A Ten-Year Experience with Ninety-Two Cases of Hirschsprung's Disease

Including Sixty-Seven Consecutive Endorectal Pull-Through Procedures

THEODORE Z. POLLEY, JR., M.D., ARNOLD G. CORAN, M.D., JOHN R. WESLEY, M.D.

From the Section of Pediatric Surgery, University of Michigan Medical School and Mott Children's Hospital, Ann Arbor, Michigan

From July 1974 through November 1984, 92 patients with Hirschsprung's Disease (congenital aganglionosis) have been treated at the University of Michigan-Mott Children's Hospital. This series includes 67 consecutive modified endorectal pull-through (ERPT) procedures in children. Fifty-one of these 67 patients had standard rectosigmoid disease and underwent a successful ERPT with only two major complications. One of these 51 children underwent a successful ERPT but died in the late postoperative period from severe congenital heart disease. Eighteen of the 92 patients suffered from total aganglionosis or long-segment disease. Sixteen of these have undergone an ERPT with no mortality or operative morbidity. The follow-up ranges from 6 months to 10 years. All of the children who have reached 3 years of age are continent. Of the remaining 23 patients, 12 were referred following an unsuccessful pull-through at another hospital. The 12 operations included five Swenson pull-throughs, five Duhamel procedures, one ERPT, and one subtotal colectomy. It was possible to redo or revise the pull-through procedures successfully in all but one patient, who required a permanent colostomy. Finally, 11 children were referred for management of a variety of complications following pull-through procedures performed at other institutions. None of these 11 patients required a reperformance of their pull-through, and all were successfully treated with lesser surgical procedures or with medical management. The excellent functional results and the low morbidity and zero operative mortality are attributed to the technical ease of performing the modified ERPT.

HIRSCHSPRUNG'S DISEASE, or congenital aganglionosis, first described in 1888 by the Danish pediatrician, Harold Hirschsprung, continues to be an important cause of intestinal obstruction in the newborn period and remains prominent in the differential diagnosis of constipation in the child.[1] Although each has been modified following its original description, three procedures (the endorectal pull-through (ERPT), the Duhamel procedure, and the Swenson abdominal perineal pull-through) continue to be used as the standard operations for the definitive treatment of this disease. At the University of Michigan-Mott Children's Hospital, we have had the opportunity to care for 92 patients with Hirschsprung's disease during the past 10 years. This series represents patients diagnosed, treated, and followed at our institution as well as those referred from other hospitals following operative management of their disease. This report includes 67 consecutive modified endorectal pull-through procedures, the results of which constitute the basis for this report.

Presented at the 105th Annual Meeting of the American Surgical Association, New Orleans, Louisiana, April 25–27, 1985.

Reprint requests: Theodore Z. Polley, Jr., M.D., Room F7516, Box 066, Mott Children's Hospital, Ann Arbor, MI 48109.

Submitted for publication: May 2, 1985.

Materials and Methods

From July 1, 1974, through November 30, 1984, 92 patients with Hirschsprung's disease have been treated at the Mott Children's Hospital. The age at presentation ranged from newborn to 33 years. Eighty-eight children were under 8 years of age (mean, 5 months) at initial evaluation. The other four patients were 33, 17, 16, and 8 years old at the time of their presentation. Seventy per cent of the patients (64) were male, representing a male to female ratio of 2.3 to 1. The expected spectrum of presenting complaints in our series included constipation, abdominal distention, vomiting, failure to pass meconium within the first 24 hours of life, and free intraperitoneal air in one patient. Fourteen patients (15%) of the entire group had associated congenital anomalies including Down's syndrome in eight.

The patient population can be most conveniently divided into four subgroups (Fig. 1). The first consists of 51 patients with standard rectosigmoid disease who underwent diagnosis, definitive operation, and follow-up at our

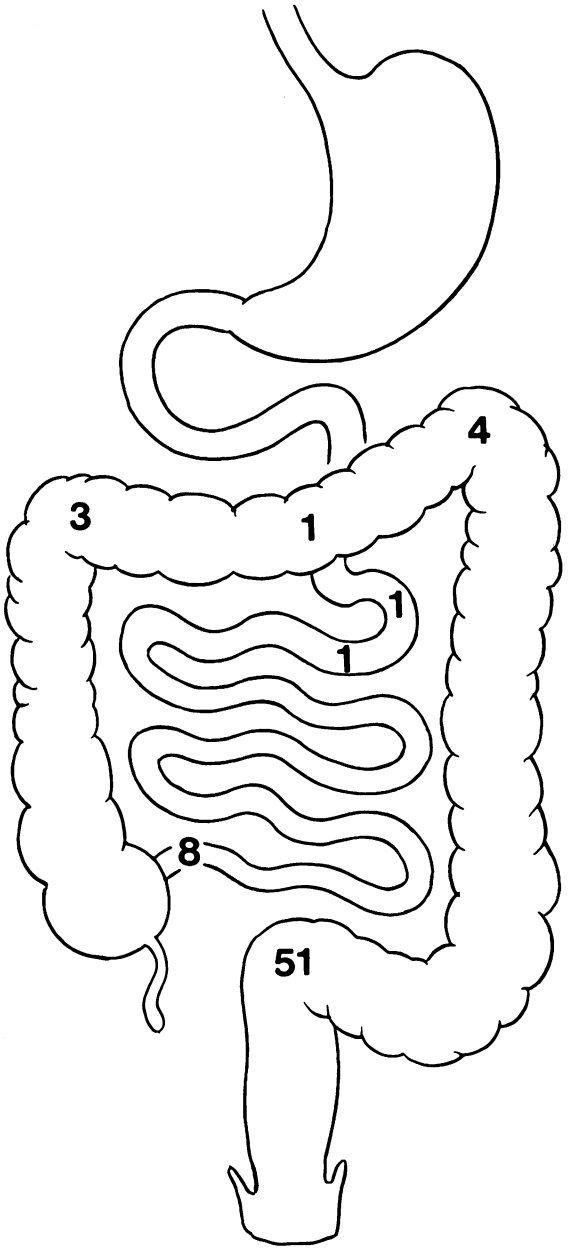

FIG. 1. Distribution of cases managed solely at the University of Michigan-Mott Children's Hospital.

institution. The modified ERPT procedure was performed in all of these patients. Follow-up in this group is complete and ranges from 6 months to 10 years. The male to female ratio in this group is 7.4 to 1.

Eighteen of the patients, who were totally managed at our hospital, suffered from long-segment disease (Fig. 1). In four of these, aganglionosis was present to the splenic flexure; in one, to the mid-transverse colon; and in three, to the hepatic flexure. In eight patients, the disease involved the entire colon, with the absence of ganglion cells extending to the distal jejunum in one of these eight children and to the proximal jejunum in another. The male to female ratio in this group was one. Sixteen of these 18 patients underwent the ERPT at an average age of 16.5 months. Follow-up, which was again 100%, ranged from 4 months to 8 years.

Twelve patients were referred to our institution following failure of their original procedure. Among these 12 patients, the original operation was the Swenson procedure in five. These five were treated with a redo Swenson procedure in three, a ERPT in one, and a rectal myotomy in the fifth. One patient had undergone an ERPT using aganglionic bowel and was treated with a redo ERPT. One patient had ganglionated colon resected at another hospital and was treated with an ERPT at our hospital. Five patients had previously undergone Duhamel procedures. Two of these were treated with a redo Duhamel procedure and three with a septectomy. One redo proved unsatisfactory and the patient was finally treated with a permanent colostomy.

Finally, a group of 11 patients were referred for management of complications following procedures done elsewhere. Six of these patients had Swenson procedures done and four had undergone an ERPT at other institutions. One child had not yet undergone a definitive procedure and was referred for nutritional management.

The details of diagnosis, preoperative preparation, operative technique, and postoperative care have been reported previously;[2,3] however, these subjects will be briefly summarized here. The diagnosis of aganglionosis is routinely made by suction rectal biopsy. There have been no false-negative or false-positive results with this simple technique.[4] In those few cases where results were equivocal, diagnosis was made by open, full-thickness rectal biopsy. Leveling colostomy is then performed with frozen section confirmation of ganglionated bowel at the site of the colostomy or ileostomy. The patient is then readmitted between 8 months and 1 year of age or when the child has reached approximately 20 pounds. In children older than 1 year, the interval between colostomy and ERPT is individualized but is usually 2 to 3 months. At this admission, preoperative bowel preparation is carried out; it consists of a clear liquid diet for 2 days, oral Neomycin® and Erythromycin® base for 24 hours prior to surgery, and rectal and colonic irrigations with 1% Neomycin for the 24 hours prior to the definitive procedure. Parenteral broad spectrum antibiotics are used during the perioperative period.

Details of the operative technique have been previously reported.[2,5] Immediately prior to the completion of the procedure, which is accomplished *via* combined abdominal-perineal approach, a silastic sump drain is placed within the rectal-muscular cuff. This drain remains in place for approximately 24 hours, as does the Foley catheter which is placed intraoperatively. Neither backup colostomy nor ileostomy is routinely used. Oral feedings are

TABLE 1. *Results Following Modified ERPT Procedure in Fifty-One Patients with Rectosigmoid Hirschsprung's Disease*

	Number of Patients	Per cent
Deaths	0	0%
Incontinence	0	0%
Enterocolitis	8	16%
Requiring myotomy for resolution	3	6%
Morbidity	8	16%
Major	2	4%
Minor	6	12%

TABLE 2. *Results Following Modified ERPT Procedure in Sixteen Patients with Long-segment or Total Colonic Hirschsprung's Disease*

	Number of Patients	Per cent
Deaths	0	0%
Incontinence	0	0%
Enterocolitis	2	13%
Morbidity	3	19%
Major	1	6%
Minor	2	13%

resumed once bowel activity has returned, generally at 48 hours. The child is discharged from the hospital on the seventh postoperative day following rectal examination.

Results

There were no operative or postoperative deaths in our series. Of the two patients who died, one had untreatable disease with the aganglionosis extending to within 20 cm of the ligament of Treitz and died of severe malnutrition 3 months later. The second child with rectosigmoid disease succumbed 1½ months following a successful ERPT from the complications of severe congenital heart disease (endocardial cushion defect).

All of the 51 patients with standard rectosigmoid disease who have undergone an ERPT and who have reached 3 years of age are continent (Table 1). Eight patients (16%) have suffered postoperative enterocolitis, which has been self-limited in five. Three of these eight ultimately required an internal sphincterotomy for management, which was successful. Three patients have undergone an enterolysis for adhesive bowel obstruction and two patients developed anal stenosis, which responded to dilatations in one and temporary colostomy and dilatation in the other. Five children developed infrequent fecal soiling in the early postoperative period which responded to a temporary enema program. Six patients (12%) suffered minor complications including a stitch abscess and minor wound infections. One patient underwent reexploration for bleeding and was subsequently found to be factor VIII deficient. Finally, two patients (4%) suffered major complications; one an anastomotic leak with peritonitis and the second a cuff abscess both requiring temporary colostomy. Both of these patients later had their colostomies closed and have subsequently had excellent results.

Of the 18 patients with long-segment disease, all but two of these have undergone a definitive ERPT (Table 2). The follow-up on this group of children ranges from 4 months to 8 years (mean 47 months). Two patients have suffered postoperative enterocolitis which resolved spontaneously; two suffered minor wound infections; and one patient developed an incisional hernia which was repaired. In one child (the first in the series) an ERPT was done using aganglionic bowel because of an incorrect pathology reading at the time of surgery. This child was subsequently treated with a Swenson pull-through when it was clear that she suffered from total colonic aganglionosis. She is currently completely continent with one to two bowel movements per day. These 16 children are totally continent and experience an average of three to four stools daily.

Twelve patients, 11 with classical Hirschsprung's disease and one with total colonic aganglionosis, were referred to our institution following unsatisfactory pull-through procedures (Table 3). Five of these patients had undergone a Swenson procedure; five, a Duhamel procedure; one, an ERPT; and one patient underwent a colectomy of ganglionated bowel. Of the five patients treated with the Swenson procedure, three had developed severe rectal strictures, one had unmanageable constipation, and one suffered an anastomotic disruption. One of these five patients was treated with rectal myotomy alone and achieved an excellent result, while four underwent redo-Swenson procedures with excellent results. The patient who underwent an ERPT of aganglionic bowel was managed with another ERPT with an excellent result. One male of 52

TABLE 3. *Summary of Twelve Patients Referred Following Initial Pull-Through at Another Institution Requiring Revision of the Pull-Through*

Initial Procedure	Indications for Referral	Treatment
Swenson (5)	Constipation (1)	Myotomy (1)
	Stricture (3)	Redo Swenson (3)
	Anastomotic disruption (1)	Redo Swenson (1)
ERPT (1)	Pull-through of aganglionic bowel (1)	ERPT (1)
Colectomy (1)	Removal of ganglionated bowel (1)	ERPT (1)
Duhamel (5)	Constipation (3)	ERPT (1)
	Constipation (1)	Septectomy (3)
	Stricture and abscess (1)	Redo Duhamel (1)
		Permanent colostomy (1)

TABLE 4. *Summary of Eleven Patients Referred Following Initial Procedure at Another Institution Not Requiring Redo Pull-through*

Initial Procedure	Indications for Referral	Treatment
Swenson (6)	Rectovesical fistula (1)	Repair (1)
	Stricture (1)	Permanent ileostomy (1)
	Incontinence (4)	Enema program (4)
ERPT (4)	Stricture (1)	Colostomy (1)
	Cuff abscess (1)	Drainage and uretero-lysis (1)
	Enterocolitis (1)	Medical management (1)
	Cuff abscess + 180° twist of pull-through (1)	Colostomy followed by colostomy closure (1)
Ileostomy (1)	Severe nutritional depletion (1)	Parenteral nutrition (1)

years, who at age 16 had undergone excision of dilated ganglionated bowel, underwent the ERPT procedure. After the ERPT, he developed a rectal stricture, which has been successfully managed with periodic dilatations. Of the five patients who previously underwent the Duhamel procedure, three presented with constipation secondary to a retained septum. The redo operation in these three patients consisted of a septectomy with excellent results. The fourth patient underwent a redo Duhamel procedure because of a severe rectal stricture and achieved an excellent result. In the fifth child, a redo pull-through was attempted but a satisfactory reconstruction was impossible and the patient now has a permanent colostomy.

Finally, there were 11 patients referred to us following pull-through procedures at other institutions who experienced complications which did not require performance or revision of a pull-through procedure (Table 4). Six of these patients originally had rectosigmoid disease and five had long-segment disease. Of these 11 patients, six had undergone Swenson procedures. One of these six suffered from a rectovesical fistula which was repaired successfully; one child with a rectal stricture underwent permanent ileostomy and two patients with fecal incontinence and two with constipation were successfully managed with an enema program.[6] Four of the 11 children were referred following an ERPT procedure. One suffered from a cuff abscess with a 180° twist of the pull-through colon. Colostomy was performed, the twist revised, and the colostomy subsequently closed. This patient is now continent and is having one to two bowel movements per day. One patient developed a cuff abscess 8 months following an ERPT and required operative drainage. One of these 11 children experienced multiple episodes of enterocolitis which were successfully managed conservatively; and one patient with a stricture was treated with permanent colostomy without attempting a redo pull-through. Finally, one child was referred with severe nutritional depletion after having undergone an ileostomy for what proved to be total colonic aganglionosis. Careful nutritional management using parenteral nutrition was successful in reversing these deficiencies.

Discussion

Swenson first introduced the abdominal-perineal resection with pull-through for Hirschsprung's disease in 1948.[7] Although he achieved great success with this procedure, similar results were not seen in other institutions because of the technical difficulties involved in doing this operation. In addition to the technical difficulties, the risk of damage to the sacral nerves responsible for fecal and urinary continence as well as sexual function with the Swenson operation stimulated the development of other pull-through procedures. Duhamel introduced the retrorectal pull-through and end-to-side anastamosis in 1956 and, although the operation continues to be used relatively frequently, it has the disadvantage of a slightly increased incidence of incontinence and the occurrence of fecalomas in the retained rectum. If the procedure is not performed properly, a retained septum may also create problems. In 1964, Soave[8] introduced the endorectal pull-through (ERPT) for Hirschsprung's disease which was modified by Boley in 1964[5] and by Coran in 1976.[2] The main advantages of the ERPT are ease of technical performance and avoidance of damage to the sacral nerves which control fecal and urinary continence because the dissection is done entirely inside the rectal cuff.

There have been several large reports of patients operated on for Hirschsprung's disease which can serve as a standard for comparison. In 1979, Kleinhaus et al.[9] published the report of the survey of the Surgical Section of the American Academy of Pediatrics (AAP) on Hirschsprung's disease. Mortality was 1.1% in 187 patients treated with the ERPT; 2.5% in 390 patients undergoing the Swenson procedure; and 1.8% in 339 patients managed with the Duhamel procedure. All of these patients had standard rectosigmoid disease. Swenson, in his 25-year follow-up study,[10] reported a 3.3% mortality rate in his series of 483 patients who underwent the Swenson procedure. The mortality rate in our series of patients undergoing the ERPT was 0%; it is 1.9% if the patient who died of complications due to cardiac disease is included. If the patients with total colonic aganglionosis with or without small bowel involvement are considered separately, the mortality rate in the Kleinhaus report was 30% for those patients undergoing the Swenson procedure and 25% for the Duhamel procedure. In patients undergoing the ERPT, the mortality rate was 0% in the AAP series, which is the same as the mortality rate in our postoperative patients with long-segment disease.

Of those patients who underwent the initial pull-

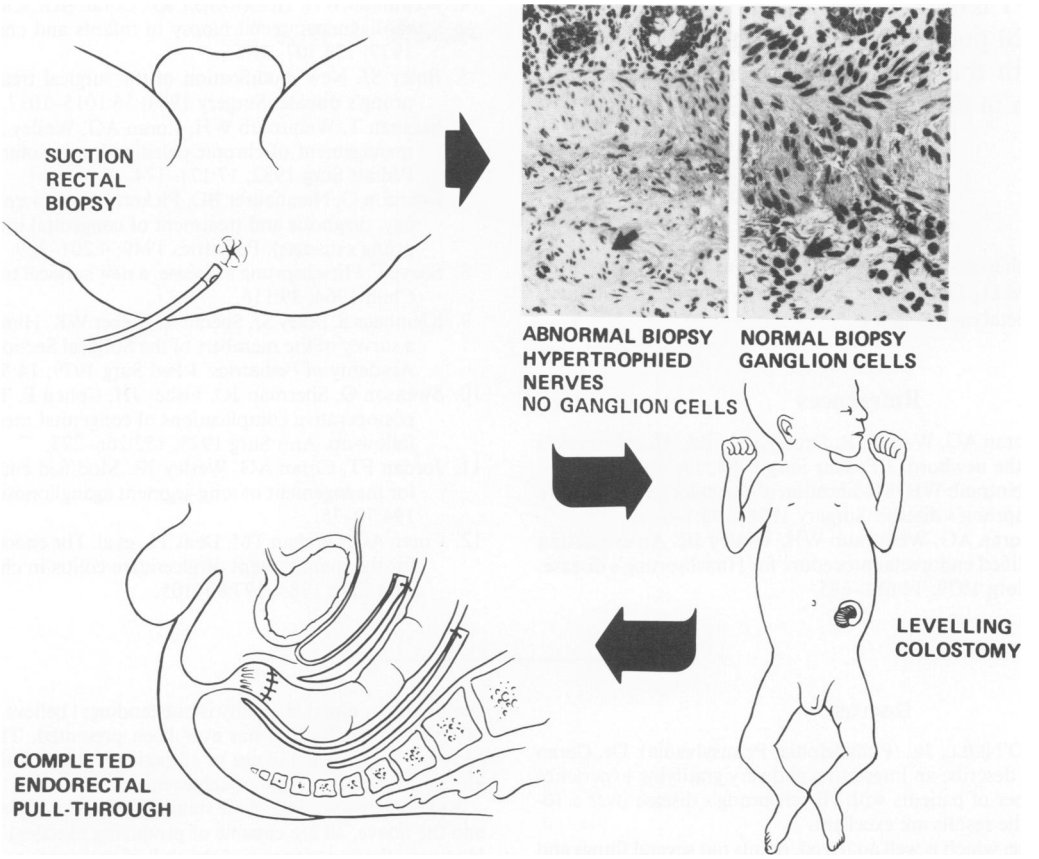

FIG. 2. Overall management of Hirschsprung's Disease at the University of Michigan-Mott Children's Hospital.

through at our hospital, none have a permanent colostomy or ileostomy and all who have reached the age of 3 years are continent. This is compared to an incidence of a permanent enterostomy of 1.4% in Swenson's series, 3.2% in those patients undergoing the Swenson operation as reported in the AAP survey, and 1.1% each in those patients undergoing the Duhamel procedure or ERPT as reported in the same survey. Likewise, the 16% morbidity rate in our series of patients with standard rectosigmoid disease compares favorably with the complication rate of 35.9% in those patients undergoing the Swenson procedure, and 17.9% each for the Duhamel procedure and the ERPT as reported in the AAP survey. In those patients with long-segment and total colonic Hirschsprung's disease in our series, the morbidity rate was 19% (3 patients) *versus* 40% following the Swenson procedures, 37% after the Duhamel procedure, and 0% after the ERPT. The postoperative enterocolitis rate in our series is 11% as compared with 15.6% following Swenson's operation, 5.9% following the Duhamel procedure, and 2.1% following the ERPT as reported in the AAP survey. In Swenson's own series, the incidence of postoperative enterocolitis was 16%. Overall, 84% of the patients in our series with standard rectosigmoid disease and 81% of those patients with long-segment aganglionosis had no complications in comparison with 52% undergoing Swenson's operation, 55% undergoing the Duhamel procedure, and 60% undergoing the ERPT again as reported in the AAP series.

There are then a number of factors which support the continued use of the ERPT in the treatment of standard, long-segment, and total colonic aganglionosis.[11] We believe that the low complication rate is related to the technical ease of performing the lower anastomosis and the advantage of doing the dissection within the rectal muscular cuff. An additional advantage of the ERPT is the ease with which it can be taught to resident surgeons. The success we have had with this procedure has encouraged us to use it in patients with familial polyposis and ulcerative colitis as well. At our institution, the ERPT has become the procedure of choice for both of these latter conditions and the results obtained in these patients are comparable to those seen in this series of patients with Hirschsprung's disease.[12]

Our approach to the infant with suspected Hirschsprung's disease is an immediate suction rectal biopsy to make the diagnosis, followed by a leveling colostomy. The

definitive ERPT is done when the child is 8 to 12 months old or about 20 pounds in weight (Fig. 2). Our 10-year experience with this approach justifies continuing this treatment plan in infants and children with all forms of aganglionosis.

Acknowledgment

The authors wish to gratefully acknowledge the assistance of Kathleen P. Heidelberger, M.D., Department of Pathology, for diagnostic interpretation of the rectal biopsy slides.

References

1. Klein MD, Coran AG, Wesley JR, Drongowski RA. Hirschsprung's disease in the newborn. J Pediatr Surg 1984; 19:370–374.
2. Coran AG, Weintraub WH. Modification of the endorectal procedure for Hirschsprung's disease. Surgery 1976; 143:1–6.
3. Jordan FT, Coran AG, Weintraub WH, Wesley JR. An evaluation of the modified endorectal procedure for Hirschsprung's disease. J Pediatr Surg 1979; 14:681–685.
4. Weintraub WH, Heidelberger KP, Coran AG. A simplified approach to diagnostic rectal biopsy in infants and children. Am J Surg 1977; 134:307–310.
5. Boley SJ. New modification of the surgical treatment of Hirschsprung's disease. Surgery 1964; 56:1015–1017.
6. Sarahan T, Weintraub WH, Coran AG, Wesley JR. The successful management of chronic constipation in infants and children. J Pediatr Surg 1982; 17:171–174.
7. Swenson O, Nieuhauser BD, Pickett LK. New concepts of the etiology, diagnosis and treatment of congenital megacolon (Hirschsprung's disease). Pediatrics 1949; 4:201–209.
8. Soave F. Hirschsprung's disease: a new surgical technique. Arch Dis Child 1964; 39:116.
9. Kleinhaus S, Boley SJ, Sheran M, Sieber WK. Hirschsprung's disease, a survey of the members of the Surgical Section of the American Academy of Pediatrics. J Ped Surg 1979; 14:588–597.
10. Swenson O, Sherman JO, Fisher JH, Cohen E. The treatment and postoperative complications of congenital megacolon: a 25-year follow-up. Ann Surg 1975; 182:266–273.
11. Jordan FT, Coran AG, Wesley JR. Modified endorectal procedure for management of long-segment aganglionosis. Ann Surg 1981; 194:70–75.
12. Coran AG, Sarahan TM, Dent TL, et al. The endorectal pull-through for the management of ulcerative colitis in children and adults. Ann Surg 1983; 197:99–105.

Discussion

DR. JAMES A. O'NEILL, JR. (Philadelphia, Pennsylvania): Dr. Coran and his coauthors describe an interesting and very gratifying experience with a large number of patients with Hirschsprung's disease over a 10-year period, and the results are excellent.

Their experience, which is well analyzed, points out several things and raises a few questions.

First of all, the Soave or endorectal pull-through operation, actually first suggested in another form by Ravitch and his group, is an effective and worthwhile approach to this disease, and despite my own personal preference for the approach that Dr. Coran has described, particularly in young children, the bulk of the experience at the Children's Hospital of Philadelphia over the last 10 years in a somewhat larger series of patients has been with the Duhamel operation. The results have been essentially identical to the Michigan results. Thus, it seems that most surgeons seem to use the procedure they feel the most comfortable with, and it is not necessarily a selection related to the patient.

Secondly, protective colostomy, as he outlined in the manuscript, is not required to prevent pelvic sepsis, and we would agree with that conclusion.

Thirdly, the incidence of enterocolitis is 16% in Dr. Coran's series and is similar to that reported in the literature. Thus, it would seem that it is not the operation but the patient that seems to make the difference, and I wonder if Dr. Coran agrees or disagrees with that.

Another question, why, when the first operation failed, and you did remedial procedures, did you repeat the same operation, particularly the Swenson? We have found, for example, that the Duhamel approach is an easier redo operation and works very well as a remedial procedure.

The authors have excellent results with continence following endorectal pull-through. We found the same thing, but occasionally see soiling for a few weeks before full continence appears. Has this been your experience as well or not?

The true mortality in this series, as mentioned, is 1.9% if the child with cardiac disease is included. Do you mean to imply that perhaps cardiac disease in a few cases should be repaired first and repair of Hirschsprung's disease deferred until a later time?

Overall, the authors have made their point very well that, in their hands with the operation as they do it, the endorectal pull-through is capable of providing absolutely superb results.

DR. E. THOMAS BOLES, JR. (Columbus, Ohio): Certainly 67 consecutive patients, including 16 with long-segment aganglionosis, with no deaths and minimal mortality is outstanding; I believe it is the best series in terms of results that has ever been presented. The results of total continence at 3 years of age in all patients reaching that age is, indeed, an extraordinarily excellent achievement.

As Dr. Coran said, the three standard procedures, Swenson, Duhamel, and the Soave, all are capable of producing excellent long-term results. However, the experience and the skill of the surgeon are the ingredients that make the difference in these patients more than the individual operation itself. Relatively high complication rates have actually been reported in all three of these procedures. Furthermore, a fourth of Dr. Coran's series were patients referred to his institution following complications of one type or another. Although in their particular hands the results with these complicated patients were also good, there is no question that, generally speaking, these secondary operations can be very difficult at times, and the overall, long-term results certainly cannot be expected to be as good as in those patients whose primary operations are uncomplicated.

Another feature in this disease that Dr. Coran did not mention is the hidden death rate among patients who never get to the surgeon. As a matter of fact, some 20-odd years ago Dr. Swenson reported this to be about 50% in the infant age group, primarily from enterocolitis and sepsis. We have been reviewing our own cases in the last 20 years. Eight of 171 patients died before diagnosis and before any type of surgical manipulation whatever, the diagnosis being made at autopsy. This is about a 5% mortality rate and obviously is minimal. I am sure that the actual mortality from undiagnosed disease is much higher.

Finally, I think that this paper is a very excellent and fitting tribute to Franco Soave, that very excellent surgeon from Genoa, Italy, who first developed and popularized this operation and who tragically died just a few months ago.

DR. JOHN R. LILLY (Denver, Colorado): Dr. Rosoff, I will limit my remarks to patients with total colon Hirschsprung's disease and, in fact, use them only as a touchstone to make some observations about patients with ulcerative colitis and familial polyposis.

In patients with total Hirschsprung's disease, Dr. Coran removes the entire colon and does an ileoanal pull-through operation without pouch formation. According to the text, he does the operation at an average patient age of one-and-a-half years, and I assume that the patient's ileum adapted relatively rapidly to its rectal location. In other communications, Dr. Coran describes an identical operation for patients with ulcerative colitis and familial polyposis. He recommends in this group, however, that the procedure be done at or after adolescence because of a belief