Original Research Article

Intestinal obstruction in cystic fibrosis: a surgeon’s perspective

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ABSTRACT

Background: Distal intestinal obstruction syndrome (DIOS) is one of the commonest gastrointestinal manifestation of Cystic Fibrosis. It occurs as a result of impaction of mucofeculent material in the ileocaecum and ascending colon. Themedical management is effective in the chronic presentation, but the optimum management in the acute presentation is still a grey area. The aim of this study is to retrospectively follow up patients presenting with features of DIOS who had surgical resection of the ileocaecum to assess the long-term outcome in terms of recurrence of symptoms.

Methods: Case notes of eight patients presenting with features of acute DIOS between 2000 and 2006 were retrieved from the Cystic fibrosis unit of a tertiary teaching hospital and analysed.

Results: Eight patients were included in this study, male: female ratio of 5:3 age range: 18-24. Acute presentation: 6, elective presentation: 2. The main presenting features were abdominal pain, constipation, vomiting and RIF mass. Six patients had ileocaecectomy; one had a small bowel resection and one an extended right hemicolectomy. Six patients had no further symptoms of DIOS four years post-surgery, two had further symptoms with one requiring further surgery.

Conclusions: Ileocaecal resection in patients presenting with acute DIOS is a viable and effective option in management.

Keywords: Cystic fibrosis, DIOS, Intestinal obstruction, Surgical management

INTRODUCTION

Cystic fibrosis (CF) is the commonest life shortening autosomal recessive disorder seen in the white population with an incidence varying between 1 in 2000 to 1 in 3000 live births. CF was recognised as a childhood condition with a high mortality rate due to respiratory complications. However, advances in the medical management of the respiratory complications of cystic fibrosis have considerably improved the quality of life and longevity of these patients who now live well into the third and fourth decades of life. This has brought forth newer challenges in the management of gastrointestinal complications in the adult cystic fibrosis patient. Distal intestinal obstruction syndrome (DIOS) or meconium ileus equivalent is one of the commonest gastrointestinal conditions in this group of patients. DIOS is a condition unique to cystic fibrosis and affects 10 to 20% of adult cystic fibrosis patients (Peckam et al). It occurs due to acute complete impaction of abnormally viscous muco-faeculent material in the terminal ileum, caecum and ascending colon. Although DIOS occurs commonly in CF patients with pancreatic insufficiency, it has also been described in patients who have normal pancreatic function (Clifton et al). Other factors contributing to DIOS include reduced intestinal water content, lower luminal acidity of the foregut, accumulation of...
intraluminal macromolecules, dehydration of the mucus layer due to altered intestinal secretion, fat malabsorption, low dietary fibre intake, anticholinergic drugs and slow intestinal transit time.\textsuperscript{2,5} It is also known to be common post lung transplantation.\textsuperscript{6}

DIOs can present acutely with signs and symptoms of small bowel obstruction and/or peritoneal irritation, or in a chronic fashion with recurrent colicky abdominal pain, bloating, nausea and anorexia. Although medical management of this condition is well documented, the optimal surgical strategy for this condition is not known. Authors retrospectively reviewed eight such cases managed successfully in the unit. The aim was to assess the presenting features, investigation modalities and surgical options in patients presenting acutely with this condition.

**METHODS**

This was a review of a prospectively collated data between January 2000 and December 2006 at the Cystic Fibrosis unit of the St James University Hospital, Leeds. A total of eight patients who presented with features suggestive of DIOs were enrolled in this study.

**Inclusion criteria**

- Patients with features of intestinal obstruction with a history of Cystic Fibrosis
- Patients older than 18 years.

**Exclusion criteria**

- Patients below 18 years
- Patients without history of Cystic fibrosis
- Patients with no documented follow up.

All patients enrolled in the study had data regarding demographics, presenting features, investigating modalities and treatment options analysed. Follow up outcome data was also collated for review.

**RESULTS**

Twelve patients were identified. However, complete data was available on only eight patients. Male to female ratio was 5:3 and age range in years was 22-36. Presenting symptoms ranged from abdominal pain, vomiting and constipation while examination revealed a mass in the right iliac fossa in five patients. Six were acute admissions while two had chronic symptoms. All patients were on pancreatic supplements. Five patients had surgery for meconium ileus as neonates while one had surgery for duodenal atresia. All patients had a Computerised Tomography Scan of the abdomen as the definitive investigation of choice. Conservative management was attempted in five patients without success. All patients underwent surgery. Six had an ileo-caecectomy, one had an extended right hemicolectomy and one had a small bowel resection. Although all patients were discharged successfully from the hospital, discharge was delayed in three patients due to respiratory complications. At follow-up, six patients had no further symptoms of DIOs. Two patients presented again, one of whom required re-do surgery (Table 1).

**DISCUSSION**

Diagnosis of DIOs requires awareness of the condition and a high index of suspicion in patients with CF. Meticulous clinical examination and appropriate investigations confirm the diagnosis in most cases.

Presenting features include right lower quadrant pain and or mass and this makes it difficult to differentiate from conditions like appendicitis, intussusception, inflammatory bowel disease, adhesional bowel obstruction or simple constipation. Patients can also present acutely with signs and symptoms of small bowel obstruction. Majority of patients in this study presented acutely with colicky abdominal pain, constipation, vomiting and right iliac fossa mass. One patient was clinically diagnosed as appendicitis but was found to have features of DIOs at surgery.

The investigative work up for both acute and sub-acute presentations includes baseline full blood count, urea and electrolyte and CRP. Plain abdominal X-rays show faecal loading in the right lower quadrant, ileal dilatation and collapsed distal bowel. It however has limited value in diagnosing this condition, and also has a poor exclusion value for other causes of acute abdomen. CT is a useful tool since it helps in the diagnosis and exclusion of other conditions. The classic feature of DIOs on CT is a small bowel filled with a homogenous mass, increasing in opacity from the duodenum to the right hemi colon suggesting increasing viscosity of intestinal content.\textsuperscript{5}

Medical management is the preferred option in both modes of presentation because of the increased operative morbidity in these patients. The increased morbidity is due to poor nutritional status, CF associated poor pulmonary function and long-term corticosteroids use.

Non-operative treatment in the chronic form of the condition involves dietetic management, adequate fluid intake, pancreatic enzyme dose adjustment and reduction of gastric acid secretion in order to improve enzyme efficacy with PPI.\textsuperscript{7,\textsuperscript{8}} Patients with mild acute exacerbation can usually be treated with laxatives, gastrografin and kleen prep.\textsuperscript{2} However, in patients who do not respond to this, there is a place for more invasive treatment which includes instillation of gastrografin (500 ml of 50%) via colonoscopy, modified ACE procedure and open disimpaction of the stool in addition to a modified ACE.\textsuperscript{9,\textsuperscript{10}} In the acute setting, the initial management involves potent analgesia, adequate intravenous fluid resuscitation, and nasogastric aspiration. Although
Shidrawi et al recommended instillation of gastrograffin at colonoscopy to resolve the symptoms, it is the authors’ view that passing a colonoscope into the large bowel in a setting of potential bowel ischaemia is a way of courting disaster. The failure of medical management in the acute setting mandates a laparotomy because of the risk of bowel ischaemia and perforation. The preferred operative option is manual disimpaction by milking the contents antegrade while administering warm isotonic saline mixed with mineral oil through a nasogastric tube. If this fails enterotomy to evacuate the inspissated faecal matter or primary ileo-caecal resection and anastomosis are required. The enterotomy is made 10 cm from the impacted column of faeces and then either closing it primarily or over a T-tube that can be used as a controlled fistula or for instillation of pancreatic enzymes until it passes freely into the colon.

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<th>Table 1: Study data.</th>
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Although Speck and Charles have suggested that primary resection and anastomosis is rarely required, the results of this study suggest that a limited ileocaecal resection with stapled side to side anastomosis is a relatively safe and viable treatment option for this condition and has a significant positive long-term outcome in terms of recurrence of symptoms.  

Although it is clear that operating on this group of patients is both an anaesthetic and surgical challenge, scrupulous preoperative workup including fluid resuscitation, acid base and electrolyte normalisation, and early involvement of the chest physician will produce a favourable postoperative outcome. Adequate fluid balance, chest physiotherapy, early mobilisation and prevention of pneumonia are essential for a favourable postoperative outcome.

Due to the advances in the medical management of pulmonary complications of CF, it is likely that DIOS would be a more frequent presentation in the adult CF patient in the future. It is therefore imperative for surgeons to have a high index of suspicion for DIOS in patients with CF and be aware of the available management strategies.

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**Ethical approval:** The study was approved by the Institutional Ethics Committee

**REFERENCES**

2. Peckam D, Morton A. Treatment of distal intestinal obstruction syndrome. Available at: www.cysticfibrosismedicine.com  

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