Congenital Aganglionic Megacolon

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The present concept of the cause of congenital aganglionic megacolon (Hirschsprung’s disease) is the result of half a century of studies beginning with the pathological reports by Tittel\(^6\) (1901) and culminating in the widespread acceptance of the experimental and clinical studies of Swenson and co-workers\(^3, 4, 5\) (1948-1949). This concept assumes the presence of an area of colon (usually rectosigmoid) in which the ganglion cells of the myenteric plexuses are congenitally absent, resulting in a lack of coordinated peristaltic activity. The dilation and hypertrophy of the proximal colon is a compensatory phenomenon. Swenson revived and applied to this problem a procedure consisting of coloproctectomy with reconstruction of the alimentary tract by a “pull-through” (1948) and has since performed over one hundred such operations. Pull-through procedures were first performed in the Los Angeles Children’s Hospital in 1949. The first patients included five children with long standing megacolon. Fourteen additional patients were subjected to resection during the subsequent five years.

The clinical material from this pediatric center was studied with emphasis on the following: (a) An evaluation of the pull-through procedure and its modifications in the treatment of megacolon and (b) a study of this disease as it appears in early infancy. The operative cases from 1949 through 1954 form the basis for the former evaluation, (a); and all neonatal cases from 1943 to 1954 were surveyed in the later study (b).

EVALUATION OF PULL-THROUGH PROCEDURE

Operative Clinical Material

Nineteen children with aganglionic megacolon were treated by coloproctectomy and pull-through reconstruction or a similar procedure (total of 23 operations) during the five-year period 1949-1954. The diagnosis was established by pathological examination of the operative specimen in all cases and confirmed by subsequent autopsy in four cases. Fifteen male and four female children were included in the operative group. The age at the time of the first hospitalization for megacolon varied from two days to 15 years (average 2.8 years). The age at the

- Twenty-one pull-through procedures for congenital aganglionic megacolon (Hirschsprung’s disease) have been performed at the Los Angeles Children’s Hospital since the adoption of the etiological concept of a distal aganglionic segment in 1949. In 14 cases the Swenson procedure as modified by Hiatt was employed, with perineal excision of the colon segment. There were four postoperative deaths and three symptomatic recurrences in this group. Three patients were treated by transabdominal resection of colon and rectum with subsequent pull-through reconstruction (Swenson). Anterior resection (State) was carried out in two cases. Three children with recurrence of symptoms following primary operation were subjected to a secondary pull-through procedure with an eventual successful outcome. The major portion of the postoperative mortality (29 per cent) in this group occurred in infants less than six months of age in whom anastomotic disruption or proximal segment infarction occurred after operation.

A study of 31 cases of congenital aganglionic megacolon in very young infants drew attention to the difficulty of establishing a diagnosis in this age group even at exploratory laparotomy. Among these infants the mortality rate was excessive, regardless of the form of therapy employed. Colostomy appeared to be the indicated surgical procedure if a conservative regimen failed to control intractable colonic obstruction during the first year of life.

History and Physical Findings

Constipation was the universal principal symptom. A history of low intestinal obstruction dating from the first six months of life was obtained in all cases. Bowel evacuation problems in the postnatal month were described in 17 children; and approximately 50 per cent of the entire group were said never to have had a spontaneous bowel movement. Children who had defecation without enema (approximately 50 per cent) usually passed a massive stool one or two times each week, particularly after the first year of life. The sand-like nonfecal character of the stools of these children was unique.

Vomiting occurred during obstructive episodes in
TABLE 1.—Congenital Aganglionic Megacolon. Results of Operative Procedures Employed (1949-1954)

<table>
<thead>
<tr>
<th>Type of Operation</th>
<th>Total No.</th>
<th>Postoperative Deaths</th>
<th>Second Operation Required</th>
<th>Ultimate Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Perineal colon resection (pull-through)</td>
<td>14</td>
<td>5</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>2. Abdominal colon resection (pull-through)</td>
<td>3</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>3. Lower anterior resection</td>
<td>2</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>19</td>
<td>6</td>
<td>4</td>
<td>1</td>
</tr>
</tbody>
</table>

eight children. Diarrhea alternated with constipation in five cases, and in four the abdominal distention was so great as to cause respiratory embarrassment. When the home care was energetic—daily effective enemata—the more striking symptoms were usually absent after the first year.

Severe malnutrition with retardation of growth was noted in three children. No definite evidence of malnutrition or retardation of any type could be detected among the remaining patients who survived infancy. Three major associated congenital anomalies (mongolism, patent ductus arteriosus and congenital hydronephrosis) were present in the cases in which autopsy was done. The physical findings associated with congenital aganglionic megacolon were unique only among the older children with low intestinal obstruction in whom the thick-walled, dilated colon could be readily palpated.

Roentgen Studies

Roentgenographic visualization with barium enema in all cases (19 children) showed dilation of the proximal colon with characteristic narrowing in the lower rectosigmoid or upper rectum to the caliber of normal bowel. In two infants this vital diagnostic feature was at first equivocal, although it became definite before the age of one year. The distal extent of the aganglionic segment was incorrectly predicted by barium enema studies in two instances, resulting in incomplete rectal resection. Following a successful pull-through procedure, x-ray studies uniformly showed a return of the remaining normal colon to normal caliber and configuration. Plain films of the abdomen did not aid in distinguishing congenital aganglionic megacolon from other forms of subacute intestinal obstruction in early infancy. Retention of a mass of barium given as an enema, retained and desiccated, was a major complication in the preoperative preparation of two children.

Operative Findings

At the initial abdominal exploration the extent and degree of colonic dilation was extremely variable. The difficulty in recognizing the disease in early infancy will be discussed in the subsequent section. Following colostomy the bowel returned to normal caliber and appearance when seen at subsequent abdominal exploration (two cases). In the older infants and children in the present series the conventional dilation from the midtransverse colon to the rectosigmoid was noted in five cases. In addition to the conventional pattern, the ascending colon was dilated in five additional patients. The sigmoid alone was the site of dilation in three children. "Skip" areas of dilation in both right and left colon were noted in one infant. In one the dilation extended into the rectum but not to the anal sphincter.

The average length of colon resected was 18.1 cm. This average included several minimal resections performed early in the series. In later years, resections were between 20 cm. and 30 cm. in length and included the major portion of the descending colon, sigmoid colon, and rectum.

Operative Procedures

Three operative techniques were employed in this series. Swenson’s procedure as modified by Hiatt was used in 14 cases. The rectum and colon were resected from below following complete intussusception of the distal bowel, and the anastomosis was made at the perineum, and inverted. There were four postoperative deaths and three recurrences in this group (Table 1).

In three cases a transabdominal resection of the dilated colon (Swenson) was performed, followed by intussusception of the distal stump and anastomosis at the perineum. One patient died after operation. There were no recurrences.

In two cases a low anterior resection (State) was performed. In only one of these was a complete left colectomy performed, however. One recurrence occurred; it was successfully treated by a pull-through procedure.

Since 1953, serial “frozen sections” have been taken at the operating table to determine the extent of the aganglionic area. At first both the proximal and distal ends of the proposed segment for resection were examined for the presence of ganglion cells in the myenteric plexuses. Recently the dissection has been carried distally to within several centimeters of the anus (region of the internal sphincter) without regard to the presence of ganglion cells in the rectal wall. There has been no recurrence of obstructive megacolon since the introduction of this technique. All patients with previous...
incomplete resections were relieved of symptoms by the resection of an additional segment of the colon or rectum.

A series of "step frozen sections" are taken at intervals of 5 to 6 cm., beginning in the dilated rectosigmoid colon. An average of six such sections were required. Two techniques for excision of tissue for "frozen section" study were employed. In one of them, a small segment of the full thickness of the colonic wall was removed, while in the other a segment of the muscular layers was excised without opening the mucosa.

An attempt was made to sterilize the colon in preparation for operating by giving antibiotics or chemotherapeutic agents by mouth. Succinyl sulfathiazole was used in early cases, later the tetracycline drugs, and in the more recent procedures neomycin, 250 mg. every six hours for three days.

Preliminary Colostomy

Ten patients with congenital aganglionic megacolon required colostomy decompression (appendicostomy in one case) because of failure of enemata to relieve obstruction. The oldest infant in this group was four months of age. In all other infants and children decompression and preparation for operation was carried out without need for colostomy.

Four of the patients died following colostomy, the oldest at four months of age. Two children are awaiting definitive operation with well functioning colostomies. Attempt was made to perform pull-through procedures following colostomy in three patients less than six months of age, and two of them died (see Table 3).

Postoperative Deaths and Complications

There were six postoperative fatalities and four additional major, nonfatal complications following these 23 operative procedures. In three cases death followed local infarction of the proximal segment of colon or was associated with apparent anastomotic disruption. The unique aspect of this group was the extraordinarily low age at which the pull-through procedure was attempted (three weeks, three months, and five months of age). There seemed to be no other common factor. Three additional complications in three children included: (a) Stricture at the site of anastomosis, (b) inadequate blood replacement, and (c) intestinal obstruction secondary to postoperative adhesive bands. The first two complications mentioned were fatal.

Obstructive symptoms recurred in four children, usually beginning within a month after the initial pull-through procedure. One died as a result of the complications of intestinal obstruction before a second resection could be performed. The three others were successfully treated by additional resection of colon or rectum. Four patients with mild anastomotic stricture responded to digital dilation and required no further operation.

ACUTE INTESTINAL OBSTRUCTION IN EARLY INFANCY

Among 31 patients with congenital aganglionic megacolon (operative and nonoperative) seen in this hospital since 1943, 26 required hospitalization for intestinal decompression during the first six months of life. The initial operative procedure or medical management was carried out in other institutions in most instances. In all of these patients, however, the eventual demonstration of an aganglionic segment was possible at operation or by repeated roentgenographic studies with barium enema. Eight of the infants had undergone exploratory laparotomy during the first 14 days of life. The preoperative history in these cases was unusual. The passage of meconium was first noted on the third or fourth day of life. The obstruction was intermittent, often temporarily relieved by enema. The predominant preoperative diagnoses were volvulus of the midgut, malrotation of the colon or intestinal stenosis.

Operative Findings

Diagnostic observations at laparotomy were meager. In one case the postoperative diagnosis was "volvulus of the sigmoid or descending colon," apparently spontaneously reduced. In three instances no abnormality at all was seen.
In two cases the possibility of aganglionic megacolon was discussed or suggested. The correct diagnosis was made at the time of exploration in only two of the eight infants. In the later four cases, mild dilation of the sigmoid colon was described.

The ultimate result of therapy in these eight cases was as follows: (a) Two patients died following a pull-through procedure performed under the age of four months; (b) two died of complications of megacolon, without operation, one at two months and one at 18 months of age; (c) one was lost to follow-up at six months of age; (d) two were successfully treated by a pull-through procedure after the age of two years; (e) one was successfully managed by a medical regimen for over four years. The mortality in this group was thus at least 50 per cent.

**DISCUSSION**

An operative approach to the problem of congenital aganglionic megacolon utilizing the pull-through principle was adopted in this hospital in 1949 and has been employed in 23 procedures during the subsequent five years. Although the basic operation has not been altered, several modifications and refinements now seem indicated.

The use of the “frozen section” technique in examination of the intestinal wall for ganglion cells in the myenteric plexuses is of great importance in establishing the site for proximal transection of the colon. When unequivocal ganglion cells are recognized by the pathologist in the proximal extremity of the resected segment, the anastomosis may be carried out without fear of recurrent obstruction. Similar studies of the distal margin are of academic interest only. Resection should be carried distally to the lower rectum in all cases without regard to the presence or absence of myenteric ganglion cells.

Both transperitoneal (Swenson) and extraperitoneal (Hiatt) resection of the dilated colon segment gave satisfactory results, and each has apparent virtues. The “step” technique for frozen section examination of the proximal colon can be adapted to either operative procedure. Biopsy specimens of the colon excised without entering the lumen are satisfactory for the recognition of ganglion cells.

In the small series presented, pull-through procedures were hazardous in the first year of life, and in general it is probably advisable to defer such operations beyond infancy. This is in striking contrast with the authors’ experience with abdominoperineal procedures for imperforate anus, which are routinely carried out in the neonatal period with a relatively low mortality.

The recognition of congenital aganglionic megacolon in the newborn may be difficult, even at laparotomy. When the condition is known, the patient should be maintained on a medical regimen (enemata). Colostomy should be performed only when obstructive episodes threaten life.

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**REFERENCES**


