Sionna Therapeutics Announces $182 Million Series C Financing to Advance Clinical Development of Novel Small Molecules in Cystic Fibrosis

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– Company positioned to have four compounds in clinical trials in 2024 including three NBD1 stabilizers and one ICL4 modulator –

– Edd Fleming, M.D., of Enavate Sciences joins company's Board of Directors –

Boston, MA, **March 6, 2024** — Sionna Therapeutics, a clinical-stage life sciences company dedicated to developing highly effective and differentiated treatments for cystic fibrosis (CF), today announced the closing of a $182 million Series C financing to support the clinical development of first-in-class small molecules designed to fully restore the function of the cystic fibrosis transmembrane conductance regulator (CFTR) protein by stabilizing the first nucleotide-binding domain (NBD1).

The Series C round, which was upsized and oversubscribed, was led by Enavate Sciences with additional new investors Viking Global Investors and Perceptive Advisors, as well as participation by all existing investors including RA Capital, OrbiMed, TPG's The Rise Fund, Atlas Venture, the Cystic Fibrosis Foundation, funds and accounts advised by T. Rowe Price Associates, Inc., and Q Healthcare Holdings, LLC., a wholly owned subsidiary of QIA. Sionna also announced today that Edd Fleming, M.D., Executive Vice President of Commercialization at Enavate Sciences, is joining its Board of Directors.

"We have deep experience in CF and a sharp focus on advancing the development of novel small molecules targeting NBD1 and complementary modulators that enable the potential for full restoration of CFTR function for most people living with CF," said Mike Cloonan, President and Chief Executive Officer of Sionna. "We are encouraged by the strong interest and validation from the excellent investors in our upsized Series C financing. This capital raise provides financial flexibility positioning us to execute our clinical development plan with funding through 2026 and multiple value-creating clinical readouts. We are also pleased to welcome Dr. Fleming to our Board and look forward to insights from his more than 30 years of experience in the health care industry."

CF is caused by mutations in the CFTR gene, which codes for an epithelial ion channel that is essential for producing healthy, freely flowing mucus in the airways, digestive system, and other organs. The most common mutation in CFTR, ΔF508, causes NBD1 to unfold at body temperature and severely impairs CFTR function.

Sionna has presented preclinical data, including data from the clinically predictive human bronchial epithelial cell (CFHBE) model, that demonstrate its NBD1 stabilizers can restore ΔF508-CFTR maturation, trafficking, and function to wild-type levels when combined with complementary modulators. A Phase 1 clinical trial of its first clinical-stage NBD1 stabilizer, SION-638, has identified doses that are generally safe and well tolerated, and target exposure (based on the CFHBE assay) was achieved at all doses, with more time above target with increasing dose.

Sionna has nominated two additional NBD1 stabilizers from its second series, SION-451 and SION-719, and plans to advance both compounds to clinical trials in 2024 pending results from ongoing Good Laboratory Practice (GLP) toxicology studies. In addition, the company is continuing to advance the development of compounds targeting complementary mechanisms including SION-109, which targets NBD1's interface with the CFTR intracellular loop 4 (ICL4) region; a Phase 1 clinical trial with SION-109 began in January 2024.

About Sionna Therapeutics

Sionna Therapeutics is a clinical-stage life sciences company dedicated to developing highly effective and differentiated treatments for cystic fibrosis (CF) by normalizing the function of CFTR, the key protein associated with disease progression in CF. Building on over a decade of extensive research on the genetic mutations associated with CF and founded in 2019, Sionna is advancing a pipeline of small molecules engineered to correct the protein defects caused by ΔF508, the most common mutation that affects the CFTR protein. The company has a first-in-class portfolio of programs directly targeting correction of NBD1, the key and unique mechanism to enable full restoration of ΔF508-CFTR function, and complementary programs targeting ICL4 and TMD1. Sionna's pipeline has the potential to deliver best-in-class efficacy and reach previously unachievable levels of long-term benefit for people with CF. For information about Sionna visit https://www.sionnatx.com/.

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