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Cystic fibrosis: evidence for gut inflammation

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Abstract

Cystic fibrosis (CF) gut manifestations are predominantly secondary to cystic fibrosis transmembrane regulator protein (CFTR) dysfunction. The CFTR gene is expressed throughout the intestinal tract. Because the intestine is difficult to assess in humans, there exists a lack of data on the underlying mechanisms of intestinal dysfunction. A more tractable approach involves the use of mouse models of CF, created by gene targeting techniques, to describe the consequences of CFTR dysfunction in the intestinal tissues, including mucus accumulation, disturbed motility, small bowel bacterial overgrowth and inflammation with altered innate immune responses, that are likely to be interrelated. We will focus on the latter. Recently, in poeple with CF, even in the absence of overt gastrointestinal symptoms, chronic intestinal inflammation and abnormal balance of the microbiota have been evidenced. Because chronic gut inflammation may be a driver for systemic inflammation, the prevention and control of intestinal inflammation represents a promising research strategy.

Key words (max 4): cystic fibrosis, gut, inflammation, immune response

Abbreviations:

CF: cystic fibrosis

CFTR : cystic fibrosis transmembrane conductance regulator

PI : pancreatic-insufficient

FC : fecal calprotectin

PA : *Pseudomonas aeruginosa*

1. Introduction

The CFTR gene is expressed throughout the intestinal tract [1] with the highest levels of CFTR mRNA expression in the duodenum. Levels of CFTR mRNA expression decrease distally to the ileum and there is a moderate expression in the large intestine. Small intestinal expression of CFTR is higher in