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CASE MANAGEMENT Unremitting Functional Constipation and Fecal Incontinence in Childhood

A 12-year-old female is referred to the Center for Colorectal and Urogenital Health at Riley Children's at Indiana University Health because of refractory abdominal pain, chronic constipation, and fecal incontinence requiring the use of pull-up diapers. Previous treatment included behavior modification toilet training, increased dietary fiber, and the use of laxatives: mineral oil, polyethylene glycol (MiraLAX®), and senna (Senokot®). Stooling issues have resulted in embarrassment and social isolation. (*continued on page 2*)

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OBJECTIVES
After reading this article, the reader should be able to:

- Define functional constipation and quantify its prevalence in children.
- Identify the common trigger for the development and persistence of functional constipation in childhood.
- Discuss the therapeutic roles of biofeedback and sacral nerve stimulation in functional constipation.
- Describe the Malone antegrade continence enema (MACE) procedure and its use in the treatment of functional constipation.
- Summarize the impact of the MACE procedure on quality of life in children with refractory functional constipation and fecal incontinence.

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On physical examination, the child is thin and has a distended, soft, and nontender abdomen. On deep palpation, firm stool is felt in the left lower quadrant. The perineum is stained with dried stool, and firm stool is palpable on digital rectal examination. Radiographic, motility, and manometry studies are performed and show a colon distended with stool and mild sigmoid dilation. Good contractions, voluntary anal sphincter relaxation, and rectal distention occur after bisacodyl stimulation. A diagnosis of functional constipation with fecal incontinence is made.

Overview of Functional Constipation in Children

Constipation is common among children, accounting for an estimated three percent of all visits to pediatricians and up to 30 percent of visits to pediatric gastroenterologists. [1] Functional or idiopathic constipation, defined as persistently difficult, infrequent, or seemingly incomplete defecation without evidence of a primary anatomic or biochemical cause and often associated with abdominal pain,[2] comprises more than 95 percent of cases of constipation in healthy children aged one year and older *(Table 1)*.[3] Children are at highest risk for developing functional constipation during three time periods: 1) following the introduction of cereals and solid food into the diet, 2) with toilet training, and 3) at the start of school. Both autism spectrum disorders and attention deficit/hyperactivity disorder appear to predispose to functional constipation, and genetics may also play a role.

"Painful defecation is a common trigger for stool withholding and contributes to the development and persistence of functional constipation," explains R. Cartland Burns, MD, associate professor of pediatric surgery at Indiana University School of Medicine and pediatric surgeon at Riley Children's Health. "When a child avoids defecating because of pain, water is absorbed from the stool, and hard fecal matter accumulates in the rectum, causing even more pain when it is passed. Ignoring the urge to defecate may initially be a conscious decision, but it eventually becomes automatic."

Fecal incontinence (encopresis) occurs in up to half of all children with functional constipation, likely the result of chronic rectal distension, which shortens the anal canal, and dilation of the internal anal sphincter.[4] This in turn causes transient relaxation of the external anal sphincter and the release of semisolid stool onto the perianal skin and clothing.

Diagnosis and Evaluation

The diagnosis of functional constipation in children relies on the Rome IV criteria, which require at least two of six symptoms describing stool frequency, hardness, size, fecal incontinence, or volitional stool retention *(Table 2)*.[5,6]

"The evaluation of a child with functional constipation refractory to standard medical management may include plain and/or contrast radiographs (i.e., barium enema) and motility studies to measure colon transit time," says Dr. Burns. "Anorectal manometry may be performed to distinguish impaired rectal sensation from decreased propulsive forces or a functional outlet obstruction, such as anismus, also referred to as dyssynergic defecation: the paradoxical contraction of the external anal sphincter during defecation."

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TABLE 1. COMMON CAUSES OF CONSTIPATION IN CHILDREN[11,12]

Type of constipation	Common causes
Functional	Idiopathic, developmental disorders
Motility-related	Hirschsprung disease, myopathy
Congenital anomalies	Anorectal malformations (e.g., imperforate anus, anal stenosis, chloaca) Spinal cord abnormalities (e.g., myelomeningocele, myelomalacia, spina bifida)
Neurologic	Cerebral palsy, congenital megacolon
Endocrine/metabolic/systemic disease-related	Hypothyroidism, renal tubular acidosis, diabetes mellitus, diabetes insipidus, hypercalemia, cystic fibrosis
Medication-related	Anticonvulsants, antipsychotics, opioids, codeine
Allergy-related	Cow's milk, celiac disease

TABLE 2. ROME IV CRITERIA FOR THE DIAGNOSIS OF FUNCTIONAL CONSTIPATION IN CHILDREN[5,6]

Infants and toddlers ≤4 years	Children with development age of ≥4 years
At least two of the following present for at least one month	At least two of the following present for at least once per week for at least one month*
• Two or fewer defecations per week	• Two or fewer defecations in the toilet per week
• History of excessive stool retention	• At least one episode of fecal incontinence per week
• History of painful/hard bowel movements	• History of retentive posturing or excessive volitional stool retention
• History of large-diameter stools	• History of painful/hard bowel movements
• Presence of a large fecal mass in the rectum	• Presence of a large fecal mass in the rectum
	• History of large-diameter stools that may obstruct the toilet
In toilet-trained children, the following additional criteria may be used	Symptoms cannot be fully explained by another medical condition
• At least one episode weekly of incontinence	
• History of large-diameter stools that may obstruct the toilet	

*Symptoms are insufficient to fulfill the diagnostic criteria of irritable bowel syndrome.

Treatment of Unremitting Functional Constipation in Children

Biofeedback

For some children with chronic functional constipation and dyssynergic defecation, a presumed learned adverse behavior, biofeedback using visual or audio displays of anorectal activity added to conventional treatment may be helpful. Long-term follow-up of 30 children aged six to 14 years found that biofeedback therapy provided in the hospital or in the hospital and at home achieved recovery in 60 percent of the participants and improvement in 33 percent *(Figure 1)*.[7]

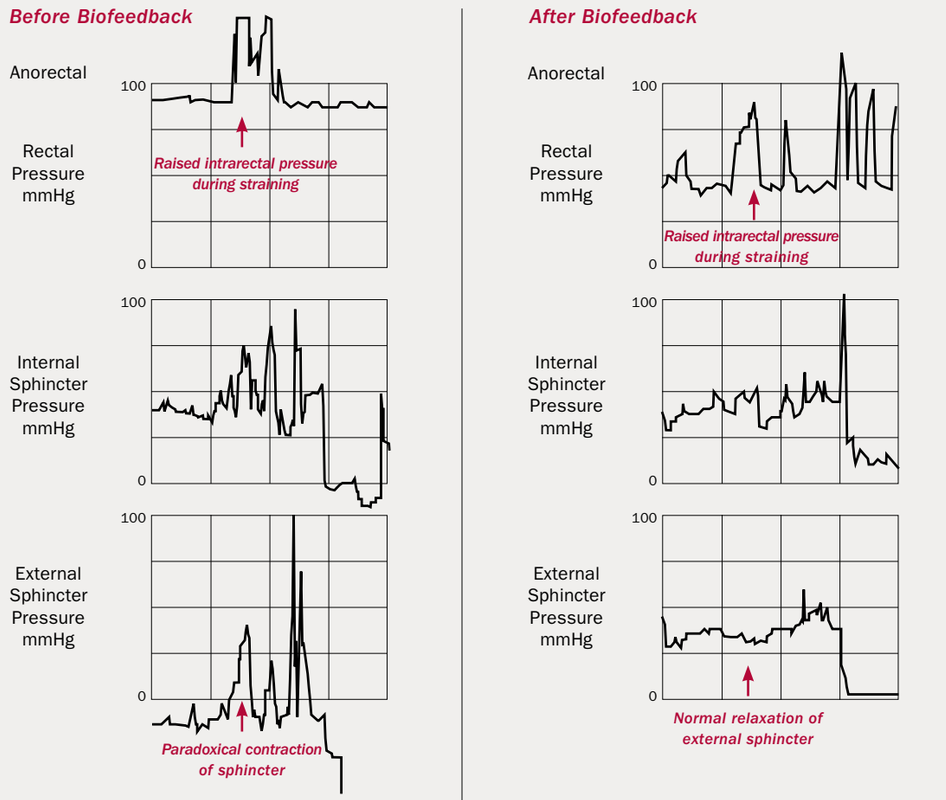
Sacral Nerve Stimulation

Another option for bowel control in children with unremitting functional constipation and fecal incontinence is implantation of a stopwatch-size device that emits a continuous, mild electrical pulse through a wire to stimulate the sacral nerves, thus strengthening the pelvic floor muscles and sphincter complex.

"Before we implant the InterStim® (Medtronic) unit, patients participate in a 14-day trial of a wearable, external version to determine if nerve stimulation is successful," Dr. Burns describes. "If it is, the surgeon inserts a permanent device under the skin in the sacral area. The patient can adjust the intensity of the electrical pulses (within physician-set limits), via a remote control."

In study of 72 adults treated with sacral nerve stimulation using the InterStim device, 89 percent reported a reduction in fecal incontinence, from 9.1 episodes weekly at baseline to 1.7 episodes at five years of follow-up, with 26 patients (36 percent) having complete continence.[8]

Figure 1. Anorectal manometry before and after biofeedback[7]



Malone Antegrade Continence Enema Procedure

When medical management fails to relieve chronic constipation and fecal incontinence, the Malone antegrade continence enema, or MACE, may be considered. This laparoscopic procedure, performed under general anesthesia via a small right lower quadrant incision, creates a cutaneous, nonrefluxing appendicostomy, with the stoma typically hidden within the umbilicus and accessed by a button *(Figure 2)*.[9] The technique allows periodic catheter access to the right colon for administration of enemas to flush stool *(Figure 3)*.

"In 2017, IU School of Medicine Divisions of Pediatric Surgery and Pediatric Gastroenterology reported on the natural history of the MACE procedure in 93 children aged 7 to 11 years with unremitting constipation and fecal incontinence followed up to 5.5 years at Riley Children's Health,"[10] says Dr. Burns. "The procedure, which was combined with sigmoid colectomy in five patients with neuropathy isolated to the sigmoid colon, was rapidly effective, with 99 percent of the children experiencing improvement at one month. At the end of the follow-up period, 83 percent had normal bowel function, and 95 percent reported improvement. Overall morbidity was 55 percent, mostly related to minor complications (e.g., surgical site infection) that resolved with time, although 12 children required an additional operation."

Figure 3. Colon flushing via MACE

The MACE tube is situated in the cecum and daily irrigations result in evacuation of the entire colon. Evacuation is typically seen within 30 minutes after flushing.

Figure 2. MACE stomal button

This laparoscopic procedure, performed under general anesthesia via a small right lower quadrant incision, creates a cutaneous, nonrefluxing appendicostomy, with the stoma typically hidden within the umbilicus and accessed by a button.

Following a discussion with the pediatric gastroenterologist and pediatric surgeon, the patient and her parents decide on the MACE procedure. After surgery, the child is hospitalized for two days to monitor healing and teach the family how to flush using the MACE button. She is discharged on a regimen of daily flushing using a saline solution that also contains polyethylene glycol and docusate (Colace®).

With practice, most patients who have undergone a MACE procedure can perform a complete colon evacuation in about 30 minutes, according to Dr. Burns, who recommends that flushing be performed in the evening to allow sufficient time and avoid rushing. A laxative and/or motility agent may be added to the saline solution, depending on the needs of the individual, with any additives adjusted or discontinued as necessary over time.

"Patients are seen at our multidisciplinary clinic* at one month, three months, and then every three months post-MACE surgery," Dr. Burns reports. "During these visits, we assess the efficacy of the procedure by asking questions about the number of accidents they've had, whether bowel habits have become predictable, and if flushing is pain-free. We also monitor long-term outcomes, including stomal stenosis, prolapse, and leakage; small bowel obstruction; recurrent impactions; and any requirement for additional surgery.

"The MACE balloon catheter is replaced at three-month intervals," continues Dr. Burns. "This simple procedure is usually done at clinic, but some parents learn to change the catheter at home."

After 18 months of successful management of bowel function and no episodes of incontinence, the patient begins to have spontaneous bowel movements without MACE flushing. Over the next several months, she is weaned from daily flushing, with the time interval between flushes gradually extended to every four days. Thirty months after surgery, the patient no longer requires the MACE. The catheter is removed, and a dressing is placed over the stoma, which closes on its own in two days.

At three-year follow-up, the child, now a high school sophomore, continues to have normal bowel function and reports participating in extracurricular activities with confidence.

"Children with unremitting functional constipation and fecal incontinence and their families experience significant social and emotional disruption, and a MACE procedure can transform their quality of life (QOL)," Dr. Burns concludes. "In our study of 15 consecutive children who underwent the MACE procedure at Riley Children's Health, the mean QOL score was 64 before surgery and 92 one year after the procedure, with every patient reporting significant improvement in each QOL category at six-month follow-up."[9]

**Comprised of pediatric surgery, pediatric gastroenterology, nursing, pediatric urology (when appropriate), and enterostomal therapy.*

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A fellow of the American College of Surgeons and a member of the American Pediatric Surgical Association and other professional organizations, Dr. Burns is the author of more than 50 peer-reviewed publications and textbook chapters and is frequently invited to lecture at national conferences.

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