

# Incomplete Distal Intestinal Obstruction Syndrome Complicated by Oligosymptomatic Intussusception

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## ABSTRACT

Distal intestinal obstruction syndrome (DIOS) is a relatively common intestinal complication in cystic fibrosis (CF), and it can eventually lead to intussusception, a less frequent complication in CF. Intussusception is classically associated with gastrointestinal symptoms that overlap those of DIOS. We describe a young woman with oligosymptomatic intussusception diagnosed after resolution of DIOS symptoms.

## INTRODUCTION

Distal intestinal obstruction syndrome (DIOS), initially described as “meconium ileus equivalent,” is characterized by acute partial or complete ileocecal obstruction by inspissated intestinal content, which occurs exclusively in patients with cystic fibrosis (CF).<sup>1,2</sup> The incidence of DIOS in CF patients is reported to be as high as 47%. DIOS was initially described in adolescents and young adults, although it can affect patients of all ages.<sup>2,3</sup> However a recent international collaboration has reported a similar incidence in the adult and in the pediatric population.<sup>4</sup> Several risk factors for DIOS were described, including, but not restricted to, severe CF genotype, pancreatic insufficiency, poor control of malabsorption of fat, dehydration, and prior history of DIOS. Recurrence is common in that a prior event of DIOS is associated with a 10 times greater risk for a subsequent episode.<sup>1</sup> Meconium ileus (MI) at birth was first described as a risk factor for DIOS, but today it is related to the risk of complete obstruction and DIOS recurrence.<sup>4</sup>

## CASE REPORT

A 17-year-old woman with CF, homozygous for the F508del mutation (class I genotype/severe phenotype), with associated pancreatic insufficiency, diabetes, pulmonary hypertension, and malnutrition, was evaluated after 4 weeks of outpatient treatment for CF pulmonary exacerbation. She complained of diffuse colicky abdominal pain and vomiting for 2 weeks. She had diarrhea for 5 days, but then returned to her regular bowel habit of soft stools 3 times daily. She had sought medical care twice and was discharged home. Physical exam revealed a weight loss of 2.3 kg in the past month, and a painful palpable mass in the right lower quadrant with no signs of peritonitis. Abdominal x-ray did not show a classic pattern of complete obstruction, but revealed a soft tissue mass overlying the right lumbar region (Figure 1). She had no previous history of MI or DIOS.

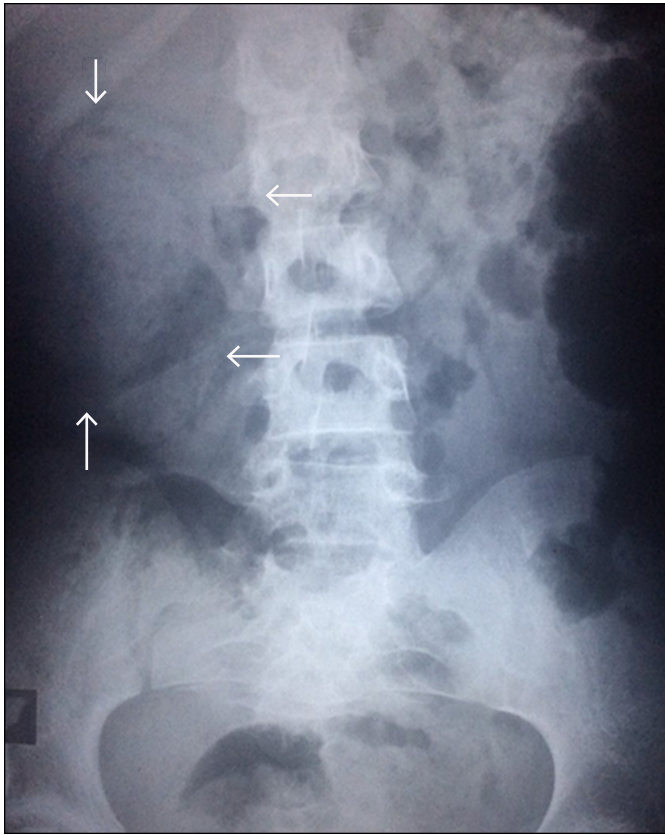
For the initial management of DIOS, polyethylene glycol (PEG) was administered orally, because the patient refused a nasogastric tube. She eliminated large amounts of feces and inspissated secretions and described relief of the abdominal pain and resolution of vomiting. The palpable abdominal mass decreased in size but did not resolve. Computed tomography (CT) revealed ileocecal intussusception associated with inspissated bowel contents (Figure 2). Bowel preparation for a colonoscopic procedure to reduce the intussusception included a

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**Figure 1.** Plain abdominal radiograph showing presence of large amounts of feces in virtually all corresponding topography of the small intestine and colonic segments, as well as the presence a soft tissue mass overlying the right lumbar region (arrows).

PEG/electrolyte solution 3 times a day and glycerin enema twice a day, over 2 days. During bowel preparation, the abdominal mass disappeared. Colonoscopy showed an enlarged colon and no intussusception at the time of the procedure, leading us to believe that the intussusception was reduced

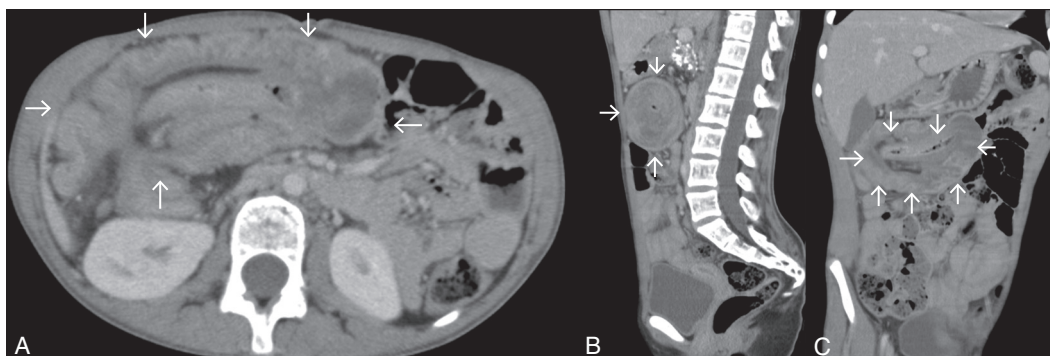
with the procedures leading up to the colonoscopy as well as gas insufflation during the study (Figure 3).

## DISCUSSION

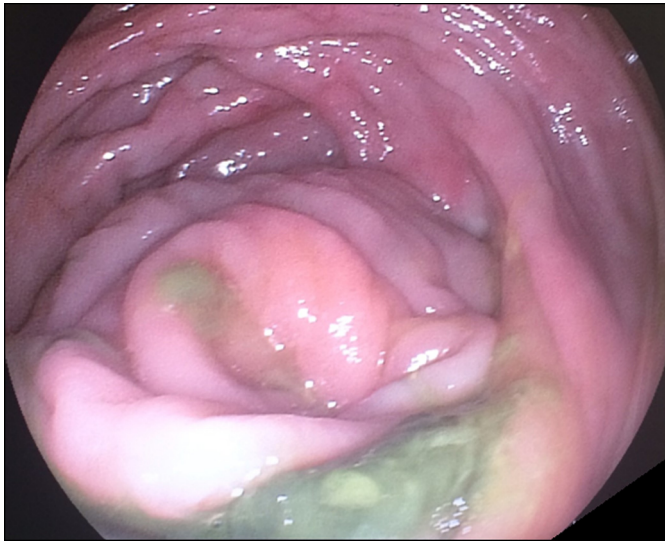
The differential diagnosis of DIOS includes chronic constipation, intussusception, intestinal obstruction of other causes, appendicitis, Crohn's disease, and fibrosing colonopathy, among others. The challenge is to differentiate incomplete DIOS versus constipation, and to avoid misdiagnosis. The ESPGHAN CF Working Group proposed that incomplete or impending DIOS should be defined as a short history (i.e., days) of abdominal pain and/or distension associated with a fecal mass in the ileocecum (without signs of complete obstruction), whereas in constipation the abdominal pain and/or distension is related to a decline in the frequency of bowel movements and/or increased consistency of stools, for weeks or months.<sup>1</sup> Typical manifestations of DIOS include crampy abdominal pain in the right lower quadrant, abdominal distension, palpable mass, flatulence, vomiting, and weight loss; bowel habits may remain unchanged, or patients may present with either diarrhea or constipation. The most common site of obstruction is the ileocecal junction.<sup>2</sup>

Plain abdominal radiographs are usually performed to support the diagnosis of DIOS and to differentiate complete versus incomplete obstruction. Other modalities of images, such as abdominal ultrasound and CT, are reserved for select cases, for differential diagnosis, and for detection of complications. CT in DIOS typically shows proximal small-bowel dilation and inspissated fecal material in the distal ileum; CT imaging is also useful to identify other causes of the symptoms, such as appendicitis or intussusception.

Management of DIOS is primarily nonsurgical, and conservative treatment is successful in the majority of cases. Most patients respond well to treatment with oral hydration and



**Figure 2.** Computed tomography imaging. (A) Axial view showing an enlarged and evident ileocecal intussusception (arrows) associated with the presence of thickened intestinal contents. The mesentery forms a crescent of tissue around the compressed innermost lumen, surrounded by the outer enveloping bowel. (B) Sagittal reconstruction showing the classic "target sign" of intestinal intussusception (arrows), with layers of fat and bowel wall is visible. Pancreatic calcifications are visible just above the intussusception. (C) Coronal view showing the longitudinal aspect of the ileocecal intussusception (arrows) as an elongated, sausage-shaped mass with visible layers.



**Figure 3.** Endoscopic view of moderately inflamed colon and increased caliber. Despite intense colonic preparation, there is a presence of mucofecaloid content thickened in the intestinal lumen.

stool softeners or laxatives.<sup>5</sup> An oral PEG solution or lavage is most frequently used.<sup>4</sup> Severe cases may require intravenous rehydration and nasogastric aspiration. Surgery is needed in a minority of cases of complete DIOS; a colonoscopic approach with local treatment (meglumine diatrizoate instillation) eliminates the need for surgical treatment in up to one third of patients who respond to conservative treatment.<sup>4</sup> Recurrence of DIOS can occur in up to 77% of patients, therefore maintenance therapy to avoid new episodes is advisable. In order to prevent recurrence we use oral osmotic laxative, PEG or lactulose, plus N-acetyl cysteine, or a lubricant laxative as mineral oil.<sup>4,6</sup>

Intussusception is a rare complication in CF, with an estimated incidence of 1%.<sup>6</sup> Compared to the general population, patients with CF have a higher risk of intussusception, and its

occurrence happens at a later age.<sup>7</sup> It is typically ileocolonic with an acute presentation, and the lead point is the mucofecaloid bolus associated with DIOS, which distends the appendix.<sup>7</sup> Signs and symptoms overlap those of DIOS, so the differential diagnosis may be difficult.<sup>6-8</sup>

## DISCLOSURES

Author contributions: NS Sandy provided the images, and wrote and edited the manuscript. EA Lomazi edited the manuscript and is the article guarantor.

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Informed consent could not be obtained because the patient is deceased and the authors were unable to contact the patient's next of kin.

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