What Causes Recurrent Abdominal Pain?

Objectives:

1. What are the red flags to look for on history for a child with Recurrent Abdominal Pain?
2. What are the red flags to look for on physical exam in a child with Recurrent Abdominal Pain?
3. What is the differential Diagnosis for Recurrent Abdominal Pain
4. What investigations would you do in a child with Recurrent Abdominal Pain?
5. What is your management in a child with Recurrent Abdominal Pain
6. What is the management for a child with constipation?

Patient Presentation A 7-year-old male comes to clinic with a 3 month history of abdominal pain that occurs off and on. The pain is periumbilical, lasts several minutes and then resolves. He says it “feels like someone is twisting me.” The episodes usually occur daily and sometimes several times per day. The pain does not wake him at night, but he wants to sit down when the pain occurs.

Pause for Discussion:

The past medical history shows that he has a normal diet and is otherwise well. The review of systems shows he has been having harder bowel movements for the past few months with occasional painful defecation. His bowel movements are every 2–4 days and occasionally clog the toilet. The rest of his review of symptoms is negative including urinary problems.

Pause for Discussion:

The pertinent physical exam shows that his growth parameters are normal. His abdominal examination shows a soft, non-tender abdomen with no organomegaly and normal bowel sounds. He has stool palpable in the left lower and upper quadrants. His rectal examination shows normal tone and soft stool in the rectal vault. The rest of his genitourinary, neurological, and orthopaedic examinations are normal.

Pause for Discussion:

The abdominal flat plate radiograph showed stool throughout the abdomen. The diagnosis of constipation was made. The patient was placed on a bowel clean-out program, and then started on a maintenance
program. He was also given information on how to use a toilet sitting program and increase the fiber in his diet.

Figure 6 – 12–27–04 – AP radiograph showing the colon to be filled with stool from the cecum to the rectum.
Resources:

1. Recurrent Abdominal Pain, Pediatrics in Review 2002;23;39 attached
2. Understanding and treating childhood bellyaches, Pediatric Annals; Feb 2004; 33, 2; attached
3. Evaluation and treatment of constipation in infants and children, NASPGHN, Journal of Pediatric Gastroenterology and Nutrition, 43:e1Ye13 attached
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Paul N. Thiessen
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http://pedsinreview.aappublications.org/content/23/2/39
Recurrent Abdominal Pain

Paul N. Thiessen, MD*

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

1. Characterize the epidemiology and classification of recurrent abdominal pain.
2. List the major clinical conditions that manifest with recurrent abdominal pain.
3. Describe the most important findings in the history and physical examination of the child who has recurrent abdominal pain that suggest an organic medical condition.
4. Outline a targeted approach to ordering investigations that will confirm or disprove suspected organic disease.
5. Delineate the prognosis of recurrent abdominal pain.

Introduction

Recurrent abdominal pain (RAP) is a frequent and troublesome complaint in childhood and adolescence, and the search for a cause and a credible approach to management can be taxing for both family and physician. The term “recurrent abdominal pain” was coined by the British pediatrician John Apley, who first published on the subject in 1958. His definition included at least three attacks of pain occurring over a period of 3 months that were severe enough to affect activities and for which no organic cause was identified. In practice, the definition may include any child or adolescent who has RAP for which the family seeks medical attention and explanation, even if the duration of the pain does not adhere strictly to the Apley definition. The definition explicitly excludes the many causes of acute abdominal pain, which lie outside the scope of this review.

There is wide variation in the threshold of severity and frequency that must be crossed before a family will seek medical attention for a child who has RAP. Inevitably, parents want a clear explanation and reassurance that no sinister causes lurk undetected, and the clinician wants to oblige but often lacks the conviction that organic causes have been excluded. The most difficult challenge for the clinician is to determine to what extent diagnostic studies should be employed before the label “recurrent abdominal pain” is applied. Whereas abdominal pain may be the chief manifestation of a large number of precisely defined illnesses, more than 90% of the time a “disease” will not be defined and the family will be left with a “functional” explanation. In spite of extensive study and a vast literature base, RAP remains an elusive symptom in search of an etiology. How to diagnose and manage this common pediatric problem constitutes the subject of this review.

Epidemiology

RAP has been reported to occur in 10% to 15% of children between the ages of 4 and 16 years. A community-based study of 500 adolescents (mean age, 15.5 y) revealed that 13% to 17% experienced weekly pain, which in 20% of cases was severe enough to affect activities. There clearly is an overlap between the “normal” population that may experience recurrent pain symptoms but not complain sufficiently to seek medical attention and others who have a seemingly similar degree and frequency of pain and do come for assessment. Many sociocultural, familial, and emotional factors determine a child’s response to pain, and these also will affect the likelihood of seeking medical attention to explain and treat the problem. Al-

Abbreviations

IBD: inflammatory bowel disease
IBS: irritable bowel syndrome
RAP: recurrent abdominal pain

*Editorial Board. Clinical Professor of Pediatrics, B.C. Children’s Hospital, Vancouver, B.C., Canada.
though Apley and others have reported RAP to be the most common pain syndrome of childhood, headaches and limb pains appear to have an equal prevalence.

In a study of 1,000 school-age children, RAP affected males and females equally up to 9 years of age. After 9 years, the incidence in females increased such that between 9 and 12 years, the female-to-male ratio was 1.5:1. The overall incidence appears to peak at 10 to 12 years. RAP is rare among children younger than 5 years of age, and an organic cause must be considered even more carefully in this younger age group. The vast literature published on this subject reveals no evidence of changes in the incidence or clinical profile of this common pediatric pain syndrome; it seems to be here to stay.

Pathophysiology

The origins of abdominal pain are complex and do not lend themselves to a single model of causation. Numerous organic disorders lead to abdominal pain; in most, the pathophysiology is related to inflammation (eg, Crohn disease) or distension or obstruction of a hollow viscus (eg, obstructive uropathy). Most studies indicate that fewer than 10% of children who present with RAP have an identifiable organic etiology.

The exact mechanism of pain remains unclear in the majority of children in whom no organic cause can be identified. The most typical pattern of periumbilical pain so characteristic of RAP appears to be visceral in origin, probably originating in the small intestine or colon. To date, no pattern of consistent motility disturbance has been identified in any subgroup of patients experiencing nonorganic abdominal pain. Emotions, cognitive processes, and other central nervous system influences may modulate the perception of pain to produce an altered awareness of the discomfort from these visceral sensations. This “visceral hyperalgesia” describes a heightened awareness of sensations that might not be perceived or expressed as pain in other children.

Models that try to relate psychological influences in a primary causal manner (emotional stress leads to RAP) are too simplistic. However, stress can cause recognized physiologic effects, such as increased cortisol levels, sympathetic tone, and tachycardia, so it is entirely plausible that it could exert physiologic effects on the gut through altered motility or some other as yet unidentified mechanism.

Oft-repeated assumptions that children who have RAP are anxious, perfectionist, socially unskilled, and self-conscious have taken on an aura of validity that is unsupported by objective evidence. Several case-control studies have failed to demonstrate significant differences for a range of measures of psychological distress between groups of children who have “functional” RAP and those who have a demonstrable organic cause for their pain. Others contradict these studies, showing that those who have RAP have higher levels of anxiety and depression than do “well” children. Illness or pain clearly causes anxiety and distress, but this must be distinguished from invoking “stress” as a source of primary causation. There are no objective methods of measuring stress, and what seems to be a source of stress for one child (eg, birth of a sibling, upcoming athletic or music competition) may be of no apparent emotional consequence for another child of the same age. As in adults, some children seem to “buckle under stress” and become anxious and emotionally distressed; others facing the same challenge become invigorated and rise to new heights of effort and achievement.

For some children, anxiety and emotional stress seem to manifest in a range of pain complaints, of which abdominal pain and headache are the two most common. Parents sometimes can date the onset of the pain to a specific time, such as the beginning of a new school year or a marriage breakup. Family dynamics and individual coping styles influence the way in which children express or even acknowledge their pain. Some families encourage their children to express pain in ways that unwittingly may reinforce the complaint.

The tripartite classification proposed by Barr may be the most helpful method of categorizing children who present with RAP. This classification includes: 1) those who have organic disease, 2) those who have a clear psychogenic etiology such as depression or school phobia, and 3) the traditional “functional” group in which neither organic disease nor a clear psychogenic etiology is manifest.

Clinical Aspects

Functional Abdominal Pain

The majority of children who have RAP are considered to have a functional etiology. The problem of defining functional RAP is daunting. In its simplest form, the concept encompasses all causes that do not have an identifiable organic etiology. Most typically, the pain occurs in episodes that are periumbilical, self-limited, unrelated to meals or activities, and rarely if ever sufficient to awaken the child from sleep. The growth pattern and findings on the physical examination are normal. The degree of interference with normal activities and school attendance may seem out of proportion to the frequency and severity of the episodes as described. It has been
observed wryly that “Organicity of pain is inversely proportional to the number of school absences.”

Irritable Bowel Syndrome

Some children who have RAP manifest many of the characteristics associated with irritable bowel syndrome (IBS), as defined in adults. The criteria for making this diagnosis are: 1) abdominal pain relieved by defecation, 2) more frequent stools at the onset of the pain, 3) altered stool form (hard or loose or watery), 4) passage of mucus, and 5) associated bloating or abdominal distension. To define this syndrome requires a degree of detail regarding bowel function that the clinician will find difficult to elicit from children, who are notoriously reluctant to reveal or discuss their bowel habits. In adults, the division is made between those who have constipation-dominant and diarrhea-dominant symptoms. There is some evidence that altered intestinal motility, mediated by peptides excreted by both gut and brain, plays a role in the etiology of IBS. There are no laboratory markers; the diagnosis rests on the history. Although some clinicians include constipation under the diagnostic category of IBS, most recognize it as a separate diagnosis.

Constipation

Many factors lead to constipation in children, the foremost of which is dietary. Modern diets are replete with highly processed starches, and many children shun fruits, vegetables, and higher-fiber foods. An unwillingness by some children to take the time to evacuate their bowels completely, coupled in some cases with a reluctance to use school washrooms, can seriously compound this problem. Sometimes the role of constipation as a major contributing factor to abdominal pain will be clear, with the parent noting that the child goes days between bowel movements and that the stool is bulky and hard. Often the pediatrician faces the problem of ferreting out this syndrome as a significant cause for RAP has waned. A wide range of racial/ethnic groups—Asian, Jewish, Mediterranean, and African-Americans—are predisposed to lactase deficiency, with incidences reported as high as 60% to 80%. Lactose ingestion will cause symptoms of bloating, loose stools, and cramping abdominal pain in those who are affected. It appears to be an uncommon cause of RAP in the absence of other gastrointestinal symptoms. The diagnosis is made most reliably by breath hydrogen testing. If this diagnostic tool is unavailable, it is reasonable to use lactase-treated milk products or a complete restriction of milk products for several weeks as a therapeutic trial. It is important to recognize that lactose intolerance results simply in carbohydrate malabsorption; it is not, per se, a cause of malnutrition or growth failure.

Helicobacter pylori Infection

The discovery of H pylori has changed the approach to diagnosis and treatment of peptic ulcer disease. Epidemiologic evidence indicates that this infection is more prevalent among those living in low socioeconomic circumstances, so infection rates are significantly higher in less developed nations. Even in developed countries, the prevalence of H pylori infection is approximately 40%. However, the great majority of affected individuals have no signs or symptoms; they have infection but no disease. The intense interest in H pylori has generated numerous tests and treatments that, unfortunately, are being used in excess of their established benefits and often counter to the best interests of the patient.

Several lines of evidence indicate that H pylori infection alone rarely is the cause of abdominal pain in children unless peptic ulcer disease is present. A meta-analysis of more than 40 published reports shows strong evidence for an association between H pylori gastritis and duodenal ulcer disease in children, but weak or no evidence for an association between H pylori infection and RAP. Serologic studies have shown that antibodies to H pylori occur with similar prevalence among children who
do and do not have RAP. In a large multicenter study from Germany, symptom assessment could not distinguish between children who had *H pylori* gastritis and those who had “functional” RAP. Symptoms improved or resolved in 87% of children in whom *H pylori* was eradicated successfully, but also in 93% of those in whom eradication failed and in 80% of those who had “functional” RAP. *H pylori*-associated peptic ulcer disease should be suspected when abdominal pain is primarily epigastric; when it awakens the child from sleep; and when it is associated with anorexia, nausea, recurrent vomiting, anemia, or gastrointestinal bleeding. Although abdominal pain is common in children, peptic ulcer disease is very uncommon; therefore, testing for *H pylori* should not be part of the preliminary evaluation of a child who has RAP.

Given the important distinction between *H pylori* infection and disease, engaging in a fishing expedition for evidence of *H pylori* infection is not an appropriate strategy for investigating RAP. Antibodies in serum or saliva may remain elevated for years after infection has resolved, making their mere presence unhelpful in initial diagnosis. The urea breath test is reliable for detecting the presence of *H pylori* infection, but many causes of esophagitis, gastritis, and peptic ulcer disease present with similar symptoms. Therefore, when the weight of symptoms suggests the presence of ulcer disease, endoscopy with biopsies is the optimal approach for confirming the diagnosis and guiding treatment. Fecal antigen tests look promising, but their role in children has not yet been defined.

**Other Causes of Peptic Ulcer Disease**

*H pylori* causes approximately 70% of primary peptic ulcer disease in children, but about 30% is idiopathic. In addition, secondary ulcer disease may be associated with nonsteroidal anti-inflammatory drug ingestion, Crohn gastritis, and other forms of erosive gastritis.

**Nonulcer Dyspepsia**

Dyspepsia is a symptom complex of epigastric pain, bloating, and discomfort that may occur with or without demonstrable acid reflux. “Nonulcer dyspepsia” is designated when these symptoms are accompanied by negative endoscopic and biopsy findings.

**Abdominal Migraine**

The association between migraine and abdominal pain remains mysterious, and many clinicians view the existence of a discreet entity of “abdominal migraine” as dubious. Because migraine is a common problem in both pediatric and adult medicine, with a prevalence reported to be as high as 5%, some children who have headache due to migraine also will experience RAP. “Abdominal migraine” usually is recognized when episodes of paroxysmal abdominal pain occur in association with nausea and vomiting, with complete recovery between episodes and sometimes with associated headache. A strong family history of migraine lends credibility to the diagnosis. Most pediatricians only accept abdominal migraine as an explanation for RAP when the patient has headaches that are conclusively migraine.

**Infestation/Infection**

The contribution of parasitic infestation to RAP is elusive. Infection with *Yersinia enterocolitica* can cause enteritis that mimics IBD, albeit usually associated with diarrhea. It is well-recognized that infestation with *Giardia* can cause diarrhea associated with abdominal cramps and pain, but diarrhea usually is the predominant complaint. The possible role of *Dientamoeba fragilis* and *Blastocystis hominis* in causing RAP in the absence of diarrhea has been raised in a small number of studies, but their role remains dubious. With the present state of knowledge, a search for an infectious etiology for RAP is not usually warranted in the absence of diarrhea.

**Gynecologic Conditions**

Many gynecologic conditions can present with RAP and must be given careful consideration, especially in postpubertal females. Early menarche, endometriosis, pelvic inflammatory disease, and ovarian cyst are important diagnostic possibilities. Many of these causes can be elucidated by ultrasonographic examination.

**Physical and Sexual Abuse**

Abuse always requires careful consideration in children who have RAP, and sensitive history taking is required to elucidate its possible role.

**Clinical Assessment**

It is heartening to reassure clinicians in the 21st century that the most powerful diagnostic tools they bring to the problem of RAP in childhood are a thorough history and physical examination. The proliferation of diagnostic technology threatens to obscure the foremost need that patients and their families be heard, not investigated. A correct diagnosis usually can be suspected following a good history and physical examination. In addition to their diagnostic roles, a complete history and physical examination will help to convince parents that their concerns are taken seriously.
A successful history places the patient and family at ease and allows them to express their concerns unhurriedly. As children advance in age, they are included in the history taking, and part of the interview of an adolescent should take place separately from the parents. It may be wise to conduct the initial interview with the parents alone; this should be discussed with the parents at the outset to reach a mutually agreeable decision. Successful interviewing involves active, empathetic listening followed by explanations given in language and terms that the family understands. The most satisfied parents are those who feel that they have been heard, and the physician who listens well earns a high degree of confidence and credibility.

The history should explore the location, nature, and frequency of the pain, along with associated symptoms. It should be acknowledged that the child’s description of the nature of the pain (eg, sharp, dull) is of limited importance in making a diagnosis because children often are unclear as to the meaning of these descriptors. The relationship of the pain to school and social/family stressors is important to elicit. A careful review of systems covers the child’s diet, bowel habits, and sleep patterns and explores the context in which the pain occurs. It is essential to define the degree to which the pain actually interferes with the child’s activities and how much school has been missed. More challenging is a thorough exploration of nonmedical factors, such as family function, school performance, and manifestations of anxiety, depression, or social maladjustment. Listen carefully for the main concern. Although abdominal pain may be the purported reason for the visit, the hidden agenda may be the child’s social isolation or school avoidance. The role of medications in both causing the pain (eg, naproxen) and in attempting to relieve the pain should be explored. The concept of recognizing “red flags” that suggest organic disease has a long and valid tradition, and the salient ones on history are noted in Table 1. It is important to recognize that firm conclusions may not be drawn at the first visit; follow-up visits may be needed.

### Physical Examination

The physical examination should be thorough, with particular attention paid to revealing extraintestinal manifestations. The assessment should begin with documentation of the height and weight; comparison to previous growth data is invaluable. Although the child should be asked, “Where does your tummy hurt?”, caution should be exercised in relying too much on a young child’s response. Instead, the clinician should examine the abdomen gently and thoroughly while speaking with the child and observe the response to palpation. The perianal area should be examined carefully for fissures, skin tags, or signs of sexual abuse. Although a rectal examination may be appropriate, it is highly upsetting to many children and should not be performed routinely. When performed, it is important to progress slowly and gently, to minimize pain and discomfort. Attention should be paid particularly to the “red flags” in Table 2.

### Investigations

Accepting that only 10% to 15% of cases of RAP are due to an organic etiology, investigations to identify organic disease should be carefully targeted. The “rule out all possibilities” approach can lead to a spiral of investigations that simply reinforces the impression that some hidden cause has been overlooked and must be unmasked, even when the clinician is convinced of the functional nature of the pain. In most cases, investigations should be limited to a complete blood count, urinalysis, and perhaps examination of a stool specimen for occult blood. In the presence of significant diarrhea, a stool for enteric culture and ova and parasite examination is indicated.

### Table 1. “Red Flags” on History of Recurrent Abdominal Pain

- Localization of the pain away from the umbilicus
- Pain associated with change in bowel habits, particularly diarrhea, constipation, or nocturnal bowel movements
- Pain associated with night wakening
- Repetitive emesis, especially if bilious
- Constitutional symptoms, such as recurrent fever, loss of appetite or energy
- RAP occurring in a child younger than 4 years of age

### Table 2. “Red Flags” on Physical Examination for Recurrent Abdominal Pain

- Loss of weight or decline in height velocity
- Organomegaly
- Localized abdominal tenderness, particularly removed from the umbilicus
- Perirectal abnormalities (eg, fissures, ulceration, or skin tags)
- Joint swelling, redness, or heat
- Ventral hernias of the abdominal wall
The role of radiographic investigations is important but limited and requires careful consideration. A single view of the abdomen can be valuable in defining the presence of significant constipation, especially when suspicion is high but the history is sketchy and results of the physical examination are inconclusive.

The value of abdominal ultrasonography as a screening tool seems very limited based on available literature. However, it can be valuable for diagnosing certain causes of abdominal pain, particularly when the origin is renal (e.g., obstructive uropathy or hydronephrosis), gynecologic (e.g., ovarian cysts), or the gall bladder. Rare gastrointestinal causes such as an enteric duplication also may be revealed by ultrasonography. It is an appropriate investigation when the pain is lateralized, when there are abnormalities on urinalysis, or when the pain localizes to the lower quadrants in a female of any age.

If IBD is considered as a possible diagnosis, erythrocyte sedimentation rate, serum protein and albumin levels, and stool for occult blood should be obtained. When this diagnosis is highly suspected, referral for endoscopic and histologic confirmation is essential. Upper gastrointestinal series and small bowel follow-through remains a valuable modality in the diagnosis of Crohn disease. Barium enema almost never is indicated. Elevated fecal calprotectin levels recently have been described as a sensitive screening test for IBD, but their role as a clinical tool remains to be elucidated.

When the pattern of pain strongly suggests peptic ulcer disease, upper gastrointestinal endoscopy with multiple biopsies is the optimal approach to diagnosis. When either IBD or acid peptic disease is given serious diagnostic consideration, referral to a gastroenterologist to assist in diagnosis and management is important.

Given the complex biopsychosocial nature of abdominal pain in children, the clinician needs to have sufficient acumen and experience to know when to pursue and when to halt investigations.

Management

Although many cases of RAP may reveal a probable diagnosis on first encounter, diagnostic certainty in others may be achieved only after several office encounters and the completion of salient investigations. The clinician should adopt an unhurried approach that allows formation of a fully informed diagnostic impression before making management decisions. Language barriers may require appropriate translation.

In the majority of cases, the diagnostic impression will be one of functional RAP. The first and admittedly most challenging task is to explain the concept of functional abdominal pain to the parents. Many parents will assume that pain that has a “nonphysical” origin implies imagined or contrived pain—that the child is “faking it.” The most convincing method of divesting the parents of this notion is to compare the abdominal pain with headache in adults. Most adults have occasional headaches, and although the cause rarely is associated with any abnormal physical findings or investigations, the pain is undoubtedly real and not imagined. When this concept has been grasped, it is important to guide parents on how to manage the problem.

The parents need to maintain a sympathetic attitude that acknowledges the pain but encourages continued activities and school attendance to the greatest degree possible. It is important to point out that young children are highly suggestible, and parents should refrain from questioning the child about the pain if the child is not complaining. The role of increased dietary fiber in alleviating the pain is unclear; only one published study (albeit a double-blind, randomized, controlled trial) has suggested its therapeutic value. It must be acknowledged, however, that the diets of many children in developed nations are lacking in fiber, and a trial of increasing fiber by dietary modification seems a prudent strategy that will do no harm. The impulse to commence a trial of empiric medication to provide symptomatic relief should be resisted.

The role of mental health professionals in the management of the child who has RAP is controversial, and many families will resist accepting their interventions. However, some children have pain that has clear markers of a psychogenic origin, which interferes repeatedly with school attendance and other activities. These children may be resistant to usual office management, and the intervention of a psychologist or psychiatrist skilled in chronic pain management can provide valuable insight and support.

The suspicion or clear diagnosis of constipation requires treatment with regular stool softeners, which may need to be preceded by an enema to ensure that the lower bowel is adequately evacuated. It should be made clear that the role of constipation in causing RAP cannot be determined with certainty until the child is having regular soft bowel movements for a period of weeks.

When the history suggests lactose malabsorption, most clinicians forego confirmatory investigations and recommend a trial of a lactose-free diet for several weeks. At the minimum, such a diet should eliminate obvious sources of lactose by using lactase-treated milk and avoiding ice cream and cheese. The problem with this approach is the role of the placebo effect; any change that
implies anticipated improvement may alter both the child’s and the parent’s perception of the pain.

Recognizing that enteric infections or infestations rarely cause RAP, isolation of suspected pathogens requires treatment with appropriate medications.

When the probability of abdominal migraine is seriously entertained, a trial of migraine prophylaxis seems appropriate. One study showed benefit from prophylactic pizotifen; cyproheptadine, propranolol, or amitriptyline also could be considered.

Some families and children persist in the belief that “something is wrong” despite all contrary evidence, a conviction that will be shaken only by consultation with a gastrointestinal specialist.

Prognosis
The degree to which “little bellyachers” become “big bellyachers” has puzzled clinical researchers for decades. In Apley’s classic follow-up study (1973), more than one third of former RAP patients continued to complain of abdominal pain 1 to 2 decades later. Fewer than 5% of the follow-up sample were identified as having an organic cause for their pain. In a follow-up of 161 patients who had RAP from the Mayo Clinic (Stickler and Murphy, 1979), nearly 25% maintained the complaint 5 to 17 years after their initial evaluation. A Danish study by Christensen and Mortensen (1975) found that more than one half of 34 former RAP patients still had chronic or recurrent abdominal symptoms as adults, and one third complained of nonabdominal symptoms, especially headaches. Two well-conducted follow-up studies of RAP patients by Walker et al (1995 and 1998) evaluated patients 5 to 6 years after initial evaluation. Those who had RAP reported significantly higher levels of abdominal pain and other somatic symptoms, averaged twice as many absences from work or school, and made significantly more mental health visits during the intervening years than the well patients. Certainly the best attitude the pediatrician can demonstrate is optimism that the pain will be conquered, acknowledging that for some this will not be the outcome.

Everyone who has dealt with RAP is left with some nagging anxiety that a significant organic diagnosis has been missed that will appear at some future time to direct an accusing finger at the hapless clinician who assumed in error that the cause was functional. This seems to be a rare occurrence. Stickler and Murphy’s long-term follow-up found evidence of “missed” organic disease in only 3 of 161 patients, and other studies have shown similar low rates.

Summary
Recurrent abdominal pain in childhood will continue to defy simplistic approaches to diagnosis or treatment. The wise clinician will make a careful evaluation based first and foremost on a thorough history and physical examination, supplemented as appropriate by prudently targeted investigations.

Suggested Reading
PIR Quiz

Quiz also available online at www.pedsinreview.org

1. You are evaluating a 14-year-old girl who has had RAP for the past 6 months. There is a positive family history for peptic ulcer disease in both the maternal and paternal grandfathers. The parents ask that you investigate the possibility of this condition in their daughter. Among the following, the best method to confirm the presence of *Helicobacter pylori* disease is:
   
   A. Assay of salivary antibody.  
   B. Assay of serum antibody.  
   C. Endoscopy with biopsy.  
   D. Fecal antigen testing.  
   E. Hydrogen breath test.

2. On taking the history of an 8-year-old child in whom functional RAP is suspected, which of the following characteristics of the pain would be most consistent with that diagnosis?
   
   A. Accompanied by bilious vomiting.  
   B. Associated with watery stools.  
   C. Awakens the child at night.  
   D. Located around the umbilicus.  
   E. Occurs in the presence of fever.

3. In an 11-year-old patient who has had RAP of 6 months' duration, which of the following laboratory results is most consistent with a functional disorder?
   
   A. Erythrocyte sedimentation rate of 7 mm/h.  
   B. Hematocrit of 28% (0.28).  
   C. Stool that is positive for occult blood.  
   D. Urinalysis showing 2+ proteinuria.  
   E. White blood cell count of 21 × 10⁹/mcL (21 × 10⁹/L).

4. You are considering a diagnosis of inflammatory bowel disease in a 15-year-old patient. The study of choice to confirm this diagnosis is:
   
   A. Barium enema.  
   B. Computed tomography.  
   C. Endoscopy.  
   D. Plain radiography.  
   E. Ultrasonography.
Understanding and Treating Childhood Bellyaches

Application of the pediatric Rome criteria permits a rapid, positive diagnosis, enabling the patient and family to understand and cope with abdominal pain.

Every child complains about a bellyache now and then. It is not always easy for a clinician to determine what is wrong or if it is dangerous. Children younger than ages 5 or 6 are unable to describe the sensations accurately. Toddlers do not separate emotional from physical distress. The young child’s bellyache may represent hunger, fatigue, or a need to use the bathroom. School-age children may wake with bellyaches on school days. Are they sick, or just anxious about an important test or threats by a bully? What about when the bellyache comes at a birthday party? Some bellyaches seem to arise from too much excitement or worry. When should we be concerned?

Dr. Hyman is professor of pediatrics and chief of Pediatric Gastroenterology at the University of Kansas Medical Center, Kansas City, Kansas. Dr. Danda is assistant professor of pediatrics for the departments of Pediatric Gastroenterology and Behavioral Pediatrics also at the University of Kansas Medical Center.

Address reprint requests to Dr. Hyman at University of Kansas Medical Center, 3901 Rainbow Blvd., Kansas City, KS 66160-7330.

Drs. Hyman and Danda have no industry relationships to disclose.
In 1999, an international working team of pediatric gastroenterologists arrived at a consensus for the symptom-based diagnosis of 13 pediatric functional gastrointestinal (GI) disorders. These diagnostic criteria became known as the Rome criteria, because the working team met in Rome. A functional disorder is characterized by symptoms that are caused by an alteration in the way the body functions rather than by a disease (an anatomic abnormality or serious tissue damage). Like a runner’s leg cramp or shivering from the cold, functional symptoms fall within the expected range of behavior for a healthy person.

For most pediatric functional GI disorders, no medical tests are necessary or desirable, because there is no test that confirms them. The diagnosis for each pediatric functional GI disorder depends on symptom-based criteria in the absence of warning signs for disease. Children old enough to give accurate pain histories may have the same functional abdominal pain disorders as adults. In both children and adults, functional GI disorders are more common than disease.

Using the Rome symptom-based criteria, clinicians are able to make a positive diagnosis for a majority of school-age children with bellyaches simply by taking a history and completing a physical examination that reveals no signs of disease. Families are grateful when clinicians are able to provide prompt answers to their concerns. They are satisfied when they hear effective reassurance, including a diagnosis with an explanation for the symptoms, a prognosis, a plan for treatment, and a promise of continued availability for reassessment if the symptoms change.

The traditional approach to childhood bellyaches for the past 50 years had been to rule out disease with a series of screening tests before labeling the condition just another “recurrent abdominal pain of childhood.” Reassurance may be provided that the symptoms will resolve, or the family may be referred to a mental health clinician, but no clear treatment plan is offered. Often, both clinician and family find this disposition unsatisfying. The clinician may be annoyed that the search for disease has wasted time and resources. The family may be frustrated because they understand that the clinician did not find anything wrong and there is no explanation for the symptoms. Each negative test may reinforce the belief that something is being missed. The more anxious the family becomes that something is being missed, the more tests are ordered. In some cases, a cycle of pain, anxiety, and medical testing may accelerate, and the child and family may become disabled.

In contrast to the traditional approach, application of the pediatric Rome criteria permits a rapid, positive diagnosis, enabling the patient and family to understand and cope with the symptoms. There are no tests for functional GI disorders. Some symptoms and signs that suggest a higher pretest probability of organic disease may justify diagnostic testing (Sidebar 1. In contrast, many factors do not help to discriminate between functional disorders and organic disease (Sidebar 2, see page 99).

The majority of this article focuses on functional dyspepsia, irritable bowel syndrome, and functional abdominal pain syndrome, the most common abdominal problems. Diagnostic criteria are given for other conditions, however. In a validation study for the pediatric Rome criteria, 72% of pediatric patients presenting with recurrent abdominal pain had symptom profiles that confirmed the Rome criteria.
consistent with the functional GI disorders. Chronic or recurrent bellyaches are common, affecting more than 10% of school-aged children and more than 10% of teens and adults. Most chronic or recurrent bellyaches are functional, meaning that the pain is real, but not due to any organic disease.

**DIAGNOSTIC CRITERIA**

**Functional Dyspepsia**

A diagnosis of functional dyspepsia can be made in children mature enough to provide an accurate pain history of at least 12 weeks, which need not be consecutive, within the preceding 12 months of persistent or recurrent pain or discomfort centered in the upper abdomen; no evidence, including upper endoscopy, that disease is likely to explain the symptoms; and no evidence that the dyspepsia is relieved by defecation or associated with constipation or diarrhea.

**Irritable Bowel Syndrome**

Children mature enough to provide an accurate pain history must experience at least 12 weeks, which need not be consecutive, within the preceding 12 months of symptoms to be diagnosed with irritable bowel syndrome. Abdominal discomfort or pain must be present and must include two of three features: relieved with defecation; onset associated with a change in frequency of stool, ie, constipation or diarrhea; or onset associated with a change in appearance of stool (ie, constipation or diarrhea). For the diagnosis of irritable bowel syndrome to be made, there must be no structural or metabolic abnormalities to explain the symptoms.

Other features that support the diagnosis of irritable bowel syndrome include lumpy, hard or loose, watery stools, passage of mucus in the stools, and bloating or a feeling of abdominal distention. Straining, urgency, or a feeling of incomplete evacuation of stool is also a feature, as is the passing of more than three bowel movements per day, or fewer than three bowel movements per week.

**Abdominal Migraine**

Children with three or more sudden spasms, or paroxysmal episodes, of intense abdominal pain lasting 2 hours to several days, with intervening symptom-free intervals lasting weeks to months, may have abdominal migraine. For diagnosis, there must be no evidence of metabolic, GI, or central nervous system structural or biochemical disease. Abdominal migraine must also have two or more of these features: headache during episodes; sensitivity of the eyes to light during the episodes; family history of migraine; headache on one side only;

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**SIDEBAR 2.**

**Factors that Fail to Discriminate Between Functional Abdominal Pain and Organic Disease in the Absence of Alarm Symptoms (see Sidebar 1).**

- Pain frequency, severity, location, and effects on lifestyle
- Pain-associated sleep disturbance
- Anorexia
- Nausea
- Episodic vomiting
- Constipation
- Diarrhea
- Headache
- Pallor
- Joint pain
- Anxiety
- Depression
- Behavior problems
- Recent negative life events
- Lab screening tests (CBC, Comprehensive Metabolic Panel, urinalysis, stool parasite analysis)
- Abdominal ultrasound
- Endoscopy
- Esophageal pH monitoring

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**Functional Abdominal Pain Syndrome**

Functional abdominal pain syndrome can be diagnosed when at least 12 weeks of continuous or nearly continuous abdominal pain in a school-aged child or teen is present. Other factors include: no or only occasional relation of pain with physiological events such as eating, menses, or defecation; some loss of daily functioning; assurance that the pain is not feigned; and insufficient symptoms for any other functional GI disorder to explain the pain.

and an aura or warning period consisting of blurred or impaired vision, inability to speak, or paralysis.

**Aerophagia**

Children must have at least two of the following symptoms for at least 12 weeks, which need not be consecutive, in the preceding 12 months to be diagnosed with aerophagia. Symptoms include air swallowing, abdominal distention, and repetitive belching or increased flatus. Typically, the child’s abdomen swells progressively throughout the day, there is passage
of flatus throughout the night as the child sleeps, and the abdomen is flat in the morning. The swollen abdomen is either asymptomatic or associated with a bellyache and reduced appetite.

Children with chronic pain may adapt to their discomfort, and verbal reports may be a more accurate measure of pain intensity than direct observation. If pain episodes last less than 5 minutes, the bellyache is unlikely to be anything to be concerned about, even if it recurs over many days. The closer the pain is to the umbilicus, the more likely it is to be functional.

Night waking does not help discriminate functional pain from disease. The anxiety associated with pain, and the tendency for pain to become a focus of attention at night, when children may cease distracting activities, may cause an inability to fall asleep or mid-night awakenings.

**PATHOPHYSIOLOGY OF CHILDHOOD BELLYACHES**

The publication of the Rome criteria sparked basic and clinical research in the functional GI disorders. Although these conditions are not dangerous, and there are not simple medical tests for them, we have learned a great deal about the subtleties in physiology that may accompany them.

Functional dyspepsia may be associated with one or more of three distinctive abnormalities: impaired gastric relaxation, poor gastric motility, or gastric hyperalgesia. The term hyperalgesia refers to pain sensations from stimuli that are not expected to cause pain, such as normal gastric contractions or normal increases in gastric pressures. Irritable bowel syndrome may begin following an enteric infection, as inflammation in the mucosa is slow to resolve, and inflammatory mediators alter motor nerve function and induce gastric or colonic hyperalgesia. Intestinal gas transit is impaired in patients with irritable bowel syndrome.

Pain refrains arise mainly from the brain. Chronic hyperalgesia in visceral afferent nerves alters pain pathways in the spinal cord and brain. Brain-imaging studies have shown that adults with irritable bowel syndrome process pain sensations differently from control subjects. Moreover, a patient’s pain experience is composed of and altered by not only noxious stimuli but also cognitive and emotional factors. For example, negative emotions such as depression and anxiety have been correlated with increased pain intensity and disability. Increased negative emotions can induce increased muscle tension and pain; likewise, increased pain experiences can trigger more anxiety or depression.

For the purpose of functional bellyaches, the brain and gut should be considered inexorably linked.

**Educating Patients and Families**

The decision to seek medical care for a bellyache arises from a parent or caretaker’s concern for the child. The parent’s threshold for concern varies with his or her own experiences and expectations, coping style, and perception of the child’s illness. For this reason, the office visit is not only about the child’s bellyache but also about the family’s conscious and unconscious fears. The clinician must not only make a diagnosis but also recognize the family’s emotional...
tune and ability to function. An effective treatment plan, therefore, must attend to the child and the family and depends upon securing a therapeutic alliance with both.

Effective communication between the clinician and the family ensures that the child’s problems are understood and treated properly. Every parent wants the answer to four questions: What is wrong with my child? Is it dangerous? Will it go away? What can we do about it? The clinician answers each of these questions at the first visit in the appropriate order. An example answer is, “Today your child’s symptoms met the criteria for a diagnosis of irritable bowel syndrome. It is not dangerous. It comes and goes, and may even change over time to a different kind of bellyache. However, there are many effective choices for treatment, and we will discuss your options to find the ones that best meet your preferences.”

The Influence of Stress on Bellyache

When a child has functional symptoms, the child and parents often hear that stress is a part of the problem. Because the term is used broadly and frequently, the word “stress” has an ambiguous meaning to most people. When used in relation to their child’s bellyaches, parents tend to interpret the word as meaning it is a psychological issue. An explanation of stress based on its biology facilitates understanding of the mind/body connection. Parents need to understand that the experience of stress includes physiologic as well as cognitive, emotional, and behavioral factors.

Stress is disruption of homeostasis, the way that the body regulates itself. This disruption can occur due to environmental factors, such as exposure to cold temperature, or in response to an event that is arousing, exciting, or threatening. For children, examples of stress-associated situations might include a birthday party, airplane ride, increased academic demands, changes in peer relationships, or a death in the family. Indeed, the bellyaches themselves are significant sources of stress for the child and family as well and should be described as such.

One key to helping families understand stress is describing the autonomic, physical response that occurs—the increase in heart rate and blood pressure, the slowing of the digestive system. Because the body responds to arousal with a slowing of digestive functions, the connection between stress and bellyaches is easier to understand.

A recent study of schoolchildren evaluated for bellyache found most children believed the bellyache was related to stress and feared the clinician might find a disease.11 In contrast, parents worried that the clinician might not find a physical cause, and feared their child’s problem would be labeled psychological. The parents may be defensive about a psychological cause for their child’s bellyache, because they fear the clinician will blame them for faulty child rearing. There may also be a cultural foundation for familial bias against psychological disorders. Clinicians not formally trained in psychology may also feel unease about making a diagnosis that would force them to refer the patient for additional services.

Factors Influencing Pediatric Gastrointestinal Disorders

No functional disorder is totally medical or totally psychological. The GI tract is connected to the brain by millions of nerves, and each system influences the other. Functional bellyaches are a result of three interacting physiologic factors: motility, sensory perception, and arousal. Motility is defined by GI tract wall movements and the transit of its contents. Sensory perception includes feelings of pain, discomfort, or nausea. Arousal is influenced by internal factors such as pain, beliefs, interpretations, and emotions, and by external factors such as physical threats or the pleasure of seeing an old friend.

An explanation for the pain is often required for effective reassurance and for understanding why drug treatment and cognitive behavioral therapy work. An example would be a bellyache that starts with a viral gastroenteritis with symptoms such as vomiting and diarrhea, which are symptoms of disordered motility. The stretching of the bowel and the inflam-
mation within the bowel wall stimulate pain nerves that travel from the GI tract to the spinal cord. In the spinal cord, the intestinal pain nerves transfer their messages to nerves that travel up to the brain. Pain or the sensation of nausea is thus delivered to the brain. Based on the child’s interpretation of these symptoms (influenced by the caregiver’s past and current response to the symptoms), the child changes his or her behavior. The child stops playing and lies down.

It is easy to understand how abnormal motility causes pain and changes behavior, but pain and emotions also change motility. If one has pain for any reason, such as putting a hand into ice water (a validated research test for pain responses), gastric emptying slows. When a person is aroused by emotion, motility changes. When a person is sad, pains grow worse; emotional discomfort accompanies physical discomfort. Thus, all three factors influence each other, and bellyaches are not all psychological.

TREATMENT OF PEDIATRIC GASTROINTESTINAL DISORDERS

Perceptions of Illness

There is often little relationship between pain severity and the degree of danger a condition poses. Thoughts, emotions, attention, and expectations influence pain. For example, an athlete injured in a competition may not notice pain because her attention is completely focused on the event. When distractions are reduced afterward, however, the pain sensations may become more intense.

We are accustomed to requiring a swift response to symptoms to reduce the danger and discomfort to the child. When a child is diagnosed with a functional disorder, however, the condition is not dangerous to the child, and the acute medical response is no longer appropriate. Rather, the goal is helping the child learn how to manage the symptoms and to behave again as a healthy child, attending school and participating in age-appropriate activities. This requires changing the perception that this is a sick child.

First, parents must be reassured that organic disease is unlikely in the absence of alarm symptoms such as persistent fever, weight loss, growth failure, bleeding, persistent vomiting, and severe diarrhea. When it fits the medical history, the Rule of Ones helps provide effective reassurance. The Rule of Ones states that if a patient has a bellyache only, it is probably functional. If there is a bellyache and a second sign or symptom, then a careful evaluation is in order. If, in addition to the bellyache, there is weight loss, fever, daily or nightly vomiting, blood in the stool, difficulty swallowing, or pain with urination, then medical tests are necessary.

The Rule of Ones does not help explain irritable bowel syndrome, functional fecal retention, or abdominal migraine, because there is more than one symptom for each of these functional GI disorders. The Rule of Ones is most helpful for explaining bellyaches in children who do not meet diagnostic criteria because of a short duration of symptoms. The Rule of Ones must always be used in combination with the promise of continuing availability of the clinician, which helps cement the therapeutic alliance between family and clinician. Handing a business card with your e-mail address to the child reinforces the collaborative nature of the patient-clinician relationship (not to mention that children often have computer skills lacking in their parents).

Parents and patients have a choice when it comes to treatment of functional dyspepsia and irritable bowel syndrome. Everyone receives the educational component. For many, a rational explanation for symptoms is all that is necessary. Others may prefer medication, cognitive-behavior therapy, or both.

Drugs

The placebo response rate in functional GI disorders is about 40%. This high response rate to placebo may reflect the importance of the education component, the clinician-patient-family therapeutic alliance, and the patient’s expectation for improvement.

Upper GI endoscopy is often recommended in adults with dyspepsia. Endoscopy serves to screen adults for gastric cancer and peptic ulcer disease. In otherwise healthy children with bellyaches, gastric cancer is unheard of and peptic ulcer disease is rare, so the risk of masking disease with drug treatment is small. In communities where pediatric endoscopy is not readily available, a diagnostic and therapeutic drug trial is justified.

A trial using a proton pump inhibitor seems appropriate for dyspepsia, due to the device’s recorded safety and efficacy. Proton pump inhibitors inhibit gastric acid secretion, so if acid is involved in the production of pain, proton pump inhibitors should be effective. When they do not reduce pain in a child or adolescent who meets symptom-based criteria supporting a functional GI disorder diagnosis, then the problem is likely to be gastric hyperalgesia.

Low-dose tricyclic antidepressant treatment for adults with abdominal pain caused by a functional GI disorder is safe and effective. Tricyclics have been used for decades in children to treat reflex sympathetic dystrophy, bedwetting, and attention-deficit disorder, but there are no published pediatric studies with tricyclics for functional GI disorders. In the authors’ experience, imipramine or amitriptyline administered at a dose of 0.2 mg/kg at bedtime, increased weekly by 0.2 mg/kg to a maximum of 1 mg/kg or 50 mg, may
reduces pain by increasing the pain threshold of sensory afferent nerves. Amitriptyline has strong anticholinergic effects and may be ideal not only for reducing pain but also for improving restful sleep and eliminating loose stools in diarrhea-predominant irritable bowel syndrome. For constipation-predominant irritable bowel syndrome, imipramine is preferable, because it is less constipating than amitriptyline.

In the authors’ experience, tricyclics are helpful in reducing the frequency and intensity of abdominal pain in about four of five children and adolescents with functional dyspepsia, irritable bowel syndrome, and functional abdominal pain syndrome. About one in 20 patients complains of hyperactivity, restlessness, insomnia, and irritability with a tricyclic, necessitating discontinuation of the drug. In the authors’ experience, gabapentin reduces pain in about half the patients who do not respond to tricyclics. Antispasmodics also have also been used for irritable bowel syndrome, but well designed studies demonstrating efficacy are lacking.

During the past decade, new drugs specifically targeting the serotonin receptors in the GI tract that mediate pain and motility have served as treatment advances. Alosetron at a dose of 1 mg once or twice a day reduced pain and diarrhea in diarrhea-predominant irritable bowel syndrome. Tegaserod at a dose of 2 or 6 mg twice a day reduced pain and increased the frequency of stools in constipation-predominant irritable bowel syndrome.

Cognitive-behavior Therapy
Cognitive-behavior therapy (CBT) consists of providing patients with active behavioral strategies such as relaxation and helping patients use the thinking part of their brain to change the way they think about and cope with pain and stress. Relaxation techniques help reduce autonomic arousal and muscle tension and have been proven to be effective treatment for children with bellyaches.

The cognitive part of therapy involves challenging patients’ passive approach to pain and empowering them to become more active in their recovery. Family sessions may also be necessary to help the family support the child in getting well by changing the way they behave around the patient. Learning these cognitive behavioral strategies can be achieved in about 6 or 8 hours of therapy.

When combined with education and relaxation, CBT is effective for irritable bowel syndrome. Disadvantages of CBT are its lack of availability in many areas, as well as reluctance by medical insurers to cover this expense. The psychologist or mental health clinician must also understand functional disorders and work closely with the physician. Families may feel abandoned if they are referred to a mental health clinician without follow-up from the physician to monitor medications and any new symptoms. Continued reassurance from the physician empowers patients to become more active in their treatment and recovery.

Diet and Dietary Fiber
Diet is important, but every child is different. It is important to eat a well-balanced diet. If it is possible to identify foods that trigger symptoms, those foods can be reduced or eliminated from the diet. In large, population-based studies, dietary fiber seems to have some long-term health advantages. In the short term, increasing fiber in a child’s diet may not be worth the effort. Getting toddlers to eat anything every day becomes a struggle, because it is developmentally appropriate for them to be testing their ability to control their environment. The same is true for adoles-

The future
In years to come, the acquisition of high-quality evidence will clarify many of the unknown factors about bellyaches. There will be modification and validation of the Rome criteria for pediatric functional GI disorders. The next working teams will meet in December 2004 to develop the Rome III criteria. Suggestions for the criteria may be sent to Dr. Hyman at hyman@kumc.edu.

Complementary and Alternative Medicine
In one randomized, double-blind, placebo-controlled, 2-week trial, peppermint oil reduced pain in 75% of those treated.

PAIN-ASSOCIATED DISABILITY SYNDROME
Visceral pain-associated disability syndrome (PADS) is defined by at least 2 months of continuous or recurrent abdominal or chest pain (or other discomfort such as nausea) sufficient to disrupt activities of daily life. Also present are impairment in normal functioning that the patient attributes to the pain, failure of previous pain-management strategies that would be expected to improve pain and enhance functioning for similar patients, and lack of an organic explanation after a thorough medical symptom assessment.
Patients with PADS experience pain that is not feigned or induced by the patient or caretakers. PADS is a term that enables clinicians to describe a subset of chronically ill patients with severe restriction in daily functioning (cannot eat, or cannot attend school), and in whom acute pain management strategies fail. Bellyaches severe enough to curtail normal activities may affect up to 20% of school-aged children but rarely cause prolonged school absence or weight loss requiring special nutritional support.

Visceral PADS most often is associated with an unrecognized functional GI disorder and a comorbid psychological stressor, such as an unrecognized learning disability, social anxiety disorder, or a family problem such as substance abuse or separation anxiety. Low perceived social or academic competence is linked to the higher levels of disability associated with functional bellyaches.

In addition, the practice of punitive medicine — ordering more invasive tests with each negative battery of studies — increases pain experiences in the patient and parental anxiety concerning whether something is being missed. The pain and arousal caused by each negative test increases the visceral hyperalgesia and promotes disability. Thus, zealous clinicians may contribute to the problem.

Treatment for PADS begins with defining the problem and providing an explanation for functional disorders and the interaction among motility, pain, and arousal that satisfies the patient and family. It is important to avoid separating psychological from physical factors; the two are so closely linked, there is no reason to dichotomize.

Treatment requires a collaboration that includes a medical clinician, mental health professional, the patient, and the family. Medical interventions, such as the use of narcotics, parenteral nutrition, and tube feedings, are systematically withdrawn, and all medical testing stops. Medication is prescribed to regulate the sleep cycle, because it is nearly always disrupted in patients with PADS. CBT and relaxation techniques improve patient coping. Family therapy is often required to reduce arousal in the family and the patient. Neuropathic pain medications such as the tricyclics, gabapentin, and clonidine are often helpful. Daily exercise that does not cause discomfort and activities that promote a sense of well-being, such as listening to music or getting a massage, are recommended. Comorbid mental health issues should be identified and treated when present. Patients and their families return weekly until the patient feels capable of coping with his or her functional disorder.

SUMMARY
Symptom-based diagnoses for most childhood bellyaches may be applied at the first visit, reducing family anxiety and healthcare spending. Primary care clinicians are able to diagnose and treat these disorders effectively. The promise of continuing availability is essential and assures that no disease will be missed.

REFERENCES
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. JPN 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...
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.

TABLE 1. Normal frequency of bowel movements

<table>
<thead>
<tr>
<th>Age</th>
<th>Bowel movements per week</th>
<th>Bowel movements per day</th>
</tr>
</thead>
<tbody>
<tr>
<td>0–3 Months</td>
<td>Breast-fed 5–40</td>
<td>2.9</td>
</tr>
<tr>
<td></td>
<td>Formula-fed 5–28</td>
<td>2.0</td>
</tr>
<tr>
<td>6–12 months</td>
<td>5–28</td>
<td>1.8</td>
</tr>
<tr>
<td>1–3 years</td>
<td>4–21</td>
<td>1.4</td>
</tr>
<tr>
<td>More than 3 years</td>
<td>3–14</td>
<td>1.0</td>
</tr>
</tbody>
</table>


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

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

*Categories 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.

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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.

* J Pediatr Gastroenterol Nutr, Vol. 43, No. 3, September 2006*
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. 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 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).

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**TABLE 3. History in pediatric patients with constipation**

<table>
<thead>
<tr>
<th>Age</th>
<th>Sex</th>
<th>Chief symptom</th>
<th>Constipation history</th>
<th>Frequency and consistency of stools</th>
<th>Pain or bleeding with passing stools</th>
<th>Abdominal pain</th>
<th>Waxing and waning of symptoms</th>
<th>Age of onset</th>
<th>Toilet training</th>
<th>Fecal soiling</th>
<th>Withholding behavior</th>
<th>Change in appetite</th>
<th>Nausea or vomiting</th>
<th>Weight loss</th>
<th>Perianal fissures, dermatitis, abscess, or fistula</th>
<th>Current treatment</th>
<th>Current diet (24-hour recall history)</th>
<th>Current medications (for all medical problems)</th>
<th>Oral, enema, suppository, herbal</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
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</tr>
</tbody>
</table>

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*J Pediatr Gastroenterol Nutr, Vol. 43, No. 3, September 2006*
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 4. Differential diagnosis of constipation

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

TABLE 5. Physical examination of children with constipation

<table>
<thead>
<tr>
<th>General appearance</th>
<th>Vital signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Temperature</td>
<td>Pulse</td>
</tr>
<tr>
<td>Respiratory rate</td>
<td>Blood pressure</td>
</tr>
<tr>
<td>Growth parameters</td>
<td>Head, ears, eyes, nose, throat</td>
</tr>
<tr>
<td>Neck</td>
<td>Cardiovascular</td>
</tr>
<tr>
<td>Lungs and chest</td>
<td>Abdomen</td>
</tr>
<tr>
<td>Distension</td>
<td>Palpable liver and spleen</td>
</tr>
<tr>
<td>Palpable liver and spleen</td>
<td>Fecal mass</td>
</tr>
<tr>
<td>Anal inspection</td>
<td>Anal wink</td>
</tr>
<tr>
<td>Position</td>
<td>Anal tone</td>
</tr>
<tr>
<td>Stool present around anus or on clothes</td>
<td>Fecal mass</td>
</tr>
<tr>
<td>Perianal erythema</td>
<td>Presence of stool</td>
</tr>
<tr>
<td>Skin tags</td>
<td>Consistency of stool</td>
</tr>
<tr>
<td>Anal fissures</td>
<td>Other masses</td>
</tr>
<tr>
<td>Rectal examination</td>
<td>Explosive stool on withdrawal of finger</td>
</tr>
<tr>
<td>Anal wink</td>
<td>Occult blood in stool</td>
</tr>
<tr>
<td>Anal tone</td>
<td>Back and spine examination</td>
</tr>
<tr>
<td>Fecal mass</td>
<td>Dimple</td>
</tr>
<tr>
<td>Presence of stool</td>
<td>Tuft of hair</td>
</tr>
<tr>
<td>Consistency of stool</td>
<td>Neurological examination</td>
</tr>
<tr>
<td>Other masses</td>
<td>Tone</td>
</tr>
<tr>
<td>Explosive stool on withdrawal of finger</td>
<td>Strength</td>
</tr>
<tr>
<td>Occult blood in stool</td>
<td>Cremasteric reflex</td>
</tr>
<tr>
<td>Back and spine examination</td>
<td>Deep tendon reflexes</td>
</tr>
</tbody>
</table>
TABLE 6. Physical findings distinguishing organic constipation from functional constipation

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

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.

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.

*Adapted from the NASPGHAN Constipation Guideline Committee.*

The information is obtained from the literature. Information mostly obtained from use of the drug in question or from clinical experience. Only information proven to be accurate by clinical trials or other proven methods is included. The safety of long-term maintenance use of many of these agents is not well established.

**TABLE 7. Medications for use in treatment of constipation**

<table>
<thead>
<tr>
<th>Laxatives</th>
<th>Dosage</th>
<th>Side effects</th>
<th>Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Osmotic</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lactulose†</td>
<td>1–3 mL/kg/day in divided doses; available as 70% solution.</td>
<td>Flatulence, abdominal cramps; hypernatremia has been reported when used in high dosage for hepatic encephalopathy; case reports of nontoxic megacolon in elderly.</td>
<td>Synthetic disaccharide. Well tolerated long term.</td>
</tr>
<tr>
<td>Sorbitol†</td>
<td>1–3 mL/kg/day in divided doses; available as 70% solution.</td>
<td>Same as lactulose.</td>
<td>Less expensive than lactulose.</td>
</tr>
<tr>
<td>Barley malt extract†</td>
<td>2–10 mL/240 mL of milk or juice</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Magnesium hydroxide†</td>
<td>1–3 mL/kg/day of 400 mg/5 mL; available as liquid, 400 mg/5 mL and 800 mg/5 mL, and tablets.</td>
<td>Infants are susceptible to magnesium poisoning. Overdose can lead to hypermagnesemia, hypophosphatemia and secondary hypocalcemia.</td>
<td>Acts as an osmotic laxative. Releases cholecystokinin, which stimulates gastrointestinal secretion and motility. Use with caution in renal impairment.</td>
</tr>
<tr>
<td>Magnesium citrate†</td>
<td>&lt;6 Years, 1–3 mL/kg/day; 6–12 Years, 100–150 mL/day; &gt;12 Years, 150–300 mL/day; in single or divided doses. Available as liquid, 16.17% magnesium.</td>
<td>Infants are susceptible to magnesium poisoning. Overdose can lead to hypermagnesemia, hypophosphatemia and secondary hypocalcemia.</td>
<td></td>
</tr>
<tr>
<td>PEG 3350</td>
<td>Disimpaction: 1–1.5 g/kg/day for 3 days Maintenance 1 g/kg/day</td>
<td></td>
<td>Superior palatability and acceptance by children. Safety studies necessary before widespread use is recommended in infants.</td>
</tr>
<tr>
<td><strong>Osmotic enema</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Phosphate enemas</td>
<td>&lt;2 Years old: to be avoided; ≥2 years old: 6 mL/kg up to 135 mL</td>
<td>Risk of mechanical trauma to rectal wall, abdominal distention or vomiting. May cause severe and lethal episodes of hyperphosphatemia hypocalcemia, with tetany.</td>
<td>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.</td>
</tr>
<tr>
<td><strong>Lavage</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Polyethylene glycol-electrolyte solution</td>
<td>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.</td>
<td>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.</td>
<td>Information mostly obtained from use for total colonic irrigation. May require hospital admission and nasogastric tube.</td>
</tr>
<tr>
<td><strong>Lubricant</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mineral oil†</td>
<td>&lt;1 Year old; not recommended. Disimpaction: 15–30 mL/yr of age, up to 240 mL daily. Maintenance: 1–3 mL/kg/day.</td>
<td>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.</td>
<td></td>
</tr>
<tr>
<td><strong>Stimulants</strong></td>
<td></td>
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</tr>
<tr>
<td>Senna</td>
<td>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.</td>
<td>Idiosyncratic hepatitis, Melanosis coli, Hypertrophic osteoarthropathy, analgesic nephropathy.</td>
<td></td>
</tr>
<tr>
<td>Bisacodyl</td>
<td>≥2 Years old: 0.5–1 suppository 1–3 tablets per dose. Available in 5-mg tablets and 10-mg suppositories.</td>
<td>Abdominal pain, diarrhea and hypokalemia, abnormal rectal mucosa, and (rarely) proctitis. Case reports of urolithiasis.</td>
<td></td>
</tr>
<tr>
<td>Glycerin suppositories</td>
<td></td>
<td>No side effects.</td>
<td></td>
</tr>
</tbody>
</table>

*Adapted from the NASPGHAN Constipation Guideline Committee.*

The information is obtained from the literature. Information mostly obtained from use of the drug in question or from clinical experience. Only information proven to be accurate by clinical trials or other proven methods is included. The safety of long-term maintenance use of many of these agents is not well established.
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

Gregory S. Liptak, MD
Syracuse, NY

Richard B. Colletti, MD
Burlington, VT

Joseph M. Croffie, MD
Indianapolis, IN

Carlo DiLorenzo, MD
Columbus, OH

Walton Ector, MD
Charleston, SC

Samuel Nurko, MD
Boston, MA

**REFERENCES**


NASCUGHAN CONSTIPATION GUIDELINE COMMITTEE


