

Effect of Lubiprostone on Nutritional Status and Pulmonary Function in Adults with Cystic Fibrosis

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Abstract

Background: Cystic fibrosis (CF) affects an estimated 30,000 people in the United States. It is caused by a mutation in the gene encoding a protein called cystic fibrosis transmembrane regulator (CFTR). This protein functions as a chloride channel in epithelial cells of multiple organ systems. The mutation results in a dysfunctional or absent CFTR channel and a decrease in chloride secretion, which results in thickened secretions in multiple organ systems including the lungs and gastrointestinal (GI) tract. This patient population commonly suffers from nutritional deficiency, and achieving and maintaining normal nutritional status is an important goal of therapy as body mass index (BMI) is positively correlated with FEV1, a measure of pulmonary function. Lubiprostone activates type 2 chloride channels (CIC-2) on the apical membrane of GI epithelial cells. Because its mechanism of action closely parallels the disease pathology, lubiprostone has the potential to provide GI benefits beyond the relief of constipation.

Objectives: The proposed pilot project is a prospective observational study to examine the effects of lubiprostone in adults with CF. The specific aims are to determine the effects of lubiprostone on: 1) nutritional markers and 2) pulmonary function in adults with CF.

Methods: Adults with CF who are currently taking lubiprostone chronically will be sought for enrollment. Study subjects will be followed for approximately 3-months with serial assessment of indicators of nutrition and pulmonary function. Nutritional markers to be measured include body weight, albumin, prealbumin, and vitamins A, D, and E. Pulmonary function will be assessed by pulmonary function tests, a survey to monitor for symptoms of pulmonary exacerbation, and monitoring of the frequency of hospitalizations and IV antibiotic use.

Expected Results: We expect to see a beneficial effect on nutritional markers, body weight, and BMI. We hope this translates into a concomitant improvement in pulmonary function.