Drug Act, orphan drug status provides incentives, including tax credits, grants and waiver of certain administrative fees for clinical trials, and seven years of market exclusivity following drug approval.

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About Pompe Disease
Pompe Disease is a rare congenital metabolic disorder in which there is a deficiency of the enzyme necessary for the breakdown of glycogen within cells, resulting in the accumulation of glycogen in cells throughout the body, especially in muscle cells. This leads to various symptoms such as muscle weakness and delayed growth. In Japan, it has been designated as a "specified rare disease" and a "specific pediatric disease," making it eligible for medical expense assistance programs and other forms of support.[i]

About Maze
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.

About Shionogi
Shionogi & Co., Ltd. is a leading global research-driven pharmaceutical company dedicated to bringing benefits to patients based on its corporate philosophy of "supplying the best possible medicine to protect the health and well-being of the patients we serve." Shionogi has discovered and developed novel antibiotics, medicines for HIV and influenza and currently markets medicines for infectious diseases and central nervous system disorders. Shionogi’s global pipeline includes research programs in infectious disease, pain/CNS, metabolic disorders, rare disease, oncology and stroke. For more information, visit https://www.shionogi.com/global/en/.

Forward-Looking Statements
This announcement contains forward-looking statements. These statements are based on expectations in light of the information currently available, assumptions that are subject to risks and uncertainties which could cause actual results to differ materially from these statements. Risks and uncertainties include general domestic and international economic conditions such as general industry and market conditions, and changes of interest rate and currency exchange rate. These risks and uncertainties particularly apply with

respect to product-related forward-looking statements. Product risks and uncertainties include, but are not limited to, completion and discontinuation of clinical trials; obtaining regulatory approvals; claims and concerns about product safety and efficacy; technological advances; adverse outcome of important litigation; domestic and foreign healthcare reforms and changes of laws and regulations. Also for existing products, there are manufacturing and marketing risks, which include, but are not limited to, inability to build production capacity to meet demand, lack of availability of raw materials and entry of competitive products. The company disclaims any intention or obligation to update or revise any forward-looking statements whether as a result of new information, future events or otherwise.

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References

[i] Pompe disease: pathogenesis, molecular genetics and diagnosis. Taverna S et al., Aging 2020, Vol. 12, No. 15.

[ii] Maze Therapeutics Announces Positive Phase 1 Results from First-in-Human Trial Evaluating MZE001 as a Potential Oral Treatment for Pompe Disease | Maze Therapeutics (mazetx.com)

[iii] Maze Therapeutics Announces FDA Orphan Drug Designation Granted to MZE001 for the Treatment of Pompe Disease | Maze Therapeutics (mazetx.com)