Growth Retardation in Cystic Fibrosis is not Directly Related to Loss of CFTR Function in Intestinal Epithelia

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Abstract

Cystic Fibrosis (CF) is a fatal, hereditary disease that is characterized by gastrointestinal, pancreatic, and pulmonary disorders. CF is caused by a mutation in the Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) gene. An inherited condition, CF causes an organism-wide defect in epithelial tissue function. Common phenotype seen in patients and mouse models with CF are intestinal obstruction and malabsorption. These observations are widely regarded as primary reason for growth retardation in CF patients. In order to examine the impact of CFTR loss in the intestines only, we created tissue specific mice that displayed wild type or mutant CFTR function in all tissues except for the intestinal epithelia. By isolating the impact of the intestinal epithelia, we saw that CFTR function in the intestinal epithelia prevents obstruction and related mortalities; however, it did not directly and solely address the issue of growth retardation in mice. On the other hand, lack of CFTR in the intestines led to intestinal obstruction and goblet cell hyperplasia; however only slight impairment in growth. These results indicate that the previously held idea of intestinal epithelium being the sole cause of growth retardation and obstruction in CF mice does not explain the observed rates of mortality and obstruction seen in practice.