Dr. Gavin Stone, Ms. Aoife Harrington & Prof Des Cox

Department of Paediatric Respiratory Medicine

Cystic Fibrosis: Distal Intestinal Obstruction Syndrome

Aim

The aim of this document is to provide a guideline for the management of children with cystic fibrosis presenting with Distal Intestinal Obstruction Syndrome

Definition of terms

DIOS; Distal Intestinal Obstruction Syndrome. NG; Nasogastric. AXR; Abdominal X-ray, CXR; Chest X-ray. CFTR: Cystic Fibrosis Transmembrane Conductance Regulator

Target patient population

This guideline applies to children with cystic fibrosis presenting with suspicion of DIOS.

Target users

This guide is directed towards health-care professionals engaged in the care of infants and children with cystic fibrosis. It is hospital policy that any patient with cystic fibrosis presenting to the Paediatric Emergency Department for evaluation is discussed with the on-call Paediatric Respiratory Fellow/Consultant. During normal working hours please contact the Paediatric Respiratory Registrar.

Available Evidence

This guideline is adapted from the Royal Brompton Hospital Clinical Guidelines: Care of Children with Cystic Fibrosis 8th Edition (1). Other guidelines from the European Cystic Fibrosis Society (ECFS)(2), European Society for Paediatric Gastroenterology Hepatology and Nutrition (ESPGHAN)(3) and Cochrane Library(4, 5) have also been incorporated.

Presentation

DIOS is a common complication in CF (paediatric lifetime prevalence of ~8% (6)). Its incidence varies widely but it principally affects those with pancreatic insufficiency. Delayed arrival to hospital after the initial onset of symptoms causes significant morbidity (2).

The definition of DIOS is an acute complete or incomplete faecal obstruction in the ileo-cecum, whereas constipation is defined as gradual faecal impaction of the total colon (3). The pathophysiology of DIOS is not fully understood, but there are multiple contributory factors including:

- CF genotype
- Pancreatic insufficiency
- Inadequate salt intake
- Dehydration
- Poorly controlled fat malabsorption
- History of meconium ileus as neonate or DIOS
- Post organ transplantation

Viscid muco-faeculent material accumulates in the terminal ileum / caecum usually leading to partial obstruction (now called "incomplete or impending" DIOS) with pain often in the right lower quadrant, abdominal fullness and a palpable mass in the right iliac fossa. Children often report having their bowels open as usual, or sometimes diarrhoea (from overflow). Bowel motion history can be inaccurate or misleading.

Important features that increase suspicion of DIOS are:

- Acute periumbilical or right lower quadrant abdominal pain
- Vomiting
- Palpable faecal mass in right lower quadrant
- Previous DIOS

Complete DIOS is when there is total bowel obstruction characterised by abdominal distension, pain (often colicky), fluid levels on AXR and vomiting, usually bilious.

Differential Diagnosis

- Constipation (commonest)
- Adhesions post abdominal surgery
- Appendicitis
- Intussusception
- Volvulus
- Fibrosing colonopathy (extremely rare)
- Biliary tract or gallbladder disease
- Acute pancreatitis
- Urinary tract infection.

Complete DIOS is rare in children, but a surgical opinion should be sought early if there is any doubt about the differential (e.g. risk of adhesions if previous surgery).

Investigations

A good history and abdominal examination is often sufficient to diagnose DIOS. A plain abdominal x-ray (AXR) may be needed to diagnose DIOS or constipation, radiation dose is up to 20x that of a chest X-ray (CXR) and should be used sparingly. Faecal loading throughout the colon, especially in the right iliac fossa suggest DIOS. Intestinal fluid levels confirm severe DIOS with obstruction; the differential diagnosis of a surgical cause of obstruction must always be considered.

If there if are doubts over the cause of abdominal pain, the following may be helpful:

- WBC, amylase, liver function tests, ESR, CRP.
- Urinalysis
- Abdominal ultrasound.
- Barium / gastrografin® enema - by specialist radiologist can diagnose and help treatment at same time

Acute Treatment

A stepwise process will always include adequate hydration. For detailed drug information, including doses please see the CHI Formulary.

Incomplete or Impending DIOS – Mild

1. Ensure;

   a. *Rehydration* – patient must be well hydrated before and during treatment.
   b. Adequate salt replacement to help terminal ileum absorption of bile acids and correct any bowel CFTR electrolyte imbalance that may be implicated in DIOS.
   c. Pancreatic enzyme replacement therapy is reviewed and adjusted if needed.
2. Movicol®/Laxido®(Macrogol)
   a. Paediatric preparation is used up to 12 years age.
   b. Doses are age dependent, usually start at 1-2 sachets daily.

3. N-acetylcysteine
   a. A disulphide bond breaker.
   b. Tastes like rotten eggs
   c. Available preparations
      i. 600mg tablet (Can be split or dispersed in water)
      ii. Intravenous solution (200mg/ml) can be given orally
         1. Usually only used in the neonatal setting and should be mixed with water to a concentration of 40mg/ml (orange or blackcurrant juice or cola may be used as diluent to mask the taste).
4. Oral gastrografin® (Amidotrizoates)
   a. Ensure adequate hydration before using gastrografin as it is highly osmotic and can cause hypovolaemia and bowel perforation. Be particularly careful in babies & infants(4).
   b. Often commenced as an inpatient - for first doses and in severe cases when IV fluids are required.
   c. Use for up to 3 days if no response in first 24 hours but not if symptoms worsen.
   d. **Contraindicated if complete bowel obstruction**.

Incomplete or Impending DIOS – Severe

For detailed drug information, including doses please see the CHI Formulary.

1. Klean-Prep® (Macrogol)
   a. Admit patient
   b. Aim is to take solution until *clear fluid is passed per rectum*.
   c. Do NOT give in the setting of bile stained vomiting.
   d. Consider giving a single dose domperidone 30 minutes before starting to increase gastric emptying
   e. For Klean-Prep® dosing please refer to the CHI Formulary.
   f. NG tube is usually required as volume is so large.
      i. Occasionally patients will prefer to drink it
      ii. More palatable when cool. Can be mixed with *clear* fruit cordial
   g. No food in 2 hours before treatment and during the 4 hours of clean prep to be able to assess for clear fluid being passed PR. If not passing (essentially) clear fluid per rectum a further 4 hours of Kleanprep® treatment can be given
   h. Beware hypoglycaemia (especially in Cystic Fibrosis Related Diabetes) and electrolyte imbalance.

Complete DIOS - Severe

For detailed drug information, including doses please see the CHI Formulary.

If there is obstruction (e.g. bilious vomiting) an NG tube is needed to empty the stomach and prevent bilious aspiration, and IV fluids are given ('drip and suck'). An early specialist opinion from gastroenterologist or surgeon may be needed, always beware of other causes of bowel obstruction or an acute abdomen.

Potential Treatments

(ensure adequate hydration)

- Rectal Gastrografin (Amidotrizoates)
    a. Rarely used.
    b. Consider if oral administration not possible or there is vomiting due to obstruction.
    c. Administered under **radiological guidance**.
    d. Consider a plain Abdominal XR at 1 hour to exclude massive dilation.
- Picolax® (Sodium picosulfate + magnesium salts)
    a. May be administered first line
    b. Heat is generated on addition to water, allow to cool before drinking.
- Colonoscopy and surgery
    a. Rarely indicated when adequate medical management instituted
    b. Potential surgical approaches; Laparotomy and enterotomy, bowel resection.

Chronic Management

The onset of DIOS may be indolent with just intermittent abdominal colicky pain, some anorexia and palpable right iliac fossa mass. Laxatives e.g. Movicol or occasionally lactulose in a young child should be continued for several months post DIOS. Make sure child has been reviewed by a dietitian.

Important considerations include(5);

- Avoid dehydration
- Ensure adequate fluid & salt intake.
- Check dose / compliance / timing of enzyme supplements(4).
- If ongoing malabsorption is documented, consider starting omeprazole.
- Diet – ensure adequate dietary roughage.
- Ensure patient has well established toilet routine (try to go after meals), even at school.
- Movicol (Paediatric if < 12 yrs) is first line treatment, lactulose may help.
- In some children, oral N-acetylcysteine may help, especially in settling abdominal pain.

Constipation

If severe should be considered as part of DIOS spectrum. However, beware of increasing enzyme doses when all that is needed is simple childhood constipation treatment. The main difference from DIOS is that constipation tends to be limited to rectum, so faecal masses are only felt in the left iliac fossa. Stool is more likely to be hard and pellet like or even painful to pass.

Treatment:

- Ensure adequate fluid intake.
- Movicol (Paediatric if <12 yrs) or Lactulose may be used.
    - Movicol dose can be adjusted up and down to produce regular soft stools.
    - Lactulose can cause stomach cramps and flatulence in large doses.

Link to Reference List