

Clinical Practice Guideline

Evaluation and Treatment of Constipation in Infants and Children: Recommendations of the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition

ABSTRACT

Constipation, defined as a delay or difficulty in defecation, present for 2 or more weeks, is a common pediatric problem encountered by both primary and specialty medical providers. The Constipation Guideline Committee of the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition (NASPGHAN) has formulated a clinical practice guideline for the management of pediatric constipation. The Constipation Guideline Committee, consisting of 2 primary care pediatricians, 1 clinical epidemiologist, and pediatric gastroenterologists, based its recommendations on an integration of a comprehensive and systematic review of the medical literature combined with expert

opinion. Consensus was achieved through nominal group technique, a structured quantitative method. The Committee developed 2 algorithms to assist with medical management, 1 for older infants and children and the second for infants less than 1 year of age. The guideline provides recommendations for management by the primary care provider, including evaluation, initial treatment, follow-up management, and indications for consultation by a specialist. The Constipation Guideline Committee also provided recommendations for management by the pediatric gastroenterologist. *JPGN* 43:e1–e13, 2006. **Key Words:** Constipation—Guidelines. © 2006 Lippincott Williams & Wilkins

BACKGROUND

A normal pattern of stool evacuation is thought to be a sign of health in children of all ages. Especially during the first months of life, parents pay close attention to the frequency and the characteristics of their children's defecation. Any deviation from what is thought by any family member to be normal for children may trigger a call to the nurse or a visit to the pediatrician. Thus, it is not surprising that approximately 3% of general pediatric outpatient visits and 25% of pediatric gastroenterology consultations are related to a perceived defecation disorder (1). Chronic constipation is a source of anxiety for parents who worry that a serious disease may be causing the symptom. Yet, only a small minority of children have an organic cause for constipation. Beyond the neonatal

period, the most common cause of constipation is functional and has been called idiopathic constipation, functional fecal retention, and fecal withholding.

In most cases the parents are worried that the child's stools are too large, too hard, painful, or too infrequent. The normal frequency of bowel movements at different ages has been defined (Table 1). Infants have a mean of 4 stools per day during the first week of life. This frequency gradually declines to a mean average of 1.7 stools per day at 2 years of age and 1.2 stools per day at 4 years of age (2,3). Some normal breast-fed babies do not have stools for several days or longer (4). After 4 years, the frequency of bowel movements remains unchanged.

Functional constipation—that is, constipation without objective evidence of a pathological condition—most commonly is caused by painful bowel movements with resultant voluntary withholding of feces by a child who wants to avoid unpleasant defecation. Many events can lead to painful defecation such as toilet training, changes in routine or diet, stressful events, intercurrent illness, unavailability of toilets, or the child's postponing defecation because he or she is too busy. Withholding feces can lead to prolonged fecal stasis in the colon, with reabsorption of fluids and an increase in the size and consistency of the stools.

The passage of large, hard stools that painfully stretch the anus may frighten the child, resulting in a fearful

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Address correspondence and reprint requests to Executive Director, NASPGHAN, 1501 Bethlehem Pike, PO Box 6, Flourtown PA 19031. (e-mail: naspgghan@naspgghan.org).

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Disclaimer: The guidance in this report does not indicate an exclusive course of treatment or serve as a standard of medical care. Variations, taking into account individual circumstances, may be appropriate.

TABLE 1. Normal frequency of bowel movements

Age	Bowel movements per week ^a	Bowel movements per day ^b
0–3 Months		
Breast-fed	5–40	2.9
Formula-fed	5–28	2.0
6–12 months	5–28	1.8
1–3 years	4–21	1.4
More than 3 years	3–14	1.0

Adapted from Fontana M, Bianchi C, Cataldo F, et al. Bowel frequency in healthy children. *Acta Paediatr Scand* 1987;78:682–4.

^aApproximately mean \pm 2 SD.

^bMean.

determination to avoid all defecation. Such children respond to the urge to defecate by contracting their anal sphincter and gluteal muscles, attempting to withhold stool (5,6). They rise on their toes and rock back and forth while stiffening their buttocks and legs, or wriggle, fidget, or assume unusual postures, often performed while hiding in a corner. This dance-like behavior is

frequently misconstrued by parents who believe that the child is straining in an attempt to defecate. Eventually, the rectum habituates to the stimulus of the enlarging fecal mass, and the urge to defecate subsides. With time, such retentive behavior becomes an automatic reaction. As the rectal wall stretches, fecal soiling may occur, angering the parents and frightening the child (7). After several days without a bowel movement, irritability, abdominal distension, cramps, and decreased oral intake may result.

Although constipation is a common pediatric problem, no evidence-based guidelines for its evaluation and treatment currently exist. Therefore, the Constipation Guideline Committee was formed by the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition (NASPGHAN) to develop a clinical practice guideline.

METHODS

The Constipation Guideline Committee, which consists of 2 primary care pediatricians, 1 clinical epidemiologist, and

TABLE 2. Summary of recommendations and the quality of the evidence

Recommendations	Quality of evidence ^a
General recommendations	
A thorough history and physical examination are an important part of the complete evaluation of the infant or child with constipation.	III
Performing a thorough history and physical examination is sufficient to diagnose functional constipation in most cases.	III
A stool test for occult blood is recommended in all constipated infants and in those children who also have abdominal pain, failure to thrive, diarrhea, or a family history of colon cancer or polyps.	III
In selected patients, an abdominal radiograph, when interpreted correctly, can be useful to diagnose fecal impaction.	II-2
Rectal biopsy with histopathological examination and rectal manometry are the only tests that can reliably exclude Hirschsprung disease.	II-1
In selected patients, measurement of transit time using radiopaque markers can determine whether constipation is present.	II-2
Recommendations for infants	
In infants, rectal disimpaction can be achieved with glycerin suppositories. Enemas are to be avoided.	II-3
In infants, juices that contain sorbitol, such as prune, pear, and apple juices can decrease constipation.	II-3
Barley malt extract, corn syrup, lactulose, or sorbitol (osmotic laxatives) can be used as stool softeners.	III
Mineral oil and stimulant laxatives are not recommended for infants.	III
Recommendations for children	
In children, disimpaction can be achieved with either oral or rectal medication, including enemas.	II-3
In children, a balanced diet, containing whole grains, fruits, and vegetables, is recommended as part of the treatment for constipation.	III
The use of medications in combination with behavioral management can decrease the time to remission in children with functional constipation.	I
Mineral oil (a lubricant) and magnesium hydroxide, lactulose, and sorbitol (osmotic laxatives) are safe and effective medications.	I
Rescue therapy with short-term administration of stimulant laxatives can be useful in selected patients.	II-3
Senna and bisacodyl (stimulant laxatives) can be useful in selected patients who are more difficult to treat.	II-1
Polyethylene glycol electrolyte solution, given in low dosage, may be an effective long-term treatment for constipation that is difficult to manage.	III
Biofeedback therapy can be an effective short-term treatment of intractable constipation.	II-2

^aCategories of the quality of evidence (95):

I: Evidence obtained from at least one properly designed randomized controlled study. II-1: Evidence obtained from well-designed cohort or case-control trials without randomization. II-2: Evidence obtained from well-designed cohort or case-control analytic studies, preferably from more than 1 center or research group. II-3: Evidence obtained from multiple time series with or without intervention. Dramatic results in uncontrolled experiments (such as the results of the introduction of penicillin treatment in the 1940s) could also be regarded as this type of evidence. III: Opinions of respected authorities, based on clinical experience, descriptive studies, or reports of expert committees.

5 pediatric gastroenterologists, addressed the problem of constipation in infants and children who had no previously established medical condition. Neonates less than 72 hours old and premature infants of less than 37 weeks' gestation were excluded from consideration. This clinical practice guideline has been designed to assist primary care pediatricians, family practitioners, nurse practitioners, physician assistants, pediatric gastroenterologists, and pediatric surgeons in the management of children with constipation in both inpatient and outpatient settings. Constipation was defined as a delay or difficulty in defecation, present for 2 or more weeks, and sufficient to cause significant distress to the patient. The desirable outcome of optimal management was defined as a normal stooling pattern, with interventions that have few or no adverse effects, and with resultant resumption of functional health.

To develop the initial evidence-based guideline, articles on constipation published in English were found using Medline (8).

A search for articles published from January 1966 through November 1997, revealed 3839 articles on constipation. The Cochrane Center has designed a search strategy for Medline to identify randomized controlled trials. This strategy includes controlled vocabulary and free-text terms such as *randomized controlled trial*, *clinical trial*, and *placebo* (9). When this search strategy was run with the term *constipation*, 1047 articles were identified, 809 of which were in English and 254 of which included children.

After letters, editorials, and review articles were eliminated, 139 articles remained. Forty-four of these were studies in special populations, such as children with meningomyelocele or Hirschsprung disease, and were eliminated. Ninety-five articles remained and were reviewed in depth. A second search strategy was used to identify articles on constipation that related to treatment, including drug therapy (75 articles), surgery (64 articles), and "therapy" (144 articles). This added 148 new articles

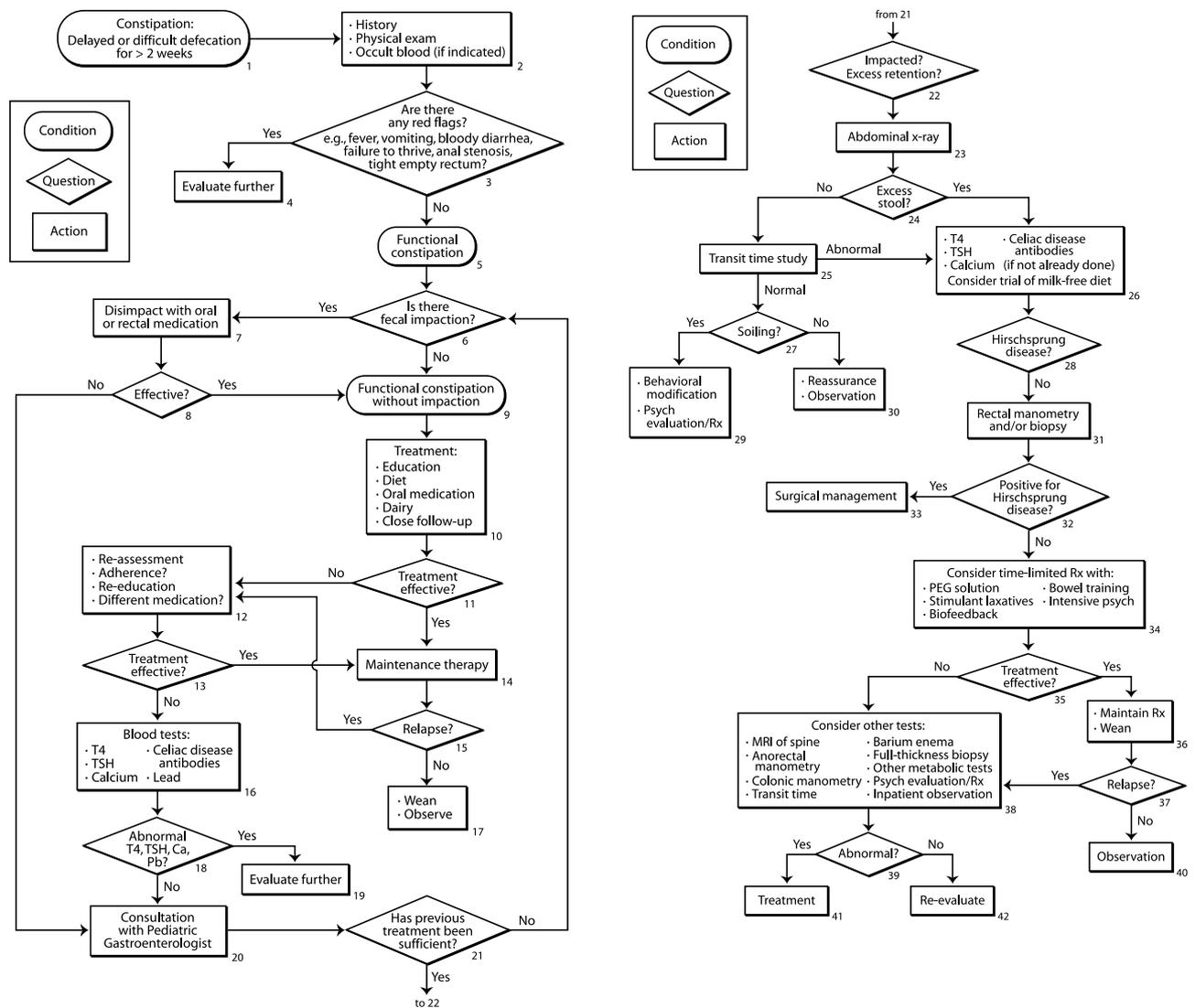


FIG. 1. An algorithm for the management of constipation in children 1 year of age and older. T4, thyroxine; TSH, thyroid-stimulating hormone; Ca, calcium; Pb, lead; Rx, therapy; PEG, polyethylene glycol electrolyte; psych, psychological management; MRI, magnetic resonance imaging.

in which the abstracts were reviewed. If the abstract indicated that the article may be relevant, the article was reviewed in depth. Seven additional articles were identified from the reference listings of the articles already cataloged. In total, 160 articles were reviewed for these guidelines.

Articles were evaluated using written criteria developed by Sackett et al. (10,11). These criteria had been used in previous reviews (12,13). Five articles were chosen at random and reviewed by a colleague in the Department of Pediatrics at the University of Rochester (New York, U.S.A.) who had been trained in epidemiology. Concordance using the criteria was 92%. Using the methods of the Canadian Preventive Services Task Force (14), the quality of evidence of each of the recommendations made by the Constipation Guideline Committee was determined and is summarized in Table 2. The Committee based its recommendations on integration of the literature review combined with expert opinion when evidence was insufficient. Consensus was achieved through nominal group technique, a structured, quantitative method (15).

The guidelines were critically reviewed by numerous primary care physicians in community and academic practices,

including members of several committees of the American Academy of Pediatrics. In addition, the guidelines were distributed to the NASPGHAN membership for review and comment and finally were officially endorsed by the society's Executive Council.

Two algorithms were developed (Figs. 1 and 2). The initial discussion is based on the algorithm for children 1 year of age and older. The second algorithm is for children less than 1 year of age. In this article, the first algorithm is discussed in detail, and the second algorithm is discussed only when it diverges from the first.

To evaluate evidence published since 1997, literature searches using the key word "constipation," limited to English language, and "All Child" (which includes children and adolescents 0–18 years of age) were performed in PubMed on May 5, 2003, August 8, 2003 and August 9, 2004. The Database of Abstracts of Reviews of Effects (DARE) and the Cochrane Database of Systematic Reviews also were searched using the key word "constipation." From this search 90 total articles were identified by this process; 27 applied to children who did not have an underlying chronic condition. The authors

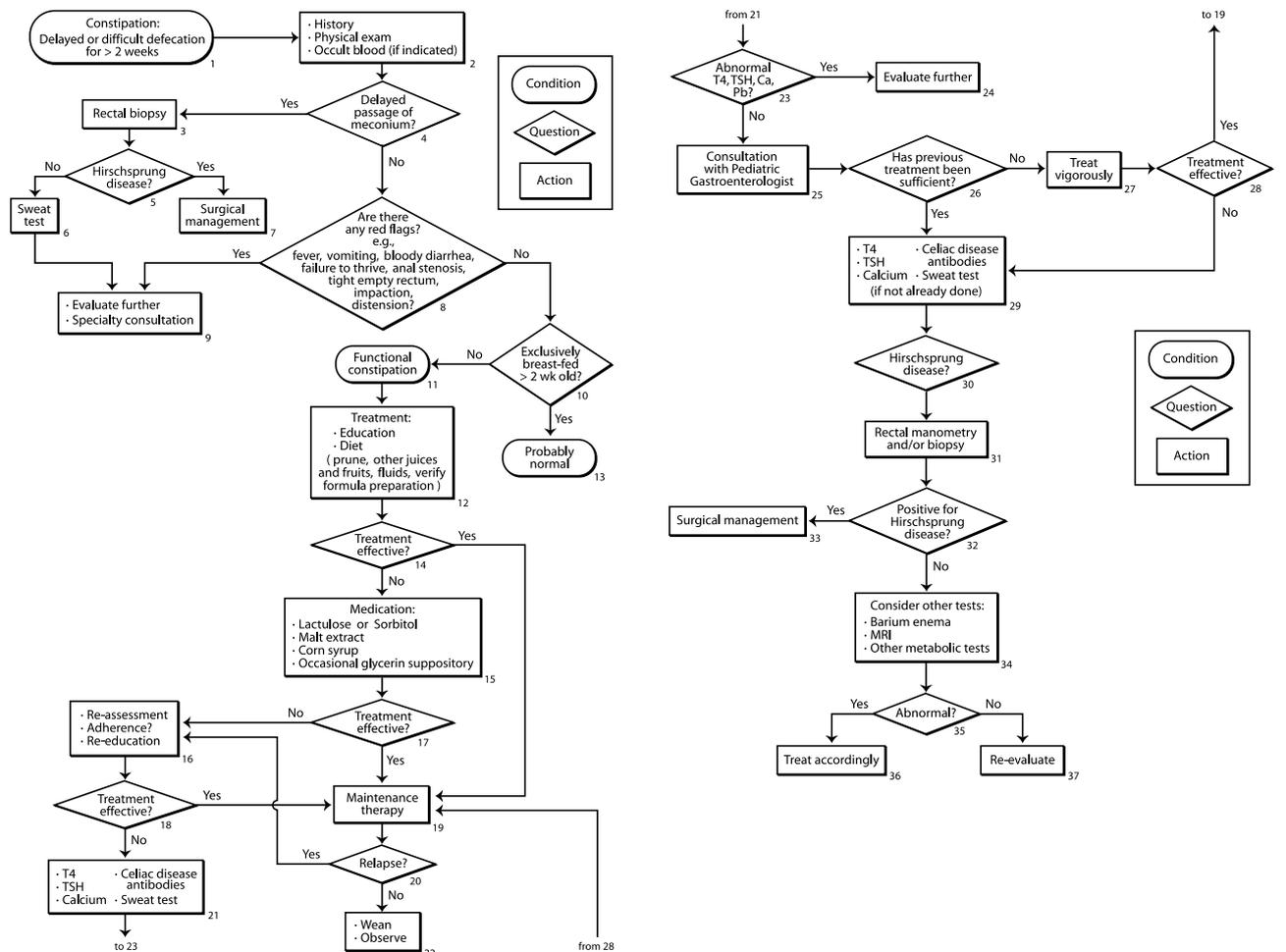


FIG. 2. An algorithm for the management of constipation in infants less than 1 year of age. T4, thyroxine; TSH, thyroid stimulating hormone; Ca, calcium; Pb, lead; Rx, therapy; PEG, polyethylene glycol electrolyte; psych, psychological management; MRI, magnetic resonance imaging.

identified an additional 8 articles during the subsequent discussions. The quality of evidence was categorized according to Fisher et al. (16). The papers were reviewed in detail and discussed by the Constipation Guideline Committee until consensus was achieved on whether the original recommendations should be modified based on the new evidence.

MEDICAL HISTORY

Based on clinical experience, a thorough history is recommended as part of a complete evaluation of a child with constipation (Table 3). There are no well-designed studies that determine which aspects of a history are pertinent. Important information includes the time after birth of the first bowel movement, what the family or child means when using the term "constipation" (17), the length of time the condition has been present, the frequency of bowel movements, the consistency and size of the stools, whether defecation is painful, whether blood has been present on the stool or the toilet paper, and whether the child experiences abdominal pain. Fecal soiling may be mistaken for diarrhea by some parents. A history of stool-withholding behavior reduces the likelihood that there is an organic disorder. Medications are an important potential cause of constipation (Table 4).

Fever, abdominal distension, anorexia, nausea, vomiting, weight loss, or poor weight gain could be signs of an organic disorder (Table 4). Bloody diarrhea in an infant with a history of constipation could be an indication of enterocolitis complicating Hirschsprung disease.

A psychosocial history assesses the family structure, the number of people living in the child's home and their relationship to the child, the interactions the child has with peers, and the possibility of abuse. If the child is in school it is important to learn whether the child uses the school restrooms and if not, why. The caregiver's assessment of the child's temperament may be useful in planning a reward system for toilet behavior.

PHYSICAL EXAMINATION

Based on clinical experience, a thorough physical examination is recommended as part of a complete evaluation of a child with constipation (Table 5). No well-designed studies have been conducted to determine the aspects of the physical examination that are most important. External examination of the perineum and perianal area is essential. At least one digital examination of the anorectum is recommended. The anorectal examination assesses perianal sensation, anal tone, the size of the rectum, and the presence of an anal wink. It also determines the amount and consistency of stool and its location within the rectum. It is recommended that a test for occult blood in the stool be performed in all infants with constipation, as well as in any child who also has abdominal pain, failure to thrive, intermittent diarrhea, or a family history of colon cancer or

TABLE 3. History in pediatric patients with constipation

Age
Sex
Chief symptom
Constipation history
Frequency and consistency of stools
Pain or bleeding with passing stools
Abdominal pain
Waxing and waning of symptoms
Age of onset
Toilet training
Fecal soiling
Withholding behavior
Change in appetite
Nausea or vomiting
Weight loss
Perianal fissures, dermatitis, abscess, or fistula
Current treatment
Current diet (24-hour recall history)
Current medications (for all medical problems)
Oral, enema, suppository, herbal
Previous treatment
Diet
Medications
Oral, enema, suppository, herbal
Prior successful treatments
Behavioral treatment
Results of prior tests
Estimate of parent/patient adherence
Family history
Significant illnesses
Gastrointestinal (constipation, Hirschsprung disease)
Other
Thyroid, parathyroid, cystic fibrosis, celiac disease
Medial history
Gestational age
Time of passage of meconium
Condition at birth
Acute injury or disease
Hospital admissions
Immunizations
Allergies
Surgeries
Delayed growth and development
Sensitivity to cold
Coarse hair
Dry skin
Recurrent urinary tract infections
Daytime urinary incontinence
Other
Developmental history
Normal, delayed
School performance
Psychosocial history
Psychosocial disruption of child or family
Interaction with peers
Temperament
Toilet habits at school

colonic polyps. Detection of a physical abnormality could lead to the identification of an organic disorder (Table 6).

A thorough history and physical examination is generally sufficient to allow the practitioner to establish whether the child requires further evaluation (Fig. 1, box 4) or has functional constipation (Fig. 1, box 5).

TABLE 4. *Differential diagnosis of constipation*

Nonorganic
Developmental
Cognitive handicaps
Attention-deficit disorders
Situational
Coercive toilet training
Toilet phobia
School bathroom avoidance
Excessive parental interventions
Sexual abuse
Other
Depression
Constitutional
Colonic inertia
Genetic predisposition
Reduced stool volume and dryness
Low fiber in diet
Dehydration
Underfeeding or malnutrition
Organic
Anatomic malformations
Imperforate anus
Anal stenosis
Anterior displaced anus (96)
Pelvic mass (sacral teratoma)
Metabolic and gastrointestinal
Hypothyroidism
Hypercalcemia
Hypokalemia
Cystic fibrosis
Diabetes mellitus
Multiple endocrine neoplasia type 2B
Gluten enteropathy
Neuropathic conditions
Spinal cord abnormalities
Spinal cord trauma
Neurofibromatosis
Static encephalopathy
Tethered cord
Intestinal nerve or muscle disorders
Hirschsprung disease
Intestinal neuronal dysplasia
Visceral myopathies
Visceral neuropathies
Abnormal abdominal musculature
Prune belly
Gastroschisis
Down syndrome
Connective tissue disorders
Scleroderma
Systemic lupus erythematosus
Ehlers–Danlos syndrome
Drugs
Opiates
Phenobarbital
Sucralfate
Antacids
Antihypertensives
Anticholinergics
Antidepressants
Sympathomimetics
Other
Heavy-metal ingestion (lead)
Vitamin D intoxicification
Botulism
Cow's milk protein intolerance

MANAGEMENT OF CHILDREN WITH FUNCTIONAL CONSTIPATION

The general approach to the child with functional constipation includes the following steps: determine whether fecal impaction is present (Fig. 1, box 6), treat the impaction if present (Fig. 1, box 7), initiate treatment with oral medication, provide parental education and close follow-up, and adjust medications as necessary (Fig. 1, box 10).

Education

The education of the family and the demystification of constipation, including an explanation of the pathogenesis of constipation, are the first steps in treatment. If fecal soiling is present, an important goal for both the child and the parent is to remove negative attributions. It is especially important for parents to understand that soiling from overflow incontinence is not a willful and defiant maneuver. Parents are encouraged to maintain a consistent, positive, and supportive attitude in all aspects of treatment.

TABLE 5. *Physical examination of children with constipation*

General appearance
Vital signs
Temperature
Pulse
Respiratory rate
Blood pressure
Growth parameters
Head, ears, eyes, nose, throat
Neck
Cardiovascular
Lungs and chest
Abdomen
Distension
Palpable liver and spleen
Fecal mass
Anal inspection
Position
Stool present around anus or on clothes
Perianal erythema
Skin tags
Anal fissures
Rectal examination
Anal wink
Anal tone
Fecal mass
Presence of stool
Consistency of stool
Other masses
Explosive stool on withdrawal of finger
Occult blood in stool
Back and spine examination
Dimple
Tuft of hair
Neurological examination
Tone
Strength
Cremasteric reflex
Deep tendon reflexes

TABLE 6. *Physical findings distinguishing organic constipation from functional constipation*

Failure to thrive
Abdominal distension
Lack of lumbosacral curve
Pilonidal dimple covered by a tuft of hair
Midline pigmentary abnormalities of the lower spine
Sacral agenesis
Flat buttocks
Anteriorly displaced anus
Patulous anus
Tight, empty rectum in presence of palpable abdominal fecal mass
Gush of liquid stool and air from rectum on withdrawal of finger
Occult blood in stool
Absent anal wink
Absent cremasteric reflex
Decreased lower extremity tone and/or strength
Absence or delay in relaxation phase of lower extremity deep-tendon reflexes

It may be necessary to repeat the education and demystification processes several times during treatment (18).

Disimpaction

Fecal impaction is defined as a hard mass in the lower abdomen identified during physical examination, a dilated rectum filled with a large amount of stool found during rectal examination, or excessive stool in the colon identified by abdominal radiography (19). Disimpaction is necessary before initiation of maintenance therapy. It may be accomplished with either oral or rectal medication (Fig. 1, box 7). In uncontrolled clinical trials, disimpaction by the oral route, the rectal route, or a combination of the 2 has been shown to be effective (Table 7) (20). There are no randomized studies that compare the effectiveness of 1 with the other. The oral approach is not invasive and gives a sense of power to the child, but adherence to the treatment regimen may be a problem. The rectal approach is faster but is invasive. The choice of treatment is best determined after discussing the options with the family and child.

Disimpaction with oral medication has been shown to be effective when high doses of mineral oil, polyethylene glycol electrolyte solutions, or both are used, (20–24). Although there are no controlled trials demonstrating the effectiveness of high-dose magnesium hydroxide, magnesium citrate, lactulose, sorbitol, senna, or bisacodyl for initial disimpaction, these laxatives have been used successfully in that role (25,26). It is recommended that mineral oil, oral electrolyte solutions, or the listed laxatives be used alone or in combination for initial disimpaction when the oral route is selected.

Rectal disimpaction may be performed with phosphate soda enemas, saline enemas, or mineral oil enemas followed by a phosphate enema (27,28). These enemas are widely used and are effective. The use of soap suds, tap water, and magnesium enemas is not recommended

because of their potential toxicity. Rectal disimpaction has also been effectively performed with glycerin suppositories in infants (29) and bisacodyl suppositories in older children.

The Committee discussed the use of digital disimpaction in chronic constipation in the primary care setting. However, there was insufficient literature on the subject, and the Committee could not reach consensus on whether to discourage or recommend its use.

Maintenance Therapy

Once the impaction has been removed, the treatment focuses on the prevention of recurrence. In the child who has no impaction (Fig. 1, box 9) or after successful disimpaction, maintenance therapy is begun. This treatment consists of dietary interventions, behavioral modification, and laxatives to assure that bowel movements occur at normal intervals with good evacuation.

Dietary changes are commonly advised, particularly increased intake of fluids and absorbable and non-absorbable carbohydrate, as a method to soften stools. Carbohydrates and especially sorbitol, found in some juices such as prune, pear, and apple juices, can cause increased frequency and water content of stools (30,31). There are conflicting reports about the role of dietary fiber, with evidence that constipated children have a lower, equivalent or higher intake of dietary fiber (32–35). Administration of glucamomannan (36) in addition to laxatives may be beneficial in the treatment of constipation. Until additional studies demonstrate the efficacy of treatment with fiber, the current findings are too weak to support a definitive recommendation for fiber supplementation in the treatment of constipation. A balanced diet that includes whole grains, fruits, and vegetables is recommended as part of the treatment for constipation in children. Forceful implementation of diet is undesirable.

Behavioral Modification

An important component of treatment includes behavior modification and regular toilet habits (37). Unhurried time on the toilet after meals is recommended. As part of the treatment of constipation, with or without overflow incontinence, it is often helpful to have children and their caregivers keep diaries of stool frequency. This can be combined with a reward system. For example, a child can use a calendar with stickers to record each stool that is passed in the toilet. The calendar can then be taken on visits with the health care provider and can serve as both a diary and a point for positive reinforcement. In cases in which motivational or behavioral problems are interfering with successful treatment, referral to a mental health care provider for behavior modification or other intervention may be helpful.

TABLE 7. Medications for use in treatment of constipation

Laxatives	Dosage	Side effects	Notes
Osmotic			
Lactulose ^a	1–3 mL/kg/day in divided doses; available as 70% solution.	Flatulence, abdominal cramps; hypernatremia has been reported when used in high dosage for hepatic encephalopathy; case reports of nontoxic megacolon in elderly.	Synthetic disaccharide. Well tolerated long term.
Sorbitol ^a	1–3 mL/kg/day in divided doses; available as 70% solution.	Same as lactulose.	Less expensive than lactulose.
Barley malt extract ^a	2–10 mL/240 mL of milk or juice		Unpleasant odor. Suitable for infants drinking from a bottle.
Magnesium hydroxide ^a	1–3 mL/kg/day of 400 mg/5 mL; available as liquid, 400 mg/5 mL and 800 mg/5 mL, and tablets.	Infants are susceptible to magnesium poisoning. Overdose can lead to hypermagnesemia, hypophosphatemia and secondary hypocalcemia.	Acts as an osmotic laxative. Releases cholecystokinin, which stimulates gastrointestinal secretion and motility. Use with caution in renal impairment.
Magnesium citrate ^a	<6 Years, 1–3 mL/kg/day; 6–12 years, 100–150 mL/day; >12 years, 150–300 mL/day; in single or divided doses. Available as liquid, 16.17% magnesium.	Infants are susceptible to magnesium poisoning. Overdose can lead to hypermagnesemia, hypophosphatemia and secondary hypocalcemia.	
PEG 3350	Disimpaction: 1–1.5 g/kg/day for 3 days Maintenance 1 g/kg/day		Superior palatability and acceptance by children Safety studies necessary before widespread use is recommended in infants.
Osmotic enema			
Phosphate enemas	<2 Years old: to be avoided; ≥2 years old: 6 mL/kg up to 135 mL	Risk of mechanical trauma to rectal wall, abdominal distention or vomiting. May cause severe and lethal episodes of hyperphosphatemia hypocalcemia, with tetany.	Some of the anion is absorbed, but if kidney is normal, no toxic accumulation occurs. Most side effects occur in children with renal failure or Hirschsprung disease.
Lavage			
Polyethylene glycol-electrolyte solution	For disimpaction: 25 mL/kg/hr (to 1000 mL/hr) by nasogastric tube until clear or 20 mL/kg/hr for 4 hr/day. For maintenance: (older children): 5–10 mL/kg/per day.	Difficult to take. Nausea, bloating, abdominal cramps, vomiting, and anal irritation. Aspiration, pneumonia, pulmonary edema, Mallory–Weiss tear. Safety of long-term maintenance not well established.	Information mostly obtained from use for total colonic irrigation. May require hospital admission and nasogastric tube.
Lubricant			
Mineral oil ^a	<1 Year old; not recommended. Disimpaction: 15–30 mL/yr of age, up to 240 mL daily. Maintenance: 1–3 mL/kg/day.	Lipoid pneumonia if aspirated. Theoretical interference with absorption of fat-soluble substances, but there is no evidence in the literature. Foreign-body reaction in intestinal mucosa.	Softens stool and decreases water absorption. More palatable if chilled. Anal leakage indicates dose too high or need for clean-out.
Stimulants			
Senna	2–6 years old: 2.5–7.5 mL/day; 6–12 years old: 5–15 mL/day. Available as syrup, 8.8 mg of sennosides/5 mL. Also available as granules and tablets.	Abdominal pain, cathartic colon (possibility of permanent gut, nerve, or muscle damage). Idiosyncratic hepatitis, Melanosis coli, Hypertrophic osteoarthropathy, analgesic nephropathy.	Increased intestinal motility. Melanosis coli improves 4–12 mo after medications discontinued.
Bisacodyl	≥2 Years old: 0.5–1 suppository 1–3 tablets per dose. Available in 5-mg tablets and 10-mg suppositories.	Abdominal pain, diarrhea and hypokalemia, abnormal rectal mucosa, and (rarely) proctitis. Case reports of urolithiasis.	
Glycerin suppositories		No side effects.	

^aAdjust dose to induce a daily bowel movement for 1 to 2 months.

The successful treatment of constipation, especially with overflow incontinence, requires a family that is well organized, can complete time-consuming interventions, and is sufficiently patient to endure gradual

improvements and relapses. Close follow-up by telephone and by office visit is recommended. Some families may need counseling to help them manage this problem effectively.

Medication

It is often necessary to use medication to help constipated children achieve regular bowel movements (Table 7). A prospective, randomized trial showed that the addition of medications to behavior management in children with constipation is beneficial (38). Children who received medications achieved remission significantly sooner than children who did not. The use of laxatives was most advantageous for children until they were able to maintain regular toilet habits.

When medication is necessary in the daily treatment of constipation, mineral oil (a lubricant) or magnesium hydroxide, lactulose, sorbitol, polyethylene glycol (PEG) (osmotic laxatives), or a combination of lubricant and laxative is recommended. At this stage in the treatment of constipation, the prolonged use of stimulant laxatives is not recommended. Extensive experience with long-term use of mineral oil (39), magnesium hydroxide (40), and lactulose or sorbitol (40) has been reported. Long-term studies show that these therapies are effective and safe (9,40,41). PEG 3350 appears to be superior to other osmotic agents in palatability and acceptance by children (42–49). Preliminary clinical data in 12 infants suggest that administration of PEG 3350 to infants is effective with no adverse effects noted (50). Further studies are needed before widespread use can be recommended in infants. The doses and potential adverse effects of these medications are found in Table 7. Because mineral oil, magnesium hydroxide, lactulose, or sorbitol seem to be equally efficacious, the choice among these is based on safety, cost, the child's preference, ease of administration, and the practitioner's experience (Fig. 1, box 14).

A stimulant laxative may be necessary intermittently, for short periods, to avoid recurrence of an impaction (Fig. 1, box 15) (51). In this situation the use of stimulant laxatives is sometimes termed rescue therapy.

Maintenance therapy may be necessary for many months. Only when the child has been having regular bowel movements without difficulty is discontinuation considered. Primary care providers and families should be aware that relapses are common and that difficulty with bowel movements may continue into adolescence. Long-term follow-up studies have demonstrated that a significant number of children continue to require therapy to maintain regular bowel movements (52,53).

CONSULTATION WITH A SPECIALIST

Consultation with a pediatric gastrointestinal specialist becomes necessary when the therapy fails, when there is concern that an organic disease exists, or when management is complex (Fig. 1, box 20). A consultant can re-evaluate the child with nonresponding constipation, exclude an underlying organic process, perform specialized tests, and offer counseling. The pediatric gastroenterologist (Fig. 1, boxes 21–23) can review

previous therapies, consider using different or additional medications or higher doses of the current medications, and reassess previous management before performing additional studies (Fig. 1, box 23).

A careful review by the primary care practitioner of the differential diagnosis (Table 4) of the organic causes of constipation may be helpful at this time to determine which laboratory tests are indicated before referral to a specialist. It is recommended that the primary care physician consider whether the children who require evaluation by a specialist should have blood tests to identify evidence of hypothyroidism, hypercalcemia, celiac disease, and lead toxicity (Fig. 1, box 16). By having these tests ordered by the primary care provider just before referral to a pediatric gastroenterologist, patients who are found to have a medical problem that requires evaluation by a different subspecialist can be referred directly to the appropriate subspecialist. For example, a child with hypothyroidism can be referred directly to a pediatric endocrinologist.

Abdominal Radiograph and Transit Time

An abdominal radiograph is not indicated to establish the presence of fecal impaction if the rectal examination reveals the presence of large amounts of stool. A retrospective study in children manifesting encopresis showed that a moderate to large amount of stool found on rectal examination has high sensitivity and positive predictive value (greater than 80%) for fecal retention determined by abdominal radiograph, even using the radiologist's subjective interpretation (54). However, the specificity and negative predictive value were 50% or less. When the systematic scoring system developed by Barr et al. (19) was used for the presence of fecal retention on radiograph, the sensitivity of moderate to large amounts of stool on rectal examination improved to 92%, and the positive predictive value was 94%. However, the specificity remained at only 71%, and the negative predictive value was only 62% (55).

This suggests that, when there is doubt about whether the patient is constipated, a plain abdominal radiograph is reliable in determining the presence of fecal retention in the child who is obese or refuses a rectal examination, or in whom there are other psychological factors (sexual abuse) that make the rectal examination too traumatic. It may also be helpful in the child with a good history for constipation who does not have large amounts of stool on rectal examination (Fig. 1, box 23). In a recent study the value of the Barr score was compared with the colonic transit time. The Barr score was shown to be poorly reproducible, with low interobserver and intraobserver reliability, and there was no correlation with measurements of transit time (55).

Some patients have a history of infrequent bowel movements but have no objective findings of constipation. The history obtained from the parents and child may not be

entirely accurate (56). In these patients an evaluation of colonic transit time with radiopaque markers may be helpful (Fig. 1, box 25) (57). The quantification of transit time shows whether constipation is present and provides an objective evaluation of bowel movement frequency. If the transit time is normal, the child does not have constipation. If the transit time is normal and there is no soiling, the child needs no further evaluation (Fig. 1, box 30). In children who have soiling without evidence of constipation, the best results have been achieved with behavior modification, but in some instances psychological evaluation and treatment may be necessary (Fig. 1, box 29). If the transit study is abnormal or fecal impaction is present, further evaluation is needed (Fig. 1, box 26). When there is objective evidence of constipation and it is refractory to treatment, it is important to consider Hirschsprung disease (Fig. 1, box 28).

Hirschsprung Disease

Hirschsprung disease is the most common cause of lower intestinal obstruction in neonates and is a rare cause of intractable constipation in toddlers and school-age children (52,58–60). It is characterized by absence of ganglion cells in the myenteric and submucous plexuses of the distal colon, resulting in sustained contraction of the aganglionic segment. The aganglionic segment begins at the internal anal sphincter, extending orad in a contiguous fashion. In 75% of cases, the disease is limited to the rectosigmoid area. The bowel proximal to the aganglionic zone becomes dilated because of the distal obstruction.

The incidence of Hirschsprung disease is approximately 1 in 5000 live births. The most common associated abnormality is trisomy 21. More than 90% of normal neonates and less than 10% of children with Hirschsprung disease pass meconium in the first 24 hours of life (61,62). Thus, a delayed passage of meconium by a full-term infant raises the suspicion of Hirschsprung disease. Hirschsprung disease can have symptoms of bilious vomiting, abdominal distension, and refusal to feed, all of which are suggestive of intestinal obstruction. Short-segment Hirschsprung disease may go undiagnosed until childhood. Affected children have ribbon-like stools, a distended abdomen, and, often, failure to thrive. In rare cases constipation is the only symptom. Fecal soiling is even more rare and occurs only when the aganglionic segment is extremely short.

Enterocolitis, the most feared complication of Hirschsprung disease, may be its initial manifestation. Enterocolitis has initial symptoms of sudden onset of fever, abdominal distension, and explosive and at times bloody diarrhea (63,64). Occurring most often during the second and third months of life, it is associated with a mortality of 20%. The incidence of enterocolitis can be greatly reduced by a timely diagnosis of Hirschsprung disease.

The mean age at diagnosis decreased from 18.8 months in the 1960s to 2.6 months in the 1980s because of

physicians' vigilance, anorectal manometry, and early biopsy. However, in 8% to 20% of children, Hirschsprung disease remains unrecognized after the age of 3 years (65,66). Physical examination reveals a distended abdomen and a contracted anal sphincter and rectum in most children. The rectum is devoid of stool except in cases of short-segment aganglionosis. As the finger is withdrawn, there may be an explosive discharge of foul-smelling liquid stools, with decompression of the proximal normal bowel. In the older child with constipation, a careful history and a thorough physical examination are sufficient to differentiate Hirschsprung disease from functional constipation in most cases.

Once Hirschsprung disease is suspected (Fig. 1, box 28), it is recommended that the patient be evaluated at a medical center in which a pediatric gastroenterologist and a pediatric surgeon are available and where diagnostic studies can be performed. Delay in diagnosis increases the risk of enterocolitis. Rectal biopsy with histopathologic examination and rectal manometry are the only tests that can reliably exclude Hirschsprung disease. Rectal biopsies demonstrating the absence of ganglion cells in the submucosal plexus are diagnostic of Hirschsprung disease (67). The biopsies, obtained approximately 3 cm above the anal verge, must be deep enough to include adequate submucosa. The presence of hypertrophied nerves supports the diagnosis. However, in total colonic aganglionosis there is both an absence of ganglion cells and an absence of hypertrophied nerves. Occasionally, suction biopsies are not diagnostic, and a full-thickness biopsy is necessary.

Anorectal manometry (Fig. 1, box 31) evaluates the response of the internal anal sphincter to inflation of a balloon in the internal anal sphincter (68). When the rectal balloon is inflated, there is normally a reflex relaxation of the internal anal sphincter. In Hirschsprung disease this rectoanal inhibitory reflex is absent; there is no relaxation, or there may even be paradoxical contraction, of the internal anal sphincter. In a cooperative child, anorectal manometry represents a sensitive and specific diagnostic test for Hirschsprung disease. It is particularly useful when the aganglionic segment is short and results of radiological or pathological studies are equivocal. If sphincter relaxation is normal, Hirschsprung disease can be reliably excluded. In the presence of a dilated rectum, it is necessary to inflate the balloon with large volumes to elicit normal sphincter relaxation. In the child with retentive behavior, there may be artifacts caused by voluntary contraction of the external anal sphincter and the gluteal muscles. Sedation, which does not interfere with the rectoanal inhibitory reflex, may be used in newborns and uncooperative children. If manometry results are abnormal, diagnosis should be confirmed with a biopsy.

Although a barium enema is often performed as the initial screening test to rule out Hirschsprung disease, it is usually unnecessary beyond infancy (69). When stool is present in the rectum to the level of the anus, the barium enema provides no more useful information than

can be obtained with a plain radiograph. However, after the diagnosis of Hirschsprung disease has been made, the barium enema may be useful in identifying the location of the transition zone, provided that laxatives or enemas have not been administered before the study to clean out the colon. The barium enema may not show a transition zone in cases of total colonic Hirschsprung disease, or may be indistinguishable from cases of functional constipation when ultra-short-segment Hirschsprung disease is present.

Other Medications and Testing

If constipation is not resolved with the treatments outlined above, and Hirschsprung disease has been excluded, other therapies may be considered (Fig. 1, box 34). Clearly, treatment may be necessary for an extended period—months or years. Stimulant laxatives can be added for short periods. There is extensive experience with senna, bisacodyl, and phenolphthalein (70,71). However, phenolphthalein is no longer available in the United States because of concerns about its carcinogenic potential.

For most children with constipation the benefits of cisapride do not outweigh the risks (72–75). The committee does not recommend its use.

Biofeedback therapy has been evaluated in multiple open-label studies in which it was found to be efficacious (76). Results in some recent controlled studies, however, did not demonstrate long-term efficacy. Biofeedback may be beneficial for the treatment of a small subgroup of patients with intractable constipation (77–79). At times intensive psychotherapy may be needed. On rare occasion, hospital admission with behavioral therapy may be necessary.

Many conditions can cause constipation (Table 4). For children who remain constipated despite conscientious adherence to the treatments outlined, other tests may be indicated (Fig. 1, box 38). Magnetic resonance imaging (MRI) of the lumbosacral spine can demonstrate intraspinal problems, such as a tethered cord, tumors, or sacral agenesis (80). Other diagnostic tests such as anorectal manometry, rectal biopsy, colonic manometry, barium enema, and a psychological evaluation can be helpful. Colonic manometry, by providing objective evidence of colonic function, can exclude the presence of underlying neuropathy or myopathy and may guide therapeutic intervention (81–83). Barium enema can be useful to exclude the presence of anatomic abnormalities or of a transition zone. Full-thickness rectal biopsy can be useful to detect neuronal intestinal dysplasia or other myenteric abnormalities, including Hirschsprung disease. Metabolic tests, such as serum calcium level, thyrocalcitonin concentration, or thyroid function tests, can detect metabolic causes of constipation (84).

For children unresponsive to conventional medical and behavioral management consideration may be given to a time limited trial of cow's milk-free diet (85–88).

ALGORITHM FOR INFANTS LESS THAN 1 YEAR OF AGE

The evaluation of infants differs in some aspects from that of older children. Even in infancy, most constipation is functional. However, when treatment fails, when there is delayed passage of meconium (Fig. 2, box 4), or when red flags are present (Fig. 2, box, 8), particular consideration of Hirschsprung disease and other disorders is necessary. Hirschsprung disease has been described in detail. In a constipated infant with delayed passage of meconium, if Hirschsprung disease has been excluded, it is recommended that a sweat test be performed to rule out cystic fibrosis (Fig. 2, box 6). Constipation can be an early manifestation of cystic fibrosis, even in the absence of failure to thrive and pulmonary symptoms.

Special consideration should also be given to breast-fed infants in the first year of life. Greater variability in stool frequency occurs among breast-fed infants than in formula-fed infants (4,89,90). Unless suspicion of Hirschsprung disease is present, management of a breast-fed infant requires only reassurance and close follow-up if the infant is growing and breast-feeding normally and has no signs or symptoms of obstruction or enterocolitis.

Some important differences in treatment of constipation in infants include increased intake of fluids, particularly of juices containing sorbitol, such as prune, pear, and apple juices, which is recommended within the context of a healthy diet. Barley malt extract, corn syrup, lactulose, or sorbitol can be used as stool softeners. Light and dark corn syrups are not considered to be potential sources of *Clostridium botulinum* spores (91). Mineral oil, stimulant laxatives and phosphate enemas are not recommended. Because gastroesophageal reflux and incoordination of swallowing are more common in infants, there is greater risk of aspiration of mineral oil, which can induce severe lipoid pneumonia (92–94). Glycerin suppositories can be useful, and enemas are to be avoided.

NASPGHAN Constipation Guideline Committee

Susan S. Baker, MD, Chair Buffalo, NY	Carlo DiLorenzo, MD Columbus, OH
Gregory S. Liptak, MD Syracuse, NY	Walton Ector, MD Charleston, SC
Richard B. Colletti, MD Burlington, VT	Samuel Nurko, MD Boston, MA
Joseph M. Croffie, MD Indianapolis, IN	

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