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Cystic Fibrosis Research News

Title:
Review of Gastrointestinal Motility in Cystic Fibrosis

Lay Title:
Review of abnormal Stomach and Intestinal Movement in Cystic Fibrosis

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What was your research question?
We wanted to summarize the key and latest research highlighting the normal movement of the bowel (motility) and abnormal movements (dysmotility). We wanted to explore theories for multiple and overlapping causes of dysmotility, different tests to diagnose it, and treatment options.

Why is this important?
Cystic fibrosis (CF) affects many organ systems including the gastrointestinal (GI) tract, commonly referred to as bowels. In fact, people with CF (pwCF) find bowel symptoms important contributors to an impairment of the quality of life. Examples of bowel symptoms are constipation, poor absorption of nutrients resulting in decreased growth, belly pain, nausea, vomiting, bloating, and even blockages of the intestines. GI dysmotility is thought to play a major role in these symptoms and this review focuses on the abnormal GI motility in pwCF to explain those symptoms and help to reach a proper diagnosis and treatment.

What did you do?
We provided a comprehensive review on GI motility in CF by conducting a careful literature search and summarizing the key research studies. We discussed the effect of malfunctioning cystic fibrosis transmembrane conductance regulator (CFTR) genes on GI motility in CF animal models and humans. We described presentations of dysmotility in CF in a comprehensive way. We reviewed newer tests to diagnose abnormal motility such as intraluminal

manometry (measurement of pressure within the bowels). We have also provided our expert opinion on the potential role of novel motility tests to establish abnormalities in CF intestines. We reviewed the role of highly effective CFTR modulators (CF protein correcting drugs) and their possible effect on improving dysmotility.

What did you find?

Factors involved in GI dysmotility in CF include abnormal bowel acidity, bowel inflammation, lack of digestive enzymes and abnormal composition of usual GI bacteria (microbiome). Muscles and nerves in GI tract may be unable to function in the same rhythm. Newer techniques such as Magnetic Resonance Imaging (MRI) and intraluminal manometry can detect abnormalities in the intestinal motility pattern in CF. We summarized the use of highly effective CFTR modulators (HEM) in pwCF and provided our hypothesis on HEM use and improvement of dysmotility.

What does this mean and reasons for caution?

For many years, scientists have theorized how CF affects bowel motility. Our review sheds light on how CFTR directly and indirectly affects abnormal bacteria, bowel acidity and bowel muscles and nerves and how these factors drive dysmotility, for example CFTR deficient mice have altered structural and functional bowels smooth muscle. HEM improve CFTR function and perhaps this can improve the abnormal bowel motility. We recognize the ambiguity of this complex topic and contradictory findings between different studies. This is also likely due to the lack of any baseline knowledge on normal motility pattern in pwCF and use of very different diagnostic techniques exploring dysmotility.

What's next?

We hope to deepen readers' understanding of abnormal motility in CF and open new areas of research to examine motility in pwCF. This could potentially help establish new treatments for DIOS (distal intestinal obstruction syndrome), gastroesophageal reflux, gastroparesis and abdominal pain to improve the quality of life of pwCF.

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