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RESEARCH ARTICLE

AN UNUSUAL CAUSE OF BOWEL OBSTRUCTION

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Abstract

Background: Although gastrointestinal issues and bowel blockages can appear at any age in individuals with cystic fibrosis (CF), acute intestinal obstructions after the neonatal stage are relatively rare. Distal intestinal obstruction syndrome (DIOS) is more likely to develop in cystic fibrosis patients who have pancreatic insufficiency, a prior history of meconium ileus, or 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 non-biliousvomitings. On examination: the abdomen was soft and distended, and a mass in RIF could be palpated, the Rest of the General physical and systemic examinations were 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 recognized complication of cystic fibrosis. It results from the accumulation of viscid fecal matter mixed with thick, sticky intestinal mucus. This mixture typically adheres to the intestinal wall—most often in the terminal ileum and caecum—causing a fixed, difficult-to-resolve obstruction.Once DIOS is diagnosed, the primary objective is prompt relief of the obstruction. Medical treatments, including osmotic contrast enemas such as Gastrografin, are often effective. Surgical intervention is generally reserved for refractory cases due to its high post-operative morbidity.

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. (1,2)

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 inspissatedfecal material.
The incidence of DIOS with complete intestinal obstruction studied in children was found to be between5 and 12 episodes per 1000 patients per year throughoutEurope. (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 in15-month-old child who was managed conservatively.

Case presentation:
Our patient was a 15-month-old baby, whowas 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 vomitings 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 as shown in (FIG-1)

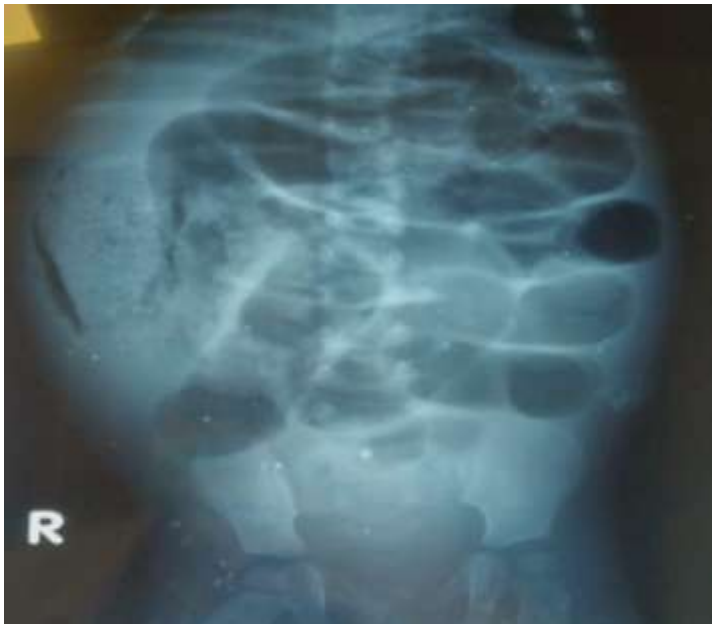


Fig-1:-Plain X-ray abdomen supine showing fecal impaction on the right side.

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 rectal 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)

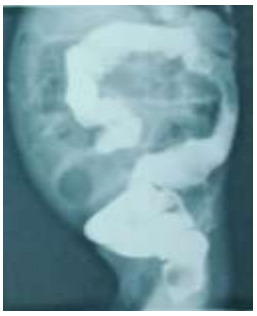
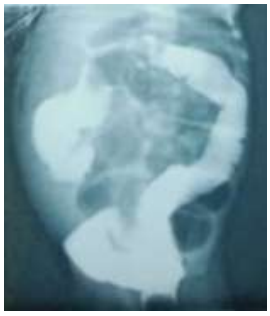
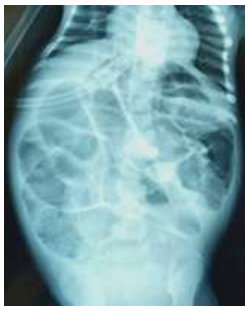
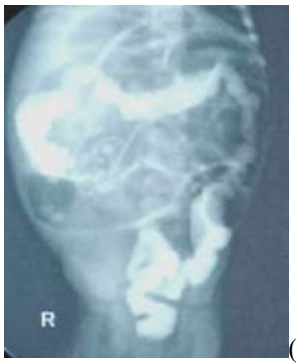
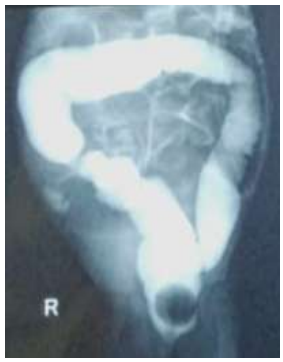
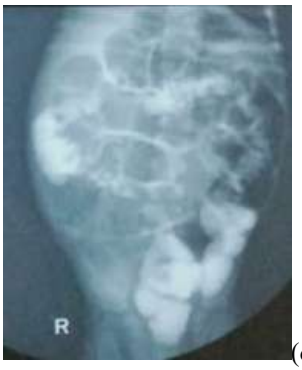
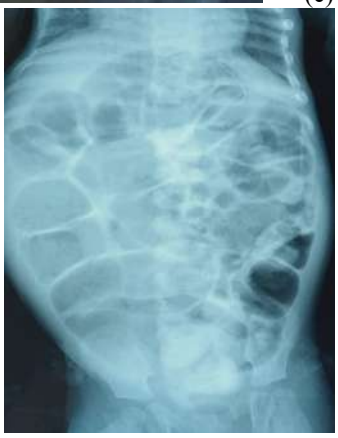
(a)  (b)  (c) 

Fig. 2:-

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

(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 well-documented gastrointestinal complication affecting individuals with Cystic Fibrosis (CF). It results from the buildup of abnormally thick stool mixed with tenacious mucus, which becomes lodged in the bowel—most often in the terminal ileum and caecum. This material tends to stick firmly to the intestinal walls and embeds within the lining structures such as crypts and villi, making it especially difficult to clear. (1,3)

Clinical Classification

DIOS may present in two distinct forms—complete and incomplete obstruction. Guidelines developed by the CF Working Group under the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition (ESPGHAN) outline clear diagnostic criteria:

Complete DIOS is marked by a full blockage of the intestine. Affected patients often show signs such as bilious vomiting, cramping abdominal pain, significant bloating, and a palpable mass in the lower right abdomen. Imaging typically reveals small bowel dilation and multiple fluid levels.

Incomplete DIOS presents more mildly. While patients may experience abdominal discomfort, bloating, and a detectable mass near the ileocecal region, symptoms like bilious vomiting and air fluid levelson X-rays are absent. (2)

Differentiating from Constipation:

Constipation can mimic the symptoms of DIOS but usually develops more gradually over weeks or months. It involves infrequent bowel movements, hard stools, and abdominal discomfort that generally improves with laxatives. Unlike DIOS, constipation doesn't typically result in acute obstruction or require urgent intervention. (5)

Epidemiology

The incidence of DIOS in individuals with CF is estimated to be around 6.2 cases per 1,000 patient years. This figure is higher than earlier studies, such as one by Andersen and colleagues, which reported an incidence of 2.5 per 1,000 patient-years in CF patients under 20 years old. (1,3)

Diagnosis

Abdominal X-rays in both complete and incomplete forms of DIOS often reveal dense stool accumulation in the lower right abdomen, specifically in the terminal ileum and caecum. Proximal bowel loops may be distended, sometimes with fluid and gas, depending on the severity of the obstruction. (3,4)

Treatment Options

Most cases of DIOS respond well to non-surgical management:

For mild to moderate cases, the preferred treatment includes oral hydration paired with osmotic agents such as polyethylene glycol (PEG). PEG is typically dosed at 2 grams per kilogram per day, with a maximum dose of 80–100 grams. Alternatively, PEG solutions can be administered at 20–40 ml/kg/hour, up to 1 liter per hour, for several hours until stool output becomes clear and symptoms resolve. (2)

Gastrografin, a contrast agent with laxative properties, may also be used. This can be given orally or through a nasogastric tube—typically 50 ml diluted in 200 ml of fluid for children under six years, and 100 ml in 400 ml for older children and adults. Subsequent doses may be reduced depending on the clinical response. (2,3)

Other therapies, such as N-acetylcysteine, have largely fallen out of favor due to limited effectiveness. While prokinetic agents are sometimes used in other bowel motility disorders, they have not proven effective in treating DIOS.

Severe Cases and Hospitalization

If the obstruction leads to persistent vomiting—especially if bilious—or does not improve with oral therapies, hospital admission is needed. Management should include intravenous fluids and decompression with a nasogastric tube. In such cases, Gastrografin enemas (typically diluted 1:4 with water) may be given under imaging guidance to reach the terminal ileum.An alternative approach is to directly instill the contrast into the caecum via colonoscopy. (1,3)

Surgical Considerations

Surgery is rarely needed and generally reserved for cases where medical treatment has failed. The first surgical step is usually bowel irrigation via a small surgical opening (enterostomy). Resection of the affected bowel, including the ileocecal region, should only be considered in extreme situations due to the higher risk of postoperative complications. (3,4)

Conclusion:-

DIOS represents a significant gastrointestinal challenge in patients with CF, particularly during adolescence and adulthood. It arises from the accumulation of thick secretions in the lower intestine and can lead to partial or complete blockage. Early diagnosis and aggressive medical therapy are usually effective. Surgical intervention is reserved for resistant cases and should be approached with caution due to associated risks.

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