



# Disorders of defecation and fecal continence

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■ Although fecal incontinence and disorders of defecation are not uncommon, these disorders are poorly understood by most physicians. Successful management requires an understanding of colorectal function, delineation of the problem and potential contributing factors, careful examination of the anorectal and pelvic floor areas, and psychosocial assessment. Specialized studies to evaluate colorectal function frequently help determine patterns of abnormality and may suggest appropriate therapeutic approaches. Therapeutic options include pharmacologic, behavioral, and surgical approaches, which often ameliorate symptoms and dramatically improve quality of life.

□ INDEX TERMS: DEFECTION; FECAL INCONTINENCE □ CLEVE CLIN J MED 1989 56:491-501

**F**ECAL incontinence and disorders of defecation affect all age groups. Unfortunately, many physicians are not knowledgeable about anorectal continence mechanisms and, in general, constipation and defecatory disorders are poorly understood. Proper evaluation of patients with these complaints requires an understanding of the normal function of the anorectal area and an awareness of currently available techniques for assessing colonic and anorectal function.

## ANORECTAL STRUCTURES AND FUNCTION

The main function of the anorectum is to store fecal wastes and allow elimination in a socially acceptable manner. The rectum serves as a storage reservoir, the pelvic floor muscles and two anal sphincters regulate defecation and retention of feces, and a sensory mechanism allows awareness of rectal filling and impending

defecation. The important structural components of this area are illustrated in *Figure 1*.

## PHYSIOLOGY OF CONTINENCE

Afferent nerves that supply the rectum allow perception of rectal distension with relatively small volumes.<sup>1</sup> Activation of stretch receptors by distension with air or stool produces a sensation of fullness or an urge to defecate while simultaneously inhibiting the internal anal sphincter (IAS), which transiently reduces the resting pressure of the anal canal (*Figure 2*). Incontinence is prevented by the contemporaneous voluntary contraction of the puborectalis muscle and the external anal sphincter (EAS), the former by narrowing the anorectal angle and the latter by increasing anal canal pressure (*Figure 1*). The rectum quickly accommodates to the fecal material through adaptive compliance, in which intraluminal pressures increase by relatively small amounts in response to increases in rectal volume.<sup>2</sup> This permits delay of defecation until an appropriate time.

Thus, continence is maintained by a number of different mechanisms. These include: the ability to sense and distinguish the nature of rectal contents (anorectal sensation); the ability of the rectum to store feces through

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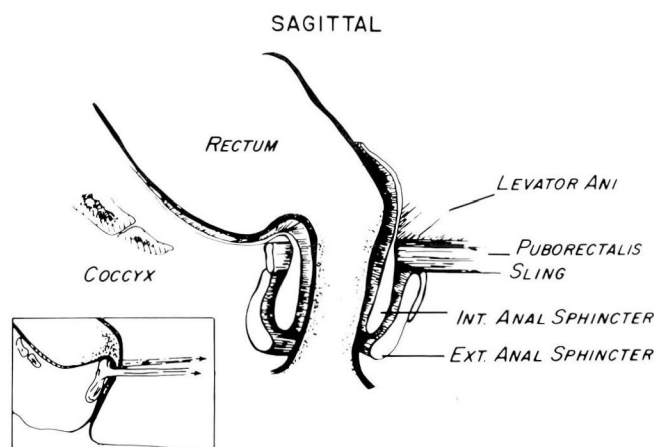


FIGURE 1. Sagittal view of the anorectum illustrating the important structural components. Inset depicts the anterior pull of the anorectum and narrowing of the anorectal angle during contraction of the puborectalis muscle (Wald A. Fecal incontinence: effective nonsurgical treatments. *Postgraduate Medicine* 1986; 80(3):123–130. ©McGraw-Hill, Inc. Reprinted by permission.)

adaptive compliance and accommodation; voluntary contraction of the puborectalis and EAS muscles at appropriate times; and motivation to make the appropriate responses and to remain continent.

#### PATHOPHYSIOLOGY OF FECAL INCONTINENCE

Most patients with fecal incontinence have demonstrable abnormalities of one or more of these important continence mechanisms (Table 1). In some clinical situations, such as surgical disruption of the puborectalis muscle or severe injury to the anal sphincter muscles, the reasons for incontinence are straightforward and easy to understand. In other conditions, the causes of incontinence are often multiple and not as easily understood.

#### Reduced rectal compliance

In patients with reduced rectal compliance, a given volume of fecal material produces higher intrarectal pressures than in patients with normal compliance; by exceeding anal sphincter pressures at earlier stages of rectal filling, rectal storage time is significantly shortened and rectal urgency occurs. Such abnormalities have been described in patients with inflammatory bowel disease,<sup>3</sup> patients with chronic rectal ischemia,<sup>4</sup>

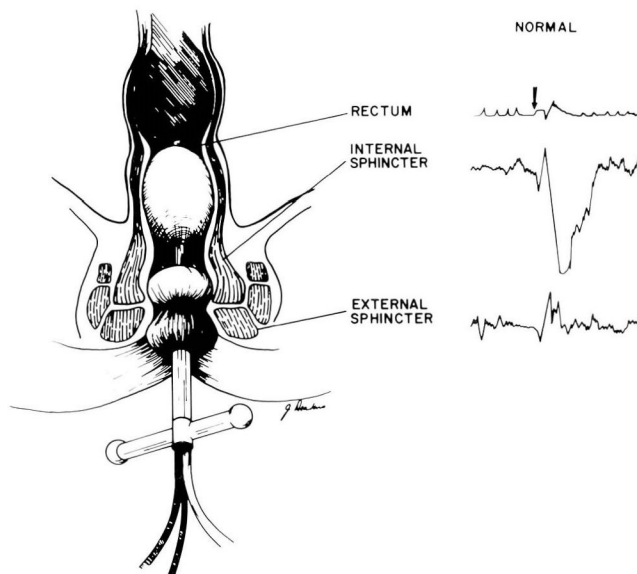


FIGURE 2. Anorectal manometer composed of three air-filled balloons is used to elicit anal sphincteric responses. Transient distension of the proximal balloon (arrow) produces reflex relaxation of the internal anal sphincter and transient contraction of the external anal sphincter. In Hirschsprung's disease, internal anal sphincter relaxation is absent. (Wald A. Abnormalities of anorectal function. [In] Cohen S, Soloway RD, eds. *Functional Disorders of the Gastrointestinal Tract*. New York, Churchill Livingstone, 1987, pp 121–138. Reprinted by permission.)

patients who have undergone pelvic irradiation,<sup>5</sup> and in association with aging.<sup>6</sup>

#### Impaired rectal sensation

Impaired rectal sensation may adversely affect continence since it removes the warning signal that defecation is impending. Rectal sensation can be impaired because of megarectum associated with fecal impaction, a condition found commonly in children with encopresis<sup>7</sup> and in physically or mentally impaired elderly individuals.<sup>8</sup> Moreover, sensory impairment may persist after disimpaction and bowel cleansing.<sup>9</sup> Another group of patients with sensory impairment are those with neurologic disease. For example, more than 50% of patients with diabetes mellitus and fecal incontinence have impaired rectal sensation but have no abnormalities of rectal compliance or change in the threshold for IAS relaxation.<sup>10</sup> In contrast, sensation is normal in continent diabetic patients, suggesting that sensory impairment is

**TABLE 1**  
ABNORMALITIES OF CONTINENCE MECHANISMS IN FECAL INCONTINENCE

| Categories of patients                         | Anorectal continence mechanisms |      |            |           |            |
|------------------------------------------------|---------------------------------|------|------------|-----------|------------|
|                                                | Reservoir                       | PRM* | Sphincters | Sensation | Motivation |
| Adults                                         |                                 |      |            |           |            |
| Anal trauma/surgery                            | —                               | —    | +          | —         | —          |
| Idiopathic (adult)                             | —                               | +    | +          | —         | —          |
| Diabetes mellitus                              | —                               | —    | +          | +         | —          |
| Proctitis                                      | +                               | —    | —          | —         | —          |
| Ileoanal anastomosis                           | +                               | —    | —          | —         | —          |
| Neurogenic (spinal cord)                       | —                               | +    | +          | +         | +          |
| Neurogenic (frontal cortex)<br>or subcortical) | —                               | —    | —          | —         | +          |
| Associated with constipation                   | —                               | —    | —          | +         | Variable   |
| Children                                       |                                 |      |            |           |            |
| Encopresis                                     | —                               | —    | —          | Some      | Variable   |
| Meningomyelocele                               | —                               | +    | +          | +         | Variable   |
| Imperforate anus repair                        | +                               | +    | +          | —         | Variable   |

+ = Abnormal; — = Normal

\*Puborectalis muscle

a key factor in fecal incontinence in that population. Rectal sensation may also be impaired in many patients with meningomyelocele<sup>11</sup> and in multiple sclerosis (unpublished observations).

### Other abnormalities of continence mechanisms

However, impaired rectal sensation often occurs with abnormalities of other continence mechanisms in patients with fecal soiling. For example, incontinent diabetic patients frequently have lower resting anal sphincter pressures<sup>12</sup> and impairment of EAS function as well.<sup>10</sup> These abnormalities may become clinically important if diarrhea develops; indeed, most diabetic patients with incontinence do have diarrheal stools, which are more difficult to control than solid stool.<sup>12</sup> Similarly, anal sphincter abnormalities are also present in patients with fecal incontinence associated with meningomyelocele and multiple sclerosis.

Abnormalities of neuromuscular pelvic floor function have recently been described in patients with "idiopathic" fecal incontinence, a disorder that occurs predominantly in middle-aged and elderly women who have no history of anorectal disease or trauma. Both histopathologic and electromyographic studies indicate evidence of denervation of both the puborectalis and EAS muscles.<sup>13</sup> This neuropathy has been postulated to occur because of repetitive and excessive pelvic floor descent during defecation over many years. This stretches the pudendal nerves supplying these muscles; resultant nerve damage weakens the muscles and, if persistent and progressive, leads to incontinence. A similar process

**TABLE 2**  
EVALUATION OF FECAL INCONTINENCE

|                                                                                                                                                                                                            |
|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| History                                                                                                                                                                                                    |
| Frequency, duration, severity (gas, liquid, or solid; rest or with exertion)                                                                                                                               |
| Pattern (diurnal, nocturnal)                                                                                                                                                                               |
| Associated symptoms (urgency, lack of warning, diarrhea, constipation, straining at defecation)                                                                                                            |
| Other relevant factors (anorectal trauma or surgery, diabetes, laminectomy, urinary incontinence, immobility, dementia, neurologic disease, multiple childbirths, inflammatory bowel disease, medications) |
| Physical examination                                                                                                                                                                                       |
| Rectal examination (prolapse, fecal impaction or rectal mass, anal deformity or disease, anal "gaping," atrophy of gluteal muscles)                                                                        |
| Neurologic examination (mental status, sacral reflexes, perineal sensation, basic neurologic evaluation)                                                                                                   |
| Initial investigation                                                                                                                                                                                      |
| Sigmoidoscopy (proctitis, tumor, melanosis coli)                                                                                                                                                           |
| Incontinence calendar                                                                                                                                                                                      |
| Presence of diarrhea (stool for culture; ova and parasites; 72-hr stool collection for weight, fat, reducing substances; barium enema examination; small bowel series; medication and diet histories)      |

may impair continence mechanisms during childbirth and lead to sphincter dysfunction in multiparous women.

### EVALUATION OF PATIENTS WITH FECAL INCONTINENCE

Evaluation includes definition of the problem, delineation of possible contributing factors, careful examination of the anorectal area, and focused neurologic testing (Table 2). Fecal impaction, rectal prolapse, or evidence of anorectal deformity may have clinical significance. Anal "gaping" on withdrawal of the examining

TABLE 3  
SPECIALIZED STUDIES FOR FECAL INCONTINENCE

| Test                | Information obtained                                                                                                              |
|---------------------|-----------------------------------------------------------------------------------------------------------------------------------|
| Anorectal manometry | Rectal sensation<br>Resting and augmented anal canal pressures<br>Internal sphincter relaxation<br>Rectoanal contractile response |
| Rectometrogram      | Rectal compliance/accommodation                                                                                                   |
| Proctogram          | Puborectalis function<br>Assessment of perineal descent                                                                           |

finger suggests sphincter denervation but, in general, digital estimation of sphincter tone and strength often correlate poorly with objective tests. Psychosocial assessment is particularly important in both pediatric and elderly patients. The evaluation can be supplemented by specialized studies performed by physicians with experience in the interpretation of such tests (Table 3).

#### Anal sphincter evaluation

A perfused catheter is used to measure resting anal canal pressures (mainly derived from the IAS) and augmented pressures obtained by asking the patient to tighten the anal canal voluntarily (derived from the EAS). A station pull-through method, in which the catheter is withdrawn in 0.5-cm intervals, provides a more accurate estimate of resting pressures than does continuous withdrawal. Relaxation of the IAS in response to rectal distension and rectal sensation can also be assessed with this technique.

#### Rectal compliance and sensation

Several parameters can be measured by progressively distending a latex balloon approximately 10 cm in length with increased amounts of air. By recording pressures at each volume of distension and constructing pressure-volume curves, a measure of rectal wall viscoelasticity and rectal size can be obtained. The maximum volume tolerated by the patient appears to correlate with defecation frequency and severity of incontinence.<sup>14</sup> Decreased compliance is associated with decreased reservoir capacity whereas increased compliance is observed in megarectum (Figure 3).

#### Pelvic floor evaluation

This can be studied with lateral radiographs of the anorectum, a technique known as proctography. One can measure the anorectal angle created by the puborectalis muscle and look for degrees of perineal floor

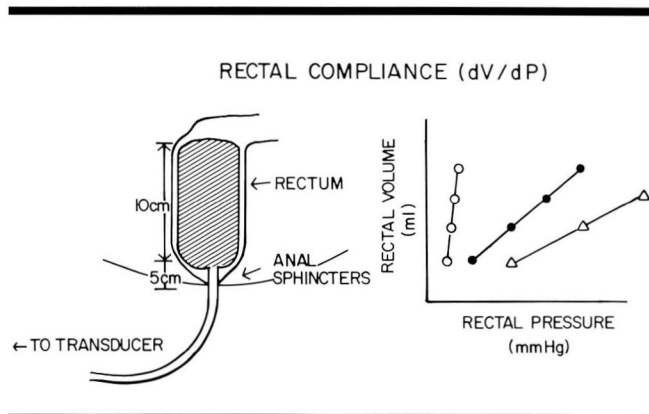


FIGURE 3. Pressure-volume curves obtained by measuring intrarectal pressures following progressive distension of a latex balloon. Normal compliance study (●) is compared to those of a patient with megarectum (○) and a patient with reduced compliance due to inflammatory bowel disease (Δ).

descent (Figure 4). It is uncertain whether this technique provides more information than does a careful digital examination and evaluation for descent and/or rectal prolapse while the patient sits on a commode chair and strains as if defecating. Nevertheless, it has been used to quantify these parameters to eliminate a certain degree of subjectivity.

#### TREATMENT

Many patients with fecal incontinence can be treated effectively with techniques that are thought to modify abnormal continence mechanisms. Therapeutic approaches consist of biofeedback or operant conditioning techniques, pharmacologic agents, and surgical procedures. Except for incontinence associated with full-thickness rectal prolapse, surgery should not be employed as initial therapy but only after nonsurgical approaches have failed.

#### Biofeedback

Biofeedback training based upon the techniques of Engel et al<sup>15</sup> is a simple and often effective treatment for incontinence associated with rectosphincteric abnormalities. The method uses a three-balloon manometry device; the recording apparatus provides information (feedback) about anorectal function so that the patient can tell whether sphincteric responses are being performed appropriately following rectal distension. During the training session, the rectal balloon is inflated with volumes of air, which the patient is taught to recognize

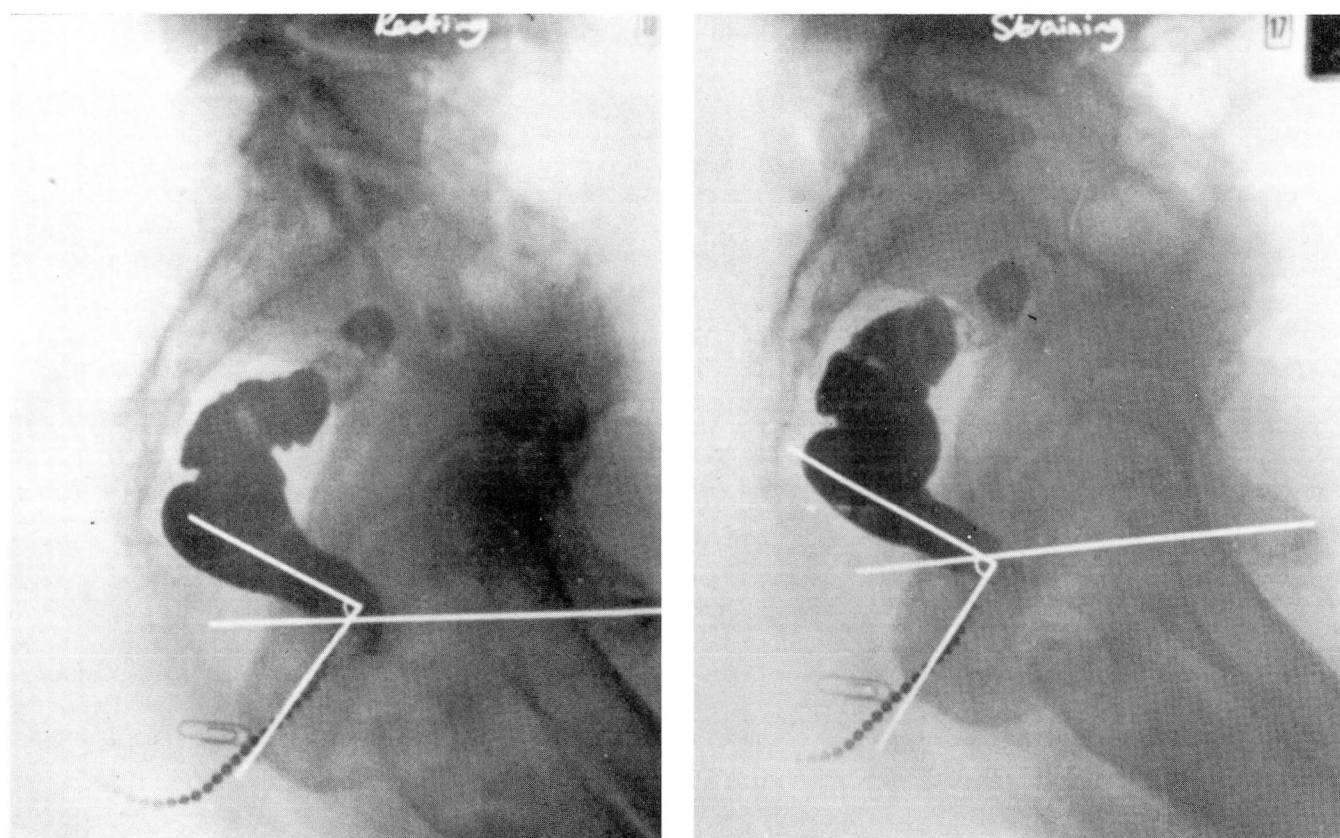


FIGURE 4. Lateral radiographs of the anorectum (proctography) in a normal subject at rest (left) and while straining (right). The anorectal angle is defined by a line drawn along the rectum and a beaded chain in the anal canal. The pelvic floor is defined by a line drawn from the symphysis pubis and the tip of the coccyx. Courtesy of Dr. N. W. Read. (Wald A. Abnormalities of anorectal function. [In] Cohen S, Soloway RD, eds. Functional Disorders of the Gastrointestinal Tract. New York, Churchill Livingston, 1987, pp 121-138. Reprinted by permission.)

as rectal distension (sensory discrimination). The patient is then instructed to squeeze the EAS whenever rectal distension is sensed; this is recorded as an upward deflection on the recorder, which the patient can immediately visualize and modify as necessary. Thus, biofeedback is a trial-and-error learning process using a visual display to monitor anal sphincter responses. Once patients master the technique, they are weaned from the instrument by blocking their view of the recording. During this latter phase, the instructor monitors performance and provides verbal feedback to guide and encourage appropriate responses. Following biofeedback, patients are instructed to practice contracting the EAS daily and to use the technique whenever they sense impending defecation (exercise training).

Prerequisites for successful biofeedback include motivation, the ability to comprehend and follow directions,

some degree of rectal sensation, and the ability to generate a squeeze pressure. Most adult patients respond after one session and reinforcement is generally unnecessary. Success has been achieved in up to 70% of patients who fulfill the above criteria, including those with incontinence due to sphincter surgery, disease, or nondeforming trauma, in most patients with "idiopathic" incontinence, and in those with diabetes mellitus.<sup>10,16,17</sup>

It should be emphasized that since neither controlled outcome nor component analysis studies have been reported, the mechanisms by which biofeedback work remain unclear. In a recent attempt to analyze the major components of biofeedback, only one of seven successfully treated patients required the procedure as originally described.<sup>18</sup> Three responded to sensory discrimination training alone, one to exercise training alone, one im-

proved without treatment, and another required contingency management (in which patients are instructed to defecate at specified intervals, and will receive cleansing enemas or suppositories after a specified interval).

Other biofeedback techniques reported to be successful have used an inexpensive pneumatic device<sup>19</sup> and an intra-anal plug and electromyometer.<sup>20</sup> These techniques do not attempt to synchronize sphincter contraction to rectal distension.

In contrast to earlier reports, other investigations have found no differences in EAS responses following successful and unsuccessful biofeedback sessions.<sup>17</sup> Notwithstanding our lack of understanding of how biofeedback works, the ease, safety, rapidity, and apparent efficacy of the procedure make it the initial choice for many patients with fecal incontinence.

### Pharmacologic agents

When fecal incontinence is associated with chronic diarrhea, symptomatic improvement often can be obtained with opiate derivatives. It is not clear whether improvement occurs because of decreased stool volume and frequency, increased fecal consistency, or because of effects on anorectal function. The few studies that have addressed this issue suggest that opiates vary in their pharmacologic actions.

In one study of patients with chronic diarrhea, both codeine and loperamide were more effective than diphenoxylate with atropine (Lomotil) in reducing fecal soiling when the drugs were given in doses that reduced stool frequency to an equivalent degree.<sup>21</sup> Another study found that, in addition to increasing stool consistency and decreasing stool volume, loperamide improved continence of rectally infused saline by approximately 50% and also increased anal sphincter pressures.<sup>22</sup> Despite some shortcomings to this study, loperamide appears to be the drug of choice when treating a patient with chronic diarrhea and fecal incontinence.

### Surgery

When incontinence is associated with gross rectal prolapse, surgical resuspension should be performed first since continence can be restored in approximately 50% of such patients. Except for this circumstance, surgical procedures should be considered only after nonsurgical interventions have proven unsuccessful.

In general, fecal incontinence due to anal sphincter damage should be treated with biofeedback techniques initially but if results are unsatisfactory, surgical repair produces good results. Other surgical approaches include repair of the puborectalis muscle (the so-called postanal

repair) and repair of the pubococcygeus muscle. The large number of procedures advocated for fecal incontinence suggests that no single one is best for all patients—nor are any easy to perform or free of complications. Very few long-term or randomized controlled studies have compared various approaches. Ideally, surgical intervention should be tailored to correct abnormal continence mechanisms as determined by objective tests of anorectal function.

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### PHYSIOLOGY OF DEFECATION

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As with continence, defecation is partly controlled by the central nervous system. During voluntary defecation, contraction of abdominal muscles and closure of the glottis increase intraabdominal pressure, colonic segmenting activity is temporarily inhibited, and fecal material is propelled toward the rectum. Simultaneously, the pelvic floor muscles relax, which leads to descent of the pelvic floor and straightening of the anorectal angle. As pressures build in the rectum, the anal sphincters relax, allowing stool expulsion. After completion of defecation, the pelvic floor ascends and anorectal angulation and anal sphincter tone are restored.

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### CONSTIPATION IN ADULTS

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Since constipation is a symptom rather than a disease, it is difficult to define precisely. Patients may complain of constipation if they defecate too infrequently, if they strain at defecation, if their stools are too hard or too small, or if there is a sense of incomplete rectal evacuation. The approach to constipated patients includes precisely defining the complaint and, in selected circumstances, employing diagnostic studies to provide objective data concerning colorectal function.

Constipation may complicate a number of medical and surgical conditions and is a side effect of many drugs (Table 4). In some patients, dietary deficiency of fluid and/or fiber may result in constipation, or symptoms may improve with fiber supplements even though fiber deficiency cannot be documented. Many patients have previously supplemented their diet with fiber, either on their own accord or in response to previous medical advice. Notwithstanding these considerations, most patients with "idiopathic" constipation probably have one or a combination of the following: an underlying disturbance of colonic motility or absorption; abnormalities of anorectal function or defecation; misperception of bowel habit or an underlying behavioral disturbance.

## SEVERE IDIOPATHIC CONSTIPATION

Many patients with severe idiopathic constipation are young to middle-aged adults, predominantly female, who complain of longstanding constipation and often experience abdominal discomfort associated with the buildup of fecal material. They are often referred after extensive studies fail to disclose structural abnormalities and after fiber supplements and laxatives do not improve their symptoms. Evaluation of colorectal function and behavioral patterns suggests that this group of patients is quite heterogeneous.

## Normal-transit constipation

By measuring transit of markers through the colon, a number of investigators have divided constipated patients into those with slow colonic transit and those with normal transit.<sup>23,24</sup> In our experience, approximately one third of patients with constipation that is refractory to medical management fall within the latter category.<sup>25</sup> Others have argued that criteria are imprecise, that transit of markers does not allow delineation of pathophysiology or the underlying pattern of colonic motility, and that such studies may be influenced by many factors.<sup>26</sup> My own view is that a normal colonic transit study refutes a stated history of infrequent defecation and narrows the differential diagnosis to a defecatory disorder or to patient misperception of bowel function. Recent studies indicate that such patients exhibit higher levels of global distress, somatization, anxiety, depression, and interpersonal sensitivity than do patients with slow-transit constipation.<sup>27</sup>

## Slow-transit constipation

By calculating the transit of markers through the different regions of the colon, Martelli et al<sup>28</sup> and Watier et al<sup>29</sup> divided patients with slow-transit constipation into those with "colonic inertia," in whom markers were delayed through the proximal colon, and those with "outlet dysfunction," in whom markers passed normally through the proximal colon but emptying from the rectum was delayed.<sup>28,29</sup> Most of the severely constipated adults that we have evaluated have exhibited the "inertia" pattern. This pattern suggests a diffuse abnormality of colonic motor function, a suggestion that has been reinforced by the finding of a number of subtle histopathologic abnormalities in the enteric nervous plexus of resected colons from such patients.<sup>30</sup> Abnormalities are not always confined to the colon; bladder dysfunction<sup>29</sup> and disturbances of esophageal and gastrointestinal motility<sup>26</sup> have been reported, suggesting that a diffuse

**TABLE 4**  
CONDITIONS ASSOCIATED WITH CONSTIPATION (PARTIAL LIST)

|                                   |
|-----------------------------------|
| Metabolic and endocrine disorders |
| Neurologic diseases               |
| Intestinal pseudo-obstruction     |
| Multiple sclerosis                |
| Parkinson's disease               |
| Paraplegia                        |
| Meningomyelocele                  |
| Spinal cord disease               |
| Autonomic neuropathy              |
| Cauda equina tumor                |
| Colonic disorders                 |
| Tumors                            |
| Intussusception                   |
| Inflammatory strictures           |
| Systemic sclerosis                |
| Diverticular disease              |
| Irritable bowel syndrome          |
| Anorectal disorders               |
| Rectocele                         |
| Anal stenosis                     |
| Anterior mucosal prolapse         |
| Anal fissure                      |
| Perianal abscess                  |
| Drugs                             |
| Opiates                           |
| Anticholinergics                  |
| Anticonvulsants                   |
| Antidepressants                   |
| Antacids (calcium, aluminum)      |
| Antiparkinson agents              |
| Diuretics                         |
| Iron                              |
| Antihypertensives                 |
| Others                            |
| Inactivity                        |
| Withholding                       |
| Situational inhibition            |
| Chronic pain                      |
| Depression                        |
| Dementia                          |
| Idiopathic                        |

disorder of smooth-muscle function exists in some of these patients.

In addition to a disturbance of colonic propulsion, some patients also have abnormalities of anorectal function, including those of sensory perception and expulsion dynamics. Several investigators noted that many patients were unable to expel a simulated stool, and did not relax the puborectalis and EAS muscles during expulsion attempts.<sup>31,32</sup> Others have reported a diminished desire to defecate<sup>32</sup> or an increased threshold of conscious rectal sensation in the absence of megarectum or altered rectal elasticity.<sup>33</sup> Patients with anorectal abnormalities may not respond to measures directed solely to the colon, emphasizing the importance of carefully evaluating anorectal function even in patients with colonic inertia (see below).

## DISORDERS OF DEFECATION

**Functional disorders**

In general, the pattern of outlet dysfunction, which is characterized by delayed transit through the rectum, usually either indicates megarectum<sup>26</sup> or suggests that abnormal defecation exists in the presence of normal colonic propulsion. In addition, patients who complain of excessive straining at defecation may have normal transit but abnormalities of defecatory function.

The pattern of outlet obstruction occurs in all age groups but is the most common pattern seen in children with idiopathic constipation.<sup>34</sup> Although constipation with soiling in children has been thought to be a psychogenic disorder, more recent theories have implicated both psychologic and physiologic factors.<sup>35</sup> For example, many children with overflow incontinence have demonstrable abnormalities of anorectal function. Due to chronic fecal retention, the rectum may dilate, rectal sensation becomes impaired, and the rectum may be unable to contract sufficiently to evacuate stool effectively. Discomfort during defecation may also lead to avoidance behavior or to an unconscious inhibition of defecation. This concept is supported by recent studies that demonstrate an abnormal defecation pattern in 40%–60% of encopretic children.<sup>36</sup> This pattern is characterized by contraction of the EAS and puborectalis muscles during simulated defecation and may represent a learned behavior since it can often be normalized using biofeedback techniques.<sup>37</sup>

Many elderly patients with constipation experience impaction because of a combination of immobility, weakness, dementia, and/or confusion, which causes them to ignore or not react to the urge to defecate.<sup>8</sup> Similar to encopretic children, megarectum is often present and may be associated with depressed rectal sensation, but abnormal defecation patterns have not been reported. The persistence of sensory impairment after bowel cleansing may predispose these individuals to recurrent fecal impactions.<sup>8</sup>

Finally, some constipated adult patients exhibit abnormal expulsion dynamics characterized by failure of the puborectalis muscle and EAS to relax during defecation attempts.<sup>31</sup> Some of these patients have difficulty in evacuating simulated stools, barium, or even saline.<sup>32</sup> Similar to children, adults with this pattern may respond to biofeedback techniques that attempt to normalize such patterns.<sup>38</sup>

**Hirschsprung's disease**

Characteristically, this disorder is associated with ob-

stipation from birth and a dilated colon proximal to a contracted, nonpropulsive segment of bowel. These findings are caused by a developmental arrest of neural crest cells that results in absence of intramural ganglion cells of both the submucosal and myenteric plexuses. The IAS is invariably involved and may be the only functional abnormality in the short-segment form of the disease.

In contrast to normal subjects (*Figure 2*), the IAS does not relax following rectal distension,<sup>39</sup> and, indeed, often contracts. Although most cases of Hirschsprung's disease are diagnosed by six months of age, a small percentage of patients have only a short aganglionic segment and may respond to medical management for constipation. Occasionally, the diagnosis may be delayed until adulthood. Characteristically, in the short-segment variants, the barium enema examination is normal and a narrow segment cannot be demonstrated. However, if manometry is performed, no IAS relaxation occurs following rectal distension. Therefore, the demonstration of IAS relaxation allows the clinician to exclude the disease with confidence.

**Anatomic abnormalities associated with constipation**

**Rectocele.** In some constipated women, radiologic assessment of rectal evacuation of barium thickened to the consistency of stool (defecography) may demonstrate redirection of expulsion forces into a weakened anterior rectal wall.<sup>40</sup> Some of these women can promote defecation by pressing on the posterior vaginal wall with their fingers. However, care must be taken when implicating a rectocele as the cause of impaired stool evacuation since rectoceles may frequently be found in subjects without constipation or defecatory difficulties; accordingly, surgical repair of the rectocele may fail to improve defecation.<sup>26</sup>

**Rectal intussusception.** Patients with rectal intussusception often complain of inability to defecate despite a great deal of straining. Defecography demonstrates the rectal wall folding into the lumen, entering the anal ring and appearing to obstruct the passage of barium during attempts to defecate.<sup>40</sup> A similar clinical picture can be seen with anterior mucosal prolapse into the anal canal. In some patients, rectal mucosa protrudes through the anus during defecatory straining and sigmoidoscopy often reveals a reddened patch of redundant anterior rectal mucosa that enters the anal canal during withdrawal of the instrument. This condition occurs mainly in women who exhibit perineal descent, low anal sphincter pressures, and who may also be incontinent.<sup>41</sup> Similar to rectoceles, intussusception is not uncommon

**TABLE 5**  
SPECIALIZED STUDIES FOR CONSTIPATION  
(IF TREATMENT FAILS)

| Test                            | Information obtained                                                                                      |
|---------------------------------|-----------------------------------------------------------------------------------------------------------|
| Colonic transit study (Markers) | Total and segmental colonic transit times                                                                 |
| Anorectal manometry             | Rectal sensation<br>IAS relaxation<br>Rectal compliance<br>Expulsion pattern                              |
| Defecography                    | Completeness of rectal expulsion<br>Anatomic abnormalities (rectocele, intussusception)<br>PRM relaxation |

in nonconstipated subjects and surgical repair frequently results in little or no clinical improvement.

#### EVALUATION OF PATIENTS WITH CONSTIPATION

As with incontinence, the evaluation should include a careful delineation of the complaint of constipation. The history should attempt to identify contributing factors such as dietary intake, medications, and other conditions known to be associated with constipation (*Table 4*). Physical examination includes a careful rectal and vaginal examination when the patient is relaxed but also during straining to look for prolapse, rectocele, excessive perineal descent, and failure to relax the puborectalis and EAS. Sigmoidoscopy, barium enema examination, and routine laboratory studies such as calcium and thyroid studies are often performed; the usefulness of the barium enema examination in chronic constipation is uncertain<sup>42</sup> and it should perhaps be reserved for constipation of short duration or a recent change in bowel habits. For constipation that is refractory to initial therapy (see below), specialized studies may be employed to define the pattern of altered colorectal function (*Table 5*).

#### TREATMENT

##### Pharmacologic agents

The unpalatable nature of bran and commercial fiber supplements and the ready availability of laxatives often results in the use and abuse of the latter. Laxatives are now classified into five groups on the basis of their mode of action—stimulant, saline, emollient, hyperosmolar, and bulk-forming. It is commonly believed that chronic use of irritant laxatives may damage the myenteric plexus of the colon<sup>43</sup> and may exacerbate constipation,

**TABLE 6**  
THERAPEUTIC APPROACHES FOR CONSTIPATION

| Categories                 | Treatment                                                              |
|----------------------------|------------------------------------------------------------------------|
| Factitious                 | Re-education, counselling                                              |
| Anatomic abnormalities     | Consider surgical correction                                           |
| Abnormal expulsion pattern | Biofeedback*                                                           |
| Colonic inertia            | Bethanechol<br>Opiate antagonists*<br>Cisapride*<br>Subtotal colectomy |
| Idiopathic childhood       | Bowel cleansing and habit training<br>Biofeedback*                     |
| Hirschsprung's disease     | Rectal myectomy                                                        |

\*Experimental

but this is arguable. An excellent review on the use and abuse of laxatives is available for those seeking more information on this subject.<sup>44</sup> When fiber and laxatives produce a suboptimal response, other approaches should be considered according to the underlying pathophysiology (*Table 6*).

In an effort to promote propulsive activity in the colon of patients with severe constipation, various drugs have been prescribed that enhance colonic contractile motor activity. Cholinergic agents such as bethanechol chloride and neostigmine have achieved variable results but are unsuccessful in most patients with colonic inertia,<sup>25</sup> perhaps because increased contractility in the colon is not necessarily associated with propulsive activity. In addition, the high incidence of uncomfortable side effects from these agents deters compliance.

The prokinetic agents—metoclopramide, domperidone, and cisapride—are another group of pharmacologic agents under investigation. Cisapride, the newest of these agents, stimulates colonic smooth muscle in vivo and enhances transit through the proximal colon.<sup>45</sup> It has achieved mixed results in treating constipation in placebo-controlled studies and is available only under experimental protocol.

Endogenous opioid compounds are among the most abundant neurotransmitters in the gastrointestinal tract and are postulated to inhibit colonic transit. Intravenous infusion of naloxone, a known opioid antagonist, has dramatically improved severe constipation in a few patients<sup>46</sup> and oral opioid antagonists are currently being developed.

##### Surgery

Surgery has been used to treat refractory and disabling constipation but extreme caution must be exercised since our understanding of colorectal motility is rela-

tively rudimentary. Indications for surgery are infrequent.

**Hirschsprung's disease.** In classic Hirschsprung's disease, the aganglionic segment usually involves only the rectum but may extend proximally. Surgical approaches involve resecting the aganglionic segment or bypassing it; of the two approaches, bypass is more easily performed and has less morbidity and mortality.<sup>47</sup> In the short-segment and ultra-short-segment forms of the disease, agangliosis is limited to the anorectal junction or anal canal. A more limited surgical procedure that consists of an internal sphincterotomy and rectal myotomy of varying length can often be successful.<sup>24</sup>

**Defecation disorders.** Caution must be exercised when contemplating surgery in patients with these disorders. Rectoceles may be repaired by a posterior colporrhaphy. However, it is essential to determine if defecation is improved by exerting pressure on the rectocele during defecography; if so, surgery is more likely to be successful. It is unclear whether rectal intussusception, rectal prolapse, or anterior mucosal prolapse actually cause defecatory dysfunction since rectopexy and similar resuspension procedures may not improve symptoms.<sup>48</sup> Experience with surgical division of the puborectalis muscle in patients who cannot relax this muscle during defecation has also been disappointing since the majority of patients report no benefit and some develop incontinence of flatus, liquid stool, and mucus.<sup>49</sup> Finally, anorectal myectomy was found to be superior to anal dilatation in a randomized controlled trial of patients with outlet obstruction.<sup>50</sup> However, only five of 11 patients

without short-segment Hirschsprung's disease were clinically improved. Better criteria for predicting successful outcome must be developed before such surgery can be recommended with confidence.

**Colonic disorders.** Slow-transit constipation with the pattern of colonic inertia undoubtedly is a heterogeneous disorder whose common feature is delayed transit through the large intestine. In carefully selected patients whose condition is refractory to medical therapy and who have disabling symptoms, subtotal colectomy with ileorectal anastomosis can be strikingly successful in ameliorating symptoms.<sup>51</sup> Most centers advocate studies of esophageal, gastric, and small intestinal motor function to ensure that no abnormalities exist. However, even if preoperative assessment of the upper gastrointestinal tract is normal, pseudoobstruction may occur after surgery.<sup>52</sup> Anorectal studies should also be performed since abnormal defecation patterns may be associated with persistence of constipation after surgery. However, abnormal anorectal studies do not preclude a successful outcome.<sup>53</sup> Most investigators believe that limited resection of the colon is not beneficial in the absence of a mechanical obstruction.<sup>51</sup> Similarly, in patients with evidence of chronic intestinal pseudo-obstruction, surgery should be avoided if small bowel involvement is demonstrated.

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