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Hirschsprung disease and fecal incontinence: diagnostic and management strategies

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Abstract

Purpose: Ideally, fecal incontinence after operative management for Hirschsprung disease should not occur. If it does, it presents a formidable challenge. The purpose of this study was to describe the causes of fecal incontinence and present our algorithm for its treatment.

Methods: We reviewed 68 patients with Hirschsprung disease and fecal incontinence referred to us after surgery at other institutions. Patients were evaluated by contrast enema and by an examination under anesthesia to look specifically for the integrity of the anal canal. They were designated as having a dilated colon and constipation or a nondilated colon and a tendency to diarrhea based on their stooling pattern and the appearance of the contrast enema. Medical management was started that included laxatives for those patients with a dilated colon and constipation. For those with a nondilated colon and tendency to diarrhea, the management included loperamide, pectin, and a special dietary regimen (constipating diet, 3 meals per day, and no snacks). Those patients who responded to medical management were retrospectively considered to have been pseudoincontinent. Those who did not respond were considered truly incontinent. The truly incontinent group was treated with enemas alone for those with a dilated colon, or enemas, loperamide, pectin, and a constipating diet for those with a nondilated colon and tendency to diarrhea.

Results: Fifty-six patients had true incontinence and 12 had pseudoincontinence. Of the true incontinent group, 27 had a dilated colon and 29 had a nondilated colon. Five of these patients had a damaged or absent anal canal (anastomosis at the anal skin) and all of them had true incontinence. In the dilated colon group with true incontinence, 23 (85%) patients were clean after treatment. In the nondilated colon group with true incontinence, 23 (79%) were successfully treated. All patients in the pseudoincontinent groups had no soiling after treatment. Of 55 in the truly incontinent group, 39 (70%) had had an endorectal (Soave type) pull-through.

Conclusion: Fecal incontinence after operative management of Hirschsprung disease represents a serious problem. Poor surgical technique may be a contributing factor in some of the cases. Successful

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management depends on the appropriate evaluation, which determines whether the incontinence is true or pseudo, and the type of colon the patient has. Each category can be well treated, leading most of the time to a clean child.

Since Orvar Swenson first described a surgical intervention for Hirschsprung disease (HD) in 1948 [1], much progress has been made in its management. Operative procedures are now less invasive and are often done in a single stage [2-5]. Despite an improved collective surgical experience, postoperative morbidity secondary to fecal incontinence persists [6-10] and can cause significant psychological and behavioral problems [11].

The reported incidence of fecal incontinence after operative management of HD is broad because of varying definitions and methods of assessment. We define continence as the ability to have voluntary bowel movements without soiling and without the use of enemas. To achieve this, there must be normal anal sensation, voluntary sphincter control, and appropriate colonic motility. Disruption of any of these factors may result in patients being partially or totally incontinent. Patients can be pseudoincontinent, meaning the physiologic mechanisms needed for continence are intact, yet the child still soils. These children have the potential for bowel control with appropriate medical therapy.

Fecal incontinence after surgical management of HD in theory should not occur because patients are born with an anatomically intact continence mechanism. In fact, all available surgical techniques were designed to preserve the anal canal, and therefore, sensation and the sphincter mechanism. Soiling can occur even when a technically adequate pull-through and colo-anal anastomosis is performed and can also occur in patients who do not have factors known be associated with higher rates of fecal incontinence, such as Down's syndrome [12,13] and total colonic aganglionosis [14-16]. This is evidence of our incomplete understanding of the pathogenesis, treatment, and management of HD.

All operations designed to treat HD produce a significant change in colonic motility because they include the resection of the rectosigmoid and varying amounts of colon. The direct consequence of resecting the rectosigmoid is the tendency to pass stool constantly throughout the day because the natural fecal reservoir has been removed. However, most patients experience constipation (hypomotility) after colonic resection. Again, this illustrates our ignorance about colonic motility disorders associated with HD. To have bowel control requires sensation, working sphincters, reliable rectosigmoid motility, as well as cooperation by the patient.

The goal of this study is to present our experience with the management of fecal incontinence after operative intervention for HD and to give practitioners a straightforward algorithm for the classification and treatment of this challenging patient population.

1. Methods

We performed a retrospective analysis of all patients with HD, presenting with soiling, who were referred to our center over a 20-year period, after operative management elsewhere. Patients were excluded if they were younger than 4 years (before the age of toilet training), had total colonic aganglionosis, had a colostomy at the time of evaluation, or required a reoperation [17] under our care. A chart review was performed to identify the technique of the initial surgery and stooling patterns. Patients were subjected to our protocol of study and management and the results were evaluated.

Our protocol included a history and physical examination, a contrast enema, and an examination under anesthesia to evaluate the integrity of the anal canal. Rectal biopsies were taken for patients with enterocolitis or constipation (reviewed previously) [17]. Patients were divided into 2 groups based on these results. Children were designated as having either a large dilated colon on contrast study and constipation, or having a normal or decreased caliber colon, frequent bowel movements, and/or a tendency to have diarrhea. The dilated colon group initially received senna-based laxatives. The laxative dose was titrated with daily radiographic monitoring of the amount needed to empty the colon of stool and was determined by trial and error. A subset of patients was able to achieve continence with laxatives only and were retrospectively determined to be pseudoincontinent (having the potential for voluntary bowel movements). The remainder of the dilated colon group did not respond to our treatment and therefore was classified as having true incontinence and received our bowel management program consisting of the administration of large-volume enemas (500-1000 mL). No response to laxatives when dealing with a patient with fecal incontinence is considered when the right amount of laxative is given (as radiologically determined) with evidence that the patient is emptying the rectosigmoid, yet the patient continues to soil.

Patients with a nondilated colon were initially treated with loperamide, pectin, a constipating diet, and limiting the number of meals to 3 per day. With this treatment, some patients were able to achieve continence and were termed *pseudoincontinent*. The remaining patients in this group in whom this management proved unsuccessful were considered to have true incontinence and therefore received

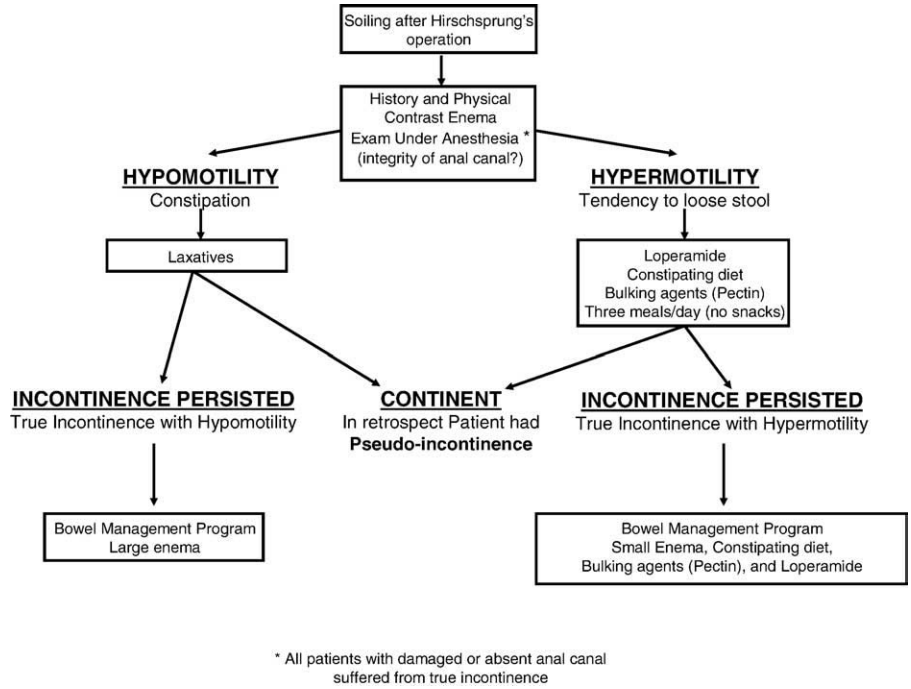

Fig. 1 Treatment algorithm for patients presenting with fecal incontinence after operative management of Hirschsprung disease.

small-volume enemas (200-500 mL), loperamide, pectin, and a constipating diet. Fig. 1 shows this algorithm.

Subsequently, every 6 to 12 months, patients with true incontinence and an intact anal canal received a trial of medical management without enemas because we felt they had the potential for bowel control based on the fact that they had an intact continence mechanism. Fecally incontinent patients with a damaged or absent anal canal remained on bowel management (enemas) with no intent of stopping them.

2. Results

A total of 195 patients were referred from outside institutions after operative management of HD. Fifty-five patients required reoperation for stricture, acquired atresia, dehiscence, megarectal pouch, fistulas, or pouchitis (previously reported) [17]. Eight patients had incomplete records and 2 patients had a permanent ostomy. A total of 130 patients presented with soiling after operative management of HD. Thirty-eight patients were lost to follow-up or were noncompliant, 22 with total colonic aganglionosis were excluded, and 2 patients were not at the age of toilet training at the time of evaluation. Sixty-eight patients comprised the study group.

Thirty-three patients had colonic dilation with infrequent bowel movements (constipation) (Fig. 2). All rectal biopsies taken in constipated patients demonstrated normal ganglion cells. This group initially received laxatives. Six patients became continent on laxatives alone and were retrospectively determined to be pseudoincontinent (encopresis). Twenty-seven patients did not respond to laxatives and were considered to have true incontinence. These patients were subjected to our bowel management program consisting of large-volume enemas (see Fig. 1) and 23 (85%) were clean without soiling after treatment (see Fig. 4).

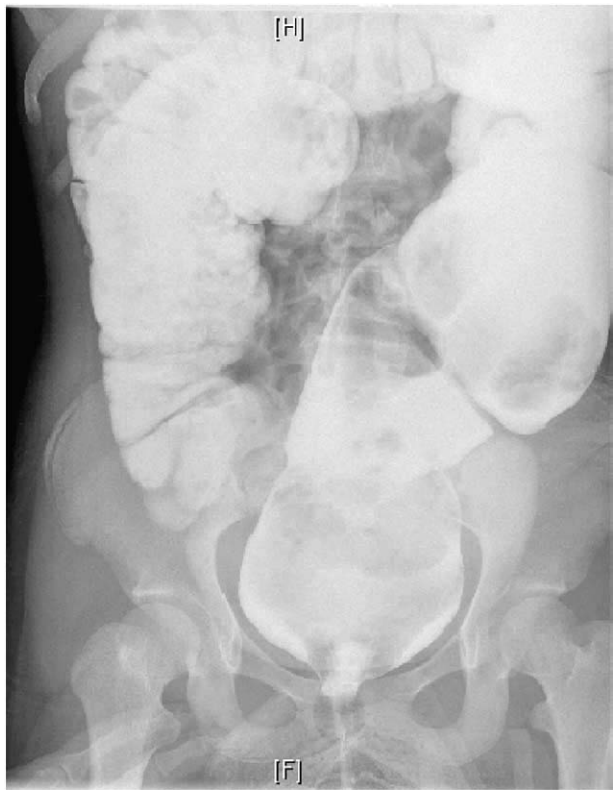

Fig. 2 Contrast enema of a patient with a dilated colon after Hirschsprung pull-through.

Thirty-five patients had a nondilated colon on contrast enema (Fig. 3) and a tendency to have diarrhea. These patients were treated with loperamide, pectin, and a constipating diet. Six patients in this group became continent and therefore in retrospect were considered to be pseudoincontinent. Twenty-nine patients had persistent soiling and were considered to be truly incontinent. A small-volume enema was added to their initial regimen (see Fig. 1), and with this, 23 (79%) became clean without soiling (see Fig. 4).

An examination under anesthesia was performed during the initial evaluation to determine the integrity of the anal canal. Five (7%) patients had loss of the anal canal in which the colonic anastomosis was performed directly at the anal skin (Fig. 5). All patients with a damaged or absent anal canal had true incontinence.

We analyzed the initial procedure for treatment of HD. Forty-four patients had a transanal endorectal pull-through and 3 had a trans-abdominal endorectal pull-through (Soave). Thirty-nine (83%) patients who had an endorectal pull-through had true incontinence and 8 (17%) had pseudoincontinence. Eleven patients had a transanal full-thickness pull-through (Swenson type). Ten (91%) patients who had a full-thickness pull-through had true incontinence and 1 (9%) had pseudoincontinence. Five patients had a Duhamel procedure. Four (80%) patients in this group had true incontinence and 1 (20%) had pseudoincontinence. Of 4 patients who had a Duhamel procedure with true incontinence, 3 were classified as having colonic hypomotility (constipation). The type of initial procedure was unknown in 5 patients.

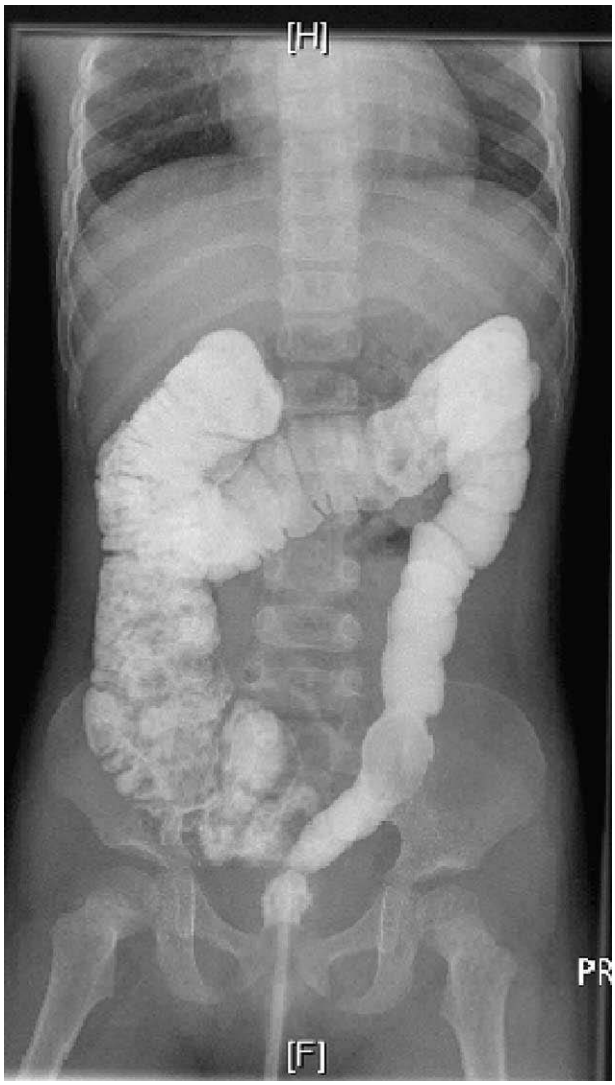

Fig. 3 Contrast enema of a nondilated colon after Hirschsprung pull-through.

3. Discussion

Postoperative fecal incontinence after operative management of HD is a devastating complication [6,7,9,10]. In this study we present our classification and treatment algorithm for this patient population (Fig. 1).

We believe that to successfully manage postoperative fecal incontinence in patients with HD, it is essential to know certain specific anatomic and functional characteristics of the patient. Correct study and classification of these patients allow the clinician to be selective and administer the right treatment modality. In our algorithm (Fig. 1), we start with conservative management; laxatives for patients with colonic dilation and loperamide, pectin, and dietary modifications for patients with a nondilated colon and tendency to have diarrhea. We only offer enemas (bowel management) to patients who are truly incontinent and fail conservative management.

Patients with an intact anal canal, who were initially considered to be truly incontinent and placed on a regimen with enemas to keep them clean, were reevaluated in 6 to 12 months to see if they could be kept clean without the use of enemas and be reclassified as pseudoincontinent. The fact that they had an intact anal canal, we believe, means they have potential for bowel control and therefore they may become continent when they grow up. Patients with an absent or damaged anal canal most likely will remain on enema therapy because of decreased sensation and sphincter function.

The exact cause of fecal incontinence after operative management of HD is obvious in those cases with an absent or damaged anal canal, but is still unknown in the other cases. The mechanisms needed to maintain continence are intact sensation, voluntary sphincter control, and appropriate colonic motility [18]. The first 2 elements (sensation and sphincter mechanism) may be damaged during the primary repair. The third mechanism (colonic motility) is substantially affected after an operation for HD. A damaged or absent anal canal is evidence of a surgical technique in which the transanal dissection was begun too low, which may result in poor sphincter function and poor or absent sensation. The anal canal can also be damaged during a Soave procedure,

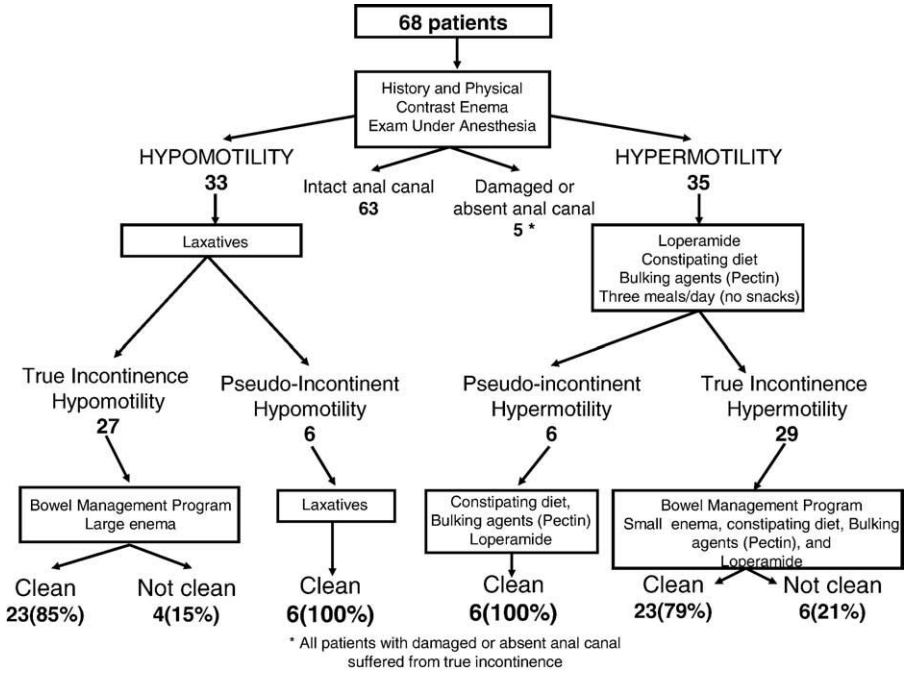

Fig. 4 Distribution of patients in each treatment group.

when the endorectal dissection from above is carried distal into the anal canal.

3.1. Anal canal sensation

Sensation depends on an intact anal canal, which allows one to perceive distention and differentiate between gas, solid, and liquid contents [19]. In our series, all patients who had loss of their anal canal had true incontinence. Preservation of the anal canal is a key feature in all techniques designed to treat HD. The operations described by Duhamel [20] result in less possibility for damage to the anal canal and may explain why true fecal incontinence in this group is rarely seen [21]. However, the Duhamel pouch can cause fecal impaction and sometimes require resection [10,17].

3.2. Sphincter mechanism

The anal sphincter may be compromised during dissection or from vigorous retraction resulting in a lax and incompetent sphincter mechanism (Fig. 6) [22-24]. It has been demonstrated that adult patients with fecal incontinence after surgery for HD have decreased external anal sphincter resting pressure compared with postoperative patients with good bowel control [25]. In our primary cases, we make extraordinary efforts to minimize sphincter stretching and avoid anal canal invasion during transanal dissection, beginning our dissection 1 to 2 cm proximal to the dentate line.

One possible criticism of our treatment algorithm is our evaluation of the anal sphincter. We determine the sphincter function indirectly from our physical examination, and contrast enema. At our institution we do not use anal manometry to directly evaluate sphincter function. Manometry might continue to add to the evaluation of postoperative fecal incontinence [23,25-27] but clinicians using this modality should be certain to document the visualized status of the anal canal because it may be a significant contributor to the etiology of incontinence.

3.3. Motility

Colonic motility is the third factor needed for continence [28]. We do not know why some patients after rectosigmoid

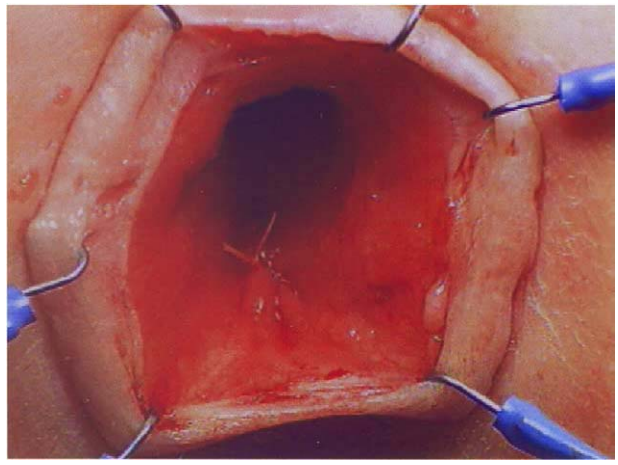

Fig. 5 Examination under anesthesia of a patient with loss of the anal canal and the dentate line. The colo-anal anastomosis was sewn to the anal skin.

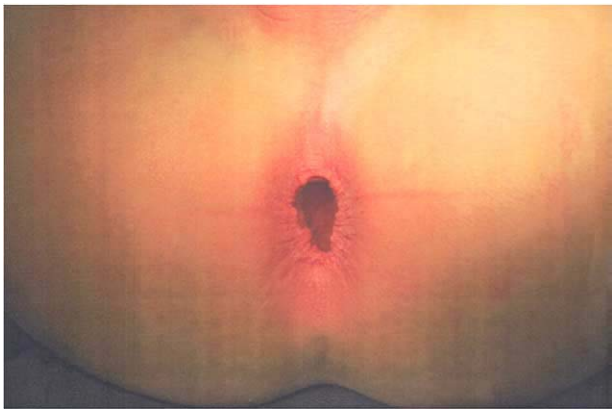

Fig. 6 Picture of an incompetent anal sphincter mechanism potentially because of excessive spreading during transanal dissection. This patient's anus remains open at all times and lacks any ability to squeeze.

resection have constipation whereas others have a tendency to have diarrhea, all this in spite of a technically correct operation and having pulled down normal ganglionic bowel. Several authors use colonic manometry to assess motility in the evaluation of patients with HD [29], and it has been shown that patients with constipation after operative management of HD have an absence of high-magnitude propagating contractions [9]. We have not found that these manometric assessments provide any additional information from what we learn from the contrast study. From the contrast enema, we are able to get a sense of the patient's colonic motility and infer that a dilated colon connotes hypomotility and a nondilated colon connotes hypermotility. Clearly, more objective testing, hopefully available in the future, will help with assessment of these patients.

The literature is unclear concerning which operative procedure for HD offers the best results in bowel control, and it seems that there is no relation between the incidence of fecal incontinence and the type of initial procedure [30]. Teitelbaum et al [7] reported good to excellent continence in children undergoing a primary endorectal pull-through for HD. Langer et al [3] reported that 19% of patients who underwent an endorectal procedure had some form of postoperative fecal incontinence. In our study, 70% of truly incontinent patients had an endorectal procedure, and most endorectal dissections in this series were performed transanally. Even when we believe that the transanal resection is a great maneuver, we are concerned that this type of operation may be leading to a tendency to start the dissection too low and may be causing too much stretching by retractors to gain visualization that can damage the anal canal. We really do not know the true morbidity of different types of operations. This article does not allow us to associate one procedure with higher rates of fecal incontinence. Here, we are analyzing a nonrepresentative sample of patients who were referred to our center after having had operations at other institutions.

We want to emphasize what we believe are the important factors of success in the management of fecal incontinence in patients after surgery for HD.

1. The clinician must first determine if the patient has a dilated or nondilated colon.
2. When giving laxatives, the dosage must be given on an individual basis and, using as a guide, the abdominal film to ensure adequate emptying. Hypermotility in Hirschprung's is counterintuitive but does exist.
3. Examination under anesthesia to determine the integrity of the anal canal is vital and adjusts the patient's expectations concerning long-term functional prognosis.
4. An appropriate bowel management program of the ideal enema, based on the type of colon, is essential for those determined to be fecally incontinent.

Currently, we do not fully understand the pathophysiology of HD and postoperative fecal incontinence. However, with the proper categorizing of patients, we feel that most can be successfully treated.

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Discussion

Dr Arnold Coran (Ann Arbor, Mich): Thank you for the presentation. A number of patients who have what appeared to be incontinence after pull-through for Hirschsprung's disease are actually suffering in many cases from low-grade, often subtle, enterocolitis and this of course overlaps with the presentation from Toronto. If that is the case or if you have that problem, I think one is required to get a rectal biopsy in order to rule out a transition zone being brought down or even an aganglionic segment. But even if the biopsy is normal, a number of these patients will respond to simple antibiotic therapy such as metronidazole or Cipro. So, in this whole group of patients, did you encounter any like that because my own experience is that is far more common than true incontinence.

Dr Martin (response): Thank you for your comment. Yes, we initially identified patients that come with problems after operation for Hirschsprung's disease, and it is our standard to get a rectal biopsy on the exam under anesthesia. A lot of the patients with enterocolitis have enterocolitis for different reasons other than problems with the initial surgery such as an atretic segment or retained aganglionic segment or a large megarectal pouch. It is our standard to give antibiotics and irrigation to treat enterocolitis, but this study is a totally separate patient population and we found that patients that had true incontinence did not have enterocolitis—two separate problems.

Dr Arnold Coran (Ann Arbor, Mich): Very nice breakdown of patients in your bowel management. Focusing on the hypomotile group, what ancillary studies did you do besides exam under anesthesia to determine whether or not that was representative of rectal achalasia or retained segments?

Then, also because large enemas were part of your treatment, what proportion of those patients went on to antegrade enemas or MACE-type adjustments?

Thank you.

Dr Martin (response): We believe that from a contrast enema alone we can tell a lot of information, contrast enema and history and physical examination, so in patients with a large dilated colon coupled with infrequent bowel movements, we found that they behave as hypomotile. We have not had experience with using achalasia studies looking at rectal function. So first, patients that are hypomotile are started on enema therapy and if we find that they are able to become clean on enemas and these patients do not have the potential to be given a subsequent trial of not having enemas, then they might be candidates for antegrade enemas at a further date.

Dr Marc Levitt (Cincinnati, Ohio): I just want to thank Dr Coran for his question and want to just add that it seems that in the patients who are not doing well with Hirschsprung's disease, there are two distinct categories, those that are soiling and those that are distended, and it is fairly rare that the two cross and that the patient has both situations. This study was specifically focusing on the patients without any distension or enterocolitis at all but were soiling. The challenge was to figure out if they were truly incontinent or pseudoincontinent. Unfortunately, many of these patients were truly incontinent and we believe it is related to either destruction of the anal canal as was discussed in a previous article or excessive stretching by the retractors, which both are, I think, surgically preventable.