Original Research Article

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Intestinal obstruction in cystic fibrosis: a surgeon's perspective

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ABSTRACT

Background: Distal intestinal obstruction syndrome(DIOS) is one of the commonestgastrointestinal manifestation of Cystic Fibrosis. It occurs as a result ofimpaction of mucofeculent material in the ileocaecum and ascending colon. Themedical management is effective in the chronic presentation, but the optimummanagement in the acute presentation is still a grey area. The aim of this study is to retrospectively follow up patients presenting withfeatures of DIOS who had surgical resection of the ileocaecum to assess thelong-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 5:3age range: 18-24. Acute presentation: 6, elective presentation:2. The mainpresenting features were abdominal pain, constipation, vomiting and RIF mass.Six patients had ileocaecectomy; one had a small bowel resection and one anextended right hemicolectomy.Six patients had no further symptoms of DIOS four years post-surgery, two hadfurther symptoms with one requiring further surgery.

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

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

INTRODUCTION

Cystic fibrosis (CF) is the commonest life shortening autosomal recessivedisorder seen in the white population with an incidence varying between 1 in 2000 to 1 in 3000 live births.1 CF was recognised as a childhood condition ahigh mortality rate due respiratory to complications. However, advances in the medical management of the respiratory complicationsof 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 gastrointestinalassociated complications in the adult cystic fibrosis patient. Distal intestinalobstruction syndrome (DIOS) or meconium ileus equivalent is one of thecommonest gastrointestinal conditions in this group of patients.²⁻⁴DIOS is a condition unique to cystic fibrosis and affects 10 to 20% of adult cysticfibrosis patients (Peckam et al). It occurs due to acute complete orincomplete impaction of abnormally viscous mucofaeculent material in theterminal ileum, caecum and ascending colon.^{2,3} Although DIOS occurscommonly in CF patients with pancreatic insufficiency, it has also beendescribed 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

intraluminalmacromolecules, dehydration of the mucus layer due to altered intestinalsecretion, fat malabsorption, low dietary fibre intake, anticholinergic drugs andslow intestinal transit time.^{2,5} It is also known to be common post lungtransplantation.⁶

DIOS can present acutely with signs and symptoms of small bowel obstructionand/or peritoneal irritation, or in a chronic fashion with recurrent colickyabdominal 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 onlyeight patients. Male to female ratio was 5:3 and age range in years was 22-36. Presenting symptoms ranged from abdominal pain, vomiting and constipationwhile examination revealed a mass in the right iliac fossa in five patients. Sixwere acute admissions while two had chronic symptoms. All patients were onpancreatic supplements. Five patients had surgery for meconium ileus asneonates 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, dischargewas 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 thismakes it difficult to differentiate from conditions like appendicitis, intussusception, bowel disease, adhesional bowel inflammatory obstruction or simple constipation. Patients can also present acutely with signs and symptomsof small bowel obstruction. Majority of patients in this study presented acutelywith colicky abdominal pain, constipation, vomiting and right iliac fossa mass.One patient was clinically diagnosed as appendicitis but was found to havefeatures of DIOS at surgery.

The investigative work up for both acute and sub-acute presentations includesbaseline full blood count, urea and electrolyte and CRP. Plain abdominal X-rayshows faecal loading in the right lower quadrant, ileal dilatation and collapseddistal bowel. It however has limited value in diagnosing this condition, and alsohas a poor exclusion value for other causes of acute abdomen. CT is a usefultool 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.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 dieteticmanagement, adequate fluid intake, pancreatic enzyme dose adjustment andreduction of gastric acid secretion in order to improve enzyme efficacy with PPI. 7.8 Patients with mild acute exacerbation can usually be treated with laxatives, gastro graffin and kleen prep. 2 However, in patients who do not respond to this, there is a place for more invasive treatment which includes instillation of gastro graffin(500ml of 50%) via colonoscope, modified ACE procedure andopen disimpaction of the stool in addition to a modified ACE. 9.10 In the acute setting, the initial management involves potent analgesia, adequateintravenous fluid resuscitation, and nasogastric aspiration. Although

Shidrawi etal recommended instillation of gastrograffin at colonoscopy to resolve thesymptoms, it is the authors' view that passing a colonoscope into the largebowel in a setting of potential bowel ischaemia is a way of courting disaster. The failure of medical management in the acute setting mandates a laparotomybecause of the risk of bowel ischaemia and perforation. The preferred operativeoption is manual disimpaction by milking the contents antegradely while administering warm isotonic

saline mixed with mineral oil through a nasogastrictube. ¹¹ If this fails enterotomy to evacuate the inspissated faecal matter orprimary ileocaecal resection and anastomosis are required. ¹² The enterotomy ismade 10cm from the impacted column of faeces and then either closing itprimarily or over a T-tube that can be used as a controlled fistula or forinstillation of pancreatic enzymes until it passes freely into the colon. ¹³

Table 1: Study data.

Age	Presenting features	Acute/ chronic	РМН	Imaging	Cons MGT	Surgical MGT	Histology	Long term
22	Colicky abdominal pain, vomiting, constipation 5/7	Acute	PEG for supplemental feeding	CT Abd	Gastrograffin enema, kleenprep	Ileo- caecectomy	Intussusception features suggestive of DIOS	No further symptoms of DIOS 4 years post op
27	Central abdominal pain, vomiting, distension 4/7	Acute	Small bowel resection for Meconium ileus as neonate	Plain AXR CT Abd	NBM, IVI, NG tube. No improvement after 5/7	Small bowel resection	Features suggestive of DIOS	No further symptoms of DIOS 4 years post op
27	Constipation, vomiting RIF pain 2/7	Acute	Meconium ileus surgery as neonate, CRF Lung transplant	CT Abd	nil	Ileo- caecectomy	Normal appendix features suggestive of DIOS in ileo- caecum	No further symptoms of DIOS 4 years post op
31	RIF mass, weight loss, iron deficiency anaemia	Elective	Appendectomy	CT Abd-	nil	Ileo- caecectomy	IleocaecalCrohns	No further symptoms of DIOS 4 years post op
26	Constipation	Acute	Meconium ileus surgery as neonate	CT Abd	Cleanprep Picolax Gastrograffin	Ileo- caecectomy	Features consistent with DIOS	No further symptoms of DIOS 4 years post op
26	Central abdominal pain, constipation vomitting	Acute	Meconium ileus IDDM, ACE procedure for recurrent DIOS	CT Abd	NBM, IVI, NG tube	Ileo- caecectomy	Features consistent with DIOS	No further symptoms of DIOS 5yrs post-surgery
25	Colicky abdominal pain, vomitting	Acute	Duodenal atresia Appendectomy volvulus modified ACE	CT Abd	Gastrograffin	Ileo- caecectomy	Features consistent with DIOS	Presented 1 year later with another episode of DIOS refractory to conservative management. Subsequently had ileal resection
36	RIF pain, weight loss, constipation		Meconium ileus surgery as neonate	CT Abd	nil	Extended right hemicolectomy	Yersinia enterocolitis	Intemittent abdominal pain

Although Speck and Charles have suggested that primary resection andanastomosis 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-termoutcome in terms of recurrence of symptoms.¹²

Although it is clear that operating on this group of patients is both an anaesthetic and surgical challenge, scrupulous preoperative workup including fluidresuscitation, acid base and electrolyte normalisation, and early involvement of the chest physician will produce a favourable postoperative outcome. Adequatefluid 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 CFpatient in the future. It is therefore imperative for surgeons to have a high indexof suspicion for DIOS in patients with CF and be aware of the availablemanagement strategies.

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Institutional EthicsCommittee

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