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RECTAL MYECTOMY FOR AGANGLIONIC MEGACOLON

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In the treatment of aganglionic megacolon, anterior resection of the rectosigmoid and variations of the widely accepted pull-through procedure have left a number of patients with unsatisfactory results. It is true that many of these patients are able to lead relatively normal lives with the use of various aids, such as special diets, laxatives, and periodic enemas. But in the more stubborn cases the result is eventually a failure, and an exasperated patient and his family seek additional relief, usually from other than the initial surgeon.

Having been exposed to my share of these cases, I have employed many methods of correcting unsatisfactory initial procedures. These methods have ranged from sphincterotomy to a redo of the original pull-through operation. Much thought has been given to this problem in an effort to provide permanent relief without resort to a major abdominal operation.

ILLUSTRATIVE CASES IN WHICH RECTAL MYECTOMY HAS BEEN USED (TABLE)

Case 1.—In August 1964 a 10-year-old boy was seen who had the typical story of constipation since birth. The diagnosis of aganglionic megacolon had been made at 1 month of age at another center, and his condition had been controlled with laxatives and enemas for 1 year.

When he was 1 year old, an anterior resection of the rectosigmoid had been performed in an attempt to relieve the constipation. Ganglion cells were present in the proximal 18 cm of the specimen and absent in the distal 7 cm. Postoperatively, the infant had a stormy course, but he was finally dismissed after about 7 weeks; the prescribed regimen included rectal irrigations and digital examinations, as well as the use of milk of magnesia and mineral oil three times daily.

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Myectomy for Aganglionic Megacolon

Patient	Age at original operation; sex	Age at myectomy, yr	Follow-up, mo	Result
Initial anterior resection				
1	1 yr; M	10	18	Excellent
Initial abdomino-perineal pull-through procedure				
2	5 mo; M	11	33	Good
3	11 mo; M	10½	8	Definite improvement
4	13 yr; F	17	7	Excellent
No previous corrective surgery; short segment				
5	F	4½	15	Satisfactory (improved)
6	F	3	9	Excellent
7	M	9	5	Much improved
8*	M	6	3	Excellent
9	F	16	10 days	Satisfactory

*This patient did undergo colostomy at 1 month; the stoma was closed when myectomy was performed.

The subsequent history included relatively good periods of bowel movements every other day, but there were also episodes of constipation, diarrhea, and abdominal distention. The parents were cooperative and conscientious in their attention to the child's toilet training and medication. In spite of this, the boy was brought back to this clinic because of poor appetite, inadequate weight gain, and infrequent stools which were usually watery or loose and foul-smelling.

X-ray studies showed pronounced distention of the remaining colon, and proctoscopic examination revealed marked dilatation starting above the dentate line. No line of anastomosis could be identified. On digital examination the internal sphincter seemed tight but not remarkably so. Although there was no emergency, the parents did feel that further surgical treatment was necessary. An operation was therefore planned.

The surgical alternatives seemed to range from a sphincterotomy to an abdominal perineal pull-through procedure of the Duhamel type. The sphincterotomy seemed inadequate treatment since there was obviously malfunctioning bowel above and the sphincter did not seem unduly constricted. The Duhamel procedure seemed to offer a better chance of cure but represented a major operation and prolonged hospitalization since it would involve resection of the rectosigmoid at a low level, closure of the rectal stump, and implantation of normal proximal colon into the posterior wall of the rectal ampulla. A compromise was decided on. This decision was based on experience with a similar situation

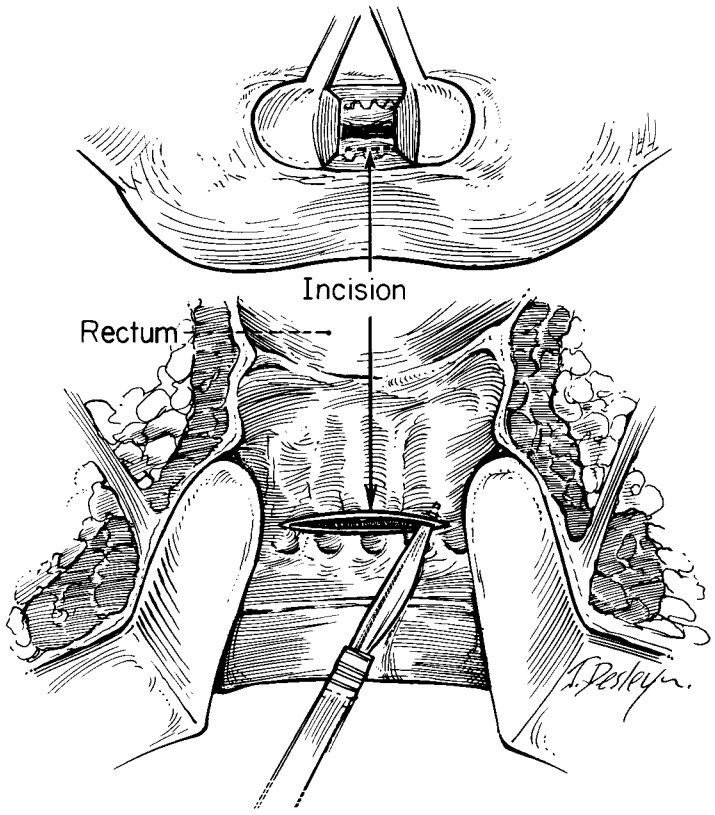

Fig. 1. Location of incision.

in an 11-year-old boy (case 2, Table) who had undergone a pull-through procedure at 5 months of age. That patient had received marked relief from division of a ring of scar tissue at the anastomosis, and longitudinal division of muscular fibers above and below the anastomotic site.

With the use of inhalation anesthesia, the anus was dilated and 2 quarts of enema fluid were siphoned off. Traction sutures were placed just above the dentate line, and a transverse incision was made between them involving the mucosa and submucosa (Fig. 1). It was then a simple matter to undermine with dissecting scissors and separate the submucosa from the underlying muscularis. A narrow strip of the muscle layer was excised for a distance of approximately 8 cm up the midline posteriorly (Fig. 2). The pathologic report was of smooth muscle with multiple large nerve fibers but no evidence of ganglion cells. Hemostasis was easily attained. Interrupted fine catgut sutures were used to close the defect

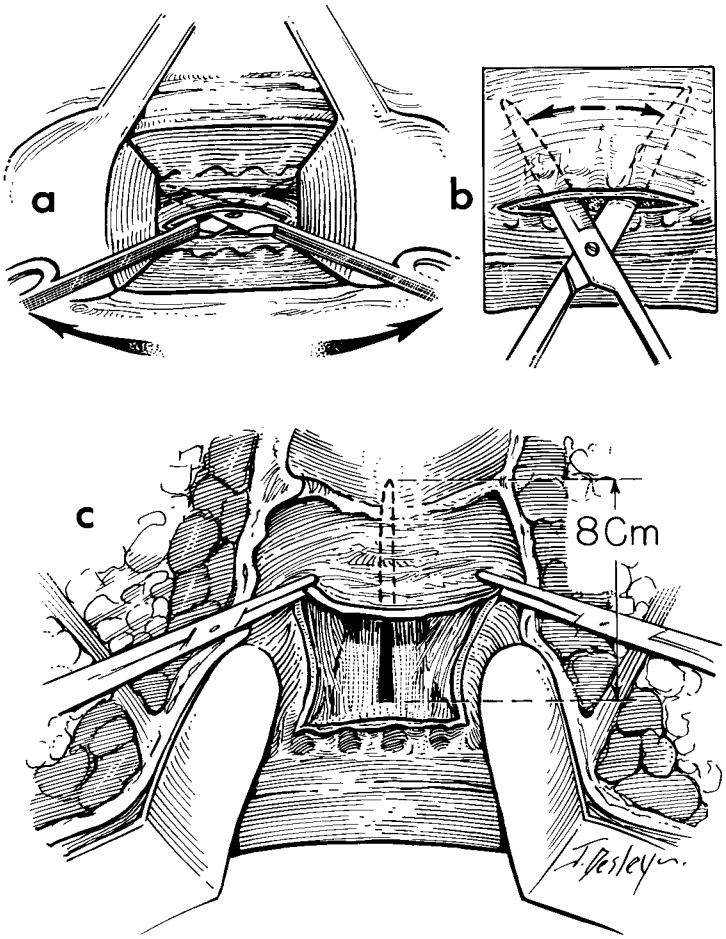

Fig. 2. Method of dissection and myectomy following identification of proper layer by frozen section.

in the mucosa. A small gauze pack was then placed in the canal.

The following morning the patient was dismissed from the hospital, and he travelled 350 miles to his home. His recovery was uneventful.

Although 2½ months was obviously not an adequate follow-up, a letter from the parents at that time stated that they were very pleased with the child's progress. He was feeling better than he had ever felt before and had not missed a day of school since the operation. He was moving his bowels at least once daily, with no difficulty and without resort to laxatives. He had gained 7 lb.

This improvement has been maintained over the past 18 months, and the parents consider him completely normal.

Case 5.—In November 1964 a 4½-year-old girl was seen at this clinic with a history of gastrointestinal difficulties since birth. During the first day of life she had sucked poorly, vomited, and had multiple small bowel movements. She had become so dehydrated that she was readmitted to the hospital at 10 days of age and remained there for 1 week. Constipation gradually developed and was severe by 2 months of age. Suppositories, enemas, laxatives, and formula changes were employed but with little success. Vomiting and abdominal distention were common, and she never had a normal bowel movement. At 13 months of age she was hospitalized for 8 days, during which time the dehydration was treated and the fecal impaction relieved. Thereafter, enemas given at least every other day were sufficient management. There was no soiling.

On examination she appeared thin, pale, and listless; her abdomen was markedly protuberant. She walked with a peculiar gait in which she leaned back and moved her legs in a rolling manner. On rectal examination a hard fecal mass could be reached with the tip of the examining finger.

She was admitted to the hospital and was subjected to extensive and intensive treatment for the impacted megacolon. After a week of cleansing, roentgenograms of the colon showed a short, constricted lower segment and a tremendously dilated proximal portion of the colon.

In view of the low-lying fecal mass on initial examination and the x-ray findings, rectal biopsy was advised. The possibility of a rectal myectomy rather than a more orthodox abdominal operation was discussed and accepted by the parents with the understanding that a major operation might be necessary in 6 months. The rectal biopsy showed aganglionosis. Rectal myectomy was performed, extending 10 cm above the pectinate line. No ganglion cells were encountered anywhere in the specimen. It was the surgeon's impression that this was as high as it was safe to excise from below. The mucosal incision was closed with three catgut sutures, and a gauze pack was placed for temporary pressure. The patient made an uneventful recovery.

At follow-up 2 months after the operation the parents felt that there was real improvement in the child's bowel function, appearance, and disposition. Some abdominal fullness was still noted on physical examination, but nothing comparable to that seen in the preoperative state. On rectal

examination there was no evidence of any constriction in the canal. The internal sphincter seemed only slightly more prominent and firm than normal, and there was soft fecal material within easy reach of the examining finger. However, the mother had been using some laxatives to maintain the daily pattern of bowel movement.

Since only time would tell if this had been a worthwhile procedure, it had been presented to the parents in just that light. A Duhamel operation had been described, and the possibility of the need for such treatment had been accepted by the parents. One year later the parents were pleased with her progress and felt that the child's personality had become much more outgoing and pleasant. With increased experience it has become obvious that the dissection in this case could have extended to a higher level.

Case 6.—The most dramatic case of this series was that of a 3-year-old girl who had roentgenographic and histologic evidence of a short segment of aganglionosis. In her mother's words, she was a normal child in 24 hours. In that patient the aganglionic segment was approximately 8 cm in length, and normal ganglion cells could be identified above this level.

Case 9.—This was a mentally retarded bed patient in whom myectomy, at the age of 16 years, extended a distance of 13 cm. The aganglionic segment represented approximately 8 to 9 cm of this distance, and the upper 4 cm showed normal ganglionic cells. It has been only 10 days since this procedure was carried out and therefore no true evaluation can be made in this case.

COMMENT

In the nine patients for whom rectal myectomy has now been employed at the Mayo Clinic, all have shown immediate dramatic improvement, except possibly one (case 5). In this patient the aganglionic segment was undoubtedly longer than x-ray studies and digital examination led us to expect. Even in this case, however, the change has been sufficiently great so that the parents believe that no further surgery is necessary.

Rectal myectomy has proved of value in at least three situations: (1) patients previously treated for aganglionic megacolon by low anterior resection; (2) patients previously treated for aganglionic megacolon by an unsuccessful pull-through procedure; and (3) patients with primary aganglionic megacolon whose history and x-ray studies indicate aganglionosis of a short terminal segment of rectum which is confirmed on biopsy.

Undoubtedly some variation of this procedure has been used by many surgeons. Bentley[1] and Dennison,[2] in Glasgow, have used it with success; in cases of failure they have been able to perform an abdominoperineal pull-through operation. In their cases, the mucosa and submucosa were incised along with the muscularis, and the defect was resutured with a running catgut suture.

The procedure is no more complicated than an adequate rectal biopsy once the technique has been mastered. It appears advisable to identify the muscular layer properly by frozen section before developing the plane of dissection cephalad. This is particularly important in the infant or small child who does not have a markedly hypertrophied muscular layer.

The small mucosal incision is now placed below the dentate line and seems to be just as satisfactory as above the line. This probably ensures partial division of the internal sphincter muscle.

Even with relatively short periods of follow-up (3 to 33 months) it is evident that dramatic relief is available with this procedure in instances when either the choice of initial operation was unsuitable or the execution was inadequate. There have been no complications.

The fact that rectal myectomy has been employed successfully as the primary procedure in five patients with aganglionic megacolon does not mean that this procedure is being offered with unqualified endorsement. Prolonged follow-up is necessary, and more cases in this highly selective category are awaited. Since dilatation of the rectum is such an essential part of this procedure, the possibility cannot be ignored that the effects of the dilatation play some role in the dramatic response of these patients immediately after operation.

SUMMARY AND CONCLUSIONS

Rectal myectomy has been employed with considerable success in nine patients who had aganglionic megacolon. This procedure is particularly of value in patients who have had a poor response to low anterior resection or a pull-through procedure; it appears to be effective too in patients with primary aganglionic megacolon who have only a short terminal segment of aganglionic colon. Relief has often been dramatic, and there have been no complications.

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