

AN UNUSUAL CAUSE OF BOWEL OBSTRUCTION

ABSTRACT:

Background: Intestinal obstruction and dysmotility occur throughout life in cystic fibrosis but rarely present as an acute intestinal obstruction beyond the neonatal period. It is commonly seen in patients with pancreatic insufficiency, a previous history of meconium ileus, and previous episodes of DIOS.

Case Presentation: A 15-month-old female patient diagnosed case of Cystic fibrosis was admitted with complaints of constipation and abdominal distension for 3 days, with a history of undocumented fever, associated with nonbilious vomiting. On examination: the abdomen was soft and distended, and a mass in RIF could be palpated, The Rest of the General physical and systemic examination was unremarkable. Initial abdominal X-rays showed persistent fecal impaction in the caecal region. The patient underwent a Gastrografin enema that relieved the obstruction partially, The Gastrografin enema repeated after 2 days resolved the obstruction completely.

Conclusion: Distal intestinal obstruction syndrome (DIOS) is a morbidity of Cystic Fibrosis. Viscid fecal material in the bowel combines with thick, sticky mucous produced in cystic fibrosis intestine. It adheres to the intestinal wall commonly the terminal ileum and caecum, making it fixed in position and difficult to remove. After making the diagnosis of DIOS, the goal is to relieve obstruction. Different medical treatments are used to relieve the obstruction. Surgical intervention is associated with high postoperative morbidity and, therefore is reserved for most refractory cases, not responding to medical management.

Keywords: Cystic Fibrosis, DIOS, Gastrografin,

Introduction:

Cystic Fibrosis (CF) is an autosomal recessive disorder that is multisystemic, progressive, and fatal. It is characterized by dysfunction of the exocrine glands (sweat, bronchial, intestinal, exocrine pancreas, etc.) which causes thickening of secretions with obstruction of canaliculi of excretory glands leading to impaired functioning. (1)

In patients with CF, meconium ileus, distal obstruction syndrome (DIOS) and constipation are a group of gastrointestinal symptoms whose severity varies. All of them result from increased viscosity of the intestinal mucus and prolonged intestinal transit time. (1)

Distal intestinal obstruction syndrome (DIOS), formerly known as meconium ileus equivalent, refers to partial or complete obstruction of the distal small bowel and right hemi-colon with meconium-like inspissated fecal material.

The incidence of DIOS with complete intestinal obstruction studied in children was found to be between 5 and 12 episodes per 1000 patients per year throughout Europe. (2) Rates for incomplete obstruction – impending DIOS – are likely to be higher. DIOS is seen more frequently in adolescents and adult populations, we present a case of DIOS in 15 months 15-month-old child who was managed conservatively.

Case presentation:

Our patient was a 15-month-old baby, who was born full-term via LSCS, she had a history of recurrent chest infections, had been diagnosed as a case of cystic fibrosis, and was on pancreatic enzyme supplementations. There was no history of meconium ileus in neonatal life. She presented with a history of absolute constipation for 3 days, abdominal distension, abdominal pain for 2 days, and non-bilious vomiting for 2 days. The patient also had a history of low-grade fever that was undocumented. On examination, she had a pulse rate of 110 b/min, R/R of 32 breaths/min, and was afebrile. On per abdominal examination, the abdomen was soft, distended, and non-tender, and there was a mass palpable in the right iliac fossa, On DRE: the rectum was empty. The rest of the General physical and systemic examinations were unremarkable. Her laboratory and radiological workup was done. Her lab workup was unremarkable, while X-rays performed showed fecal impaction on the right side of the colon at ileocecum (FIG-1)



FIG-1: x-ray of abdomen supine showing fecal impaction on the right side as shown with an arrow.

The patient was kept NPO, Stomach decompression with a nasogastric tube was done, IV fluids, and antibiotics were started, and there was no need for blood transfusions. Her washouts were done but the patient was unable to pass flatus or feces, To relieve obstruction Gastrograffin enema was performed, which relieved obstruction partially. (FIG 2 a,b,c)

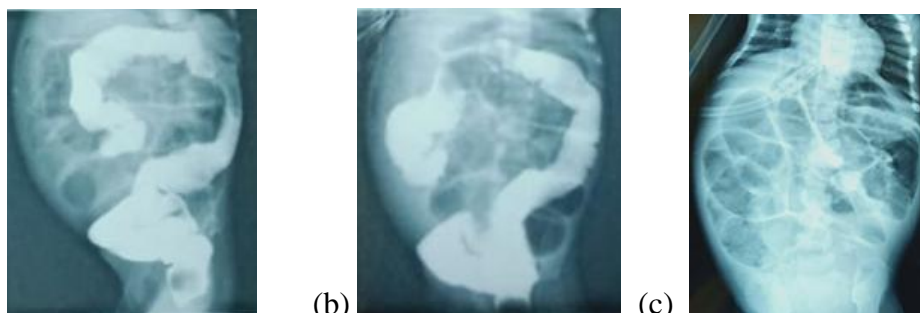
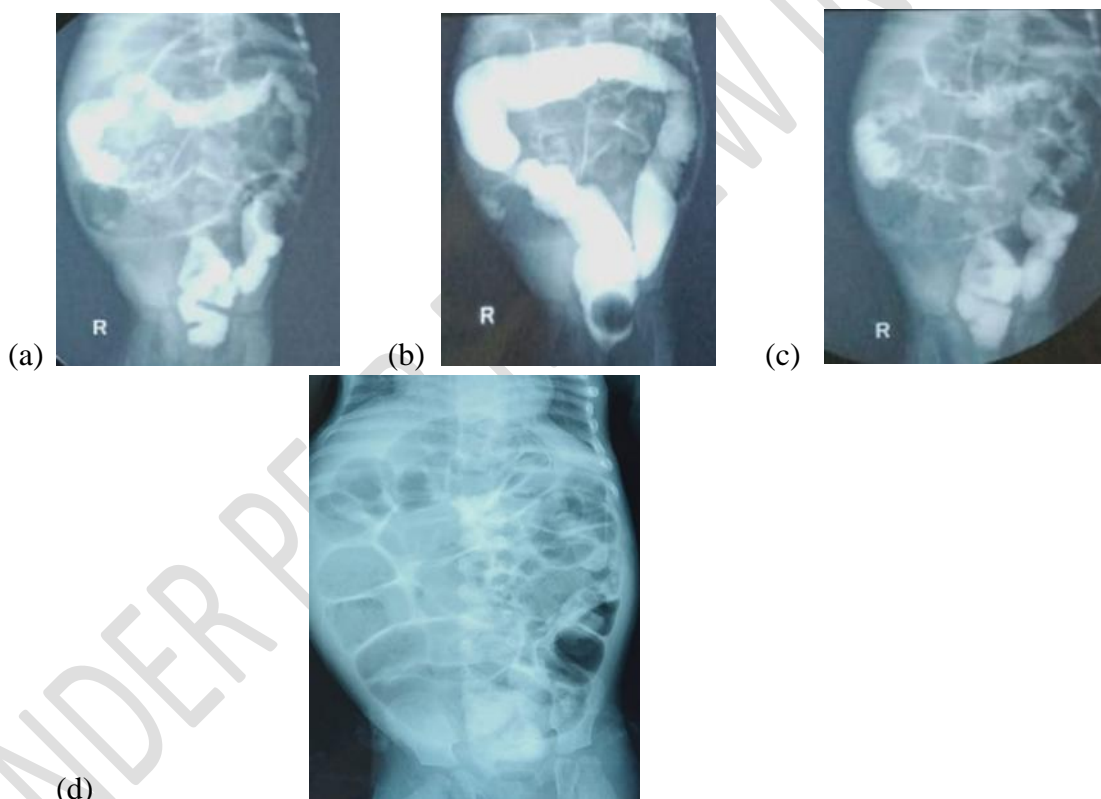


FIG 2

The patient remained vitally stable but the obstruction was not relieved, the Gastrografin enema was repeated after two days, which relieved the obstruction completely. (FIG 3 a,b,c,d)



The patient was able to pass stool, thus oral feedings were gradually established, she was then discharged on washouts and pancreatic enzyme supplementations and advised to follow up in OPD.

Discussion:

Distal Intestinal Obstruction Syndrome (DIOS) is a common complication in patients with Cystic Fibrosis (CF). It is characterized by the accumulation of viscid fecal material within the lumen of

the bowel, combined with sticky mucoid intestinal material, that is connected to the intestinal wall of the terminal ileum and caecum. It is discipale to the crypts and villi and is difficult to remove. (3)

DIOS is defined as an acute intestinal obstruction that may be incomplete or complete. According to Consensus guidelines issued by the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition (ESPGHAN) CF Working Group, Complete DIOS is total intestinal obstruction characterized by acute symptoms such as bilious vomiting with fecal ileocecal mass, pain, and/or bloating, and/or fluid levels in the small intestine seen on an abdominal radiograph.

Incomplete DIOS is in contrast defined by acute abdominal pain, bloating, and fecal ileocecal mass but is not associated with bilious vomiting or air-fluid levels.

(3)

Another important differential diagnosis of DIOS is constipation, which is also common in patients with CF. Constipation is characterized as the decreased frequency of bowel movements with increasing hardness of stools over weeks or months that can be associated with pain and/or bloating, relieved with the use of laxatives. (5)

The incidence of DIOS in CF patients has been defined to be 6.2/1,000 patient-years. This is a higher percentage than that reported by Andersen et al. who used the same definition and reported an incidence of 2.5/1,000 patient-years in CF patients younger than 20 years of age. (2)

In both varieties of DIOS, plain abdominal X-ray will show fecal impaction in the terminal ileum and cecum with proximal intestinal dilation with or without the presence of liquid and air.

Most of the time, episodes of DIOS and constipation are treated conservatively with laxatives and washouts.

Patients with incomplete DIOS are commonly treated with oral rehydration combined with stool , softeners (osmotic laxatives containing polyethylene glycol (PEG).

PEG is given at the dose of 2 g/kg/day, with a maximum dose of 80–100 g/day, or as a ready-to-use iso-osmotic PEG solution, at the dose of 20–40 ml/kg/h up to a maximum of 1 L/h over 8hours. This treatment aims to achieve fecal effluent consisting of clear fluid, with resolution of pain, abdominal distension, and vomiting.

Another alternative is sodium meglumine diatrizoate (Gastrografin) which can be administered orally or by nasogastric tube, at a dose of 50 ml diluted in 200 ml of water or juice for children who are <6 years old and 100 ml diluted in 400 ml for older patients on a day 1, and half doses on subsequent days. (2)

The use of N-acetyl cysteine has been superseded by the above medications when given orally. Prokinetics have shown some efficacy in pseudo-obstruction and postoperative ileus. However, there is no evidence to support their use in DIOS.

When DIOS presents with severe intestinal obstruction such as bilious vomiting, or when washout therapy fails, the patient should be hospitalized, and IV rehydration and nasogastric aspiration should commence. Gastrografin can be used by enema (100 ml diluted four times with water). The advantage is that it is radio-opaque, so the procedure of choice is to give it as retrograde lavage with hydrostatic pressure under direct vision until the terminal ileum is reached.

An alternative approach described in the literature is the instillation of diatrizoate in the caecum by colonoscopy.

With early aggressive medical management, surgery is seldom required. Laparotomy with washout via enterostomy should be tried before considering a more aggressive approach such as resection of the ileocaecum.

Conclusion:

Distal intestinal obstruction syndrome is one of the many presentations of cystic fibrosis, mostly in adolescent and adult patients. It results from the accumulation of thick sticky mucoid secretions in ileocecum, which may cause complete or partial intestinal obstruction. There are different modalities for the treatment. Surgery should be the last resort, as it is associated with higher morbidity and mortality.

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