

# Management of Patients With Neurological Disease

by Dr. John Bach

## **Page 70 Gastrointestinal Hypomotility**

Complaints of malaise, anxiety, epigastric discomfort, and vomiting by patients with myopathies may be due to gastrointestinal hypomotility. When abdominal distention, pain, constipation, and vomiting are severe and mechanical obstruction is excluded by abdominal radiography, contrast studies, and colonoscopy, intestinal pseudoobstruction is diagnosed. Manometric and cineradiographic findings of abnormal intestinal motility confirm-enhancing agents such as metoclopramide (Reglan). In one study, 15 of 16 patients with myotonic dystrophy had delayed gastric emptying of a solid meal, and 10 had delayed emptying of a liquid meal. Esophageal emptying was also markedly delayed in 15 of 16 patients. No relationship was found between gastrointestinal symptoms and severity of skeletal muscle weakness.

Occasionally gastroparesis and dilatation necessitate acute decompression by passing a nasogastric tube. A gastrostomy tube, when present, can be burped. Occasionally, when the distention is in the large intestine and megacolon is severe, persistent and unrelieved by mobilization or passing a rectal tube, colostomy becomes necessary. Although one patient described in the literature did not fare well postoperatively and a perioperative complication rate of 8.2-42.9% was noted for patients with myotonic dystrophy, we have had patients who survived for years after colectomy with ileorectal stapled anastomosis. This finding applies particularly to patients requiring mechanical ventilation. One patient with myotonic dystrophy and gastric distention developed gastric volvulus with pyloroantral obstruction that was successfully treated by emergency gastrectomy.

Symptoms of gastrointestinal hypomotility are often associated with congestive heart failure and left ventricular ejection fractions (LVEFs) below 15%. Acute episodes of gastric and intestinal distention and tenderness, vomiting, diarrhea, and tachycardia can result in dehydration, impaired diaphragm excursion, and hypoventilation, and, if not caused by it, can lead to cardiopulmonary failure. Hypokalemia and hypoglycaemia, which commonly occur during respiratory tract infections, can result in or exacerbate abdominal distention. It has been noted that such episodes can follow anesthesia, complicate chest colds, and be associated with emotional factors and fatigue. Intestinal obstruction, pseudoobstruction, malabsorption, and volvulus can also occur in patients with NMD and can cause or contribute to ventilatory insufficiency. Nausea, retching, vomiting, and tachycardia also may be associated with uncomplicated ventilatory insufficiency.

Mechanical ventilation, especially with positive end-expiratory pressure (PEEP), increases intrathoracic pressure and decreases venous return by reducing the systemic-venous pressure difference. This effect decreases cardiac preload and right atrial pressure and results in decreased cardiac output, particularly for patients with cardiomyopathies who receive beta-blocker and angiotensin-converting enzyme inhibitor medications. Splanchnic blood flow decreases in parallel with PEEP-induced reductions in cardiac output. This decrease can lead to gastrointestinal mucosal damage and altered gastrointestinal motility.

## **Constipation**

Constipation is common in NMD and can cause abdominal distention, which, in turn, can cause ventilatory failure. One must be aware, however, that nausea, vomiting and abdominal pain and distention can be a sign of anatomic abdominal obstruction or cardiac failure and, thus, should be evaluated before assuming that symptoms are due to simple constipation.

For treating constipation, high fluid and fruit intake is encouraged, but laxatives are often required. Bulk-forming laxatives are derived from agar, psyllium seed, kelp, plant gums, and cellulose. They facilitate passage of intestinal contents and reflexively stimulate bowel wall activity. They exert a laxative effect in 12-24 hours. The components of Metamucil (G.D. Searle & Co., Chicago IL), a bulk-forming vegetable laxative, are psyllium and dextrose. Metamucil is usually provided in dosages of 30 mg (two table spoons) per day. Its use can result in highly fibrous and bulky stools that may be increasingly difficult to evacuate as bowel weakness progresses. It also necessitates additional fluid intake, which can be difficult for people with dysphagia.

Salt laxatives include Fleet Phospho-Soda (Fleet, Inc., Lynchburg, VA), magnesium citrate, and Milk of Magnesia (Roxane Laboratories, Inc., Columbus, OH). These laxatives cause water to be retained in the intestines. Up to 20% of ingested magnesium can enter the blood stream and may cause confusion and coma in people with poor kidney function. Likewise, excessive sodium intake can be hazardous for people with edema, high blood pressure, or heart disease. Dehydration can also be problem with the use of saline laxatives.

Osmotic agents, such as saline laxatives, also cause water to be retained with the stool. Polyethylene glycol (MiraLax, Braintree Laboratories, Braintree, MA) is a synthetic glycol with a high molecular weight. It increases the water retention of stools and thereby softens them and increases the frequency of bowel movements. It has no effect on the absorption or secretion of glucose or electrolytes, and no tachyphylaxis is associated with its use. However, prolonged use requires monitoring of serum electrolytes, and some patients are allergic to polyethylene glycol. It can be used either daily at doses of about 17 gm (1 tablespoon of powder in 8 ounces of water) to effect daily bowel movements or every second to fourth day in larger doses (about 65 gm) for patients wanting to avoid constipation but not wanting daily bowel movements. Some of our patients with muscular dystrophy and severe constipation have done quite well with prolonged use of this medication.

Surface active agents increase the wetting efficiency of intestinal water and tend to soften stools. The most commonly used medication in this class is bioctyl sodium sulfosuccinate gels (Colase, Bristol-Myers Squibb Co., Princeton, NJ). Two gels (200 mg), taken once a day, can make stool more slippery and facilitate bowel evacuation. Colase can help prevent constipation but does not appear to have any effect on existing constipation. There are no side effects.

Mineral oils soften and lubricate stools. Examples include liquid petroleum and plant oils such as olive oil. Large doses should not be taken with food because they can retard stomach emptying. Since large doses can cause oil to leak out of the anus, doses should be divided or taken only before bedtime. Long-term mineral oil use can also lead to oil deposits in the lungs (and, thereby, lipid pneumonia) or lymph nodes, liver, and spleen, where they can cause chronic inflammation.

Stimulant laxatives can stimulate both small and large intestines. They increase the activity of the intestinal wall muscles by irritating the mucus lining or by stimulating local nerve reflexes to increase wall muscle activity. This effect may cause griping, increased mucus secretion, evacuation of fluid produced in the bowel, and loss of potassium. Anthraquinone laxatives include rhubarb root powder and aloe vera, substances that act to irritate the bowel wall, as well as cascara and senna. These substances act only on the large intestine (colon). The way in which they increase bowel activity is not entirely clear. Senokot (Purdue Frederick Co., Norwalk, CT) is one of the most common preparations of senna. It can be provided as granules, in tablet form, as a syrup, or as a suppository. Rhubarb and senna cause excretion of chrysophanic acid in the urine. Depending on the acid content of the urine, this substance colors the urine either yellowish-brown or reddish-violet.

Herbal Laxatives include Herb-lax (Shaklee Corporation, San Francisco, CA), a senna leaf powder preparation with numerous other organic laxative substances, including rhubarb root powder. Another example is Aloe Vera Herbal Stimulant Laxative (Nature's Way Products, Inc., Springville, UT). One Herb-lax tablet includes 175 mg of senna powder. Four or more tablets can be used at bedtime. Aloe Vera capsules include 200 mg of aloe vera resin, 50 mg of aloe barbadensis (aloe vera leaf), and other ingredients, including fennel and beet root. One or two tablets of these products usually produce a bowel movement in 6-12 hours.

Phenolphthalein was once the main ingredient of many preparations. It is a colorless, odourless substance that primarily stimulates the large intestine, but the small intestine also may be stimulated to some degree. Its mechanism of action is not known. Because it can cause serious systemic effects, products containing phenolphthalein have been withdrawn from the market. Bisacodyl (Dulcolax tablets or suppositories, Ciba-Geigy Corp., Summit NJ) is related to phenolphthalein but continues to be widely used. Although its action on the small intestine is negligible, a soft, formed stool is usually produced 6-8 hours after oral ingestion or often after 15-30 minutes when taken as a suppository. Griping, diarrhea, and from the use of suppositories, rectal burning can occur.

In addition to bisacodyl, glycerine, and senna concentrates, carbon dioxide-releasing compounds can also be used as laxative suppositories. These substances are primarily effective in evacuating the lower bowel. They are easier to administer and aesthetically and psychologically more acceptable than enemas and, therefore, should be used before resort to enemas.

Tap water, salt water, soap suds, vegetable oils, and milk enemas add bulk to the descending colon and rectum, thereby stimulating the intestines and initiating the defecation reflex. Soap suds and hydrogen peroxide also produce a bowel movement by irritating the bowel. Enemas, however, should not be overused. Excessive use of enemas can result in fluid and electrolyte imbalances, worsening constipation, and colonic perforation. The recipient should be horizontal. If an enema is applied to someone who is sitting, only the rectum will be cleared. The container holding the fluid should also be above the hips to allow free but not forced entry of the fluid. Properly introduced, a pint of fluid will cause evacuation if it is retained until lower abdominal cramping is felt.

Besides surface active laxatives, gastrointestinal prokinetic agents such as Reglan can be helpful (see above). There is one report of a patient with SMA whose constipation improved with administration of erythromycin.

Not having daily bowel movements can be especially desirable because, once begun, completion of the movement may take hours. The patient may have to remain in bed for hours to await completion. This problem can be alleviated by the use of a Hoyer or Easy Pivot Lift (Rand-Scot, Inc., Fort Collins, CO). Such a lift facilitates bowel evacuation by hoisting the user over a commode. The feet on the floor force the thighs into the abdomen as a substitute for abdominal muscle contraction, and with the buttocks in the most dependent position over a commode, gravity helps complete the bowel movement up to 10 times faster (according to some of our patients) than when they are lying in bed.

### **Consensus Statement for Standard of Care in Spinal Muscular Atrophy Gastrointestinal Dysfunction**

Children with spinal muscular atrophy suffer from the following gastrointestinal problems: gastroesophageal reflux, constipation, and abdominal distension and bloating. Gastroesophageal reflux is an important determinant of mortality and morbidity in patients with spinal muscular atrophy. It can be associated with silent aspiration and results in pneumonias and, at times, life-threatening events.<sup>60</sup> Frequent “spitting up” or vomiting after meals, complaints of chest or abdominal discomfort, bad breath, or obvious regurgitation of feeds may indicate gastroesophageal reflux. Some children may refuse feeds when they develop discomfort with swallowing, placing them at risk for undernutrition. High-fat foods delay gastric emptying and increase the risk of gastroesophageal reflux. Constipation is a frequently reported problem and is likely multifactorial in origin (ie, abnormal gastrointestinal motility, reduced intake of dietary fiber, inadequate fluid intake, low muscle tone of the abdominal wall). Infrequent bowel movements can lead to abdominal distention and bloating. In children dependent on their abdominal muscles to assist with respiration, desaturation or respiratory distress in association with attempted bowel movements may occur.