CASE REPORT

GASTROGRAFFIN USE IN DISTAL INTESTINAL OBSTRUCTION SYNDROME OF CYSTIC FIBROSIS

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Background: Meconium ileus equivalent or the Distal Intestinal Obstruction Syndrome (DIOS) is an entity that is frequently seen in patients with cystic fibrosis (CF). In the gastrointestinal tract cystic fibrosis may produces symptoms of malabsorption and constipation. The latter may be associated with complications such as impaction and intestinal perforation. Meconium ileus and DIOS are an emergency and requires to be treated aggressively to ensure that consequences such as perforation do not occur. **Method:** Traditional modes of management of DIOS have included the use of laxatives, Acetylcysteine and Gastrograffin enemas. We are describing the use of oral gastrograffin in our patient seen at the Aga Khan University Hospital where small bowel obstruction was refractory to treatment. Oral gastrograffin was used once diluted in 4 times the volume of water or fruit juice with half doses given on day 2 and 3. **Results:** Oral gastrograffin use was followed by relief of obstruction in this patient. **Conclusion:** Gastrograffin use orally or rectally may be helpful in the treatment of refractions distal intestinal obstruction syndrome in cystic fibrosis.

Keyword: Gastrograffin, Intestinal, Syndrome, Cystic fibrosis

INTRODUCTION

Meconium ileus equivalent or the Distal Intestinal Obstruction Syndrome (DIOS) is an entity that is frequently seen in patients with cystic fibrosis (CF). The defect in CF lies in the cystic fibrosis transmembrane receptor (CFTR) that results in defective passage of salt and water, manifesting as pulmonary and pancreatic insufficiency. In the gastrointestinal tract the disease may produces symptoms of malabsorption and constipation with complications such as impaction and intestinal perforation. Meconium ileus is a surgical emergency and requires to be treated aggressively to ensure that consequences such as perforation do not occur.

Manifestations range from mild abdominal pain usually localized in the right lower abdominal quadrant, to severe pain with abdominal distension and air-fluid levels on abdominal X-ray.

Traditional modes of management have included the use of laxatives, Acetylcysteine and Gastrograffin enemas. We are describing the use of oral gastrograffin in our patient where small bowel obstruction was refractory to treatment

CASE REPORT

FZ is a 3 year old patient with CF and with pulmonary and pancreatic insufficiency. She presented to the emergency room with severe abdominal pain, respiratory distress and fever. Her parents noted that she had not passed any stool since 2 days prior to presentation and had had several episodes of vomiting as well. Her emesis was noted to be bilious and foul smelling.

On examination she was found to have severe abdominal distention with tenderness on palpation. The abdomen was tense with visible bowel loops and hypoactive gut sounds. She had coarse crackles on auscultation of the chest, dry mucous membranes and poor skin turgor.

An X ray abdomen revealed multiple dilated bowel loops with several air fluid levels that suggested small bowel obstruction.

Laboratory investigation demonstrated hyponatremia with a serum sodium of 131 mEq/litre, potassium 3.3 mEq/litre, serum albumin of 2.7 mg/dl and normal serum calcium, magnesium and phosphorus. A complete blood count was normal except for a hemoglobin of 9 g/l.

She was rehydrated with intravenous fluids and received saline enemas for disimpaction. The saline enemas were not successful in relieving the impaction and were replaced with rectal acetylcysteine instillation. There was no evacuation of stool despite repetition of rectal acetylcysteine. Oral acetylcysteine was then instilled via an nasogastric tube. Her condition did not improve and the abdominal distention persisted. She was then given a gastrograffin enema which resulted in the passage of a tiny amount of stool. This was not effective in clearing the obstruction despite repetition. The gastrograffin was then orally administered via an nasogastric tube and lead to successful disimpaction and relief of obstruction.

DISCUSSION

Meconium ileus equivalent or the Distal Intestinal Obstruction Syndrome is an entity frequently seen in

patients with cystic fibrosis. This disorder is one of the most common potentially lethal autosomal recessive diseases associated with pulmonary and pancreatic insufficiency[1,2]. The defect lies in the cystic fibrosis transmembrane receptor (CFTR) and results in defective passage of salt and water and hence the development of thick and viscid secretions. In the intestine this manifests as constipation and frequently impaction. This is particularly so during hot weather when there is significant salt and water loss. While pancreatic insufficiency plays a role, DIOS has also been seen in pancreatic sufficient patients and other factors, such as enhanced viscosity of intestinal mucus, or prolonged intestinal transit time may also be important.

Severe impaction may lead to intestinal obstruction, similar to that seen with meconium ileus and may progress to intestinal perforation. This is a surgical emergency and requires aggressive management.

Data from 10 studies demonstrates the mean prevalence rate of DIOS to be 8.3% [3]. A single center study from Denmark however obtained an overall incidence of 5 episodes per 1000 patient/years[4]. A recent study by Dray et al describes a prevalence of 18%, based on the occurrence of at least one DIOS episode in 31 out of 171 adult CF patients[5]. The majority of those patients experienced the first episode after the age of 18 year, and approximately half of the patients had more than one episode. All the patients with DIOS were pancreatic insufficient (PI) and had disease associated with a severe genotype.

As DIOS nowadays is recognized earlier, and in a less advanced stage, in many patients a few phosphate enemas in combination with (increasing the dose of) oral laxatives [6] can be sufficient. To this purpose mineral oil, magnesium-sulphate or a low dose of a balanced electrolyte solution (e.g. Golytely or Transipeg) [7,8] is generally used. The use of oral N-acetyl-cysteine has declined over the years as its use may be associated with hepatotoxicity.

There has been a struggle to define non-invasive modalities for the management of DIOS. A study aimed to assess non-operative strategies for the management of meconium ileus treatment failures evaluated the use of enemas of perflubron, surfactant, Tween-80, Viokase , Gastrograffin, Golytely, DNAse, N-acetylcysteine and normal saline in mice. The authors described that in their in vivo experiments in mice Gastrograffin and surfactant enemas were the most effective in relieving constipation[8]. A survey of 66 paediatric radiology departments in the USA and Canada also demonstrated an overall higher success rate with Gastrograffin enema use [9].

Gastrograffin enemas have been used clinically to relieve obstruction when other modalities have failed. We are describing the use of oral gastrograffin in our patient where the small bowel obstruction was very refractory to treatment. In this case the oral gastrograffin served to resolve the obstruction and prevented surgical intervention.

Gastrograffin contains a mixture of sodium amidotrizoate and meglumine amiditrizoate. It has been used to treat meconium ileus in infants where it serves to resolve the impaction due to its high osmotic activity. The latter facilitates the movement of fluid into the intestinal lumen resulting in the softening and evacuation of hardened stool [10-12]. It can be given by enema to infants for non-operative treatment of uncomplicated meconium ileus in the absence of, e.g. volvulus, gangrene, perforation, peritonitis or atresia, all of which require immediate operation. The use of a large syringe and soft rubber catheter, are recommended for instillation. The buttocks can be taped tightly together to minimize leakage but a balloon catheter should not be used. The procedure must be carried out slowly and only under fluoroscopic control. Injection should stop as soon as it is seen to enter the ileum. Owing to its high osmolarity, it may cause the loss of a large amount of fluid into the intestines. An intravenous drip must therefore be set up before the enema is given and a suitable parental fluid should be infused as required. If the gastrograffin is not expelled during the first hour after removal of the rectal catheter, an X-ray should be taken to ensure that over distension of the bowel has not occurred.

Gastrograffin can be used orally through a nasogastric tube. Following oral administration only about 3% of the amidotrizoic acid is absorbed from the stomach and intestines [13, 14]. This portion is eliminated mainly via the kidneys. The half-life of the drug in patient with normal renal function is 30 to 60 minutes and is increased in those with renal impairment [15, 16]. In infants and young children (weighing under 10 kg), the hypertonic solutions of contrast media may draw excessive amounts of fluid into the intestine, possibly resulting in hypovolemia, which may cause a shock-like state. Dilution of contrast solution is therefore recommended for use in pediatric patients [17].

With oral or rectal use the most common side effects are diarrhea, nausea or vomiting [15]. Gastrointestinal obstruction may precipitate diatrizoates and may cause gastrointestinal mucosal irritation, erosion, and bleeding. Pediatric dosing suggested is gastrograffin at 15-30ml for children <15 kg in weight, 50ml for children <25kg and 100ml for >25kg in weight. The agent is used once diluted in 4 times the volume of water or fruit juice

with half doses given on day 2 and 3, either rectally or oral. Rectal administration can be repeated twice within 24-hrs [18].

Cystic fibrosis is a disorder with complex medical and surgical problems. Meconium ileus equivalent or the distal intestinal obstruction syndrome may rapidly progress to a life threatening condition with intestinal perforation. Patients with pancreatic insufficiency in CF are therefore required to be managed very carefully for constipation so that problems such as these do not arise. In the event that DIOS does occur, careful yet aggressive management is warranted to avoid a laparotomy and surgical intervention.

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