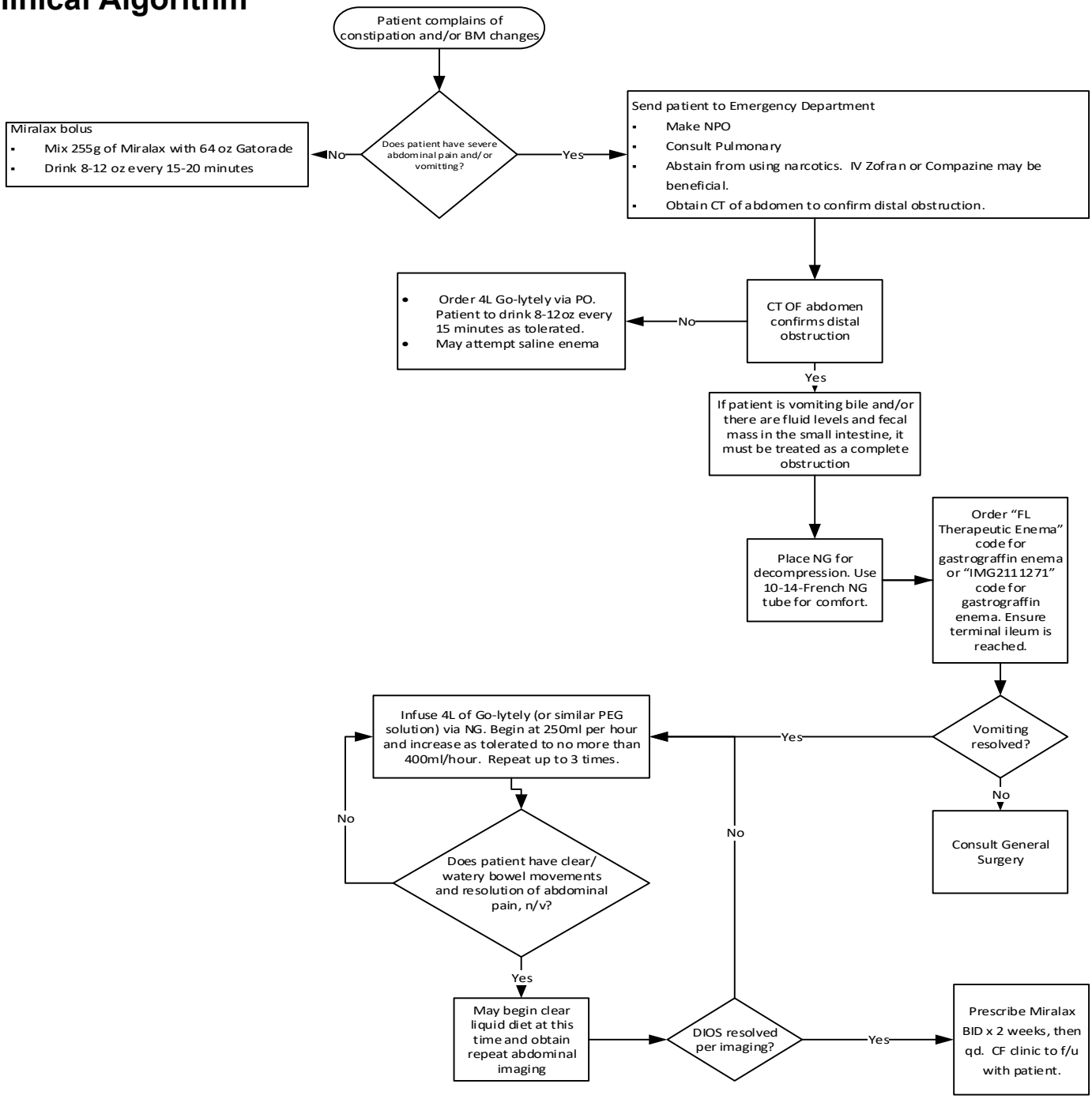
Management of Constipation and DIOS in Cystic Fibrosis, Inpatient and Outpatient

Updated: September 27, 2024

Clinical Algorithm



Clinical Pathway Summary

CLINICAL PATHWAY NAME: Management of constipation and distal intestinal obstructive syndrome (DIOS) in Cystic Fibrosis (CF)

PATIENT POPULATION AND DIAGNOSIS: Adult cystic fibrosis patients

APPLICABLE TO: Corewell Health Adult Cystic Fibrosis Care Team, Corewell Health Lung Transplant Team, Corewell Health Inpatient (all facilities)

BRIEF DESCRIPTION: People with cystic fibrosis are at high risk for distal intestinal obstructive syndrome due to large amounts of mucus and sticky stool in their intestines, intestinal dysmotility and dehydration. A person with CF may be at even higher risk if they have had an episode of DIOS in the past or have had one or more bowel resections.[1] Distinguishing between constipation and DIOS is crucial to prevent bowel perforation or unnecessary surgery. Constipation in CF is characterized by abdominal pain, distention, gradual decrease in bowel movements over several days to weeks and can typically be treated with laxatives.[2] DIOS is characterized by vomiting of bile and/or fluid in the small intestine as visualized by an abdominal film, a large amount of stool in the ileocecum, and abdominal pain or distention.[2] Treating DIOS with a gastrograffin enema allows stool to be broken up and pass through the bowel. Treating a patient with a complete obstruction with laxatives before the stool mass is removed may increase the risk of perforation.

OPTIMIZED EPIC ELEMENTS (if applicable): CF order set TBD

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Clinical Pathways Clinical Approach

TREATMENT AND MANAGEMENT[3]:

The first step in treatment is determining if the patient has constipation or DIOS. If a patient calls the outpatient clinic or presents to the ED with severe abdominal pain and bowel changes (constipation can also present as diarrhea from overflow of stool), move directly to step II.

I. If the patient is constipated but not having severe abdominal pain or vomiting, the patient may be treated with a large bolus of 64 ounces of Gatorade mixed with 255 grams of Miralax. This should be consumed at a rate of about 8-12 ounces every 15-20 minutes.

II. If the patient has symptoms of DIOS (severe abdominal pain, bowel changes and/or vomiting), they should be sent to the emergency department and made NPO until DIOS is ruled out. The patient may be given IV Zofran or Compazine, but use of narcotics should be avoided which may worsen the obstruction. A CT of the abdomen should be obtained to confirm a distal obstruction.

   A. Even if the CT is read as “incomplete obstruction”, if the patient is vomiting bile and/or there is fluid in the small intestine prior to a distal fecal mass, the patient should be treated as though they have a complete obstruction, and the subsequent steps should be followed in order.

      1. Place NG for decompression. This should ideally be performed by a skilled clinician to avoid multiple attempts. A 10-14-French NG is best for ease of placement and comfort to the patient.
      2. Place order for a “FL therapeutic enema” or code “IMG2111271” should be placed for a gastrograffin enema. A gastrograffin enema is a rectal lavage with hydrostatic pressure under direct vision by an experienced radiologist until terminal ileum is reached. <u>It is important to break up the stool via this method before giving oral solution to prevent vomiting or bowel perforation.</u>

        a. If the gastrograffin enema is unsuccessful or if vomiting persists, consult general surgery.
    3. Begin infusion of 4L of Go-lytely (or similar poly-ethylene glycol solution) via the NG. Initiate a rate of 250ml per hour and increase as tolerated to no more than 400ml per hour via the NG.
        a. This step can be repeated 3 times if needed.
    4. If the patient has clear, watery bowel movements and resolution of abdominal pain and nausea, then the patient may be advanced to a clear liquid diet and a repeat CT of the abdomen should be obtained.
        a. If the scan continues to reveal a stool burden, repeat the infusion of Go-lytely up to 3 times.
        b. If the scan shows resolution of DIOS, the NG may be removed and diet can be advanced to soft.
    5. Order Miralax to be taken twice daily for two weeks, then patient to remain on one dose daily until told otherwise. CF team will follow-up with patient once discharged.

B. If the CT scan of the abdomen reveals stool burden but does not show fluid levels in the small intestine and there is no nausea/vomiting, the provider should order 4L of Go-lytely by mouth with a goal of 8-12 oz every 15-20 minutes. A saline enema may also be beneficial. Then proceed with steps A4 and A5.

Pathway Information

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EXPERT IMPROVEMENT TEAM (EIT): N/A

CLINICAL PRACTICE COUNCIL (CPC): Specialty/Digestive

CPC APPROVAL DATE: July 17, 2023

OTHER TEAM(S) IMPACTED: Pulmonary, Interventional Radiology, Lung Transplant Team, Gastrointestinal/Digestive, General Surgery, Emergency Department

References

1. Abraham JM, Taylor CJ. Cystic Fibrosis & disorders of the large intestine: DIOS, constipation, and colorectal cancer. *J Cyst Fibros.* 2017;16 Suppl 2:S40-S49.
2. Houwen RH, van der Doef HP, Sermet I, et al. Defining DIOS and constipation in cystic fibrosis with a multicentre study on the incidence, characteristics, and treatment of DIOS. *J Pediatr Gastroenterol Nutr.* 2010;50:38-42.
3. Abraham, JM, Sabharwal, S, Grand, R, Mascarenhas, M. Recommendations of the task force on distal intestinal obstruction syndrome. *Cystic Fibrosis Foundation, Bethesda, MD, USA*; 2016.