The Surgical Management of Hirschsprung's Disease

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Etiology, Early History

Aganglionic megacolon, or Hirschsprung's disease, is characterized by constipation dating from birth. The symptoms of constipation or obstruction are due to a lack of parasympathetic innervation resulting from congenital absence of nerve ganglia of the myenteric and submucosal plexuses of Auerbach and Meissner. The aganglionic segment begins at the anus and extends proximally for a varying distance. In most cases of Hirschsprung's disease, the aganglionic segment involves only the rectum and the lower sigmoid colon. Occasionally, the aganglionic segment may involve most or all of the colon and, rarely, the lower small intestine.

The first description of Hirschsprung's disease is usually credited to a Dutch surgeon, Frederici Ryuschii, who published a detailed report in Latin in the seventeenth century (Benson et al., 1962; Jayle, 1909). Hirschsprung's Classical Paper was delivered before the Berlin Congress for Children's Diseases in 1886. He considered the dilated colon to be the primary cause, although he did mention that the rectum was not dilated and seemed slightly narrower than the normal rectum and that the mucosa of the colon was ulcerated, inflamed and edematous (Hirschsprung, 1887).

The true etiology remained generally poorly understood in spite of correct observations of several authors dating from 1901, indicating the primary defect. Swenson and Bill (1948) have described a sphincter-saving curative proctectomy removing the aganglionic segment of the colon. This operation was a significant milestone in the treatment of this previously incurable disease. However, it is associated with a high instance of postoperative complications. Several authorities have expressed concern regarding long-term complications of proctectomy for this as well as other diseases (Clausen and Davies, 1963) and have expressed a preference for preservation of the rectum in this histologically benign condition (Duhamel, 1960; Martin and Altemeier, 1962). A low anterior abdominal resection has been preferred by some (State, 1952). Others have proposed a rectosigmoid myotomy similar to the Ramstedt procedure and Heller operations (Martin and Burden, 1927). Duhamel (1960) has described a procedure which consisted of excluding the rectum, leaving it in place, and establishing an oblique, end-to-side anastomosis at the skin level. This procedure avoided the complications of proctectomy and, at the same time, the anastomosis was low enough to avoid residual symptoms of Hirschsprung's disease. There were some complications relating to accumulation of stool in the blind rectal stump. Also, the sensory urge to defecate, derived from the tense, filled rectum, was lost, since the rectum was completely bypassed by the procedure. This operation was further modified by lengthening the anastomosis as a long, side-to-side attachment of the colon to the rectum (Martin and Altemeier, 1962). Soave (1964) has recently described

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an operation which preserves the muscular wall of the rectum but removes the mucosa and does a pull-through procedure, bringing the ganglion-containing colon through the muscular sleeve of the rectum, which is kept in place.

Operative Procedure

The operation which we now employ is a modification of the above procedures (Martin and Caudill, 1967). The rectum is left in place but is completely included in the fecal stream, so that the sensory function is preserved. Dissection within the pelvis is minimal, thus reducing the risk of injury to the small nerve fibers going to the bladder and to the ejaculatory mechanism. The anastomosis can be performed at a low level without disturbing the internal sphincter.

The operation (Fig. 1) is carried out with the patient in the lithotomy position. The abdomen is opened through a generous, left paramedian rectus-retracting approach. The rectum is divided just above the peritoneal floor, with the anastomosis clamps being placed obliquely, preserving a longer segment of rectum anteriorly. Resection of the aganglionic sigmoid and descending colon is then carried out proximally to a level where adequate numbers of ganglion cells are identified microscopically on frozen section by the pathologist. Following resection, the end of the colon is then closed and inverted with interrupted silk sutures. The ends of the sutures are left long so that they may be grasped with the clamp to be subsequently inserted through the rectum.

The presacral space is next opened and dissection carried downward in the midline posterior to the rectum, using the finger to facilitate dissection until the level of the pelvic diaphragm is reached.

The surgeon then proceeds to the perineal portion of the operation. The anus is gently dilated and the rectum thoroughly irrigated with sterile saline solution followed by a mild antiseptic. Employing the Bovie electrosurgical unit, a transverse incision is made around the posterior half of the rectal wall at the apex of the anal crypts. The wall of the rectum posteriorly is dissected gently from the levator muscles, and the presacral space previously dissected from above is then entered. A long, curved hemostat is inserted from below into the presacral space, and the lower end of the colon is grasped and drawn through the wound in the posterior rectum. Several fine catgut sutures are placed about the circumference of the opening of the posterior rectal wall, and an open, end-to-side anastomosis is created from the end of the colon to the posterior wall of the rectum. After the sutures closing the end of the colon have been removed, a long spur-crushing clamp of the Mikulicz variety is inserted, with one prong in the rectum and one in the colon, and a clamp inserted the full length of the rectal stump.

The surgeon then returns to the abdominal part of the operation, and an open anastomosis is carried out betweeen the end of the rectal stump and the side of the adjacent

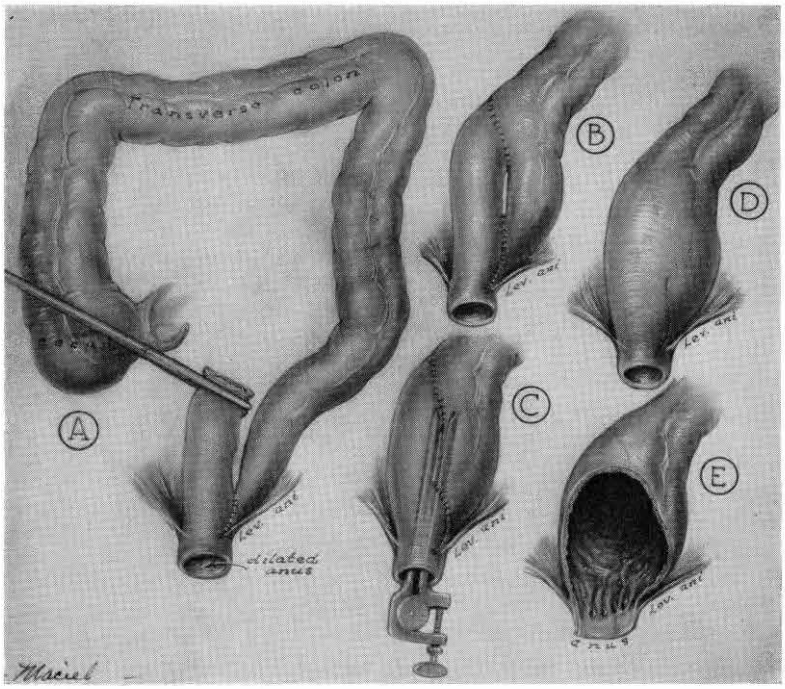

Fig. 1—A. Following resection of the aganglionic portion of the colon, the proximal end is placed in the presacral space and the end of the colon anastomosed to the posterior wall of the rectum 0.5 cm proximal to the mucocutaneous junction through an incision in the posterior wall of the rectum made transversely through the dilated anus.
B. Anastomosis from the proximal end of the rectum to the side of the adjacent colon is created, employing the open anastomosis technique.
C. The remaining colo-rectal septum is obliterated by means of a spur-crushing clamp which is inserted from below by an assistant while the surgeon is completing the proximal anastomosis. This permits placement of the clamp under direct vision before the anterior row of the anastomosis has been completed. The clamp is tightened in position and obliterates the remaining colo-rectal septum within a period of three to five days.
D. The appearance of a completed anastomosis.
E. The interior of the completed anastomosis.

colon. After the posterior half of the anastomosis has been completed, the spur-crushing clamp, which was previously inserted through the perineal approach, is inserted further by the assistant and placed under direct vision by the surgeon, so that the clamp includes the entire length of the septum to assure its complete ablation. The anterior half of the anastomosis is completed in the usual manner. If possible, the peritoneal floor is repaired above the anastomosis. A proximal diverting colostomy has generally been established previously and is considered advisable in most cases.

Results

This operation has been employed in 17 children with Hirschsprung's disease. Their ages were from one to four years. All have been followed up at regular intervals, with the longest follow-up being four years. One child developed mild pelvic cellulitis, fever and leukocytosis following discharge from the hospital. The cellulitis cleared promptly with oral antibiotics. Three children developed mild abdominal distention and diarrhea, interpreted as enterocolitis, one to three months following closure of the colostomy. Their symptoms cleared following anal dilatation. Mild constipation in two children has required occasional medication. None have had problems related to accumulation of stool in the rectum. The degree of bowel control has been most gratifying. The children have from one to three bowel movements daily, and their underclothing remains clean during the interim. Clinically, the results of this operation have been satisfactory.

Summary

The operative technique which we have employed for 17 children with Hirschsprung's disease is presented. The procedure offers certain advantages over the original Duhamel operation and over the classic Swenson operation. It can be recommended as a safe operation for Hirschsprung's disease, and, in our experience, has given satisfactory results.

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