

History

As previously mentioned, it is important to differentiate between FI associated with stool retention and non-retentive FI as the evaluation and management between these two entities is different. Patients with non-retentive FI do not have the above-mentioned obstructive symptoms, fecal loading on physical examination, or radiographic findings. Patients may have small volume incontinence throughout the day, larger volume stool accidents post-prandially, or they may complain of worsening FI following a laxative trial. Patients may report more accidents overnight when they are not utilizing the voluntary sphincters. The perianal examination may be notable for perianal rash/excoriation, patulous or “open” sphincter, visualization of the rectal mucosa upon inspection, and asymmetrical reggae with abnormally gaping anal sphincter and poor tone and squeeze on digital rectal examination (93,94).

Histopathology

A histological evaluation is important to exclude residual aganglionosis and transition zone pull-through. If the original pathology slides from resection margins are not available, then full thickness rectal biopsy should be performed at the time of the rectal exam under anesthesia if unable to discern with certainty on history and radiographic studies between FI associated with stool retention and non-retentive FI. Patients with transition zone pull-through or retained aganglionic segment rarely if ever will present with non-retentive FI.

Anatomy

The anatomical evaluation should start with a rectal exam under anesthesia by an experienced surgeon to assess the integrity of the anorectum. The primary aim is to evaluate the anal sphincter complex comprised of the IAS and the external anal sphincter (EAS). A patulous or “open” anus, low intra-anal tone on digital examination, and asymmetrical reggae indicate compromise of the anal sphincter complex likely from excessive stretching or during the rectal dissection at time of endo-rectal pull-through, previous sphincterotomy or myectomy (11,32,93). Excessive stretching of the sphincter complex and IAS myotomy at time of the procedure have been implicated in low intra-anal pressures and squeeze pressures on ARM in children with HD who have defecation problems (95). The presence of scar tissue within the IAS and EAS on anal endosonography has been associated impaired sphincter muscle function (low intra-anal pressures and squeeze pressures on ARM) (96) and with clinical symptoms of FI in children (97). The secondary aim of the rectal exam under anesthesia is to evaluate the anal canal with specific attention to the location of the anastomosis (ie, proximity to the dentate line, anoderm, skin) and the integrity of the dentate line (11,32,93,94,98). Damage to the dentate line may disrupt sensation and potentially increase risk of FI given the poor ability of the patient to discern between gas, liquid, or solid stool (93,94,98) (Fig. 6). In their pediatric cohort, Bischoff et al (94) reported that nearly 52% of patients with HD evaluated for FI had a damaged anal canal and 21% had a patulous anus who presented with refractory FI.

Management

Postsurgical patients with HD who present with FI and are found to have compromised sphincter complex and anal canal on rectal exam under anesthesia have impaired sphincter function and sensation respectively; they have decreased potential for bowel control (11,87,93,94). Conversely, patients who have a normal sphincter complex and anal canal on rectal exam under anesthesia without evidence of mechanical/histological abnormalities and without evidence of stool retention are expected to have the

potential for bowel control. The goals of treatment are predictable bowel movements, social continence, and independence (63). Treatments range from diet, behavioral modification, and antimitility agents that modify stool consistency and transit to small volume retrograde and antegrade enemas that facilitate colonic emptying (11,63,76,77,85,87,93).

Constipating diet and soluble fiber (pectin, psyllium, gum) can be used to increase stool consistency and bulk. In a randomized placebo-controlled study, Bliss et al (99) reported that the use of fiber supplementation effectively reduced FI by half. It is important to ensure that the soluble fiber formulations do not contain sugar alcohol additives as these can cause an osmotic laxative effect. Coarse or large particle insoluble fiber can bulk the stool but should be avoided as it can make the stools looser and worsen FI (100). Patients should be encouraged to have timed toilet sitting, specifically after each meal and to try to limit the amount of snacking in between meals, with the aim of decreasing the gastrocolonic reflex and colonic contractions post-prandially to a minimum throughout the day. If there is lack of clinical improvement, referral to a Neurogastroenterology & Motility Center should be considered to assess anorectal and colonic function.

Role of a Neurogastroenterology & Motility Expert

Evaluation

Abnormal anorectal and colonic function should be suspected in patients with non-retentive FI who have a normal anal canal and preserved sphincter complex on rectal exam under anesthesia and yet continue to have FI despite optimization of medical therapy. ARM in patients with non-retentive FI helps to investigate the physiologic integrity of the anal sphincter complex, IAS, and EAS. This evaluation is important because the intra-anal pressures at rest are comprised of the IAS (70%–80%), the EAS (15%), and the hemorrhoidal cushion (5%) (101,102). Low intra-anal pressures at rest during manometry with the ability to generate normal squeeze pressures indicate compromise of the IAS with preserved EAS function. However, low intra-anal pressures in conjunction with an abnormal squeeze indicate compromise of the IAS and the EAS (Fig. 7). Low intra-anal pressures at rest have been described in postsurgical patients with HD who present with FI (65,95,103). Conversely, there are studies that have not found a correlation between intra-anal pressures and FI (96,104). Pediatric studies are limited by heterogeneous patient populations, variable methodologies, and definitions for continence. Moreover, these studies highlight that FI in postsurgical patients with HD may be multifactorial including aberrations in anorectal and colonic function (65,103,104).

Abnormal colonic function should be suspected in patients with continued FI who (a) have normal anorectal function and are not responsive to medical therapy or (b) have abnormal anorectal function (with or without compromise of the anal sphincter complex or dentate line) and are not responsive to medical therapy. In patients with FI abnormal CM findings include migration of HAPCs through the neorectum to the anal sphincter (51,105) and increased frequency of HAPCs during fasting and in the post-prandial state indicative of colonic hyperactivity (105,106) (Fig. 6C). It is important to identify these patterns on CM since management does not involve the use of laxatives and in fact the use of laxatives may worsen FI. Treatments aim to reduce the frequency and amplitude of HAPCs (anticholinergics and antidiarrheal).

In health, HAPCs originate in the proximal colon, propagate aborally, and terminate at the level of sigmoid colon without traversing the rectum. In patients with HD, resection of