

# Children's Services

# Management of Distal Intestinal Obstruction Syndrome in Paediatric Patients with Cystic Fibrosis

Guideline and Management

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### **Guideline Statement**

Subsequent to the implementation of the Scottish Government's National Delivery Plan for Specialist Children's Services, NHS Lanarkshire delivers Paediatric Cystic Fibrosis (CF) care to the local population in partnership with The Royal Hospital for Children Glasgow (RHCG). This guideline has been developed to provide direction to staff in relation to the management of Distal Intestinal Obstruction Syndrome (DIOS) in this group of children.

# **Background**

Distal intestinal obstruction syndrome (DIOS) is a condition which is unique to Cystic Fibrosis and is one of the many abdominal manifestations of CF.

Patients with cystic fibrosis may require pancreatic enzyme supplements (most commonly Creon) with milk or foods in order to digest fat and protein in their diet. Even with optimal pancreatic enzyme replacement therapy, only up to 85% of all ingested fat can be successfully absorbed. Occasionally, stool fat content increases resulting in the accumulation of faeces and sticky mucus, forming a mass in the distal part of the small intestine.

It is not simple constipation and therefore the principles of management are different.

NHS Lanarkshire bases its management on the same principles as The Royal Hospital for Children Glasgow. Our practice is in line with the West of Scotland Cystic Fibrosis standards of care.

# **Aims**

The aims of this document are:-

- To promote timely recognition and diagnosis of DIOS
- To provide clear guidance for staff on the management of DIOS
- To promote consistently good practice

# **Short Life Working Group**

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# Management

#### **Assessment**

Patients present with abdominal pain and/or distension and may have a palpable (faecal) mass on abdominal examination. Symptoms of total, acute intestinal obstruction are rare.

#### Investigations

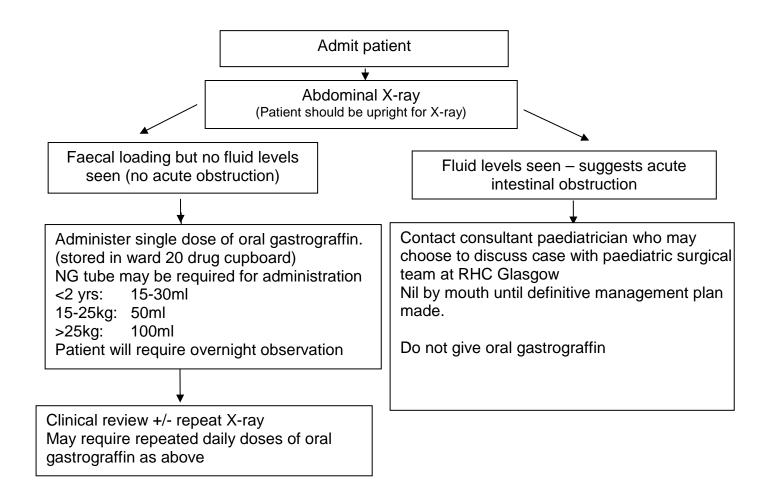
A good history and abdominal examination

An abdominal X-Ray is mandatory in order to exclude intestinal obstruction or perforation.

The typical finding in uncomplicated DIOS is gross faecal loading.

### Management

When DIOS is suspected, the child should attend hospital for assessment. Management will be dictated by X-Ray findings as follows:



## REFERENCES

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