

NCC Pediatrics Continuity Clinic Curriculum: Constipation Faculty



Goals & Objectives: To understand the differential diagnosis & management of constipation in the pediatric patient:

- Name 8 diagnoses in the differential diagnosis of constipation.
- Identify at least 5 "red flags" in the history of a patient presenting with constipation.
- List the medications (oral and rectal) used in constipation and their indications.
- Write out the behavioral and dietary management of constipation.

Pre-Meeting Preparation:

Please read/review the following enclosures:

- "Constipation" (PIR, AUG2020)
- Patient Resources: Parent Handout; Stool Diary; Management Plan

Conference Agenda:

- *Review* Constipation Quiz
- Complete Constipation Cases
- <u>"Hands-on" Activity:</u> Using a finger inserted into a balled fist, simulate the DRE findings of the following conditions: *Hirschsprung's, neurologic dysfunction, functional constipation, and normal anal tone.*

Post-Conference: Board Review Q&A

Extra-Credit:

- <u>Childhood Defecation Disorders</u> (*IFFGD*, 2006)—parent-friendly review
- Constipation (PIR, 2015)
- Prevalence, Symptoms, & Outcome of Constipation in Infants & Toddlers (JPeds, 2005)
- Evaluation & Treatment of Constipation: Recs from NASPGHN (CPG, 2014)
- <u>Evaluation and Treatment of Constipation in Infants and Children</u> (American Family Physician, 2014, alternate review article)
- Functional Constipation and the Gut Microbiome in Children: Preclinical and Clinical Evidence (Frontiers in Pediatrics, 2021)
- <u>Non-pharmacologic approach to pediatric constipation</u> (Complementary Therapies in Medicine, 2021)
- Constipation Action Plan

Constipation

Neal S. LeLeiko, MD, PhD,* Sarah Mayer-Brown, PhD,⁺ Carolina Cerezo, MD,* Wendy Plante, PhD⁺

*Department of Pediatrics, [†]Department of Psychiatry and Human Behavior, Hasbro Children's Hospital/Rhode Island Hospital, Warren Alpert School of Medicine at Brown University, Providence, Rl

Practice Gaps

- Pediatricians should be aware of the presenting signs and symptoms of constipation in children of different age groups.
- Pediatricians should understand the importance of addressing alarm signals, or "red flags," before entertaining any functional gastrointestinal diagnosis.
- 3. Pediatricians should understand that the management of constipation may require a behavioral approach.

Objectives After completing this article, readers should be able to:

- Understand the importance of distinguishing functional from organic causes of constipation.
- 2. Understand the relative roles of the Rome IV criteria for functional constipation-related disease and the *Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition* criteria for encopresis.
- 3. Understand the role of the pediatric psychologist in managing problematic patients with constipation.
- Appreciate the limitations of current research on pediatric constipation and how this affects treatment.

Abstract

Constipation in otherwise healthy infants and children is a common problem despite confusion about how to precisely define constipation and constipation-related disorders. Constipation may, rarely, be a sign or symptom of a more serious disease or a diagnosis defined only by its symptoms and without any structural or biochemical findings. In the latter case it is classified as a functional gastrointestinal disorder (FGID). FGIDs are defined as disorders that cannot be explained by structural or biochemical findings. The Rome Foundation has standardized diagnostic criteria for all FGIDs. The Rome criteria are based on the available research as well as the clinical experience of the Foundation's assembled experts. The most recent report, Rome IV, described clinical criteria and diagnostic tools and encouraged more rigorous research in the area of FGIDs. The true incidence and prevalence of constipation is difficult to know because it may be treated at home using home remedies or diagnosed at a visit to a primary care

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ABBREVIATIONS

APA	American Psychiatric Association
DSM-5	Diagnostic and Statistical Manual of
	Mental Disorders Fifth Edition
FC	functional constipation
FGID	functional gastrointestinal disorder
FI	fecal incontinence
NFI	nonretentive fecal incontinence
PEG	polyethylene glycol

provider or to a subspecialist pediatric gastroenterologist. The most recent attempts to define the prevalence of all pediatric FGIDs have been made using the Rome IV criteria. The defined FGID entities that may be associated with the complaint of constipation are infant dyschezia, functional constipation, and nonretentive fecal incontinence. The term *encopresis*, omitted from Rome IV, is defined by the American Psychiatric Association (APA) in the *Diagnostic and Statistical Manual of Mental Disorders*, *Fifth Edition*. The 3 Rome-defined (constipation-related) entities and the APA entity of encopresis are the focus of this review.

INTRODUCTION

Constipation is commonly defined by the following symptoms: hard stools, painful stools, and stools that are difficult to pass, infrequent, or incomplete, dry, or hardened. For patients presenting with the symptoms of constipation, what matters is the alleviation of symptoms and not what we call this condition. However, for those focused on studying the problem, a consistent and rational categorization of these entities is necessary.

Practically speaking, the pediatric provider must distinguish between patients who require specialized evaluation (and potentially specialized medical/surgical intervention) and those with functional gastrointestinal disorders (FGIDs), who should be managed with a focus on fecal elimination using reassurance and no or minimal medical intervention. Toward that goal, patients who exhibit alarm signals, or "red flags" (Table I), should be evaluated appropriately and should not be considered "functional" until physical and biochemical normality are evident.

The Rome Foundation has standardized diagnostic criteria for all FGIDs. The original Rome criteria were based on the available research as well as the clinical experience of the Foundation's assembled experts. Those experts originally gathered in Rome to classify FGIDs in adults. Subsequently, pediatric experts were included in the Rome meetings, and specific criteria for FGIDs occurring in childhood have been identified. The evidence base underlying the development of the specific criteria has been expanding but is yet to have the rigor that most clinicians would generally demand for known biologically defined diseases. The most recent report, Rome IV, (2) described clinical criteria and diagnostic tools and encouraged more rigorous research in the area of FGIDs.

The Rome Foundation uses symptom-based definitions for FGIDs in an effort to minimize unnecessary testing (Table 2). Recognizing that not all clinical situations allow for the same level of confidence in the interpretation of symptoms, Rome IV added more clinical flexibility to its FGID definitions by adding the following statement: "After appropriate medical evaluation the symptoms cannot be attributed to another medical condition." The aim is to allow the physician to perform selective or no testing to establish the presence of an FGID. (4)

SYMPTOM-BASED DESCRIPTIONS

Infant Dyschezia (Ages 0–9 Months)

Infants with dyschezia appear completely well in the office. Parents may report isolated episodes of distressing straining with crying, perhaps with the baby's face transiently turning deep red, in an apparent effort to defecate. The symptoms may last for 10 minutes but usually are resolved on their own by 20 minutes. The result is sometimes a soft stool, but other times it may be associated with failure to pass a stool during the episode described. Failure to pass a stool leads the parents to believe that the baby is experiencing constipation. Babies are completely better after these acute episodes. These episodes can be very distressing for parents. The onset of these symptoms is generally in the first few months of life, and they typically do not last for more than a month. The diagnosis of infant dyschezia is restricted to infants 9 months or younger (Table 3).

Babies with infant dyschezia are generally well nourished and have no red flags. The occurrence of vomiting, especially if accompanied by episodes of drawing up of the infant's legs, or episodes followed by lethargy or seeming exhaustion may require ruling out intussusception. Diarrhea with or without blood is an alarm signal.

Parents often report resorting to rectal stimulation or the use of a glycerin suppository to provide relief for their infant. These interventions should be discouraged (see later herein). Pelvic floor movement, abdominal muscular contractions, and relaxation of the anal sphincters must all be coordinated to achieve successful expulsion of the stool. The apparent cause of infant dyschezia is the newborn's inability to coordinate the voluntary and involuntary body movements necessary to expel stool. Implicit in this explanation is that this is a developmental skill that will be learned in time. Infants with infant dyschezia should be considered to have not yet learned this process, which, if unimpeded by illconsidered interventions, should be mastered by 9 months of age.

Functional Constipation in the Infant, Toddler, and Older Child (Ages 0–18 Years)

Although it is difficult to know the true prevalence of functional constipation (FC) in childhood, across a range of cultures and ages, it seems to be approximately 12% to 18% (3)(4) (Table 2). FC may begin at any time in childhood, but the highest incidence seems to be associated with toilet training. Boys are reported to have a greater incidence of soiling. (4) There is consensus agreement that the following 3 factors may predispose to FC:

- Presumed dietary inadequacy suggesting that too much or too little or the wrong components or combination of components in the feed is responsible. For example, the inclusion of fiber-containing foods has strong advocacy among some, but definitive evidence of its role in FC is lacking; milk has been implicated as a cause of FC, but recent reviews express skepticism, at least about the frequency with which milk is a culprit; children with autism or other severe emotional disturbance may have extremely restricted and unusual diets.
- Cognitive decisions on the part of the infant, toddler, or child that may be self-limiting, requiring no or minimal intervention.
- Cognitive decisions on the part of the infant, toddler, or child that may require some form of intervention to treat the resulting FC. For example, providing a note requesting that a school-age child be allowed to use the nurse's bathroom may diminish the apprehension of using the students' toilet.

Although parents (and patients) tend to report hard painful bowel movements, they sometimes report loose stools, which may represent the overflow of watery stool past a bolus of hard stool (paradoxical diarrhea). Voluntary withholding has a significant role at the time of toilet training and may play a role throughout the school years. It is considered that unpleasant experiences associated with defecation (ie, pain or needing to take a break from enjoyable activities) reinforce stool retention and promote a vicious cycle. Social circumstance (ie, discomfort with using the school toilet) may also promote stool withholding. When stool is withheld, water absorption is increased, making the stool harder and drier and more difficult to pass, with or without pain. This, in turn, creates a vicious cycle by promoting voluntary withholding. When there is an accumulation of stool in the rectum, there is a reflex decrease in gastric emptying, which may be associated with different symptoms, including upper gastrointestinal tract symptoms, such as abdominal distention, abdominal pain, loss of appetite, and nausea. The frequency, order of occurrence, and exact physiology of these symptoms is unclear. In the absence of warning signs, such as those in Table 1, the diagnostic criteria for FC should be considered (Table 4).

Primary care physicians' evaluations should include a comprehensive history and physical examination and must exclude alarm signals (Table 1). Of note, it is frequently suggested that urinary tract symptoms are more common in children with FC. van Summeren et al (5) noted in their review of urinary tract or "bladder" symptoms that there was substantial variability in reports of co-occurrence with FC. They concluded that "we cannot make any definitive statements on the prevalence of bladder symptoms in children with FC." They did conclude that symptoms referable to the urinary tract clearly occurred with "significant frequency" in children with FC and that children with FC are more likely to have [urinary tract] symptoms than are children with FC.

Clearly, symptoms related to the urinary tract are red flags that need to be assessed and resolved regardless of any real or hypothetical relationship to FC. (5) When the functional nature of the problem is considered, the Rome IV criteria for FC should be applied. Similarly, when appropriate, the *Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5)* (6)–defined diagnosis of encopresis may need to be considered (Table 5).

The presence of a large fecal mass in the rectum is one of the criteria for FC. Diagnosis of fecal impaction has historically been made based on appropriate history and digital rectal examination. The role of the digital rectal examination is now unsettled. Some physicians are comfortable with the use of digital rectal examination to determine the presence of fecal impaction in the rectum. Other physicians regard this as an unnecessary, uncomfortable (at best), and overly invasive examination for constipation. The joint report of their routine examination for constipation. The joint report of the North American and European societies of pediatric gastroenterology, hepatology, and nutrition (7) recommended a digital rectal examination if the diagnosis of FC is uncertain, ie, if only one of the (then) Rome III criteria was present, in the presence of specific alarm signals/red flags, and in the presence of intractable constipation. More recently it has been suggested that performing a digital rectal examination likely provides little additional information for the examiner. (8)

The same joint report (7) also suggested that the routine use of abdominal radiography has no role in the diagnosis of FC but that it is of use in a child in whom fecal impaction is suspected but physical examination is unreliable or not possible. Some consider that the role of the abdominal radiograph is to confirm a successful cleanout in previously impacted children as well as when a digital examination is impractical for whatever reason and when there seems to be a discordance between symptoms and examination findings. There seems to be only a minimal role for using abdominal radiography in the emergency department for diagnosis.

The overriding goal of evaluation is for the health-care provider to assure herself or himself, so that the provider can with confidence assure the parents and patient that the constipation is functional and will respond to the measures offered if followed.

Nonretentive Fecal Incontinence (Ages 4–18 Years)

Nonretentive fecal incontinence (NFI) is distinguished from FC by having normal numbers of stools and, if tested, normal colonic transit times (which are generally prolonged in FC). Patients with NFI do not have to be disimpacted because they do not have large accumulations of stool as a mass in their rectum. They can, however, be expected to have either very significant psychosocial problems or a significant neurologic lesion. Diagnostic criteria are listed in Table 5.

Encopresis

The DSM-5 continues to use the term *encopresis* with the diagnostic criteria listed in Table 5. Encopresis is considered an elimination disorder, along with enuresis. The 2 diagnoses—encopresis and enuresis—may coexist.

Although the actual symptoms associated with the American Psychiatric Association (APA) diagnosis of encopresis are distinct, so are those associated with NFI as defined by the Rome Foundation in Rome IV. For clarity, the criteria for each of these diagnoses are presented side-by-side in Table 5.

For the clinician, it may be helpful to appreciate that the Rome criteria focus on constipation as the main feature that distinguishes FC and NFI. The defining feature of encopresis is soiling. Distinctions between NFI and soiling are not always as clear as one might expect. It may be helpful to distinguish children who are soiling undergarments because of poor perianal hygiene after stooling from those who experience leakage (ie, overflow incontinence) and those who have partial or complete bowel movements in their undergarments.

Briefly, the APA *DSM*-5 diagnosis "encopresis—without constipation and overflow incontinence" seems to match the Rome IV diagnosis of NFI. However, the APA *DSM*-5 diagnosis "encopresis—with constipation and overflow incontinence" does not match the Rome IV diagnosis of FC because the problem of incontinence is not included in FC, and it does not match the diagnosis of NFI because there is fecal retention (ie, constipation). It, therefore, has its own category, ie, "encopresis—with constipation and overflow incontinence."

FGID TREATMENTS

Treatment of Infant Dyschezia

If the diagnostic criteria listed in Table 3 are met and the diagnosis of infant dyschezia is made, it is best to advise nothing more than comforting the baby during crying episodes. Gentle efforts at consoling the infant, including holding, cuddling, and massaging the abdomen, may relieve the stress on the baby and/or parents. Rectal stimulation has the risk of conditioning the infant to wait for stimulation to defecate. Depending on the vigor of the stimulation, there is the risk of physically hurting or of causing other harm. Changing infant formula (or moving away from breastfeeding) will not speed the process of resolving infant dyschezia. Laxatives do not have a role in the treatment of these infants. Reassurance is necessary because time will eventually resolve the problem.

Treatment of FC

There is substantial agreement that the first step in treating the child with FC is adequate disimpaction. Without adequate cleaning out of retained stool, we believe that successful treatment is unlikely. In practice there is wide variability in the actual approach that practitioners use to accomplish this. Particularly challenging patients may need to be hospitalized to be cleaned out. This can be accomplished at a time that accommodates both staffing and parents' work schedules.

Treatment can be laborious, and we cannot urge strongly enough that patient/parent education is critical. We also suggest that practitioners provide a handout of their own preferred approach that reflects their experience and comfort level to reinforce their verbal teaching. Because systematic reviews (7)(9) suggest a satisfactory response at 12 months of only 50%, it is even more important that the

TABLE 1. Alarm Signals (or "Red Flags") and Examples of Potential Associated Disorders that May Have Constipation as a Symptom

SYMPTOM OR FINDING	EXAMPLES OF POSSIBLE CAUSES OR ASSOCIATIONS		
Passage of meconium >48 h Constipation in the first month after birth Family history of Hirschsprung disease	Hirschsprung disease		
Family history of autoimmune disease such as celiac disease, type 1 diabetes	Celiac disease		
Family history of cystic fibrosis	Pancreatic insufficiency Meconium ileus equivalent Chronic pancreatitis		
Chromosomal abnormality (ie, Down syndrome)	Hirschsprung disease (~2.6% of patients with Down syndrome [1]) Intestinal malformation (atresia) Intestinal web		
Physical asymmetry	Intestinal malformation		
Ribbon stools	Rectal narrowing or atresia with fistula		
Blood in the stool in the absence of anal fissures	Inflammation (NEC), milk allergy, polyp, intussusception		
Weight loss or inadequate weight gain	Hirschsprung disease Numerous medical conditions		
Lagging growth	Hirschsprung disease Chronic illness		
Bilious vomiting	Intestinal obstruction		
Severe abdominal distention	Intestinal obstruction; ileus; severe obstipation Hirschsprung disease		
Episodes of inconsolable crampy abdominal pain (especially if followed by sense of exhaustion)	Intussusception Volvulus or torsion		
Urinary tract symptoms	Urinary tract disorder including obstruction or infection		
Abnormal thyroid gland/function	Hypothyroidism		
Abnormal position of the anus	Imperforate anus with fistula Anterior displacement with malpositioning of colon		
Absent anal or cremasteric reflex Decreased lower extremity strength/tone/ reflex	Spinal cord lesions (including sacrococcygeal teratoma)		
Sacral dimple Tuft of hair on spine Gluteal cleft deviation			
Dilated colon +/- ureters	Pseudo-obstruction disorders including neuropathies		
Medication consumption	Iron intake, diuretics (dehydration), antispasmodics, calcium-containing medications, aluminum antacids, opioids, SSRIs and TCAs, unknown drugs, and herbals		

Continued

TABLE 1. (Continued)

SYMPTOM OR FINDING	EXAMPLES OF POSSIBLE CAUSES OR ASSOCIATIONS
Restricted diet	Low fiber Unbalanced intake
Physical activity	Too sedentary, any chronic illness

NEC=necrotizing enterocolitis, SSRI=selective serotonin reuptake inhibitor, TCA=tricyclic antidepressant.

parents and (older) patient be educated to understand what is involved with their problem and the elements necessary for successful treatment. Potential medical therapies for treating fecal impaction are listed in Table 6.

Polyethylene glycol (PEG) 3350 is an agent that is soluble in water but minimally absorbed and seems to be inert (Table 7). It works by retaining water in the intestine and producing softer stools. It is commonly available over the counter and is sold under several different names. As with most of the commonly used agents for inducing bowel movements in children, it has not been approved for use in children. It is, however, widely used. A recent randomized, multicenter study (11) found that PEG 3350 was more effective than lactulose in achieving cleanout in FC. In addition, there were fewer adverse effects noted (principally, bloating and abdominal pain). The literature suggesting that the use of PEG 3350 is superior to lactulose is not settled because dosing and other conditions are not always comparable. It is reasonable to consider that the evidence remains inconclusive, and practitioner comfort and experience should guide the choice. Common variations in bowel disimpaction include several weeks of daily dosing as tolerated (ie, avoiding paradoxical diarrhea, especially with the use of PEG 3350). Some practitioners include the use of an oral laxative (10) (see Table 8 for often used laxatives), which may be continued for up to 2 to 4 weeks.

If the oral route is rejected or unsuccessful, the use of daily enemas may be substituted. In our experience, there is a disparity in knowledge and comfort regarding the use of enemas in both parents and medical providers. This should be taken into consideration before prescribing enemas. Some parents and providers will find biscodyl suppositories to be a more standardized and acceptable alternative to enemas. Their correct method of insertion can be demonstrated on the clothed child in the left lateral position on the examination table. The rectal suppositories melt in 10 to 15 minutes at body temperature and generally induce a bowel movement within 30 minutes to I hour; their effect is complete after 2 hours (minimizing the effect on the child's schedule). They seem to be safe and effective and can be repeated daily, over 3 days, to accomplish the cleanout. Similar to all of the laxatives used, systematic safety studies in children are lacking, and caution is always warranted.

In general, it seems that most experts prefer nonstimulant laxatives, such as PEG 3350, lactulose, and Milk of Magnesia. These tend to be the gentlest, when used at an appropriate dose, with the goal of slowly softening any fecal mass and ultimately enabling passage. Biscodyl suppositories are expected to work within I to 2 hours.

Failure to achieve disimpaction on an ambulatory basis requires hospitalization. There is anecdotal data to suggest that hospitalizations for nasogastric tube placement and nasogastric lavage have increased greatly during the past decade; regardless, it is a too frequent necessity. The procedure involves placing the nasogastric tube in the hospital and instilling a PEG 3350 solution at the initial rate of 10 mL/kg per hour and gradually increasing to 20 and then up to 40 mL/kg per hour (if vomiting ensues, then the rate must be adjusted accordingly) (Table 7). For the impacted child it can take more than 24 hours before the effluent is clear. In instances where there is lack of clarity regarding successful cleanout, an abdominal radiograph to confirm the lack of stool left in the colon may help. Manual disimpaction is so rarely necessary that few become proficient. If necessary, it should be performed under general anesthesia (or deep sedation) to minimize patient trauma. Colonic perforation is a well-known complication of digital disimpaction, and those who perform the procedure should be prepared to deal with that serious sequelae.

Once a successful cleanout has been accomplished, it is necessary to maintain the "clean" rectum to allow the distended rectum to regain tone (Table 8).

Based on very limited evidence, it is reasonable to suggest that "normal" fluid intake and a fiber-rich diet help maintain

TABLE 2. Prevalence of Specific Constipation-Related Disorders Using Rome IV Criteria

DIAGNOSIS	INFANT <1 Y (N = 58)	TODDLER 1 TO <4 Y (N 238)	CHILD/ADOLESCENT (≥4 Y) (N = 959)
Infant dyschezia	<1%		
Functional constipation	12.1%	18.5%	14.1%
Nonretentive fecal incontinence			0.2%
Any functional gastrointestinal disorder ^a	37.9%	21.4%	25%

^aFunctional gastrointestinal disorders include, in addition to infant dyschezia, functional constipation, and nonretentive fecal incontinence, infant regurgitation, infant colic, cyclic vomiting syndrome, rumination, functional diarrhea, functional dyspepsia postprandial distress syndrome, functional dyspepsia epigastric pain syndrome, functional dyspepsia unspecified, irritable bowel syndrome, functional abdominal pain not otherwise specified, aerophagia, abdominal migraine, functional vomiting, functional nausea.

Adapted from Robin SG, Keller C, Zwiener R, et al. Prevalence of pediatric functional gastrointestinal disorders utilizing the Rome IV criteria. J Pediatr. 2018;195:134–139. (3)

normal bowel function. The efficacy of using fiber remains very controversial, and when we recommend fiber, we usually suggest that the entire family be included in a diet regimen that calls for adding 2 to 3 tablespoons of unprocessed bran flakes per day. This routine has both strong advocates and detractors. Between the cleanout and establishing a regular bowel routine it may be a good idea to offer maintenance dosing of PEG 3350, 0.4 g/kg per day that should be tapered (after I–3 months) as symptoms resolve and a more acceptable bowel habit ensues. Although not approved for use in children, lactulose is used by many as an alternative to PEG 3350, especially in younger infants.

Relapses are so common that they must be anticipated. When they occur, the clinician must assess the acceptability of the original plan and decide whether a different or more rigorous maintenance regimen will be necessary after the next cleanout/disimpaction. Relapses may also be attributed to factors affecting the individual child (eg, refusal to take medications, purposeful stool withholding) and/or parent factors (eg, lack of understanding about regimen). In these cases, the assistance of an experienced and sympathetic nurse or, on occasion, referral to a behavioral psychologist may help improve adherence to the prescribed regimen.

The approach to treating the relapse is to repeat the successful cleanout. Generally, on completion of the cleanout, a more prolonged maintenance therapy period seems appropriate. In general food allergy testing, including milk allergy, is not routinely recommended, and further blood work, in the absence of alarm signal symptoms, is not warranted. For concerns about Hirschsprung disease, rectal biopsy remains the gold standard, but I suction rectal biopsy can be expected to yield a definitive diagnosis in only 65% of patients, and a second biopsy will increase the yield to only 75%. (12) In patients with Down syndrome, where the risk of Hirschsprung disease is reported to be 2.62%, (I) it may be prudent to refer to a pediatric surgeon for full-thickness biopsy after I inadequate suction rectal biopsy. As with rectal biopsy, barium enema and anorectal manometry should not be routinely considered unless there are red flags concerning for Hirschsprung disease (Table I).

Treatment of NFI and Encopresis—Without Constipation and Overflow Incontinence

In these conditions there is no evidence of constipation on physical examination or by history. Children with NFI (and those with encopresis without constipation and overflow incontinence) have normal colonic and anorectal motility. They have normal numbers of stools and are not constipated. NFI is associated with significant emotional distress and impulsive behavior, and there are often psychological comorbidities, including learning and behavioral

TABLE 3. Rome IV Diagnostic Criteria for Infant Dyschezia

MUST INCLUDE IN AN INFANT <9 MO OF AGE:

- 1. ≥10 min of straining and crying before successful or unsuccessful passage of soft stools
- 2. No other health problems

TABLE 4. Rome IV Diagnostic Criteria for Functional Constipation

MUST INCLUDE ≥2 OF THE FOLLOWING OCCURRING AT LEAST ONCE PER WEEK FOR A MINIMUM OF 1 MO WITH INSUFFICIENT CRITERIA FOR A DIAGNOSIS OF IRRITABLE BOWEL SYNDROME:

1. ≤2 defecations in the toilet per week in a child with developmental age of ≥4 y
2. ≥1 episode of fecal incontinence per week (after toilet training)
3. History of retentive posturing or excessive volitional stool retention
4. History of painful or hard bowel movements
5. Presence of a large fecal mass in the rectum
6. History of large-diameter stools that can obstruct the toilet
After appropriate medical evaluation, the symptoms cannot be explained by another medical condition

Adapted from Hyams JS, Di Lorenzo C, Saps M, Shulman RJ, Staiano A, van Tilburg M. Functional disorders: children and adolescents. Gastroenterology. 2016;S0016-5085(16)00181-5. (2)

disorders. (13) Although history of physical and sexual abuse may be associated with NFI, high-quality data on this subject are lacking, and associations may be conjectural. The approach to treating these children requires co-treatment by a physician or pediatric nurse practitioner to manage medically related issues and a pediatric psychologist to manage emotional and behavioral factors contributing to NFI.

Treatment of Encopresis—With Constipation and Overflow Incontinence

In this condition, there is evidence of constipation on physical examination or by history. Children with encopresis with constipation and overflow incontinence seem likely to have prolonged colonic transit. In that sense they share the physical characteristics of the child with FC. They may share the behavioral concerns of the patient with NFI. As with NFI and encopresis without constipation, the approach to these children can be optimized with co-treatment by a physician or pediatric nurse practitioner who manages the constipation and other medically related issues and a pediatric psychologist who manages the behavioral issues that are contributing to or maintaining constipation and fecal incontinence.

BEHAVIORAL APPROACH TO CHILDREN WITH FC, NFI, AND ENCOPRESIS

Psychosocial correlates and consequences of childhood constipation and fecal incontinence have been understudied. In general, the child with only FC is rarely a concern for the psychologist unless there are significant other comorbid conditions. Children with fecal incontinence have been shown to have more mood concerns, disruptive behaviors, attentional problems, social problems, and school difficulties than children without fecal incontinence (14)(15)(16) Furthermore, these children experience poorer quality of life than children with other gastrointestinal disorders, such as inflammatory bowel disease and reflux. (17) It seems that the presence of fecal incontinence in children is related to significant decreases in their quality of life compared with children with FC alone. (18) Not surprisingly, both FC and fecal incontinence also affect family functioning. Parents report increased parent-child conflict about toileting, (19) as well as a high degree of parental stress, (13) which may be related to the burden of cleaning up after accidents and children's lack of candor about soiling. (20)

Although most children who experience soiling do not have psychosocial problems in the clinical range, (14) even subclinical anxiety, attentional problems, and oppositional behaviors can interfere with toileting and families' abilities to adhere to their treatment regimen (eg, anxiety about painful stools, refusal to stop playing to use the bathroom). Therefore, referral for multidisciplinary treatment may be helpful for families when high anxiety, poor executive functioning, oppositional behavior, or parent-child conflict have been identified, as well as for the approximately 40% of children with FC in pediatric practice who do not improve after 2 months of medical management. (21) Physicians should also consider referring children with FC and developmental disabilities and children with autism spectrum disorder because these children have an increased risk of toileting difficulties (22)(23) and may benefit from behavioral treatment in conjunction with medical

TABLE 5. Rome IV NFI vs DSM-5 Encopresis

ROME IV DIAGNOSTIC CRITERIA FOR NFI	DSM-5 DIAGNOSTIC CRITERIA FOR ENCOPRESIS
 Children have a developmental age of ≥4 y 	• Chronological age is \geq 4 y (or equivalent developmental level)
 Defecation into places inappropriate to the social context 	Repeated passage of feces into inappropriate places (eg, clothing, floor) whether involuntary or intentional ≥1 such event occurs each month for a ≥3 mo
• No evidence of fecal retention	 The diagnosis of encopresis should specify whether: Without constipation and overflow incontinence: There is no evidence of constipation on physical examination or by history Subtype – Feces are likely to be of a normal form and consistency, and soiling is intermittent. Feces may be deposited in a prominent location. This is usually associated with the presence of oppositional-defiant disorder or conduct disorder or may be the consequence of anal masturbation. Soiling without constipation seems to be less common than soiling with constipation. With constipation and overflow incontinence: There is evidence of constipation on physical examination or by history Subtype – Feces in this type are characteristically (but not invariably) poorly formed and leakage can be infrequent to continuous, occurring mostly during the day and rarely during sleep. Only part of the feces is passed during toileting, and the incontinence resolves after treatment of the constipation
 After appropriate medical evaluation, the symptoms cannot be explained by another medical condition 	 The behavior is not attributable to the physiologic effects of a substance (eg, laxatives) or another medical condition except through a mechanism involving constipation

DSM-5=Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition, NFI=nonretentive fecal incontinence.

treatment of constipation and fecal incontinence. Before making a referral for behavioral treatment, it is important for physicians to assess whether children are already receiving behavioral health services, and if so, whether toileting has ever been a focus of treatment. Families may continue to work with their mental health provider while also participating in shortterm treatment for FC and fecal incontinence with a pediatric psychologist or other mental health provider with expertise in this field.

Behavioral treatment of FC and fecal incontinence includes a variety of components, including education, monitoring stool frequency and regimen adherence through use of a diary, initiation of a toileting routine that includes scheduled toilet sits, and rewards for appropriate toileting behavior. (24)(25) Pediatric psychologists will reinforce education about gastrointestinal system functioning, factors that contribute to and maintain constipation, and rationale for medications prescribed by the gastroenterologist. It is important to include children in these conversations to increase their understanding and motivation, as well as to reduce stigma and embarrassment about toileting issues. Families will then begin to track the frequency of bowel movements and soiling accidents, as

well as any times that children sit on the toilet. These data are helpful in creating a structured toileting schedule. Toilet sits are typically timed to occur after meals to take advantage of the gastrocolic reflex and/or around times that children typically experience fecal incontinence. Scheduled sits should be time limited (ie, no more than 5-10 minutes) to reduce the chance that children will become resistant to completing these. Children should be comfortable and relaxed while sitting on the toilet. Therefore, children are encouraged to sit with their feet supported, and psychologists may work with children to learn diaphragmatic breathing strategies to help relax pelvic floor and gluteal muscles.

Pediatric psychologists will work with caregivers to create a reinforcement plan to increase children's motivation to complete all steps of their toileting plan (eg, cooperating with scheduled sits, defecating in the toilet, reducing fecal incontinence). Rewards may include stickers, a small toy, access to electronics, or one-on-one time with a caregiver. Of note, only rewarding children for "clean underwear" (ie, not soiling) may inadvertently reinforce children's tendency to withhold their stool, subsequently leading to further constipation. In combination

ROUTE	THERAPY	DOSING
Oral	Polyethylene glycol 3350 mixed as 17 g/8 oz of water, juice, or other liquid	>3 y of age 1–1.5 g/kg per day for 3 d, may be continued for up to a week. A large daily dose of up to 100 g/d for 1–2 d; lower doses for longer periods, up to 12 wk, have been used.
	Magnesium citrate	4 mL/kg per day – generally given on 2 consecutive evenings
	Lactulose	1 mL/kg, twice a day, for up to 12 wk then tapered over 4 wk
Rectal	Normal saline enema	10 mL/kg, generally given on 2–3 consecutive evenings
	Sodium phosphate enema	≥12 y: 4.5 oz enema on 2–3 consecutive evenings
		5-11 y: 2.25 oz on 2-3 consecutive evenings
		2-4 y: 1 oz on 2-3 consecutive evenings
	Mineral oil enema	≥12 y: 4.5 oz on 2–3 consecutive evenings
		2-11 y: 2.25 oz on 2-3 consecutive evenings
	Biscodyl suppositories	1-2 suppositories per day for 2-3 consecutive evenings, may be repeated in 1-2 wk

TABLE 6. Medical Therapies for Fecal Disimpaction at Home

Note that systematic dosage and safety studies on these medications have not been rigorously performed in the pediatric age range. Adapted from Colombo JM, Wassom MC, Rosen JM. Constipation and encopresis in childhood. Pediatr Rev. 2015;36(9):392–401. (10)

with tangible reinforcers, caregivers are taught to give children more attention for appropriate toileting behaviors and less attention for undesired behaviors (eg, withholding, soiling to help decrease battles around toileting).

Treatment may also include other components, depending on the children's presenting problems. Other targets of treatment may consist of increasing adherence to prescribed medications with structured scheduling and/or reward systems, decreasing bathroom or toiletrelated anxiety through graduated exposure, coordinating with schools to initiate toileting accommodations (eg, access to a private bathroom, prompted sit after lunch/ snacks), and expanding the variety of foods with fiber that children will eat through exposure therapy and behavior modification.

The goal of treatment is to increase defecation in the toilet, decrease fecal incontinence, and eventually wean off of medications when medically appropriate. Older research suggested that behavior therapy did not improve children's outcomes above and beyond medical treatment. (7)(26) However, there is a growing body of literature that has examined models of multidisciplinary treatment that suggests that combined treatment improves children's prognosis. For instance, Freeman and colleagues (27) conducted a systematic review and found that children experienced approximately 2.81 fewer soiling episodes per week when behavioral therapy was included in treatment. Furthermore, combining medical and behavioral interventions demonstrated a reduction in fecal incontinence, abdominal pain, and use of diapers in an integrated care clinic. (25) Not only is combined treatment possibly more efficacious than

TABLE 7. Polyethylene Glycol 3350 for Inpatient Bowel Disimpaction

MIX 255 G IN 64 OZ OF BALANCED MULTI-ELECTROLYTE SOLUTION (OR 32 G IN 8 OZ)

Bowel cleanout (oral)	Administer at approximately 8 oz per hour until stool is clear
Bowel cleanout (nasogastric tube)	Administer at ~10 mL/kg per hour to start and increase as tolerated by 10–20 mL every 1–2 h to a maximum of ~40 mL/kg per hour

THERAPY TYPE	THERAPY	DOSING	
Osmotic laxatives	Polyethylene glycol 3350	0.5–1.5 g/kg per day (start low and titrate up with a maximal dose of 17 g/day)	
	Lactulose	1–3 mL/kg per day in 2 divided doses	
	Magnesium hydroxide	<2 y: 0.5 mL/kg per day	
		2–5y: 5–15 mL/day (before bedtime or divided)	
		6–11 y: 15–30 mL/day (before bedtime or divided)	
		≥12 y: 30–60 mL/day (before bedtime or divided)	
Stool softeners/lubricants	Docusate	5 mg/kg per day (400 mg maximum)	
	Mineral oil	1–3 mL/kg per day divided into 2 doses	
Stimulant laxatives	Senna	1–2 y: 2.2–4.4 mg/day as 1 or 2 divided doses	
		>2-6 y: 4.4–6.6 mg/day as 1 or 2 divided doses	
		>6–12 y: 8.8–13.2 mg/day as 1 or 2 divided doses	
		>12 y: 17.6–26.4 mg/day as 1 or 2 divided doses	
	Biscodyl	3–12 y: 5–10 mg/day	
		>12 y: 5–15 mg/day	
Chloride channel activators	Lubiprostone Linaclotide	New prescription medications approved only for adults with very limited pediatric (off-label) experience	

TABLE 8. Maintenance Therapy for Chronic Constipation

Adapted from Colombo JM, Wassom MC, Rosen JM. Constipation and encopresis in childhood. Pediatr Rev. 2015;36(9):392-401. (10)

medical treatment alone, but behavior therapy plus laxatives has been shown to actually reduce the need for medical appointments (28) and is less expensive than standalone medical treatment. (29) Therefore, we recommend that pediatricians and pediatric gastroenterologists continue to follow their patients with constipation closely to monitor and manage medications while families also work with a pediatric psychologist to increase the success of treatment.

CONSTIPATION AND THE INTESTINAL MICROBIOME

Recent studies (30) of the microbiota inhabiting the intestines of patients with chronic constipation have suggested that changes may occur (dysbiosis) that may be related to constipation. It is tantalizing to suspect that changing the gut bacteria may one day become a therapy.

As of the writing of this review we have encountered many parents who can ill afford the cost of various unproven probiotics, prebiotics, and synbiotics. These potential therapies, even fecal microbiome transplant, are tantalizing but totally unproven. They are likely to become a major section of future versions of this review, but for now they cannot be considered efficacious.

Summary

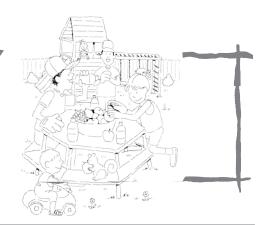
- Based on expert opinion, case reports, and reasoning from first principles (evidence quality "D"), (2) our view of functional gastrointestinal disorders (FGIDs) has been greatly enhanced. The work performed by the Rome Foundation has been modified, and their previous insistence that to diagnose an FGID there needed to be no evidence of organic disease has appropriately been updated to allow the diagnosis if "after appropriate medical evaluation the symptoms cannot be attributed to another medical condition." This is important progress because it allows the clinician flexibility regarding the amount (if any) of testing deemed appropriate. It becomes easier for us to understand and to accept that FGIDs can coexist with other organic disorders, such as inflammatory bowel disease.
- Based on expert opinion, case reports, and reasoning from first principles (evidence quality "D") and on some observational studies (evidence quality "C"), the American Psychiatric Association definition of encopresis has been partially aligned with the Rome criteria diagnosis of nonretentive fecal incontinence, (2)(6) and we can look forward to greater harmony in these definitions in the future.

- Based on expert opinion, case reports, and observational studies (evidence quality "D" and "C"), (6)(7)(13)(25)(27) the importance of the biobehavioral model continues to expand. Furthermore, the biobehavioral model will be further advanced as the field of neurogastroenterology matures and future basic science and clinical research begins to clarify how the brain-gut interactions affect the FGIDs. The role of the gut microbiome and its effect on gut signaling and its consequences will no doubt yield many new questions and, we hope, a few answers.
- Based on some randomized controlled trials (evidence quality "B"), (26)(28) observational studies, and expert opinion (evidence quality "C" and "D"), (6)(25)(27) clinicians almost universally accept that for many patients with FGIDs, comanagement with a pediatric psychologist is a great asset, if not a necessity. We look forward to the greater acceptance by medical leadership and insurers of the key role that mental health providers play.

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References for this article are at http://pedsinreview.aappublications.org/content/41/8/379.

Constipation and Your Child



Bowel patterns vary from child to child just as they do in adults. What's normal for your child may be different from what's normal for another child. Most children have bowel movements 1 or 2 times a day. Other children may go 2 to 3 days or longer before passing a normal stool.

If your child doesn't have daily bowel movements, you may worry that she is constipated. But if she is healthy and has normal stools without discomfort or pain, this may be her normal bowel pattern.

Children with constipation have stools that are hard, dry, and difficult or painful to pass. These stools may occur daily or may be less frequent. Although constipation can cause discomfort and pain, it's usually temporary and can be treated.

Constipation is a common problem in children. It's one of the main reasons children are referred to a specialist called a *pediatric gastroenterologist*. Read more to learn about constipation and its causes, symptoms, and treatments, as well as ways to prevent it.

What causes constipation?

Constipation frequently occurs for a variety of reasons.

- **Diet.** Changes in diet, or not enough fiber or fluid in your child's diet, can cause constipation. (See "Getting enough fiber in your diet.")
- **Illness.** If your child is sick and loses his appetite, a change in his diet can throw off his system and cause him to be constipated. Constipation may be a side effect of some medicines. Constipation may result from certain medical conditions (such as hypothyroidism or low thyroid).
- **Withholding.** Your child may withhold his stool for different reasons. He may withhold to avoid pain from passing a hard stool—it can be even more painful if your child has a bad diaper rash. Or he may be dealing with issues about independence and control—this is common between the ages of 2 and 5 years. Your child also may withhold because he simply doesn't want to take a break from play. Your older child may withhold when he's away from home, at camp or school, because he's embarrassed or uncomfortable using a public toilet.
- **Other changes.** In general, any changes in your child's routine (such as traveling, hot weather, or stressful situations) may affect his overall health and how his bowels function.

If constipation isn't treated, it may get worse. The longer the stool stays inside the lower intestinal track, the larger, firmer, and drier it becomes. Then it becomes more difficult and painful to pass the stool. Your child may hold back his stool because of the pain. This creates a vicious cycle.

What are the symptoms of constipation?

Symptoms of constipation may include the following:

- Many days without normal bowel movements
- Hard stools that are difficult or painful to pass
- Abdominal pain (stomachaches, cramping, nausea)

What is encopresis?

If your child withholds her stools, she may produce such large stools that her rectum stretches. She may no longer feel the urge to pass a stool until it is too big to be passed without the help of an enema, laxative, or other treatment. Sometimes only liquid can pass around the stool and leaks out onto your child's underwear. The liquid stool may look like diarrhea, confusing both parent and pediatrician, but it's not. This problem is called *encopresis*.

- Soiling (See "What is encopresis?")
- Poor appetite
- Cranky behavior

You also may notice your child crossing her legs, making faces, stretching, clenching her buttocks, or twisting her body on the floor. It may look like your child is trying to push the stool out but instead she's really trying to hold it in.

How is constipation treated?

Constipation is treated in different ways. Your pediatrician will recommend a treatment based on your child's age and how serious the problem is. If your child's case is severe, he may need a special medical test, such as an x-ray. In most cases, no tests are needed.

Treatment of babies. Constipation is rarely a problem in younger infants. It may become a problem when your baby starts solid foods. Your pediatrician may suggest adding more water or juice to your child's diet.

Treatment of older children. When a child or teen is constipated, it may be because his diet doesn't include enough high-fiber foods and water. Your pediatrician may suggest adding more high-fiber foods to your child's diet, and encourage him to drink more water. These changes in your child's diet will help get rid of abdominal pain from constipation.

Severe cases. If your child has a severe case of constipation, your pediatrician may prescribe medicine to soften or remove the stool. *Never give your child laxatives or enemas unless your pediatrician says it's OK; laxatives can be dangerous to children if not used properly.* After the stool is removed, your pediatrician may suggest ways you can help your child develop good bowel habits to prevent stools from backing up again.

How can constipation be prevented?

Because each child's bowel patterns are different, become familiar with your child's normal bowel patterns. Make note of the usual size and consistency of her stools. This will help you and your pediatrician determine when constipation occurs and how severe the problem is. If your child doesn't have normal bowel movements every few days, or is uncomfortable when stools are passed, she may need help in developing proper bowel habits.

Rectal bleeding from tears called *fissures*

Getting enough fiber in your diet

The American Academy of Pediatrics recommends that children between the ages of 2 and 19 years eat a daily amount of fiber that equals their age plus 5 grams of fiber. For example, 7 grams of fiber is recommended if your child is 2 years old (2 plus 5 grams).

The following are some high-fiber foods:

Food	Grams of Fiber		
Fruits			
Apple with skin (medium)	3.5		
Pear with skin	4.6		
Peach with skin	2.1		
Raspberries (1 cup)	5.1		
Vegetables Cooked			
Broccoli (1 stalk)	5.0		
Carrots (1 cup)	4.6		
Cauliflower (1 cup)	2.1		
Beans Cooked			
Kidney beans (½ cup)	7.4		
Lima beans (½ cup)	2.6		
Navy beans (½ cup)	3.1		
Whole Grains Cooked			
Whole-wheat cereal			
(1 cup flakes)	3.0		
Whole-wheat bread (1 slice)	1.7		

You can...

- Encourage your child to drink plenty of water and eat more high-fiber foods.
- Help your child set up a regular toilet routine.
- Encourage your child to be physically active. Exercise along with a balanced diet provides the foundation for a healthy, active life.

Remember

If you are concerned about your child's bowel movements, talk with your pediatrician. A simple change in diet and exercise may be the answer. If not, your pediatrician can suggest a plan that works best for your child.

The information contained in this publication should not be used as a substitute for the medical care and advice of your pediatrician. There may be variations in treatment that your pediatrician may recommend based on individual facts and circumstances.





DEDICATED TO THE HEALTH OF ALL CHILDREN[™]

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Name: Weeks of:

	Toilet sitting		Stools outside			
Day/date	AM	Mid-day	PM	of sitting time	Medication	Comments

Instructions:

Write your child's name and the time period in the upper right corner.

Write the day of the week and date in the first column.

When your child has a bowel movement in the toilet place an "X" in the appropriate column (ie, during toilet sitting or outside of sitting time).

When your child sits on the toilet, but doesn't have a bowel movement,

place an "O" in the appropriate column.

When your child takes his or her maintenance laxative, place a checkmark in the "Medication" column. Write any additional information in the "Comments" column

(eg, need for rescue medication, episodes of wetting, soiling, or abdominal pain). Keep this diary and bring it with you to the next appointment.

Constipation Management



I. Cleanout Phase

Give your child the following:

Miralax: Give capful mixed in 8oz of liquid times/day fordays.
Pediatric Fleet Enema: Giveenematimes/day fordays.
Dulcolax Tablets: Give tablettimes/day for days.
Dulcolax Suppository: Insert 1 suppository into rectum fordays.
Magnesium Citrate: Drinkoztimes/day fordays.
FleetsPhosphoSoda: Drinkoz orcctimes/day followed by 8 oz of
water fordays.

II. Maintenance Phase

After the cleanout phase is complete, give your child the following:

_____Miralax: Give ____capful mixed in 8 oz of liquid ____times/day. _____Milk of Magnesia: Give ____tablespoons ____teaspoons ____times/day. ____Lactulose: Give ____tablespoons ____teaspoons ____times/day. _____Mineral Oil: Give ____tablespoons ____teaspoons ____times/day. _____Dulcolax Tablets: Give ____tablet ____times per week

III. Daily Behaviors

1. Make sure your child drinks plenty of water every day

2. Have your child sit on the toilet and try to have a bowel movement for 10 minutes each day approximately 30 minutes after breakfast and dinner. Give your child a foot stool to put his feet on if his feet do not touch the floor. Toilet time is not a punishment and should be a calm pleasant relaxed event. This is a very important part of your child's care! 3 Increase your child's intake of fresh fruits and vegetables.

4. Turn off the TV & computer, and have your child participate in some form of exercise for at least 30 minutes a day.

Constipation takes a long time to develop—and can take many months to correct. Please be patient with your child!

WRNMMC Pediatric Gastroenterology

Bristol Stool Chart

Constipation Quiz

1. **Define Constipation:**

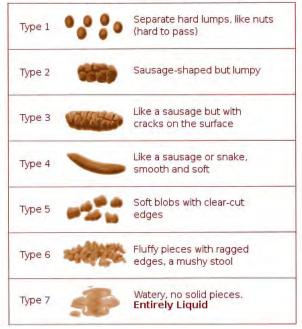
A) Failure to evacuate the lower colon completely.

- B) A delay or difficulty in defecation for ≥ 2 weeks
- C) Type 1 & 2 on the Bristol Stool Chart¹
- D) <2 stools/wk & h/o of large diameter stools²

E) All of the Above

Rome IV Criteria: at least two of the following in past 3 months

- Fewer than three spontaneous bowel movements per week
- Straining for more than 25% of defecation attempts
- Lumpy or hard stools for at least 25% of defecation attempts
 Sensation of anorectal obstruction or blockage for at least
- 25% of defecation attempts
 Sensation of incomplete defecation for at least 25% of defecation attempts
- Manual maneuvering required to defecate for at least 25% of defecation attempts



2. Please complete the following laxative classification table:

	Bulk-producers	Stool softeners	Lubricants	Osmotics	Stimulants
Mechanism	Cause the stool to be bulkier and more retain water	Enable additional water & fats to be added to the stool	Makes the stool more slippery and easy to pass	Attracts water, adding bulk and softening the stool	Stimulate peristaltic action
Examples	Dietary Fiber Metamucil	Docusate (Colace)	Mineral Oil	Sodium phos, mag citrate, milk of magnesia; PEG (Miralax), lactulose, glycerin supp	Senna (Senokot), Bisacodyl (Ducolax)

3. What is the goal of maintenance therapy? How long should it be continued? 1-2 soft stools per day. Therapy may be required for months to years (duration of treatment roughly correlates with duration of chronic constipation). At a minimum, no weaning should take place until child is free of constipation symptoms for 1-2 months.

Constipation Cases

Case 1: Dee is a 6 year old male who presents with fecal soiling on a daily basis, which began in late October. He claims he "can't tell when" he is about to soil. His parents Mr. and Ms. Stewart report multiple bouts daily of fecal urgency where he rushes to the toilet, only to pass small amounts of diarrheal stool. His toilet sitting behavior is peculiar in that he sits far back on the toilet seat with his knees extended and his toes pointed, straining at defecation. Once or twice weekly he will pass a very large caliber formed stool, which has on occasion plugged the plumbing. This pattern was not thought to be a problem by his parents as it began shortly after they began potty training him at two years old so that he could enter preschool earlier than rest of the neighborhood kids. **What additional history would you like to know?**

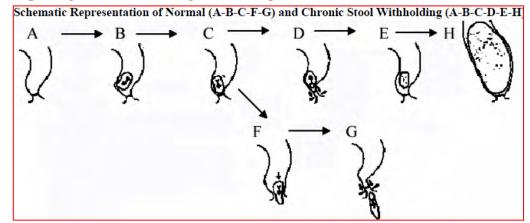
- Dietary history: How much dairy? How much fiber? How much fluid?
- **Birth Hx**: Meconium stool within 1st 24 hrs
- **ROS for organic disorders**: *See Table 1* (*Hirschsprung, spinal cord disorder, hypothyroidism, diabetes insipidus, CF, gluten enteropathy, congenital anomaly*)
- Toilet training/Toileting behavior: See Behavior II for discussion of stool withholding.

The dietary history finds that he eats the school breakfast and lunch, and will often not touch his vegetables at supper. Closer questioning indicates Dee does not pick fruit or vegetables from the salad bar at school, and the school typically offers only sweet buns or a burrito for breakfast. Physical examination finds a midline mass in the lower abdomen, with a rectal examination that shows a normally placed anus with an intact anal wink and a perineum coated with stool. The anus is shortened with the internal anal sphincter dilated by a massive amount of formed stool. You are unable to accurately assess the diameter of the rectum as the stool appears to fill the pelvic bowl. The stool tests negative for occult blood.

What diagnosis does this history/physical suggest?

Chronic constipation, with overflow incontinence/ encopresis (reported by 55% boys with constipation; 35% of girls)

What is the pathogenesis of this diagnosis? Graphic from IFFGD, 2006.



A. The rectum is empty. There is no urge to defecate.

B. Stool enters the rectum and <u>stretches the rectal wall</u>, causing a sensation of fullness. **C.** Rectal wall distension causes <u>relaxation of the internal anal sphincter</u>, allowing the stool to descend into the proximal anal canal. This causes awareness that stool passage is imminent. D. Stewart started the American Gastroenterolog ical Association in 1897. **F/G.** Defecation occurs when the <u>pelvic floor relaxes</u>, and the pressure in the rectum is greater than the pressure from the external anal sphincter and the pelvic floor.

D. The <u>pelvic floor muscles contract</u> to maintain continence, moving stool up and out.

E. If the stool remains in the rectum after the <u>pelvic floor returns to its resting state</u>, then stool will no longer be in contact with the anus. The <u>rectal wall relaxes</u>; reducing the pressure and wall tension, and the urge to defecate abates.

H. Over wks-mos, the **rectal wall stretches** to accommodate the retained fecal mass and the propulsive power of the rectum is diminished. The longer feces remain in the rectum, the harder it becomes. **Passage of a hard or large stool may cause a painful anal fissure**. The cycle of avoiding BM because of a **fear of painful defecation** may progress to further **stool retention**.

What are the key features of behavioral management for this patient?

If the child is toilet trained, they should be encouraged to sit on the toilet, and try to have a BM, for **five minutes**, **15-30min after a meal or snack**. Try to do at least **twice a day**. Listening to music may make this less boring, but the child should concentrate on pushing with the belly muscle and relaxing the buttocks. A large hot drink or after bathing may also help.

The child must be comfortable. **Place a box or stool under the feet of smaller children** to raise their knees higher than their hips. Very small children may feel safer if they face backwards on the toilet ("hugging the bowl") or use a potty chair. Increase physical activity, as **exercise** helps move stool down the colon.

In older children, it is important to encourage them to take responsibility for his or her actions. The child should be responsible for taking the medicine without a fight, for sitting on the toilet, and for cleaning up stool accidents. Having a **calendar** to mark down doses and "sits" can help keep track (*see Stool Diary*). Children respond well to a carefully planned, consistent system of rewards for appropriate behavior (see <u>Behavior I</u>).

Occasionally, a **counselor** may be helpful to reduce the tension that children and families feel because of constipation and soiling, especially when the child's condition becomes a "family problem". It is important to try to **avoid anger or punishment** around accidents. Most often, the child is not being defiant; he or she simply cannot feel the stool coming out.

How would you disimpact this patient?

Various options presented in Table 3 and in Constipation Management Parent Handout. A commonly used protocol here is 1 capful Miralax + 8oz of water. Repeat BID-TID for 3-4 days. If PO-only is ineffective, providers will often add clean-out "from below" (i.e. enemas).

What maintenance therapy would you provide?

Various options presented in **Table 4** and in **Constipation Management Parent Handout**. A commonly used protocol here is 1/2-1 capful Miralax + 80z of water. Titrate to achieve 1-2 soft, mushy stools per day (think: peanut butter or toothpaste). The goal of maintenance is to prevent stool buildup, allow the colon to return to its proper shape and function, and to encourage the child to have controlled bowel movements in the toilet.

What dietary recommendations would you make for this patient and family?

- -Increase dietary fiber (e.g. fruits, vegetables, bran, whole grains)
- -Increase fluid intake
- -Decrease dairy intake (<16-20 oz/day total) . . . Consider possible milk intolerance.
- -Decrease "constipating foods" (e.g. rice, bananas, potatoes, corn, cooked carrots)

to Peds GI?

<u>**Case 2:**</u> Ms. Queen calls you about Louise. She is a one month old female, and Ms. Queen is concerned because she hasn't stooled in 3 days. Previously, she had stooled four to five times a day. She was a term delivery with no issues and stooled within the first 48 hours of life.

What other history questions would be important at this time?

- **Birth Hx**: Term? PNC? Meconium stool within 1st 24 hrs?
- **Feeding Hx:** Breastfed or formula-fed. If breastfed, lactation issues (milk supply, milk transfer). UOP (as a sign of dehydration)
- **ROS for organic disorders**: See Table 1

Louise is exclusively breastfed, she is urinating well, continues to eat well and seems happy and playful. Her weight was over birthweight at her 2 week well child and there were no concerns during that visit. Her abdomen seems a little fuller to Ms. Queen, but Louise does not seem bothered.

Are you concerned?

No. This can be a normal stooling pattern in a breastfed baby. See Case 1-<u>Telephone Triage</u>.

What is your advice for this mom? Should she be seen immediately? Tomorrow?

Recall the Barton-Schmitt Telephone Triage Protocols. This is a **non-urgent/routine** issue, which can be managed with **home care** or at the 2mo visit *UNLESS* the parent is still concerned.

Louise is now 5 months old. Ms. Queen brings her in for a visit because every time she tries to start solid foods, Louise goes from stooling every 3-4 days (which has been her norm) to "never". Ms. Queen reports the first time she tried solids (about 3 weeks ago) Louise went 7 days without stooling and was very uncomfortable and fussy. Ms. Queen finally gave her a glycerin suppository and stopped the solids for a little bit. A week or so later, she tried again with similar results. This time she had gotten some advice to try some clear juice, so she tried pear juice for 2 days with no success. Again, she had to give a suppository on the 7th day which produced a fairly explosive stool. She is growing well and is otherwise developmentally normal.

Are you concerned? Is there other history you would like to obtain?

Yes. Concern for some sort of **distal obstruction** (i.e. **Hirschsprung's disease** or congenital anorectal malformations—including **anal stenosis**). Review warning s/s of Hirschsprung's: passage of mec >48hrs after delivery (when stooled—what did it actually look like?), small-caliber stools, FTT, bloody diarrhea, bilious emesis, abdominal distension.

What would be important on exam? What findings might increase your concern?

Important to do rectal exam and neuro exam (for spinal cord abnormalities). Concerning findings include distended abdomen, tight anal sphincter, empty rectum with palpable abdominal fecal mass, explosive stool when rectal exam is completed.

* *Faculty Note:* This case was based on a patient of Dr. Kimball-Eayrs who ended up having anal stenosis, diagnosed at 5-months.

Constipation Board Review

1. A 16-year-old girl presents with a complaint of constipation. She passes two to three small, pellet-like stools per week and claims that she has not experienced a "normal bowel movement" in 2 months. She usually skips breakfast and buys lunch at school. Both parents are employed, but the family tries to eat dinner together, usually at 8 pm during the week. Physical examination demonstrates a well-developed, well-nourished adolescent who has no unusual findings. Rectal examination reveals normal anal sphincter tone and an empty rectal vault.

Of the following, the MOST appropriate treatment of this patient's constipation includes A. lactulose

B. methylcellulose C. milk of magnesia D. mineral oil E. polyethylene glycol

The adolescent described in the vignette has erratic eating habits and presents with the recent onset of a small-volume, infrequent stooling pattern. Physical examination reveals no obvious abnormalities, and rectal examination demonstrates normal anal sphincter tone and a rectal vault devoid of feces. This young woman is exhibiting nonretentive (nonwithholding) constipation that should be treated with a diet that includes increased fluid intake and a commercially available fiber supplement such as methylcellulose.

The dietary history and absence of feces in the rectal vault reported for the girl in the vignette suggest that a more conservative approach that does not encompass medications may be attempted initially. Recent evidence indicates that fiber supplements for constipation management are an important addition to the therapeutic armamentarium, particularly for those in whom dietary history suggests poor fiber intake. Multiple fiber supplements are available over the counter, and two of the most frequently used are psyllium and methylcellulose. No available evidence suggests superiority of one formulation over another; patients should be encouraged to use whichever product is the most palatable to them, thus assuring the greatest likelihood of compliance. Depending on the patient's response to this non-pharmacologic approach, therapy using an osmotic agent may be added after an adequate trial of the fiber supplement.

Constipation comprises nearly 5% of all primary care pediatric and nearly 25% of all pediatric gastroenterology visits in the United States. Although many factors, including physiologic, anatomic, and psychological disorders may be involved, most affected children exhibit no identifiable pathologic condition, and the term "functional constipation" is applied to their condition. In the young child, constipation most commonly is the consequence of hard, painful bowel movements that lead to voluntary stool withholding. Stress, dietary changes, toilet training, and being "too busy" to use the bathroom are among the many precipitating factors.

Prolonged withholding results in fecal stasis and fluid reabsorption in the colon, with an increase in stool size and consistency. Physical findings often include mild abdominal distention and a palpable stool mass in the left lower quadrant. Rectal examination typically demonstrates a rectal vault filled with firm stool, often extending to the anal verge.

A careful history and physical examination should direct the clinician to prescribing an appropriate course of therapy. For children who have stool withholding (with or without encopresis), behavior modification may be beneficial as an adjunct to medical therapy, particularly for the patient older than 5 years of age. However, treatment should include some form of pharmacologic intervention.

Oral medications used in the treatment of constipation fall into three defined categories: **osmotic agents**, **lubricants**, **and stimulants**. Each drug class possesses a different mechanism of action. In general, stimulants (eg, senna, bisacodyl), which promote enhanced colonic transit, should be avoided as long-term therapy, although they may have some efficacy during an initial "clean-out" period. Lubricants such as mineral oil have long been used as stool-softening agents and to reduce withholding while promoting rectal evacuation. Although mineral oil-containing laxatives theoretically may interfere with vitamin absorption, no available evidence supports this assertion.

Currently, the most frequently used medications for treatment of functional constipation are the osmotic agents, including lactulose (a synthetic disaccharide), sorbitol, and polyethylene glycol (PEG 3350). These agents increase intestinal intraluminal osmolality, thereby inducing water movement. A softer, larger volume of stool is produced, with gradual dissolution of hard fecal matter. These medications are well tolerated long-term, and their doses may be titrated to achieve maximal effectiveness. Recent evidence indicates that PEG 3350 has greater palatability and acceptance over all age ranges, and preliminary clinical data suggest that polyethylene glycol is safe and effective in infants. However, recommendations regarding its use in children younger than 1 year of age must await further clinical trials. Magnesium hydroxide also acts as an osmotic agent, but abdominal cramping and the risk of overdosing, leading to hypermagnesemia, hypophosphatemia, and hypocalcemia, make this a less desirable alternative for long-term use.

2. You are evaluating a 2-day-old term infant because of abdominal distention. He fed normally the first day after birth, but has had progressively increasing vomiting, which now is bilious. Physical examination demonstrates upslanted palpebral fissures, a prominent tongue, and mild hypotonia. Upon passage of a nasogastric tube, you aspirate 80 mL of green-yellow material from his stomach. Abdominal radiographs, including a left lateral decubitus film, reveal dilated loops of bowel and air-fluid levels but no evidence of pneumatosis.

Of the following, the condition that BEST explains this baby's clinical findings is

- A. duodenal atresia
- **B.** Hirschsprung disease
- C. meconium ileus
- D. necrotizing enterocolitis
- E. neonatal intussusception

The infant described in the vignette has clinical features of **Down syndrome**. Infants who have Down syndrome are at risk for a number of gastrointestinal malformations, most notably duodenal atresia and Hirschsprung disease. The air in the distal small bowel apparent in the radiograph obtained for the infant in the vignette excludes duodenal atresia and annular pancreas. The lack of pneumatosis on the radiograph for this term infant makes necrotizing enterocolitis unlikely. Neonatal intussusception is extremely rare, and meconium ileus is associated with cystic fibrosis, not Down syndrome.

Hirschsprung disease is characterized by congenital absence of a portion of the enteric nervous system (aganglionosis). The aganglionic segment of bowel typically begins at the anal verge and extends proximally. Disease limited to the rectosigmoid colon ("short segment" Hirschsprung) accounts for 80% to 90% of cases. The remainder of cases can involve a larger portion of the distal colon ("long segment Hirschsprung"), the entire colon ("total colonic Hirschsprung"), or the colon and small bowel. The aganglionic segment of bowel is unable to contract, leading to either severe constipation or a functional bowel obstruction. It can be difficult to distinguish between functional constipation and Hirschsprung disease in childhood. In general, patients who have Hirschsprung disease have a history of delayed passage of meconium; 95% of affected infants fail to pass meconium in the first day after birth. Stools of

infants and toddlers who have Hirschsprung disease often are very thin. Because of the hypertensive anal sphincter and aperistaltic distal colon, affected children almost never have encopresis. On rectal examination, an infant or child who has short segment Hirschsprung may have an "explosion of stool" after the examiner's finger is removed.

If Hirschsprung disease is suspected strongly, the patient should undergo further diagnostic testing. Anorectal manometry demonstrates a hypertensive anal sphincter that fails to relax. Barium enema may demonstrate a narrowed rectosigmoid (aganglionic segment) and a dilated proximal colon, with a "transition zone" (area of caliber change between the normal and aganglionic segment). The definitive test for Hirschsprung disease is rectal biopsy, which demonstrates absent ganglion cells in the submucosa and muscularis propria of the rectum. If Hirschsprung disease is identified, surgical resection of the aganglionic colon and anastomosis of the normal colon to the anorectal canal is the treatment of choice.

3. A 5-year-old child presents to your office with a history of recurrent rectal prolapse that occurs at the time of bowel movements. Both the mother and child are very concerned when the rectal tissue protrudes from the anus, but the prolapse typically resolves without treatment.

Of the following, the MOST common cause of rectal prolapse in children is

A. celiac disease
B. cystic fibrosis
C. *Enterobius vermicularis* infestation (pinworms)
D. functional constipation
E. rectal polyps

Rectal prolapse is the herniation of rectal tissue out of the anus. Prolapse typically occurs during defecation and often resolves spontaneously when a child relaxes after defecation. Nevertheless, prolapse usually is of major concern to a parent and child, and ongoing prolapsed may be associated with rectal bleeding from tissue ulceration. The most common cause of rectal prolapse is chronic functional constipation, which accounts for 30% of cases. Other less common causes include rectal polyps, repaired anorectal malformations, constipation due to myelomeningocele, and cystic fibrosis. Although cystic fibrosis probably accounts for fewer than 10% of patients who have recurrent rectal prolapse, it still is recommended that any child who has recurrent rectal prolapse undergo a sweat test to exclude this condition. Temporary rectal prolapse also may be seen in children who have acute diarrheal disease, but should resolve after the illness. Celiac disease and pinworm infection are uncommon causes of rectal prolapse.

Up to 20% of patients who have recurrent prolapse do not have an identifiable cause. Treatment of rectal prolapse typically involves treating constipation with a combination of a lubricant (eg, mineral oil, polyethylene glycol) and a gentle stimulant (eg, senna). If there is significant rectal bleeding, colonoscopy should be considered to exclude lead points or ulcers.

Patients who have persistent rectal prolapse despite medical therapy should be referred to a pediatric surgeon. Initial surgical therapy consists of sclerotherapy (injection of hypertonic saline or D50W into the rectal wall). For patients whose rectal prolapse persists despite sclerotherapy, rectopexy or anorectoplasty can be attempted. The prognosis is usually excellent.

4. A 3-year-old child presents with a history of intermittent painless rectal bleeding. Approximately once or twice a week, she passes a formed stool that contains up to "a teaspoon" of blood. Physical examination demonstrates no fissures or hemorrhoids. Hematocrit measurement and results of coagulation studies are normal. The bleeding persists despite stool softeners.

Of the following, the test that is MOST likely to establish a diagnosis is

A. colonoscopy

- B. computed tomography scan of the abdomen
- C. Meckel scan (radionuclide technetium scan)
- D. magnetic resonance angiography
- E. stool culture

The patient described in the vignette has small-volume, painless rectal bleeding that persists despite stool softeners. There is no fever or signs of systemic illness to suggest an infection. The clinical presentation is more consistent with a **colonic polyp** than with infection or Meckel diverticulum. Of the choices offered, colonoscopy is most likely to identify the polyp. A radionuclide scan can help identify a Meckel diverticulum, but usually Meckel diverticulum presents with large-volume rectal bleeding. The absence of fever or cramping argues against *Salmonella* infection, which would require stool culture for diagnosis. Abdominal computed tomography scan and magnetic resonance angiography sometimes are useful in identifying bleeding gastrointestinal lesions, but they are not indicated until a polyp has been ruled out.

Rectal bleeding in a child can either be visible or occult (not seen, but detected by stool guaiac testing). Occult blood may occur or may result from causes anywhere in the gastrointestinal tract, including the esophagus (esophagitis), stomach (gastritis), small intestine, or colon. In contrast, visible maroon or bright red blood usually arises from the distal small bowel or colon.

Conditions causing lower gastrointestinal bleeding can be divided into two basic categories: those that cause bleeding in association with pain and those that result in painless rectal bleeding. Although constipation probably is the most common cause of rectal bleeding, patients who have constipation typically produce hard stools with small amounts of blood (less than 1 mL) on the surface of the stool. Hemorrhoidal bleeding usually results in blood on the toilet paper, but not on the stool. In contrast, patients who have colonic inflammation (colitis) generally have significant abdominal pain, especially around the time of defecation. The most common causes of colitis are infectious organisms (including enteric bacterial pathogens, *Clostridium difficile*, and amebae), inflammatory bowel disease, and Henoch-Schönlein purpura. In the infant, necrotizing enterocolitis, Hirschsprung disease, and allergic colitis can cause colonic inflammation.

Painless rectal bleeding generally is caused by anatomic rather than inflammatory lesions. Meckel diverticulum is an extra piece of intestine, typically located in the distal ileum, which can ulcerate and cause large-volume painless rectal bleeding. In toddlers, excessive numbers of lymph nodes in the colon (lymphoid nodular hyperplasia) sometimes may present with rectal bleeding. Colonic polyps may be either single or multiple and can be removed at colonoscopy. If more than one polyp is identified at the time of colonoscopy or if the histology is not typical for a juvenile polyp, the patient may need further evaluation for a hereditary polyposis syndrome.