Emergency colonoscopy for distal intestinal obstruction syndrome in cystic fibrosis patients

Recent advances in the treatment of cardiopulmonary complications in patients with cystic fibrosis (CF) have improved median survival and resulted in gastrointestinal complications becoming more evident. The distal intestinal obstruction syndrome (DIOS) develops due to the accumulation of viscid mucophaeulent material in the terminal ileum and right hemicolon. Reduced intestinal water content, lower luminal acidity of the foregut, and accumulation of intraluminal macromolecules contribute to the development of DIOS. Typically, patients develop progressive symptoms and signs of small bowel obstruction. A plain abdominal x-ray will reveal dilated loops of centrally placed small bowel, with faecal loading at the terminal ileum and right hemicolon (see fig 1). Most cases will respond to the aggressive use of laxative agents administered orally and rectally, intravenous hydration, pancreatic supplements, and N-acetylcysteine (Parvolex) and gastrografin (meglumine diatrizoate) orally or per rectum. Surgical decompression has been reserved for refractory cases but carries a high operative mortality. Colonic resection is contraindicated in patients with significant malabsorption, as colonic butyrate metabolism contributes significantly to caloric intake in these patients.

The following experience using emergency colonoscopy as a relatively noninvasive technique suggests surgery may often be avoided in this high risk cohort. Colonoscopy may also help distinguish DIOS from colonic strictures due to fibrosing colopathy, intussusception, and colonic malignancies, all recognised complications in CF patients.

**PATIENTS AND METHODS**

We reviewed our experience of 550 patients at the Royal Brompton Hospital Cystic Fibrosis Unit between January 1991 and January 1999. One hundred and eight patients (20%) were found to complain of isolated or repeated episodes of abdominal pain, distension, and constipation. Eleven patients (2%) (five males, six females) with a mean age of 27.6 years (range 18–42) developed sixteen episodes of DIOS with radiographic evidence of small bowel dilatation refractory to medical therapy. Each DIOS episode was precipitated by an intercurrent respiratory infection, dehydration, and relative immobility, associated with poor dietary intake. Five patients were heart/lung recipients and two patients were awaiting transplantation. All patients had significant cardiopulmonary comorbidity and were assessed as high risk candidates unsuitable for surgery.

An experienced colonoscopist carried out emergency colonoscopy once the patients became refractory to standard medical therapy. The procedures were technically demanding but were well tolerated (duration 60–120 minutes). Patients underwent benzodiazepine sedation and opiate analgesia under continuous pulse oximetry monitoring. Most patients were profoundly hypovolemic with capillary oxygen saturations of 70–80% on 8 l/min via nasal cannulae prior to the procedures. Standard Olympus CF200 colonoscopes (KeyMed, UK) were used. A total of 500 ml of 50% gastrografin was instilled into the lumen at the limit of the examination. Continued intravenous hydration and vigorous laxative therapy were maintained following each procedure to ensure a complete response.

**RESULTS**

The caecum was visualised in 13/16 episodes, including terminal ileoscopy on three occasions and limited to the ascending colon for the remaining episodes. Despite the patients’ poor cardiopulmonary function, the procedures were well tolerated and no procedure related complications were noted. A satisfactory result, determined by resolution of symptoms and radiographic changes, was observed in 14/16 episodes. One patient required four colonoscopies over a period of 16 months for recurrent DIOS episodes. Another patient ultimately underwent a colectomy and ileorectal anastomosis. Two patients died of multiorgan failure with persisting intestinal obstruction. Four further patients have since died of unrelated causes.

**DISCUSSION**

The incidence of DIOS in our CF patients was similar to that reported by other workers. All but two of our patients (82%) with refractory DIOS were on pancreatic supplements but pancreatic exocrine deficiency is not always a prerequisite. As reported in other series, the development of neonatal meconium ileus did not appear to be a predictor of the future development of DIOS in our CF patients.

Gastrografin is an osmotically active hypertonic agent that stimulates colonic peristalsis. The use of oral and rectal gastrografin in the diagnosis and treatment of DIOS has been reported by several workers but this is the first report of the use of gastrografin instillation at colonoscopy for the treatment of DIOS in CF patients.

In our experience, this technique proved safe and was relatively well tolerated in this cohort. Emergency colonoscopy, undertaken early in patients with progressive symptoms who prove refractory to medical therapy, is a novel and effective modality of therapy and avoids the need for surgical intervention.

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Figure 1 Plain abdominal x ray identifying the features of the distal intestinal obstruction syndrome in a patient with cystic fibrosis.

Note the dilated loops of small bowel, with shadowing in the right iliac fossa indicating ininspiated mucosal faecalulent material contents within the terminal ileum.
REFERENCES