

Posterior Sphincterotomy and Rectal Myotomy in the Management of Hirschsprung's Disease

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COORDINATED and effective peristalsis in the gastrointestinal tract apparently depends upon the integrity of ganglion cells and nerves of the myenteric plexus. This is particularly true in the distal colon and rectum in which their absence seriously interferes with the normal mechanism of defecation.^{9, 12} Distention of the rectosigmoid results in increased tone of the distal aganglionic rectum and internal sphincter which is the fundamental disturbance in Hirschsprung's disease. The normally innervated proximal colon responds to this type of functional obstruction by dilatation and hypertrophy. Chronic constipation, protuberant abdomen, disturbances in growth and nutrition, with possible diarrhea, enterocolitis, volvulus, or perforation signify the pathologic and functional changes of Hirschsprung's disease in a child. In recent years, with better understanding and earlier recognition, Hirschsprung's disease is most commonly diagnosed in the neonate with abdominal distention, delay in meconium evacuation and radiologic findings of large bowel obstruction.²²

Congenital aganglionosis also constitutes a disease which may involve almost any length of the gastrointestinal tract, with variation in symptomatology that may be

independent of the involved aganglionic segment.^{5, 8, 14} Most commonly involved are the rectum and rectosigmoid, and emphasis has properly been placed upon identification of the proximal extent of the disease. Many of the poor results associated with the surgical management are secondary to incomplete resection of the diseased aganglionic segment.

At the other end of the spectrum are patients who have a very short aganglionic segment; here recognition is often difficult. In the younger age group these patients are more likely to present with mild chronic constipation rather than the classic picture of Hirschsprung's disease. Barium enema x-ray may suggest the presence of a short, narrow segment but may not indicate precisely the extent of the disease and, in some instances, a constricting segment may not be demonstrable.⁶ Thus, although the diagnosis may be suspected by history as well as by barium enema, confirmation can be made only by histologic examination. As a consequence of these vagaries of the disease, 80 years after Harald Hirschsprung's original and classic description and 20 years after Swenson introduced a surgical treatment based upon its pathologic physiology, the evaluation and management of patients with Hirschsprung's disease remains a challenge.

This paper reports our experiences with patients in whom a posterior sphincterot-

Presented at the Annual Meeting of the Southern Surgical Association, December 8-10, 1969, Hot Springs, Virginia.

TABLE 1. *Clinical and Pathologic Findings in Patients Treated for "Short Segment" Hirschsprung's Disease*

Patient	Ganglion Cells	Onset	Abnormal Barium Enema	Age at Operation	Weight %	Height %	Subsequent Treatment	Other Conditions	Follow-up	Results
1. W. S. ♂	Absent to 6 cm.	Birth	+	4 mo.	25%	3%	Swenson proced. at age 2	None	3 yr.	Immediate—poor Ultimate—good
2. M. S. ♂	In paramuscular & fibro-adipose tissue only	6 mo.	+	10 mo.	10-25%	25-50%	Dulcolax suppository	Hirschsprung's disease?	1½ yr.	Fair to good
3. C. B. ♂	Several	2 mo.	+	14 mo.	3%	3%	Cathartic on alternate days	None	1 yr.	Fair to good
4. A. R. ♂	Absent	3 mo.	+	2 yr.	3%	3%	Enema occasionally 1st yr.; none, none	Mental retardation	2½ yr.	Good
5. T. W. ♀	Absent	Birth	+	2½ yr.	25%	10-25%	None	None	4 yr.	Excellent
6. T. G. ♀	Absent to 4 cm.	Birth	+	3½ yr.	10-25%	3%	None	None	9 mo.	Excellent
7. G. B. ♂	2 groups—prox. seg.	Birth	+	3½ yr.	10-25%	10-25%	Enemas initially, none past 2 yr.	Achondroplastic dwarf	2 yr.	Good
8. F. G. ♂	Absent	Birth	+	9 yr.	50%	10%	Occasional laxatives	Mental retardation	1½ yr.	Fair
9. L. L. ♂	Absent	Birth	mega-rectum	11 yr.	75-90%		Improved but required Swenson proced. after 1 yr.	Pica, megaloureter, megacystica	2 yr.	Poor
10. L. C. ♂	Absent	Birth	+	15 yr.	10-25%		Enemas & suppositories	None	1 yr.	Poor (x-rays show persistent narrow segment)
11. R. W. ♂	Degenerating ganglia, fibrosis of smooth muscle	6 yr.	mega-rectum	18 yr.	90%		Enemas & suppositories	Megacystica, hyposthetic rectum	9 mo.	Poor (continued fecal accumulation)

TABLE 2. *Clinical Findings in Patients Treated By Sphincterotomy and Rectal Myotomy Following Swenson's Procedure*

Name of Patient	Age at Swenson's Procedure	Symptoms	Age at Operation	Subsequent Treatment	Other Conditions	Follow-up	Results
12. M. F. ♀	1 yr.	Recurrent constipation after 6 mo.	1½ yr.	Colostomy for recto-vaginal fistula	Mongolism	2½ yr.	Poor nutrition
13. J. S. ♂	1 yr.	Constipation, enterocolitis	2 yr.	None	Sibling with Hirschsprung's disease	1 yr.	One episode of obstruction at 9 mo. requiring anal dilatation
14. N. S. ♂	2 yr.	Enterocolitis	3½ yr.	None	Sibling with Hirschsprung's disease	4½ yr.	Good with normal development
15. B. B. ♀	1½ yr.	Constipation, abdominal distention, fecal impaction	18 yr.	Daily Dulcolax suppository	None	8 mo.	Fair but requires suppositories for fecal accumulation & impaction

omy and rectal myotomy were carried out because of a suspected short aganglionic segment of rectum. In selected patients with chronic constipation this procedure was performed for both diagnosis and treatment. In patients having already undergone Swenson's operation, it was used as an adjunctive means of therapy.

Patients and Methods

(a) Clinical Findings

Eleven patients with chronic constipation but without evidence of "long" aganglionic segments of colon on barium enema x-ray, underwent sphincterotomy and rectal myotomy through a posterior approach (Table 1). Four patients with Hirschsprung's disease previously treated by Swenson's proctosigmoidectomy underwent similar operative procedures because of continued symptoms of either chronic constipation or intermittent large bowel obstruction or both (Table 2).

Of the 11 patients in the first group, all had chronic constipation of varying degrees. Patients in the older age range were more likely to have the classic symptoms or findings of Hirschsprung's disease but to be in good nutritional state. Fecal soiling was not observed and in all but one

patient constipation dated from birth or developed within the first 3 months of life. This is in contrast to the so-called "functional constipation" or "psychogenic megacolon," in which the onset more often occurs at the commencement of toilet training. In the infant, abdominal distention was common, as was some degree of undernutrition. Not infrequently there would be remission of symptoms with the child being managed by cathartics, enemas, and suppositories. Two patients with definite histories of delay in the passage of meconium proved to have aganglionic segments of sufficient length that ultimately required proctosigmoidectomy (Patients 1 & 9). None of the patients less than 6 years of age had a history of fecal impaction or enterocolitis. Despite the presumed short segment of aganglionic bowel, there was not the same type of episodic constipation, diarrhea, and bacterial enterocolitis that is sometimes seen after Swenson's procedure.²⁰

Patients' ages varied from 4 months to 18 years, with most being younger than 4 years. The incidence of associated disease seemed high, viz., two had mental retardation, one was an achondroplastic dwarf, one had associated pica (clay) and two had atonic bladders (one with associated

megaureter). There was no familial history. There were nine males and two females; two of the males later proved to have long segment involvement.

Surgical treatment was undertaken in these 11 patients to identify those whose constipation might be secondary to a short aganglionic segment. This was performed with the realization that there are a number of individuals with severe chronic constipation ("pseudo-Hirschsprung's disease") in whom the histologic findings vary from normal ganglia to hypoganglionosis.⁴

(b) Evaluation of "Short Segment Disease" by Barium Enema X-ray

The examination of the distal rectum to demonstrate a short distal aganglionic segment by barium enema x-ray requires a particular technic and even then may be difficult to evaluate. Cathartic cleansing of the colon is not attempted, although a small cleansing enema may be used. The barium mixture is suspended in isotonic saline to avoid water intoxication. A soft catheter or infant enema tip is gently inserted into the rectum without using a retention balloon of any type. In small infants appropriate restraints are employed. An AP survey film of the abdomen is made prior to the instillation of barium. For the contrast study, the patient is turned to a high right anterior oblique position in relation to the screen and the barium allowed to flow in very slowly. Spot films are routinely exposed in different degrees of obliquity as progressive filling of the rectum is accomplished. Once the dilated rectum and sigmoid are opacified it is unnecessary to fill the remaining colon.

Films that include an AP and lateral view are made and these are repeated after evacuation of the barium. All films are immediately reviewed to ensure technical perfection. If additional special views are required, they can be made at this time. A 24-hour and occasionally a 48-hour film is procured in order to evaluate retention

of barium. An alternative method of examination of the rectum and anus (anogram) is by the use of a thick barium mixture.

In the evaluation of these patients, the following measurements were made: 1) the width of the distal rectal segment proximal to the anal canal in both AP and lateral films; 2) the thickness of the presacral space to indicate hypertrophy of the rectal wall; 3) the greatest AP diameter of the rectum, and 4) the retention of barium on the postevacuation films, immediate and delayed. Comparable studies were performed postoperatively in most individuals. The findings in these patients were compared with normal children of the same age range.

Excluding the four patients who had had prior Swenson procedures and one with an obvious megarectum, all barium enema x-rays were interpreted as abnormal. Figure 1 shows the method and Table 3 the results of mensuration. In the normal group there was only one patient with a presacral space greater than 0.5 cm.; in contrast, the narrowest presacral space in the patients under study was 0.5 cm. The patient with rectosigmoid involvement had a presacral space of 0.2 cm. Although these observations are limited in number, this measurement appears to be of diagnostic value if no associated inflammatory lesion is present. The width of the rectum as measured on the lateral study is of less value and correlated better with the duration of the disease. There was a definite decrease in size of the lumen of the distal rectum and anal canal (transverse and sagittal measurements) in patients with aganglionic segments of bowel. The postevacuation films showed prolonged retention of the barium and both 24 and 48-hour films are of value. Another change noted but not measured was the posterior angle at the distal rectum. The smooth obtuse angle noted on the lateral films in normal individuals (Fig. 1a) due to the puborectalis

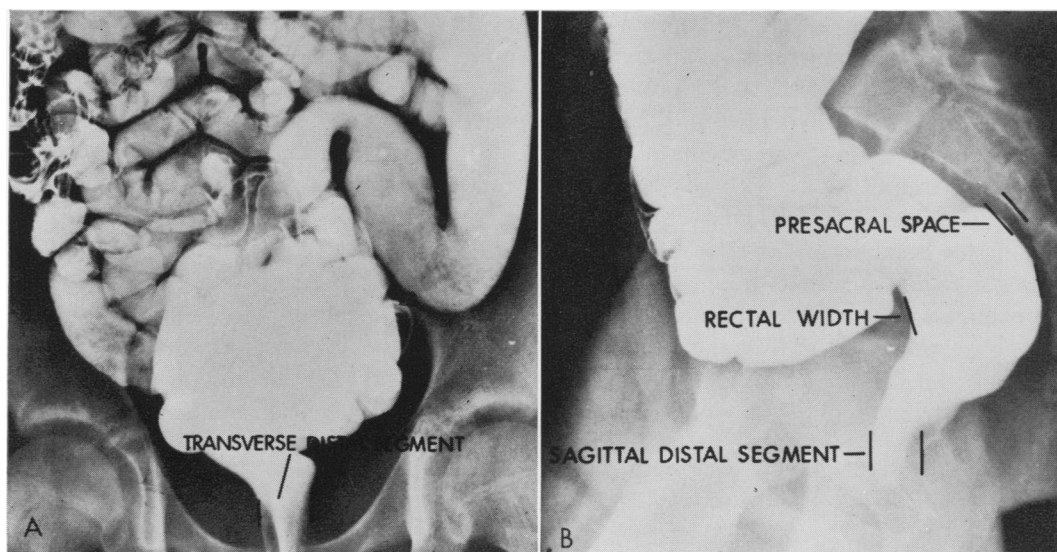


FIG. 1A and B. The normal rectum in this 4-year-old boy reveals a rectal width of 4 cm. with the distal segment showing a transverse diameter of 3.1 cm. and sagittal diameter of 3.2 cm. The presacral space posterior to the rectum measures 0.6 cm. Compare with Figure 5.

“sling”¹¹ was much more acute in patients with achalasia (Fig. 6a).

(c) Operative Procedures

The operation of posterior sphincterotomy and rectal myotomy was initially described in 1967,²¹ and has been modified only slightly since that time. A midline posterior incision is made between the tip of the coccyx and the anus immediately proximal to the mucocutaneous junction with division of the underlying ano-coccygeal

fascia and exposure of the puborectalis muscle (Fig. 2). One finger in the rectum facilitates identification of this muscle as well as the internal sphincter and muscularis propria of the proximal rectum. The puborectalis muscle is retracted distally to expose the internal sphincter which cannot be sharply distinguished at its transition into the smooth muscle of the distal rectum. Commencing at the cephalad margin of the external sphincter, parallel incisions are made 1–2 cm. apart—depending upon the circumference of the bowel—through the internal sphincter along the long axis of the rectum. They are carried proximally for the suspected length of the aganglionic segment. In the infant or young child resection of the coccyx has usually not been required to remove a 4 cm. segment of muscularis propria; in the older child or young adult, resection of the coccyx and occasionally a portion of the lower sacrum (Kraske approach) gives a more adequate exposure, permitting excision of an 8–10 cm. segment. The proximal extent of the incisions will depend upon the radiologic findings as well as the gross findings at

TABLE 3. Radiologic Findings in Normal Patients and Patients with Short Segment Hirschsprung's Disease

	Normal (cm.)	Short Segment Hirschsprung's Disease (cm.)
Presacral space	0.2–1.1 (Mean—0.46)	0.5–1.9 (Mean—0.95)
Rectal width	3.8–6.7 (Mean—4.6)	2.7–11.0 (Mean 5.8)
Distal segment		
Transverse	3.1	1.4
Sagittal	3.2	2.5
Retention	None	+

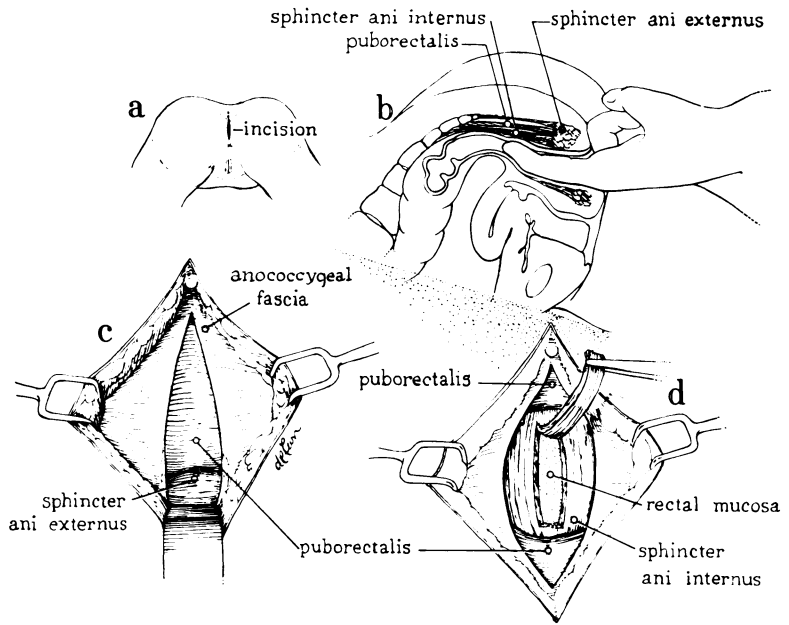


FIG. 2. a-d. Surgical approach and technic of sphincterotomy and rectal myotomy in the "short segment" Hirschsprung's disease.

operation.* The smooth muscle is carefully elevated from the underlying lamina propria and rectal mucosa between the parallel incisions. This muscle strip is severed distally and removed in a cephalad direction by sharp and blunt dissection, being guided by the finger in the rectum. Both longitudinal and circumferential layers of the muscularis propria are resected, maintaining the integrity of the rectal mucosa. Upon removal of the muscularis, the mucosa herniates in a manner similar to that seen accompanying the Heller or Ramstedt procedure. Appropriate preoperative preparation of the bowel allows immediate closure of the mucosa if it is perforated during dissection. In excising this segment of muscle, care needs to be exercised to maintain appropriate orientation of the specimen to permit an accurate evaluation as well as the precise location of any ganglion cells. A disposable muscle biopsy clamp has recently been described and seems ideal for this purpose.¹⁷ Examination of the

specimen by frozen section may facilitate differentiation of smooth and skeletal muscle as well as the proximal extent of the aganglionic segment.

Upon removal of the segment of smooth muscle, the puborectalis is returned to its normal position and the ano-coccygeal ligament is approximated with fine wire. Closure of the skin with fine subcuticular silk or wire sutures avoids the problem of maceration sometimes associated with percutaneous sutures. Should the mucosa be penetrated, antibiotic agents are administered and wound drainage instituted by either a suction catheter or small Penrose drain for 24 hours. A normal diet is permitted on the day following operation, with defecation usually occurring spontaneously within 48 to 72 hours.

Results

Of 11 patients operated upon for short segment disease, eight proved to have no ganglion cells and three had some ganglion cells demonstrable. In none of the three, however, was the complement thought to be normal. In the eight patients in whom

* In one patient a definite zone of hypertrophied muscle 3 cm. above the internal sphincter was clearly distinguishable.

no ganglia could be demonstrated, two had excellent results, i.e., normal bowel function without cathartics or enemas (Table 1—Patients 5 & 6). Two patients had good results, requiring an occasional cathartic or suppository but with no episodes of obstruction, impaction, or diarrhea. One child had a fair result, being improved but requiring a regular schedule of suppositories or cathartics. Three patients were unimproved. One of the latter was demonstrated to have a long aganglionic segment resulting in inadequate myotomy (Patient 1). He required proctosigmoidectomy because of persistence of symptoms. In retrospect, involvement of such a long segment should have been suspected on the basis of an onset in the neonatal period and moderately severe symptoms. The initial x-rays, however, did not clearly distinguish the extent of the disease. The other two patients (9 & 10) had short aganglionic segments by radiologic examination. The reasons for failure to improve are obscure. They may have had a transition zone with a poor propulsive proximal segment (see Discussion).

Of three patients in whom some ganglion cells were present, two had fair results. The etiology is not clear and they may represent individuals with hypoganglionosis. In one patient, the ganglion cells appeared in the extramuscular areolar tissue which may be an abnormal innervation. One patient who was unimproved was considered to have a functional type of constipation. However, "degenerated ganglion cells" and fibrosis of the smooth muscle of the sphincter were observed. He also had a lack of normal sensation in the anorectal canal. This patient, although not falling into any specific disease entity, must be considered to have an abnormal distal rectum and anal sphincter and may be benefited by proctosigmoidectomy.

In no instance was a patient worsened by the operative procedure.

Four patients were treated because of

continued symptoms following Swenson's operation. Two siblings have been essentially well, with normal bowel habits. One had two episodes of acute enterocolitis requiring hospitalization before sphincterotomy. He continues to maintain a semi-soft stool in his rectum but has no other evidence of obstruction. This patient's brother has had one episode of abdominal distention $4\frac{1}{2}$ years after sphincterotomy. A third patient with mongolism developed a recto-vaginal fistula 6 months after the procedure and currently has a colostomy. The fourth patient had her primary operation at 18 months. Sphincterotomy and rectal myotomy were performed at age 18. Although improved, she continues to accumulate large masses of feces and has required hospitalization for evacuation of the colon. This relatively poor result may be related to failure of normal propulsion in the proximal colon rather than failure of the sphincterotomy to give relief of obstruction. This patient's nutritional status and overall development are normal.

Complications peculiar to this procedure are wound infections and fistulas. The previous anastomoses in patients having undergone Swenson's procedure makes mucosal dissection difficult and predisposes to entering the bowel lumen. In 15 such patients, six had perforations of the mucosa during dissection. In these six patients, temporary fistula occurred in one and a superficial wound infection in another. In the remainder, the wounds healed primarily. One patient (M. F.) developed a recto-vaginal fistula 6 months after sphincterotomy. There had been no previous perforation and the reason for this late complication was not clear.

Case Reports

Case 1. W. S. A boy was evaluated at $3\frac{1}{2}$ months of age because of delay in passage of meconium and chronic constipation requiring cathartics, suppositories, and enemas. His development was otherwise normal (weight within 25 percentile and height at 3%). Barium x-ray ex-

amination of the colon disclosed a narrow distal segment with proximally dilated bowel. Posterior sphincterotomy and rectal myotomy was carried out at age 4 months. Ganglion cells were not present in the surgical specimen. The patient improved but required a Dulcolax suppository twice weekly. After two years he again developed severe constipation. Barium enema xray disclosed a long narrow segment which had not been initially appreciated (Fig. 3), and a Swenson procedure was performed. His immediate postoperative course was complicated by a breakdown of the anastomosis, requiring a transverse colostomy. This colostomy was closed at 2 months. Follow-up evaluation one year later disclosed daily spontaneous bowel movements but with encopresis.

Comment. In retrospect, the onset at birth should have suggested a length of aganglionic segment that might not be corrected by myotomy. On the other hand, this relatively simple operation provided the diagnosis as well as opportunity to be improved without colonic resection.

Case 2. M. S. This boy was seen at age 10 months with a history of chronic constipation requiring cathartic and enemas. His physical examination was normal. Barium enema x-ray suggested narrowing of the distal segment (Fig. 4). Because of his unexplained constipation a sphincterotomy and rectal myotomy were performed. Since some ganglionic cells were present in the

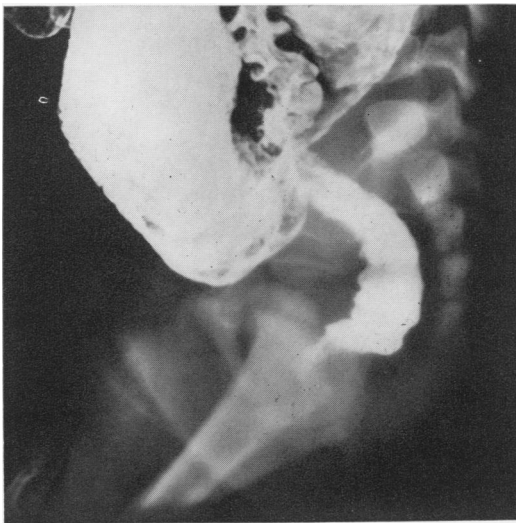


FIG. 3. This barium enema performed 18 months after myotomy disclosed a relatively long aganglionic segment and required proctosigmoidectomy for correction (Patient 1).

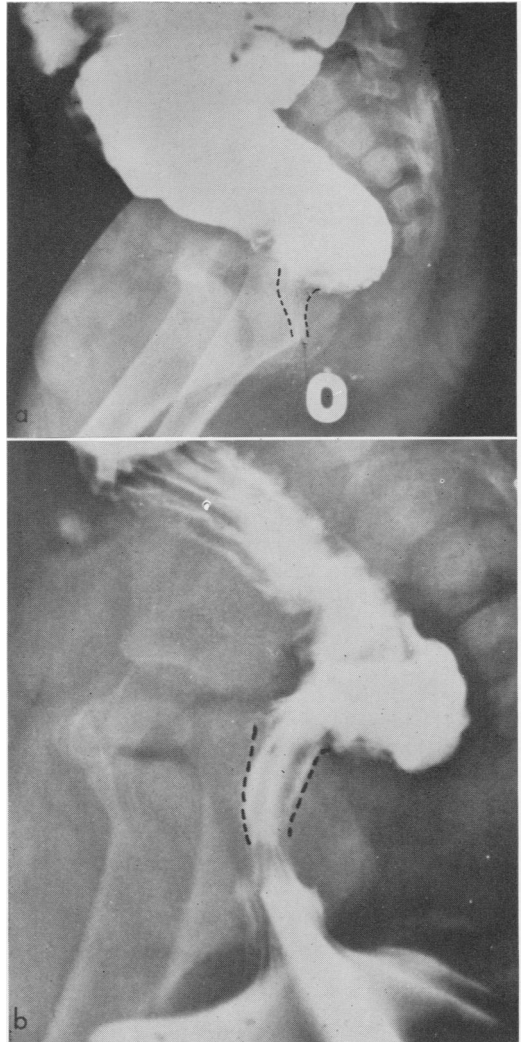


FIG. 4. The barium enema reveals narrowing of the distal rectal segment (a) and a more normal appearance (b) following sphincterotomy and rectal myotomy (Patient 2).

paramuscular and fibro-adipose tissue, a diagnosis of Hirschsprung's disease could not be established. The patient has done fairly well during the subsequent 18 months, requiring a suppository to induce defecation. Films procured one year postoperatively disclose widening of the previous narrowed segment (Fig. 4).

Comment. This may be an example of a variant of aganglionosis with faulty innervation. In this patient this procedure offered a more precise diagnosis and probably contributed to his improvement.

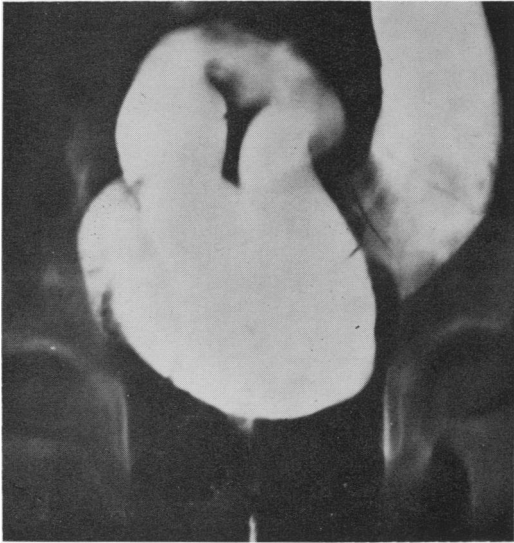


FIG. 5. Radiologic findings in this 2½-year-old girl indicated a narrow distal segment (Patient 5).

Case 5. T. W. A 2½-year-old girl was seen because of a history of chronic constipation since birth. Her development was otherwise normal, with weight and height being within the 25th percentile. On barium enema x-ray the distal rectum appeared to be narrow (Fig. 5). Following a posterior sphincterotomy and rectal myotomy the patient has done well with daily spontaneous bowel movements. She has been seen at follow-up for 4 years.

Comment. No ganglion cells were seen in the surgical specimen and this patient was apparently an ideal candidate for this operative procedure.

Case 6. T. G. At age 3 this girl had a history of chronic constipation since birth. She had required frequent laxatives and enemas. Barium enema x-ray showed a narrow distal segment (Fig. 6a, b). The postoperative course following a sphincterotomy and rectal myotomy was uncomplicated. Nine months following operation she had an excellent result with daily spontaneous bowel movements, an excellent appetite, and weight gain. Barium enema x-ray was normal with prompt evacuation of the barium within 2 hours (Fig. 6c, d).

Comment. This patient apparently had a very short aganglionic segment of bowel since ganglion cells were seen at the proximal margin of the 4 cm. surgical specimen.

Case 8. A 9-year-old boy had a history of chronic constipation since birth and mild abdominal distention. He was mentally retarded. Barium enema x-ray disclosed a very short narrow and marked increase in presacral space (Fig. 7). Posterior sphincterotomy and rectal myotomy was carried out. Postoperative course was uncomplicated. Evaluation at 18 months disclosed a history of spontaneous bowel movements on alternate days. There was mild abdominal distention but no accumulation of feces.

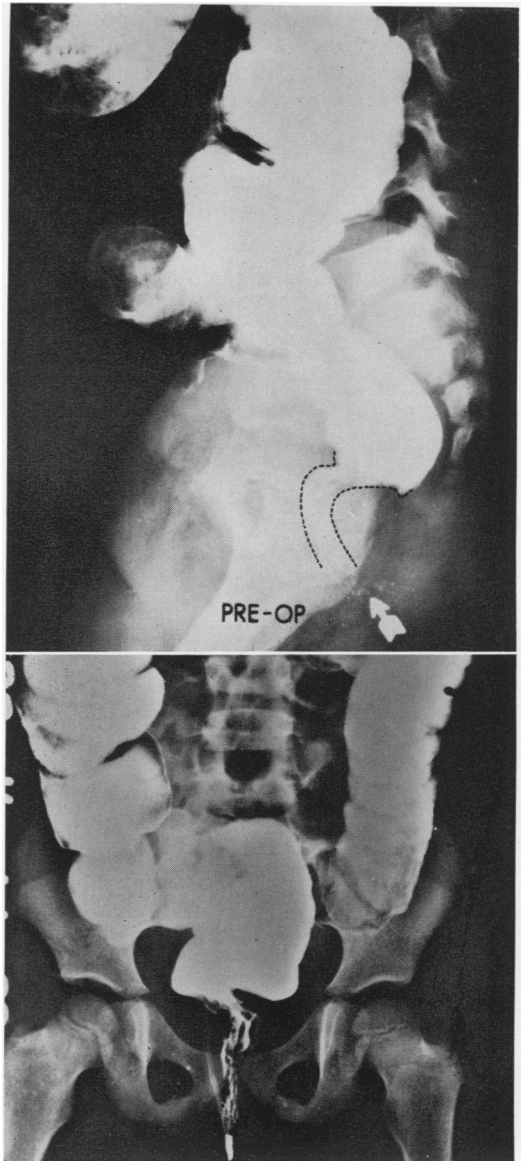


FIG. 6 A & B. The initial radiologic examination disclosed an abrupt transition from a rectum of normal calibre to a narrow segment.

Comment. This patient is improved although certainly not normal.

Case 9. L. L. This boy was 11 years old when first seen, with a history of chronic constipation and abdominal distention since shortly after birth. He had been managed with cathartics and enemas. Barium enema x-ray disclosed a very short (2 cm.)

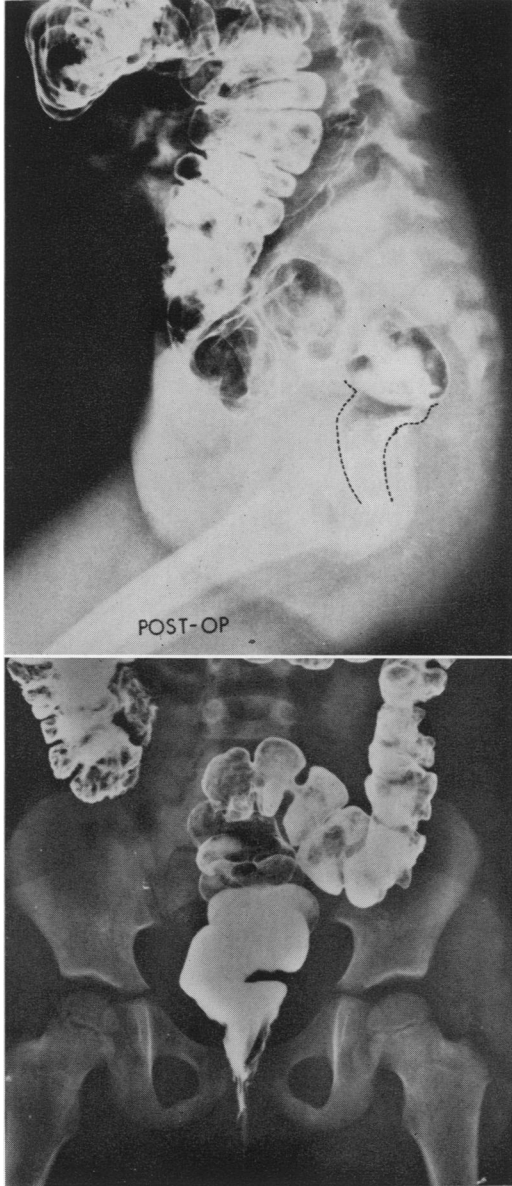


FIG. 6 C & D. Nine months following operation a barium enema was regarded as normal with good evacuation of the barium at 2 hours (Patient 6).²

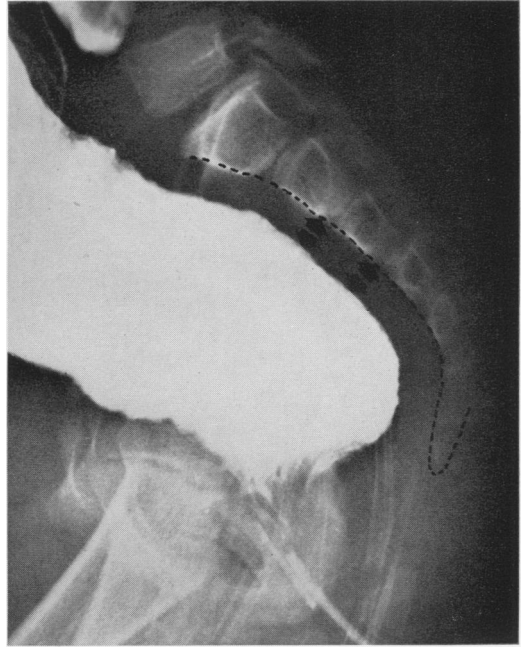


FIG. 7. A very short narrow segment with conspicuous increase in the presacral space was noted in this 9-year-old boy with chronic constipation since birth (Patient 8).

narrow segment (Fig. 8a). Intravenous pyelograms showed bilateral megaloureter and mild hydronephrosis. Following sphincterotomy and myotomy, this patient has continued to have chronic constipation with recurrent fecal impactions. Postoperative films at 3½ months show a widening at the site of myotomy but continued retention of barium (Fig. 8b). Complicating his course is a history of clay eating.

Comment. This child may have a longer aganglionic segment than anticipated or a transition zone which does not have normal propulsion. How much the pica contributes is unknown.

Case 11. F. W. An 18-year-old man was evaluated because of a history of chronic constipation since age 6, requiring cathartics and enemas. He also has urinary retention when constipated. Barium enema x-ray disclosed primarily a megarectum without a definite distal narrow segment (Fig. 9). A cystometrogram revealed an atonic bladder. Following posterior sphincterotomy and rectal myotomy he has continued to require suppositories and enemas. Pathologic examination disclosed degenerated ganglion cells in one area and definite fibrosis of smooth muscle.

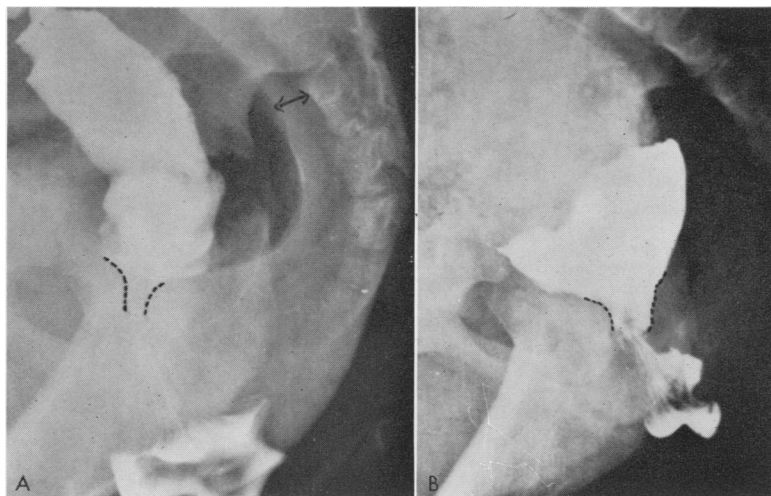


FIG. 8. Radiologic findings in an 11-year-old boy with megarectum and a very short aganglionic segment (A). Although the site of narrowing was corrected by sphincterotomy and myotomy this patient continues to have retention of feces as well as barium (B) (Patient 9).

Comment. This patient has an abnormal rectum and internal sphincter. The etiology, however, is unknown. It is probable that he will benefit from excision of the sigmoid colon and rectum.

Discussion

The complexities of diagnosis and treatment of aganglionic megacolon are illustrated in the management of patients with

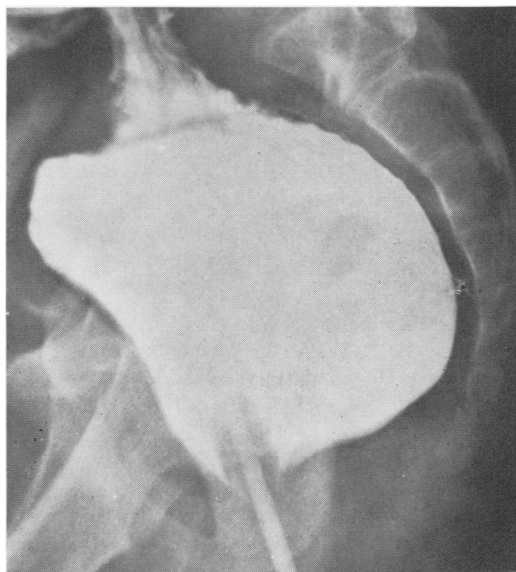


FIG. 9. Barium enema x-ray of megarectum without a definite distal narrow segment (Patient 11).

“short segment disease.” Since the diagnosis of Hirschsprung’s disease is so dependent upon anatomic findings, cognizance of the normal development and location of ganglion cells in the distal rectum and internal sphincter is essential. There is evidence that in the development of myenteric ganglia, neuroblasts, most likely of vagal origin, migrate in a cephalocaudal direction throughout the alimentary tract during the first 12 weeks of gestation.¹⁶ Any interference with such migration results in a segment of gut without ganglion cells, the length of involvement depending upon the time of “insult” during embryogenesis. Although migration of neuroblasts within the alimentary canal is complete at 12 weeks, there is progression of maturation in a cephalocaudal direction from early embryonic life throughout childhood. In the infant, the most distal internal sphincter, extending 1–1.4 cm. proximal to the anal valves (mucocutaneous junction) is without intramural ganglia.¹ This hypoganglionic zone may extend an additional 1–1.5 cm. in the child beyond one year. In association with this area of hypoganglionosis, there may be a number of non-myelinated nerves appearing similar to “hypertrophied nerve trunks” seen in the typical aganglionic segment of Hirschsprung’s disease.

Intramural ganglion cells are normally present 2–3 cm. proximal to the anal valves and number between 17–18 per sq. mm. The finding of ganglion cells in three patients (2, 3, 11) but without normal numbers suggests variations in the patterns of innervation midway between aganglionosis and a normal myenteric plexus. Whether this finding is related to bowel dysfunction will require more sophisticated methods of study, e.g., histochemical technics to identify both adrenergic and cholinergic fibers, their number and distribution, and, finally, clinical studies of recto-anal reflexes.

As evidenced from symptoms and results of treatment in these patients, as well as the observations of others, functional disturbances in bowel motility cannot always be correlated with the extent of the aganglionic bowel.^{5, 8, 14} Some differences in obstructive symptoms may be accounted for by variations in innervation and the relative number of ganglia (Table 4). Using histochemical technics it has been possible to demonstrate a variation in the number of cholinergic (acetylcholinesterase⁺), adrenergic nerves, and intramural ganglia in different segments of the bowel in patients with Hirschsprung's disease.^{2, 6, 10} The most distal aganglionic bowel characteristically has an increased number of cholinergic

fibers. The proximal segment of aganglionic bowel contains fewer than normal cholinergic fibers. This increased density of cholinergic nerves in distal bowel is in keeping with an origin from the sacral outflow and distribution in cephalad direction.⁵ The transition zone cephalad to this aganglionic segment may be of variable length, containing reduced numbers and size of intramural ganglia. Acetylcholinesterase⁺ (cholinergic) nerves are infrequent in this zone. Proximal to this zone, the colon has a normal complement of ganglia and nerves. Catecholamine fluorescent technics also demonstrate a variation in the number and distribution of adrenergic nerves.^{2, 6} However, patterns were less consistent and only a generalization can be made, viz., that the more proximal aganglionic bowel contains fewer adrenergic nerves than the distal.

Coordinated intestinal peristalsis appears to be more complex than the simplified concept of "parasympathetic-motor" and "sympathetic-inhibitory" action on smooth muscle. Changes in integrative activity secondary to malfunction of one or more of these components of the myenteric plexus provide a better explanation of the variation in symptoms and response to treatment than does the presence or absence of ganglion cells. Effective popul-

TABLE 4. *Distribution of Components of Myenteric Plexus—Hirschsprung's Disease*

Segment	Ganglion Cells	Cholinergic Nerves (Acetylcholinesterase +)	Adrenergic Nerves (Catecholamine Fluorescence)	Integrative Function
Distal aganglionic	0	++++	++	Tone ↑, aperistaltic
Proximal aganglionic	0	+	+	Tone ↑, aperistaltic
Transition zone	+	+—	+	Tone ↑, minimal propulsion
Normal colon	+++	++	++	Normal peristalsis modified from Garrett ⁸

sion is dependent upon a normal component of cholinergic nerve fibers as well as ganglion cells. The increased number of cholinergic nerve fibers in the distal rectum would account for the increased smooth muscle tone characteristic of this disease. The absence of ganglia and resulting loss of adrenergic nerve function prevents inhibition and relaxation of this segment of bowel. The resulting functional obstruction may be further aggravated by proximal transition zone in which ganglia and cholinergic fibers, though present, are reduced in number, thereby producing an ineffective propulsive segment. Symptoms, then, not only depend on the extent of aganglionic bowel but also upon the relative number and activity of cholinergic nerve fibers. These morphologic findings would explain not only the different clinical course but, in addition, the response to operative procedures designed to remove the aganglionic bowel or divide the circular muscle.

Patients with no ganglion cells and a great number of cholinergic nerve fibers may have severe symptoms early in life. The patient harboring a short aganglionic segment, with little increase in cholinergic fibers and a short transition zone, might be expected to have mild symptoms and would be expected to benefit from division of the tonically contracted segment of bowel. Conversely, the patient with a long transition zone with a short aganglionic segment, containing few cholinergic fibers, might also have mild symptoms but would not be materially improved with sphincterotomy and myotomy, due to the lack of an effective proximal propulsive segment. The failure of some patients to be relieved completely of symptoms after proctosigmoidectomy may be due not only to the length of the remaining aganglionic segment but also to the lack of normal number of cholinesterase⁺ fibers and ganglion cells in the proximal colon.

With these considerations, the operation

of myotomy becomes logical and reasonable, provided the aperistaltic zone is not unduly long and that there is a normal proximal propulsive segment. Unfortunately, these characteristics cannot always be fully identified by history, physical findings, or radiologic examination. The ideal candidate for myotomy has a short aganglionic segment, severe inhibition with an abrupt transition zone and a normally innervated proximal bowel. This would be suggested by radiologic findings of a short segment, immediate proximal dilatation and hypertrophy with increase in width of presacral space, and histologic findings of an abundance of cholinergic fibers in the aganglionic bowel.

The concept of correcting an area of achalasia by myotomy is not new, being proposed by Hurst¹⁰ and others, e.g., Martin and Burden¹⁵ and Saegesser.¹⁸ More recently, Bentley,³ Lynn,¹³ Gennaro and Bacon,⁷ and Thomas²¹ described successful results using this approach. The difference in their observations as compared with those of earlier authors probably represents patient selection based on a better understanding of the disease.

Chronic constipation in infancy and childhood is frequent and symptoms as well as physical and radiologic findings may not always identify an etiology. In Hirschsprung's disease, emphasis has been placed upon abnormalities of the myenteric plexus as an index of disease. Furthermore, at present, only gross neurological deficits, e.g., absence of ganglion cells, are recognized although variations of symptoms may be associated with the histologic findings of scanty or abnormal ganglion cells. Obviously, a better understanding will depend upon the availability of more sophisticated technics with documentation of abnormalities in rectoanal reflex mechanisms as well as changes in smooth muscle function secondary to alterations in cholinergic and adrenergic innervation. The approach recommended in this paper aids in identifying

a few patients with specific etiologies in whom a simple operative procedure may constitute both diagnosis and treatment. Furthermore, lack of improvement does not compromise the performance of a more extensive procedure later.

Summary

Experiences with sphincterotomy and rectal myotomy through a posterior incision have been reviewed in 15 patients undergoing this procedure because of probable short segment Hirschsprung's disease or residual symptoms following Swenson's procedure. The aganglionic segment may be suspected by carefully performed barium enema x-rays. This operative procedure may be used both as a method of diagnosis and treatment. In patients in whom there were no ganglion cells and the involved segment was relatively short and in whom the proximal segment had normal propulsion, results were good. In the event the aganglionic segment was longer than anticipated or the transition zone provided ineffective peristalsis, the performance of a more extensive procedure was in no way compromised.

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DISCUSSION

PRESIDENT YEAGER (Baltimore): I would like to comment on one unfortunate circumstance we had in our own city in reference to depending upon the presence or absence of ganglion in making this diagnosis. There was an infant operated on who had a Swenson pull-through procedure following

a diagnosis of Hirschsprung's disease. About 12 to 14 years later, when he was undergoing psychiatric evaluation, he was told that he had never had Hirschsprung's disease. Review of the slides showed ganglion cells. They had not been identified previously because of two factors—poor staining and, let's say, an inexperienced pathologist.