

2800 Meridian Parkway, Suite 195 Durham, North Carolina 27713 (919) 313-1180 (phone)

Contact: Paul Boucher Parion Sciences, President 919-475-9600 pboucher@parion.com

Parion Sciences Announces \$3 Million Award from Cystic Fibrosis Foundation Therapeutics to Accelerate the Development of Novel CF Treatment

Expanded Collaboration Supports Phase 2 Clinical Trial Initiation of First-In-Class ENaC Inhibitor for the Treatment of Cystic Fibrosis

Durham, NC (September 16, 2014) – Parion Sciences today announced it has received a \$3 million award from Cystic Fibrosis Foundation Therapeutics (CFFT) to support the development of P-1037, Parion's new investigational treatment for cystic fibrosis (CF). This award will accelerate the initiation of a phase 2 trial for P-1037 in people with CF. Parion intends to begin trial enrollment in early 2015 and will include people with CF regardless of an individual's genetic mutation. P-1037 inhibition of sodium channels in the airways is expected to promote fluid secretion and re-hydrate the mucus layers, thus restoring airway clearance, reducing infection and improving lung function.

"The Parion team appreciates all of the support from the Cystic Fibrosis Foundation as we advance our novel programs," stated Paul Boucher, President of Parion Sciences. "With this funding award, Parion is rapidly advancing our ENaC inhibitor, P-1037, toward initiating our phase 2 clinical trial in cystic fibrosis patients. Additionally, CFFT's extension of our CFTR program validates the promising progress made by our research team".

Under a separate award, CFFT has expanded its support of Parion's CFTR corrector research program by providing an additional \$967,000 in funding, for a total investment in CF research of approximately \$1.7 million.

About ENaC Inhibitors and P-1037

Epithelial sodium channel (ENaC) inhibitors are designed to block the sodium channels on the airway surfaces. In pulmonary diseases where there is a build-up of excessively concentrated mucus, such as cystic fibrosis and chronic obstructive pulmonary disease, preclinical models have demonstrated that blocking the ENaC channel promotes fluid secretion and re-hydrates the mucus layers. Hydration of mucosal surfaces restores airway clearance, reducing infection and improving lung function. P-1037 is a novel, long acting ENaC inhibitor that demonstrated a superior safety profile versus other known ENaC inhibitors in both pre-clinical and Phase 1 studies.

About CFTR Correctors

CF is caused by mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) protein. The most prevalent disease-causing mutation in CFTR, F508del, causes the CFTR protein to be misfolded, ultimately resulting in the loss of CFTR function, dehydration of the mucus membranes, and impairment of pulmonary, GI, and pancreatic function.

Parion is developing a novel series of small molecule CFTR correctors targeted at restoring the proper folding and stability to the FF508del CFTR protein. The Parion CFTR correctors are being developed as oral agents intended to improve CFTR function in all organs affected by CF.

About Parion Sciences

Parion Sciences is a development stage biopharmaceutical company dedicated to research, development and commercialization of treatments to improve and extend the lives of patients with innate mucosal surface defense deficiencies of the eye or airway. Parion has a diverse pipeline of preclinical and clinical candidates for the treatment of these diseases via distinctive mechanisms of action and approaches. Parion is at the forefront of ENaC development and is leveraging our scientific expertise in epithelial biology to expand our platforms and novel chemical compounds into new indications to treat mucosal defects. Parion has received support and grant funding from the National Institutes of Health and the Cystic Fibrosis Foundation Therapeutics, Inc. For more information, please see our website at <u>www.Parion.com</u>.

About the Cystic Fibrosis Foundation

The Cystic Fibrosis Foundation is the world's leader in the search for a cure for cystic fibrosis. The foundation funds more CF research than any other organization and nearly every CF drug available today was made possible because of foundation support. Based in Bethesda, MD, the foundation also supports and accredits a national care center network that has been recognized by the National Institutes of Health as a model of care for a chronic disease. The CF Foundation is a donor-supported nonprofit organization. For more information, go to www.cff.org.