Liquidia Initiates Phase 3 Clinical Trial of LIQ861 in Patients with Pulmonary Arterial Hypertension

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RESEARCH TRIANGLE PARK, NC

Liquidia Technologies, Inc., a late-stage clinical biopharmaceutical company focused on improving the performance of medicine by precisely engineering drug particles, today announced the initiation of a Phase 3 clinical trial evaluating LIQ861 for the treatment of pulmonary arterial hypertension (PAH). LIQ861, developed using Liquidia's proprietary PRINT® technology , is a powder formulation of treprostinil designed for deep-lung delivery using a disposable, dry powder inhaler (DPI). PAH is a chronic, progressive disease caused by the hardening and narrowing of the pulmonary arteries, which often results in heart failure. Previously approved by the U.S. Food and Drug Administration (FDA) in oral, nebulized and parenteral formulations, treprostinil is a synthetic analog of prostacyclin, a vasoactive mediator deficient in patients with PAH yet essential to normal lung function to regulate vessel tone.

> "We are committed to improving the daily experience of those living with PAH by advancing the clinical development of LIQ861," said Neal Fowler, Chief Executive Officer at Liquidia Technologies. "LIQ861 shows the potential of our PRINT® technology to optimize safety, efficacy and convenience of therapeutics. The initiation of our trial marks an important milestone for Liquidia, for those living with PAH and for the PAH community at large. We believe that we are well positioned to advance LIQ861 through Phase 3 clinical development."

Liquidia expects to enroll at least 100 patients with PAH across multiple U.S. sites in the open-label Phase 3 clinical trial, with the first patient enrolling within the next few weeks. Primary endpoints of the trial are long-term safety and tolerability of LIQ861. Topline data are expected in 2019. In March 2017, Liquidia completed a Phase 1 clinical trial of LIQ861 in healthy volunteers in which the drug was well tolerated at all doses, with a proportional response in pharmacokinetics. Liquidia anticipates reporting additional data from this Phase 1 trial at scientific conferences in 2018. Based on feedback from the FDA, Liquidia believes that the Phase 3 clinical trial will support a potential U.S. regulatory approval of LIQ861.

"While treatment options exist for PAH, there is still a strong need for innovation to make it easier for patients to self-administer some of the more complex therapies," said Nicholas Hill, MD, Chief Pulmonary, Critical Care & Sleep Division and Professor of Medicine at Tufts University School of Medicine. "Leveraging Liquidia's PRINT® technology to enable deep-lung delivery of inhaled treprostinil, LIQ861 offers the convenience of a dry powder inhaler, which greatly simplifies the delivery of inhaled treprostinil and I believe may likely improve the lives of such patients."

ABOUT PULMONARY ARTERIAL HYPERTENSION

PAH is a chronic, progressive disease characterized by abnormally high blood pressure in the pulmonary arteries between the heart and lungs of an affected individual. Over time, this can cause increased strain on the heart, leading to right-heart failure. Symptoms of PAH include shortness of breath, dizziness and fatigue, which grow more severe as the disease progresses. PAH represents Group I within the Pulmonary Hypertension WHO clinical classification system and is one of five such groups.

ABOUT LIQUIDIA TECHNOLOGIES

Liquidia Technologies is a late-stage clinical biopharmaceutical company that is focused on improving the performance of medicines by precisely engineering drug particles. Liquidia's proprietary PRINT® technology is designed to optimize the safety, efficacy or route of administration of a wide range of therapies by engineering uniform drug particles in a virtually unlimited number of compositions, sizes and shapes. Currently, Liquidia is developing two of its own product candidates using PRINT® particles: LIQ861 for the treatment of PAH and LIQ865 for the treatment of post-operative pain. Our lead product candidate, LIQ861, currently being evaluated in a Phase 3 clinical trial, is designed to improve the therapeutic profile of treprostinil by enabling deep-lung delivery and higher dose levels than current therapies by using a convenient, disposable dry powder inhalation device. LIQ865, currently being evaluated in a Phase 1 clinical trial, is designed to deliver sustained release particles of the non-opioid bupivacaine, a local analgesic, to treat post-operative pain. In addition to developing its own product candidates, Liquidia collaborates with leading pharmaceutical companies to apply its PRINT® technology across existing drugs, new chemical entities (NCEs) and biologics. Liquidia is based in Research Triangle Park, North Carolina. For more information, please visit www.liquidia.com.

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