Reata Enrolls First Patient in the MOTOR Study, a Phase 2 Study of the Safety, Efficacy, and Pharmacodynamics of RTA 408 in the Treatment of Mitochondrial Myopathies

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IRVING, Texas, July 8, 2015 – Reata announces the enrollment of the first patient in MOTOR, a Phase 2 dose-ranging study examining the safety, tolerability, and efficacy of RTA 408 Oral Capsules for the treatment of patients with mitochondrial myopathies (MM).

MOTOR is a multi-center study planned for approximately 52 patients with MM. MM is a collective term for a group of individual rare diseases associated with mitochondrial DNA mutations. These defects cause respiratory chain deficits and impaired energy production. Most patients with MM share a similar phenotype with skeletal muscle weakness and fatigue. They also may have additional symptoms due to impaired energy production in other organ systems and often have reduced lifespans. Approximately 20,000 people in the United States are believed to have a form of MM. There are currently no approved therapies for MM.

The primary efficacy endpoint is the change in peak workload (Watts/kg) during exercise testing. The secondary endpoint includes a patient's 6-minute walk distance. The study is also exploring the change in peak oxygen utilization during maximal exercise testing and changes in the Fatigue Severity Scale.

"Emerging translational research demonstrates that activation of Nrf2 (the target of RTA 408) can improve mitochondrial function and cellular energy production. These observations underlie our hypothesis that RTA 408 may improve exercise capacity and quality of life in patients with mitochondrial myopathies," noted Dr. Colin Meyer, Reata's Chief Medical Officer. "We are hopeful that RTA 408 will benefit mitochondrial myopathy patients, and we appreciate the guidance and support that Reata has received from the UMDF and the MM patient community."

"We are very excited this trial will be underway with its first patient," said Charles A. Mohan, Jr., Executive Director and CEO of the United Mitochondrial Disease Foundation. "Coordination, communication and collaboration between our industry partners and our patients to promote and support clinical trials is the only way we will accelerate the development of diagnostic tools, therapies, and potential cures for mitochondrial disease. We are pleased with the role Reata has taken in this endeavor and honored to call them a partner. We must all remember; no patients no trials, no trials no treatments nor cures."

For more information on this study, visit: https://clinicaltrial.gov/ct2/show/NCT02255422.

About RTA 408 and Bioenergetics Effect

RTA 408 activates the body's anti-oxidative pathways through transcription factor Nrf2 and is able to improve mitochondrial function. In mouse models of bioenergetic disease, RTA 408 demonstrated the ability to increase glucose uptake, fatty acid oxidation and oxygen consumption, direct signs of healthier cellular metabolism (http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3003357). RTA 408 analogs have also demonstrated in mouse models of oxidative stress the ability to induce genes related to mitochondrial biogenesis through the activation of Nrf2, which showed signs of potentially improving muscle function (http://www.ncbi.nlm.nih.gov/pubmed/21457778).

About Reata Pharmaceuticals, Inc.

Reata Pharmaceuticals, Inc. is a privately held company aiming to translate innovative research into breakthrough medicines for difficult diseases that have significant unmet needs. Reata is the leader in developing a novel class of drugs with potent transcription-regulating activity, called antioxidant inflammation modulators (AIMs). AIMs activate Nrf2, promoting the production of numerous antioxidant, detoxification, and anti-inflammatory genes, and inhibit NF-κB, a gene that regulates many pro-inflammatory proteins. The pharmacology of the AIMs mimics that of endogenous prostaglandin

metabolites that are responsible for the orchestrated resolution of inflammation. The anti-inflammatory, cytoprotective and energy metabolism effects of AIM pharmacology have been documented in more than 250 scientific papers and are potentially relevant to a wide range of diseases.

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