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CASE REPORT

Distal intestinal obstructive syndrome (DIOS): a gastrointestinal complication of cystic fibrosis in adults

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Abstract
Distal intestinal obstructive syndrome (DIOS) is a gastrointestinal complication of cystic fibrosis caused by increased viscosity of intraluminal contents leading to partial or complete bowel obstruction. DIOS is often misdiagnosed or mistaken for surgical bowel obstruction or appendicitis. This can lead to unnecessary surgical intervention when DIOS should be treated medically with aggressive bowel regimen. The aim of this report is to present a case of a young female with cystic fibrosis presenting with DIOS and discuss the clinical recognition and management of DIOS.

Keywords Distal intestinal obstructive syndrome · Bowel obstruction · Cystic fibrosis

Introduction

Cystic fibrosis (CF) is genetic disease caused by autosomal recessive mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) protein. CFTR is a key regulator in the secretory function of the epithelium of the respiratory and intestinal tracts. The respiratory manifestations of CF are well known features of the disease. However, the gastrointestinal manifestations are recognized less frequently.

CF can cause obstructive gastrointestinal disease, including meconium ileus in children and distal intestinal obstructive syndrome (DIOS) in adults [1]. DIOS occurs when viscous fecal material obstructs the terminal ileum, cecum or ascending colon. DIOS occurs in 10–15% of CF patients [1] and is more common in those with homozygous CFTR mutation [2].

It is often a recurrent process, with tenfold increased risk of developing recurrence after the first episode [1].

The clinical presentation is typically acute and often mimics appendicitis. Symptoms include abdominal pain and bloating, bilious vomiting, and inability to pass stool or flatus [1, 2]. A cecal mass may be palpable on exam [1, 2]. The diagnosis is made clinically, but can be confirmed with abdominal imaging. Radiologic findings are consistent with partial or complete intestinal obstruction, including bowel dilation and air-fluid levels [2]. This report presents a case of a young female with CF and recurrent DIOS to highlight the identification of DIOS in this patient population and discuss the appropriate management.

Case description

A 29-year-old female with CF, pancreatic insufficiency, and meconium ileus at birth, presented to the emergency department with a 1 day history of left-lower quadrant (LLQ) pain, abdominal distension and vomiting. Her last bowel movement was 5 days prior and she was unable to pass flatus. She had tried some bisacodyl suppositories without relief. Of note, she has had 3 similar episodes in the past. She had no surgical history nor history of tobacco, alcohol or recreational drug use. Her medications included inhaled budesonide, inhaled albuterol, inhaled dornase alfa, montelukast, pancrelipase, and lansoprazole.

At presentation, she was hemodynamically stable with blood pressure 112/63, heart rate 88 bpm, respiratory rate 18 bpm and oxygen saturation of 98% on room air. Physical exam revealed mildly distended abdomen with LLQ tenderness to palpation and hypoactive bowel sounds. There was no rebound tenderness, guarding or masses. Laboratory workup including complete metabolic panel and complete blood count was unremarkable, except for leukocytosis to 16.7 K/μL. An abdominal x-ray was

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non-diagnostic, however, contrast-enhanced CT abdomen/pelvis demonstrated multiple fluid filled, dilated loops of small bowel (Fig. 1).

She was admitted to the medical service without surgical consultation. A nasogastric tube was placed and she was treated with polyethylene glycol (PEG) solution. It was noted that during a prior episode of DIOS she required nearly 8 L (2 gallons) of PEG solution to resolve her symptoms, therefore, large volume of gastrointestinal lavage was anticipated during this presentation. Her obstruction began to resolve with the passage of small amount of stool within approximately 24 h of treatment. PEG administration was continued at 1 L every 8 h to complete a total of 7.4 L, at which time her obstruction was completely resolved. Abdominal XR at this time demonstrated significant reduction in stool burden. She was started on daily PEG powder to prevent recurrence.

Fig. 1 Coronal computed tomography of the abdomen/pelvis showing multiple fluid-filled dilated loops of small bowel (red arrows), consistent with DIOS

Discussion

The pathogenesis of DIOS is complex and involves multiple processes that contribute to increased viscosity of luminal contents (Fig. 2). The primary mechanism is mediated by disordered chloride secretion causing less fluid secretion into the bowel lumen. Bile handling in the ileum also plays a role. Bile is normally reabsorbed via a CFTR-mediated cotransporter [4]. In CF, less bile is reabsorbed and consequently, less fluid is secreted by the bile cotransporter [4]. Additionally, CF patients commonly suffer from concomitant pancreatic insufficiency. This leads to poor fat reabsorption and, therefore, increased presence of viscous fat in the bowel lumen [4]. Fat malabsorption then triggers the "ileal brake", which slows gut transit time in CT patients [1]. The accumulation of dehydrated fecal material in addition to slowed transit time leads to partial or complete intestinal obstruction.

Distinguishing DIOS from general bowel obstruction or other mimickers is important because the management is very different. If this distinction is not recognized, patients may be subjected to unnecessary surgical intervention. The majority of cases resolve with medical management. First-line treatments include administration of osmotic laxatives such as PEG or sodium meglumine diatrizoate (Gastrografin®) in addition to adequate hydration [1, 4]. According to recommendations from the European Cystic Fibrosis Society in 2011, the dosing of PEG should be started at 2 g/kg/day with maximum rate of 1 L/h or Gastrografin® 50 mL diluted in 200 mL of water on day 1, followed by 25 mL daily until resolution [4]. Other studies have used higher doses of Gastrografin®, up to 100 mL in 400 mL of water per dose [2]. This patient was treated with PEG at a rate of approximately 1 L/h, however, required a total dose of 4 g/kg/day. She had been successfully treated for a prior episode of DIOS with large volume of PEG and again had high requirement for full resolution of her obstructive symptoms. The exact reason for this is unclear but is likely due to the severity of her recurrent disease and the extremely high viscosity of her gastrointestinal secretions. Alternative regimens for medical management include lubiprostone or linaclotide [1, 4]. Oral *N*-acetyl cysteine has also been used as an adjunct therapy given its success as an inhaled mucolytic agent for the respiratory components of CF [1]. In some cases, nasogastric aspiration is needed for decompression. One study reported that 91% of cases resolved with medical management within 48–72 h [3]. If medical management fails to relieve obstruction, surgical intervention is warranted. When managed properly the presence of DIOS does not increase mortality in CF patients [2].

The aforementioned treatment strategies for medical management are relatively safe. With aggressive intestinal

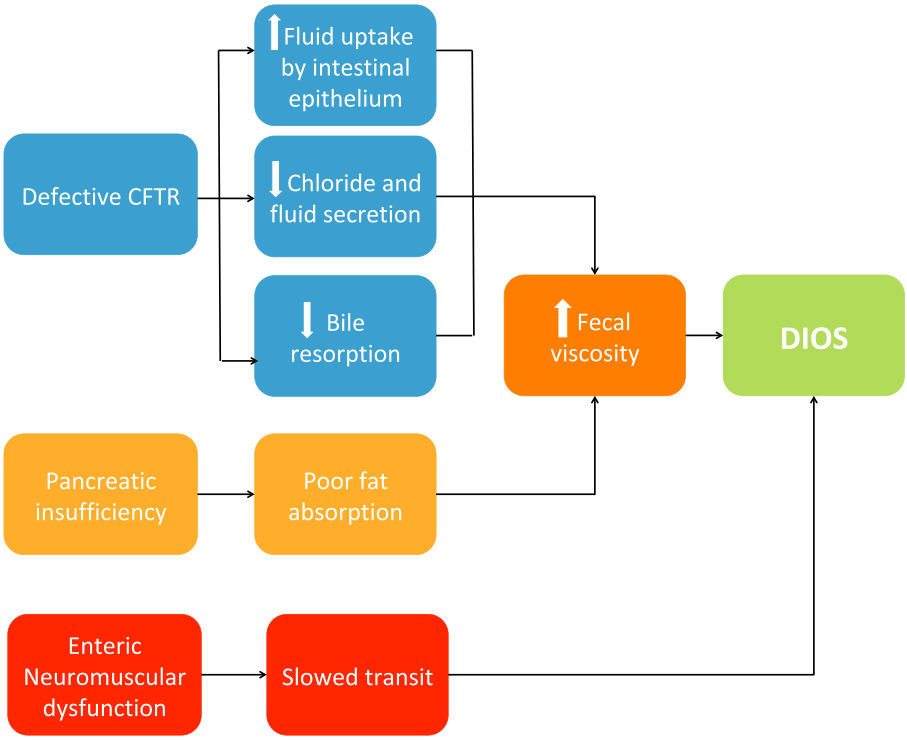

Fig. 2 Pathogenesis of DIOS in CF patients

lavage, there is risk of electrolyte abnormalities and dehydration. For this reason, serum chemistry and clinical volume status should be closely monitored. With ingestion of Gastrografin® or PEG solution there is also risk of vomiting or aspiration of the solution, leading to potential pulmonary complications [2]. In the setting of CF, any insult to the diseased lungs can be devastating.

Because DIOS has a propensity to recur, prevention with daily bowel regimen is key. Although no guidelines are available on prevention of DIOS, a commonly used regimen is polyethylene glycol 1–3 times daily [1].

Management of DIOS is an important aspect of care in CF patients. As the average age of CF patients increases, it is increasingly important to be able to identify and manage DIOS and the other gastrointestinal complications of CF.

Compliance with ethical standards

Conflict of interest Author has no financial conflicts of interest related to this work.

Human and animal rights All procedures followed have been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

Informed consent Informed consent was obtained from all patients for being included in the study.

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