**Title:** A Long-Term Prospective Observational Safety Study of the Incidence of and Risk Factors for Fibrosing Colonopathy in US Patients with Cystic Fibrosis Treated with Pancreatic Enzyme Replacement Therapy: A Harmonized Protocol Across Sponsors. Protocol No.: CFFC-OB-11

**Principal Investigator**: Brian O'Sullivan, M.D.

**Sponsor:** AbbVie, Aptalis Pharma US, Inc. (a subsidiary of Aptalis Pharma Inc.), and Janssen Research & Development, L.L.C.

**Purpose of Research**

It is not known exactly what causes FC. The purpose of this study is to determine the number of patients affected by FC and to learn more about what might cause it, including use of pancreatic enzyme supplements. The first reports of FC were in the 1990's and were linked with the use of high-dose pancreatic enzyme supplements. Dosing guidelines for pancreatic enzyme supplements were then put in place, and use of high-dose supplements was stopped. FC still occurs rarely in some patients with Cystic Fibrosis, so it is important to monitor the use of pancreatic enzyme supplements and other standard treatments in patients with Cystic Fibrosis to learn more about FC. The U.S. Food and Drug Administration (FDA) has asked the manufacturers of pancreatic enzyme supplements to study the incidence of FC in cystic fibrosis patients to see if there may be a link between taking pancreatic enzyme medications and developing FC.

The study will be conducted for a total of 10 years at up to 150 Cystic Fibrosis Care Centers and their partners. This includes all patients with active Cystic Fibrosis enrolled in the patient registry (Port CF), which is approximately 25,000 patients. Any patient with Cystic Fibrosis and with suspected FC will be invited to participate in this study. It is estimated that approximately five (5) cases of suspected FC will be reported per year across this entire Study.

**Inclusion Criteria**

1. Enrolled in Port CF.
2. Diagnosis of CF.
3. Receiving medical care from a CFF-accredited care center providing data to Port CF with IRB approval for this study.