Megasigmoid: A Source of Pseudoincontinence in Children With Repaired Anorectal Malformations

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● **Three children with a history of anorectal malformation repairs were referred to the authors for evaluation and management of fecal incontinence. Their ages ranged from 5 to 7 years. On examination, all the children had fecal impaction and localized dilatation of the rectosigmoid colon. Medical treatment was tried but failed to control the symptoms, and the patients frequently had to be hospitalized for disimpaction. To correct this problem, the authors resected the dilated sigmoid colon, anastomosing the nondilated descending colon to the rectal ampulla, which was preserved to serve as a reservoir. Postoperatively, constipation was cured in all patients. In addition the patients became fecally continent postoperatively, which was an unexpected bonus. The authors believe that localized dilatation of the rectosigmoid should always be considered whenever a child is having intractable constipation after repair of an anorectal malformation and that sigmoid resection may be considered as a therapeutic alternative. Segmental dilatation of the sigmoid colon may be a source of fecal pseudoincontinence and, therefore, should be ruled out when the surgeon is evaluating patients with fecal incontinence.**

INDEX WORDS: Sigmoid colon, segmental dilatation, resection; intractable constipation; megasigmoid; imperforate anus.

FECAL INCONTINENCE after repair of anorectal malformations remains a challenging problem in pediatric surgical practice. In this study, we present the results of sigmoid resection in three children suffering from fecal incontinence, intractable constipation, and a dilated rectosigmoid after anorectal malformation repairs. These children became fecally continent after the operation. The operation was originally performed with the goal of improving a severe problem of intractable constipation; this goal was achieved. However, fecal continence was an unexpected benefit secondary to the surgical procedure. In retrospect, what seemed to be a severe problem of fecal incontinence was actually overflow incontinence secondary to constipation, which is frequently seen in children with anorectal malformations. Sigmoid resection has been used for the treatment of idiopathic constipation in the adult age group,[1-7] as well as for treatment of idiopathic segmental dilatation of the colon.[8-16] In children, Powell et al[17] and Cloutier et al[18] described cases of megarectum and megasigmoid occurring after anorectal malformation repairs; in their patients, they performed resection of the dilated bowel (including the rectum) with a pull-through procedure; the constipation was relieved, whereas bowel control remained a problem in most of them. In our patients, the sigmoid resection did not include the rectum, which was preserved to serve as a reservoir.

To the best of our knowledge, sigmoid resection with preservation of a rectal reservoir has not been previously performed in children with a dilated rectosigmoid after anorectal malformation repairs.

MATERIALS AND METHODS

The patients' data are summarized in Table 1. These patients, were originally referred to us with the diagnosis of fecal incontinence and their clinical study showed that they also suffered from severe constipation. They were born with malformations whose clinical results after repair are expected to be good (probably low malformations in two patients and a rectourethral fistula in another one.) Interestingly, all of them underwent primary operations elsewhere during which their rectum was not resected (perineal and sacroperineal approaches).

Clinical examination showed large fecal masses in the sigmoid colon in all patients. Anorectal strictures were excluded by rectal examination. Hypaque enemas showed a dilated, aperistaltic sigmoid colon, behaving like a floppy sac. Proximal to the dilated bowel, a normal-sized descending colon with good peristalsis was observed (Fig 1). Aganglionosis was previously excluded by histological examination of rectal biopsies in all patients. In spite of repeated courses of laxatives and 2 daily enemas, the patients still had to be hospitalized periodically for fecal disimpaction.

The problem was severe enough to give these children a very unhappy existence. Consequently, we decided to perform a resection of the dilated sigmoid with preservation of their rectum and a descending colon-to-rectum anastomosis.

Operative Findings

The sigmoid colon was found to be enormously dilated with a very thick wall. No taeniae were noted in the dilated portion of the colon. The serosal vessels were found to be tortuous and prominent (Fig 2). The dilated colonic segment was resected in all cases. We always kept the distal line of resection above the level of the peritoneal reflection in order to preserve a rectal reservoir. Proximally, the resection was right above the upper limit of the dilated bowel. An end-to-end anastomosis was performed between the descending colon and the dilated rectum (Fig 3).

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Table 1. Summary of Results

Patient No.	Sex	Age at Operation (yr)	Original Malformation and Operation	Symptomatology	Results Postsigmoid Resection
1	M	5	Probably rectourethral fistula; sacroperineal operation	Fecal incontinence, fecal impaction	No constipation, no soiling, daily voluntary BM
2	M	11	Probably low defect; perineal repair	Fecal incontinence, fecal impaction	No constipation, no soiling, daily voluntary BM
3	M	7	Probably low defect; perineal repair	Fecal incontinence, fecal impaction	No constipation, occasional soiling, daily voluntary BM

Abbreviation: BM, bowel movement.

Postoperative histological examination of every resected specimen showed ganglion cells in the submucosal and myenteric nerve plexuses with hypertrophy of both muscle layers.

RESULTS

The follow-up period ranged from 6 months to 3 years. Patients and their parents were happy with the results of the operation when specifically asked. In all cases, constipation was cured and none of the patients had postoperative fecal impaction. In addition, the patients originally referred for management of fecal incontinence became totally continent after the operation. Postoperative hypaque enemas showed normal caliber of the descending colon with adequate motility (Fig 4).

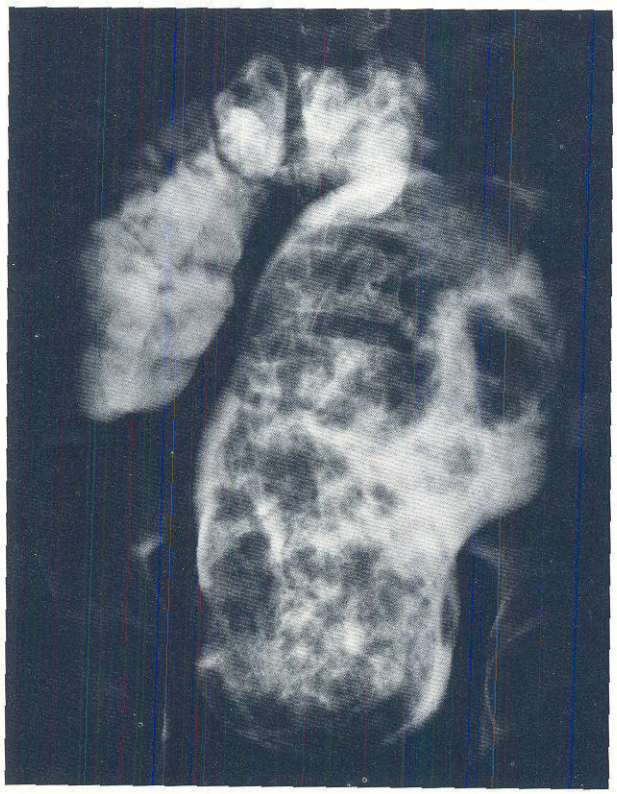

Fig 1. A preoperative hypaque enema showing an enormously dilated sigmoid colon with a normal-sized proximal descending colon.

DISCUSSION

Interestingly, all the patients in this series suffered from severe constipation and had a history of an operation for the treatment of imperforate anus in which the rectum was preserved (sacroperineal pull-through or perineal anoplasty). To explain this, we believe that the most distal portion of the rectum in patients with imperforate anus has a primary hypomotility disorder. We based our thinking on the results of the postoperative evaluation of those patients of our own[19] who suffer from constipation. One could be tempted to postulate that this constipation is a consequence of an intraoperative denervation resulting from the mobilization of the rectal pouch in order to gain length. For this to be true, one would expect to find more severe constipation in those patients with higher defects, but this has not occurred in our experience. In fact, constipation seems to be more severe in cases with lower anomalies.[19] Tapering the bowel could also be considered a causative factor. However, in comparing those patients in our series who underwent tapering with those who did not, we

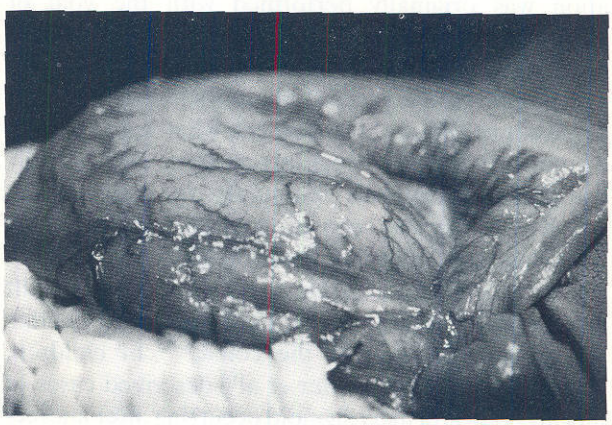

Fig 2. Dilated sigmoid colon found at laparotomy with a normal-sized proximal descending colon.

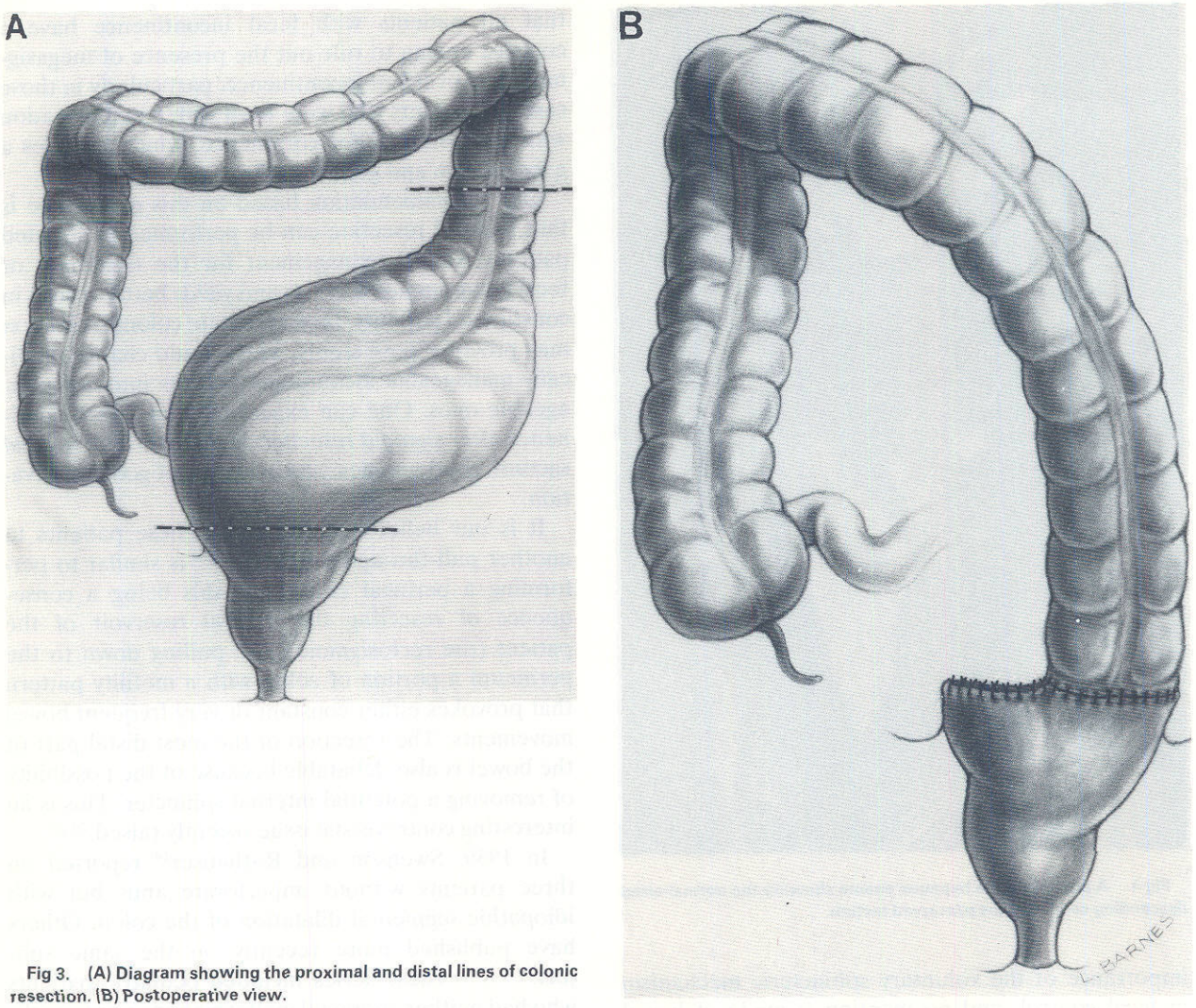

Fig 3. (A) Diagram showing the proximal and distal lines of colonic resection. (B) Postoperative view.

found that this maneuver does not seem to be responsible for the constipation.

On the other hand, it has been our experience that patients referred to us for the management of fecal incontinence who have had abdominoperineal pull-through operations with endorectal dissection, or any other maneuver that includes the resection of the rectosigmoid, usually do not suffer from constipation. In fact, they frequently have loose stools and have to be managed with a constipating diet and/or drugs. These patients have lost their natural reservoir (rectum), which is an important part of the mechanism of continence; they behave like patients having a perineal colostomy, and they are very difficult to manage. We still believe, therefore, that the rectum must be preserved to avoid the problem of loose stools, which may render these patients nonmanageable.

The goal of the operation performed in our patients was not only to remove the intraperitoneal aperistaltic portion of the sigmoid to treat constipation but also to preserve a rectal segment, which has a rather poor motility and acts, therefore, as a reservoir that helps to avoid fecal incontinence.

We believe that the dilatation of the rectosigmoid in these three cases was not due to mechanical obstruction, for anorectal strictures were excluded in all patients prior to sigmoid resection.

The original purpose of the sigmoid resection was only to make their constipation problem more manageable. To our surprise, the operation not only solved the problem of constipation but also rendered them fecally continent. In retrospect, we concluded that these patients were suffering from overflow soiling or what we call "pseudoincontinence" due to severe constipation. The literature referring to the management of fecal incontinence emphasizes the

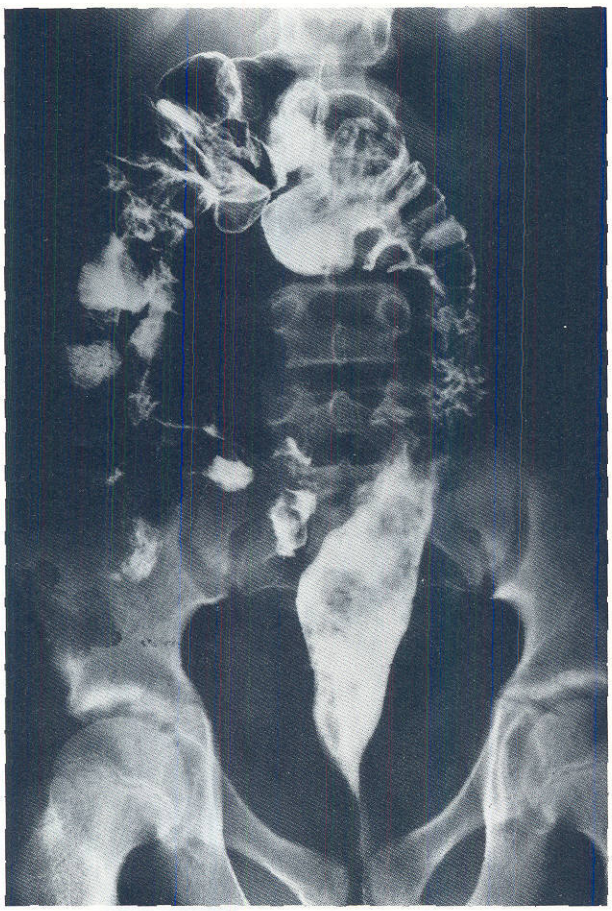

Fig 4. A postoperative hypaque enema showing the normal-sized descending colon and the preserved rectum.

importance of the voluntary sphincteric mechanism in fecal control, and no mention is made of bowel motility. We are convinced that in order to enjoy normal bowel control, it is essential to have normal colonic and rectosigmoid motility. There are good clinical examples that demonstrate this. Patients who have undergone a total colectomy and ileoprocto anastomosis frequently have a hard time controlling their stools. These patients have no reservoir, they have innumerable bowel movements per day, and sometimes they suffer from severe perineal rash in spite of having normal sensation and normal sphincters. The other clinical example is represented by patients with normal sphincters who suffer from severe idiopathic constipation and overflow incontinence (constant soiling). This is similar to our patients. Not treating their constipation problem but rather performing one of the conventional operations to improve the sphincter mechanism would not only not help them but also could actually worsen their condition. Thus, it is our specific recommendation that all patients with fecal incontinence have a contrast enema to rule out the presence of megasigmoid and overflow incontinence, particularly in those cases of patients who were born with a malformation with the potential for continence, which includes a good sacrum and good muscles.

Our recommendation based on this experience is that sigmoid resection can be performed in selected patients. Bowel management for the treatment of fecal incontinence is easier and yields better results in constipated patients. Indiscriminate colonic resection may provoke loose stools, which could change medically manageable incontinent patients into nonmanageable ones. One can expect only improved continence after sigmoid resection in patients with normal sacrum, good muscles, and evidence of good innervation.

It is our belief that subjecting these patients to another pull-through operation[17,18] is similar to performing a perineal colostomy, this being a consequence of resecting the natural reservoir of the patient (the rectosigmoid) and pulling down to the perineum a portion of colon with a motility pattern that provokes either constant or very frequent bowel movements. The resection of the most distal part of the bowel is also debatable because of the possibility of removing a potential internal sphincter. This is an interesting controversial issue recently raised.[20-25]

In 1959, Swenson and Rathauser[14] reported on three patients without imperforate anus but with idiopathic segmental dilatation of the colon. Others have published more recently on the same subject.[8-13,15,16] These series included pediatric patients who had neither anorectal malformations nor aganglionosis; they had only a primary segmental colonic dilatation. Proximal and distal to this dilated bowel segment, the intestine was normal in caliber and motility. The resection of the segmental dilatation proved to be curative. Even when this is a different disease, there is some similarity with our cases in that the resection of a nonfunctional dilated bowel solved the problem of constipation.

The exact etiology of the problem of the dilated rectosigmoid after the repair of anorectal malformations remains unknown. Interestingly, it is well known to the pediatric surgical community that very dilated blind bowel pouches in cases of intestinal atresia suffer some form of bowel motility disturbance and must, therefore, be resected.[26-31] Tepas et al[30] reported on the loss of ATPase activity in the muscle layers of the distended proximal loop in cases of intestinal atresia. Extrapolating this fact to the subject of anorectal malformation could erroneously in-

duce one to recommend the resection of the rectum. We still believe, however, that at least a part of the rectum should be preserved as a reservoir. In addition, many of our patients with a preserved rectum do not suffer from constipation.[19] We obviously need an explanation of why some patients develop a more severely dilated sigmoid colon than others.

Perhaps we could get more valuable information about the nature of the problem by applying more accurate and objective methods of evaluation of the bowel motility or else by using more sophisticated histological and histochemical techniques in the study of the resected specimens.

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