Case report

Treatment of resistant distal intestinal obstruction syndrome with a modified antegrade continence enema procedure

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Abstract

We report a case of a patient with CF who had a long history of recurrent distal intestinal obstruction syndrome. She had been treated with conventional treatment including gastrografin, n-acetyl cysteine, Klean prep and Picolax.

She underwent a modified antegrade continence enema procedure. She currently irrigates her conduit every 2–3 days. She has had no further symptoms of distal intestinal obstruction syndrome.

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1. Case report

A 17-year-old woman with cystic fibrosis presented to the Leeds Adult Cystic Fibrosis Unit at regular intervals with severe abdominal pain due to distal intestinal obstruction syndrome (DIOS). Abdominal X-rays at the time of presentation showed evidence of faecal loading in the caecum and the ascending colon. She had been diagnosed as having cystic fibrosis (CF) during the neonatal period and genetic screening has shown her to be homozygous for the ΔF508 mutation. She was born with duodenal atresia and had a laparotomy for this shortly after birth. She underwent a further laparotomy in 1997 for obstruction secondary to adhesions. During this laparotomy she had adhesiolysis and an appendectomy.

Prior to a modified antegrade continence enema (ACE) procedure, the patient’s height was 1.56 m, weight 48.5 kg and body mass index (BMI) 19.9 kg/m². She was pancreatic insufficient and was taking pancreatic enzyme replacement therapy in the form of Creon 10000 (Solvay Healthcare). She was not diabetic, had not received nasogastric or gastrostomy feeding, but did take nutritional supplements orally as part of her calorie intake.

Assessment of absorption was not carried out quantitatively but faecal fat microscopy was negative for both neutral and split fats. Faecal chymotrypsin was 633 mcg/g (normal>120 mcg/g). This had improved from 3 years previously when fat microscopy indicated neutral fat +++, split fat ++. At this time faecal chymotrypsin was <60 mcg/g suggesting poor adherence to enzyme therapy. Dietetic education was implemented at that time and she had undergone regular review.

The reported dose of pancreatic enzyme replacement therapy was less than 2100 IU lipase/kg/meal and approximately 4800 IU lipase/kg/day, a level that is well within the recommended range being neither excessively high nor low for a patient of this age. In addition the patient was receiving regular omeprazole (AstraZeneca) as an adjunct to enzyme therapy.

The patient was chronically infected with Pseudomonas aeruginosa and received regular courses of home and hospital intravenous antibiotics. Her lung function was stable with a baseline FEV₁ and FVC of 1.63 L/s (56% predicted) and 2.8 L (92% predicted), respectively.
The patient had been presenting with episodes of DIOS for over 10 years but the frequency of admissions had increased significantly. In February 2001 she presented with DIOS that was successfully treated with both gastrografin (Schering) orally and via enemas. Over the next few months the patient had repeated admissions to hospital with recurrent symptoms of DIOS. She was treated with gastrografin (Schering), n-acetyl cysteine (Celltech), Klean Prep (Norgine), and Picolax (Ferrings Pharmaceuticals). A barium enema was undertaken which revealed no colonic strictures or obstructing lesions. The patient was also started on a low residue, low fat diet. In spite of all these interventions the patient’s symptoms continued unabated.

Following extensive discussion with the patient and counselling by the specialist stoma nurses, a modified ACE procedure was undertaken in April 2002. The terminal ileum was used to create the irrigation conduit. The proximal end of the ileum was anastomosed to the ascending colon (Fig. 1). This was necessary as the patient’s appendix was unavailable to be used as a conduit. If her appendix had been present then a laproscopic ACE procedure could have been undertaken. The patient made an unremarkable post-operative recovery and was discharged home.

She is currently irrigating using tap water every 2–3 days depending upon her dietary intake. Following irrigation she has good bowel movements. The patient is currently making good progress and has had no further episodes of DIOS.

2. Discussion

DIOS develops in patients with CF due to the accumulation of viscous mucous and faecal material in the terminal ileum, caecum and ascending colon [1]. DIOS results in symptoms of recurrent abdominal pain, bloating, nausea and anorexia, and can lead to small intestine obstruction. The incidence of DIOS in patients with CF is about 15% [2]. Initially DIOS was only described in pancreatic insufficient patients with CF, however it has been since described in patients with normal pancreatic function [3,4]. Approximately 11% of all patients with CF have no symptoms of malabsorption [2].

Conventional treatment of DIOS involves rehydration (intravenous fluids if necessary) and the use of oral gastrografin (Schering) [1]. Alternatively the intestines can be flushed out using a balanced electrolyte solution such as Klean-Prep (Norgine) [3] which may need to be given orally or via a nasogastric tube. In the more severe case, DIOS can be successfully treated using gastrografin (Schering) directed into the lumen of the ascending colon by means of either an enema [1] or colonoscopy [5]. In a minority of patients conventional therapy may fail and results in repeated admissions, chronic abdominal symptoms and a reduction in quality of life.

It has been suggested that the pro-kinetic drug, cisapride (Janssen-Cilag) can be used to improve the symptoms of chronic DIOS, although it does not abolish the need for intestinal lavage [6]. The use of Cisapride (Janssen-Cilag) has been discontinued in many CF centres following the suspension of the product licence due to concern about prolongation of the QT interval and ventricular arrhythmias.

A number of studies have suggested that DIOS occurs more frequently in patients on inadequate pancreatic enzyme replacement [7]. Assessment of pancreatic enzyme therapy and knowledge of enzyme titration should be assessed by an experienced CF dietitian [8]. This is essential when patients present with recurring DIOS. Reduction of gastric acid or acid suppression as a means of improving enzyme efficacy with a proton pump inhibitor has been reported and proton pump inhibitors are often used [7].

Faecal chymotrypsin is low in untreated pancreatic insufficient patients with CF and can be useful for monitoring enzyme therapy with low values suggesting inadequate therapy possibly due to poor adherence [8]. Whilst levels had been low in this patient in the preceding years there had been improvement in adherence to therapy and the patient’s knowledge of enzyme titration through education.

In addition to assessment of enzyme therapy, assessment of malabsorption and/or steatorrhoea is valuable. The 72 h faecal fat collection is the gold standard for assessing fat absorption however faecal fat microscopy has been shown as a useful tool to detect steatorrhoea [9]. Fat microscopy shows some correlation between microscopic grading and severity of steatorrhoea and has been validated by comparison with quantitative measurements [10].

The ACE procedure was initially described for the treatment of intractable constipation or faecal incontinence in children [11,12]. It has since been used to treat adults with both constipation and faecal incontinence [13]. The underlying diseases that have necessitated an ACE procedure include anorectal malformations, Hirschsprung’s disease, spina bifida, spinal injuries, severe perineal injury, neuropathic disorders, cerebral palsy, and idiopathic constipation [14]. The procedure involves bringing the appendix out onto the abdominal wall to create a conduit resulting in a small stoma. Modifications to the ACE procedure have been undertaken where the appendix is not available using...
tubularised caecum, ileum or defunctionalised caecum to create the irrigation conduit [14]. In a retrospective review of the ACE procedure using either ileum or appendix to create the irrigation conduit there was no difference in compliance or continence rates [15].

There is a possibility that the disruption of the ileo-caecal valve by the modified ACE procedure is of importance in this patient. The terminal ileum is important in the development of DIOS. However in this patient the faecal material was in the caecum and ascending colon. We feel that irrigation of the terminal ileum via the conduit is unlikely, but the only way to determine whether this occurs is to undertake a contrast study which would involve radiation exposure to the patient and would be of academic interest only.

To our knowledge this is the first report of a modified ACE procedure being used for the treatment of DIOS in a patient with cystic fibrosis. The procedure provides an alternative approach when conventional treatment has been unsuccessful or is not tolerated.

References