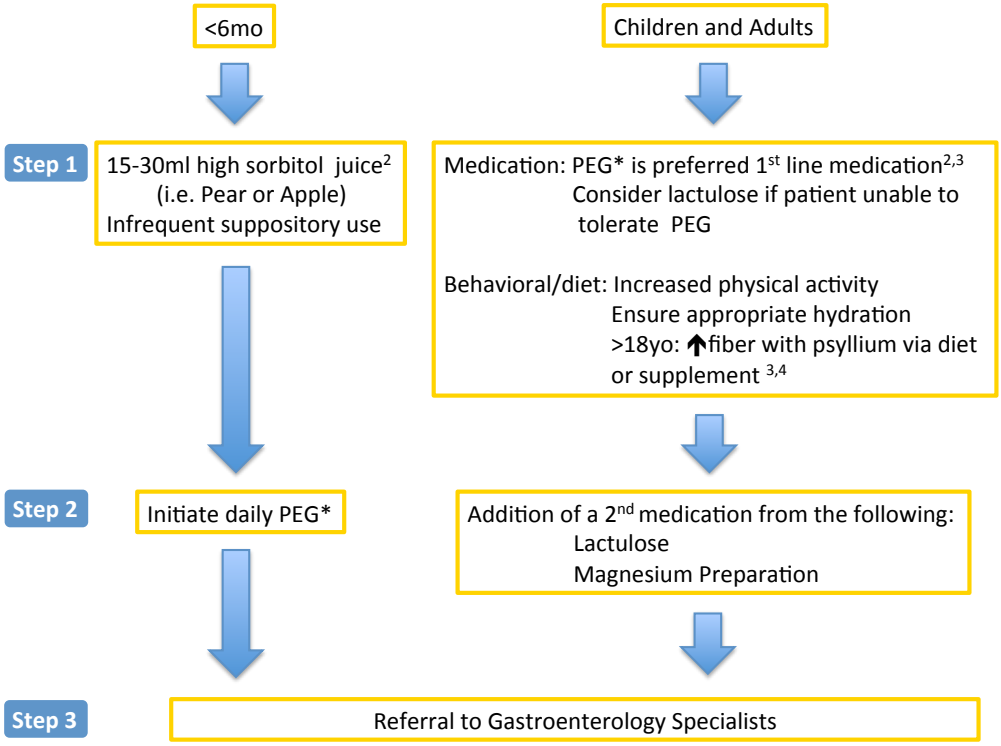
Management of Constipation in Children and Adults with Cystic Fibrosis

Constipation is a common reason for gastrointestinal problems in patients with cystic fibrosis (CF). Symptoms may include flatulence, poor appetite, abdominal pain, infrequent or difficult to pass stool, straining, hard/lumpy stools and prolonged time to stool. Daily bowel movements do not exclude the possible diagnosis of constipation. Untreated constipation can present in a number of ways including but not limited to: bloating, distention, bloody stools, poor weight gain and "overflow" diarrhea.

Diagnosis is made clinically and imaging is usually not warranted. Minimal evaluation includes a stooling history and physical exam, which may show palpable fecal mass in the pelvis (but its absence does not rule out constipation). Perirectal visual inspection can rule out fissures, abscess or rectal abnormality; if present, refer to a GI specialist. In most cases constipation can be managed in the outpatient setting with a plan that includes laxative use, dietary habits and behavioral modifications (see chart below).

Although often categorized together, distal intestinal obstruction syndrome (DIOS) and constipation are distinct diagnoses that are managed differently. DIOS is defined as complete or incomplete intestinal obstruction with fecal mass often in the ileocecum. Constipation with symptoms of obstruction, such as acute abdominal pain with nausea and vomiting warrant imaging to rule out DIOS. Although surgery is seldom indicated, DIOS requires a more intensive treatment regimen, often with inpatient hospital monitoring and advanced imaging[1].

Here we present a basic stepwise approach for providers caring for CF patients suspected of having constipation adapted from the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition (NASPGHAN), American College of Gastroenterology (ACG) and American Gastroenterological Association (AGA) guidelines[2,3,4].



*See dosing table on next page

Other Considerations:

For some patients, gastroenterology referral may be warranted after failing step 1.

Avoid opiate use. Start concurrent bowel regimen if opiates are necessary.

Both pancreatic insufficient and sufficient patients are at risk for constipation.

There is no evidence to suggest that increased pancreatic enzyme supplementation increases constipation risk.[5]

Prepared for the CFF by Dr. A Jay Freeman and Dr. Monika Kowalczyk

Summary of Medications Commonly Used to Treat Constipation in Children and Adults with Cystic Fibrosis

Generic Name	Trade Name Examples	Dosage	Comments/Side Effects
Glycerin	Pedia-Lax	1 pediatric suppository (only in <6months)	No more than 1x per week
Sorbitol in fruit juice	Apple juice Prune juice Pear juice	15-30 mL once or twice daily	First line therapy in patients < 6 months Abdominal cramps, bloating, flatulence
Polyethylene Glycol (PEG)	Miralax Colyte	<6 months: 1 tsp in 4oz fluid daily <10kg: 1/2 cap in 4 oz of fluid daily 10-22kg: 1 cap in 8oz of fluid daily >22kg and adult: 1 cap in 8oz of fluid up to three times daily	First line therapy in patients > 6 months May see incontinence due to potency Dosing frequency may vary from patient to patient
Lactulose		<6 months: not recommended <10 kg: 1-2g once daily 10-22 kg: 1-2g once daily (max 40g) >22kg to adult: 15-30 mL once or twice daily	Abdominal cramps, bloating, flatulence Decreased Na^+ and increased glucose levels
Magnesium	Milk of Magnesia	<10kg: not recommended 10-22kg: 5 mL daily before bed >22kg and adults: 15-30 mL once or twice daily -or- 2-4 tablets once or twice daily	One to two doses may be sufficient Should not be used continuously > 14 days Caution in renal insufficiency (magnesium toxicity)

References:

1. Colombo C et al. Guidelines for the diagnosis and management of distal intestinal obstruction syndrome in cystic fibrosis patients. *J Cyst Fibros.* 2011 Jun;10 Suppl 2:S24-8.
2. Tabbers MM et al. Evaluation and treatment of functional constipation in infants and children: evidence-based recommendations from ESPGHAN and NASPGHAN. *J Pediatr Gastroenterol Nutr*. 2014 Feb;58(2):258-74.
3. Chang L, Lembo A, Sultan S. American Gastroenterological Association Institute Technical Review on the pharmacological management of irritable bowel syndrome. *Gastroenterology.* 2014 Nov;147(5):1149-72.e2.
4. Ford AC et al. American College of Gastroenterology Monograph on the Management of Irritable Bowel Syndrome and Chronic Idiopathic Constipation. *Am J Gastroenterol* 2014; 109:S2 – S26.
5. Baker SS et al. Pancreatic Enzyme Therapy and Clinical Outcomes in Patients with Cystic Fibrosis. *J Pediatr*. 2005 Feb; 146(2):189-93.