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ORIGINAL ARTICLE

THE RESULTS OF TWO-STAGE SURGICAL MANAGEMENT OF HIRSCHSPRUNG'S DISEASE IN A 10-YEAR PERIOD

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Abstract

Background-Hirschsprung's disease has several surgical treatments, including curative surgery which consists of total or subtotal resection of the aganglionic segment followed by a pull-through of the normally innervated intestine. This study intends to assess the aftermath of two-stage endorectal pull-through procedures.

Methods-Medical records of 62 patients (46 males, 16 females), who were treated for Hirschsprung's disease during a 10-year period, were reviewed. Age, sex, extent of aganglionosis, primitive surgical procedure, age at colostomy, age at definitive procedure, number of stages, immediate and late postoperative complications, postoperative causes of death and time of the first postoperative defecation were retrieved in all patients.

Results- Results showed that 61 patients had colostomy or ileostomy as the primitive procedure. Two-stage Soave Boley procedure was done in 54 cases; in 32 (52.4%) patients the Soave-Boley procedure was followed by appendectomy and tube cecostomy and in 22 (36%) patients, Soave-Boley pull-through was done without appendectomy and tube cecostomy. The results of the definitive operations were promising in 90.1% of the patients who gained normal defecation. Early complications were met in 9 cases (14.7%) and late complications were present in 14 cases (22.9%). The mortality rate was 4.9% (3 patients); the first patient had sepsis following ileostomy, the second patient suffered from enterocolitis 6 months post-operatively and the third patient died four days after treatment.

Conclusions- By eliminating one of the stages in the surgical procedure, superior results with less complications are achieved. This in turn precludes the use of costly staplers. Altogether, the Soave-Boley procedure has proved to be a cost-effective and desirable procedure in comparison to other techniques in the treatment of Hirschsprung's disease.

Keywords • Hirschsprung's disease • Soave-Boley • congenital megacolon

Introduction

Hirschsprung's disease (HD) is a malformation of the distal bowel, characterized by the absence of ganglion cells beginning at the internal anal sphincter and extending proximally for various lengths.[1] The diagnosis of HD is based on radiolgraphic studies, anorectal manometery findings and histological examination of rectal wall biopsies.[2] Curative surgery consists of total or subtotal resection of the aganglionic segment followed by a pull-through of the normally innervated intestine.[1,3] The two-stage endorectal pull-through procedure with primitive leveling colostomy is easy to perform and avoids all pelvic disection by passing through rather than removing the abnormal bowel.[10,11] The advantages of this procedure are that one stage of colostomy closure is eliminated and that the use of stapler device is not required. Therefore, it is a cost effective proccedure to perform. The results of 62 patients with HD, treated by two-stage methods in a 10-year period, are reported.

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Materials and Methods

In this retrospective and descriptive study, data was obtained from patient's records in a 10-year period. Sixty-two patients diagnosed with HD, were admitted to two children hospitals (Amircola and Mofid Children's Hospitals), and were treated by the same surgeon. The data collected included: clinical presentation, age at the time of diagnosis, types of colostomy and their complications, types of definitive surgery and their immediate and late complications together with the results of the treatment and the mortality rates of the patients. All patients were diagnosed by a full-thickness posterior rectal biopsy. Primitive leveling colostomy in a divided sigmoid loop with mucous fistula had been performed in the left lower quadrant of the abdomen. Diagnosis of the most distal part of ganglionic bowel was made by anatomic changes of this level and the frozen section. Definitive surgery (endorectal pull-through) was performed 3 months after ostomy and at least, at the 6 th month of life. Before definitive surgery two pathology reports were checked: first, posterior rectal biopsy and second, biopsy of functional level ostomy. In the remote cases anastomotic coverage was done by appendectomy and tube cecostomy, but in the last four years, endorectal pull-through was done without proximal coverage. Absorbable (mostly 2.0 chromic catgut) was used for colo-anal anastomosis; no calf drainage was utilized.

Results

During the 10-year period (1990-1999), 62 patients diagnosed with HD were treated at two university pediatric hospitals by one surgeon. Age distribution at the time of diagnosis is shown in Table 1. One fourth of the patients were symptomatic at neonatal age and almost 50% of the them were diagnosed before the end of the first year of life. Mean age of patients was 9 months, ranging from 3 days to 12 years.

Table 1. Age of patients at the time of diagnosis of Hirschsprung's disease.

Age	Frequency (%)
Below one month	16 (25.8)
1-6 months	8 (12.9)
6-12 months	6 (9.6)
1-2 years	11 (17.7)
2-6 years	13 (20.9)
Above 6 years	8 (12.9)
Total	62 (100)

Table 2. Clinical presentation of the patients with Hirschsprung's disease.

Presentation	Frequency (%)
Chronic constipation	40 (64.5)
Delayed passage of meconium and distension	9 (14.5)
Icterus	1 (1.6)
Obstruction (abdominal pain, distension, vomiting, obstipation)	10 (16.1)
Perforation (peritonitis)	2 (3.2)
Total	62 (100)

Forty-six cases (74.1%) were males and 16 (25.8%) were females.

The relative frequency of different clinical presentations are shown in Table 2. In 40 cases (64.5%), the chief complaint was chronic constipation; 9 cases (14.5%) had delayed passage of meconium and abdominal distention at neonatal age. Ten cases (16.1%) presented with intestinal obstruction and in two cases (3.2%) features of acute abdomen and peritonitis along with colonic perforation were observed during laparatomy. In our sample, eleven patients (17.7%) had associated anomalies; four patients displayed signs of mental retardation, two had Down's syndrome and the other two suffered from cerebral palsy. In two patients (3.2%) there were positive family history of HD; in one case, paternal and in the other, sibling affliction was noted. In 57 patients (91.9%), the aganglionic segment was limited to the rectosigmoid; in the other patient, the segment was the descending colon and in the other two, the transverse colon. In two (3.2%) cases, the total colon was involved.

Ostomy was done in 61 patients; in one patient, definitive surgery was performed without colostomy. Fifty-six patients (91.8%) had sigmoid-leveling end-colostomy in the LLQ, three patients had transverse colostomy, two of which were placed in RUQ and one in LLQ. Ileostomy was performed and placed in LLQ in two cases. Complications of ostomies were met in three of the patients; two were dysfunctional due to hypoganglionosis and aganglionosis that needed revision of the ostomy. Sixty-one patients (98.3%)

Table 3. Types of definitive operations performed.

Type of definitive operation	Frequency (%)
Soave-Boley + appendectomy + tube cecostomy	32 (52.4)
Soave-Boley	22 (36)
State	3 (4.9)
Duhamel	3 (4.9)
Duhamel-Martin	1 (1.6)
Total	61 (100)

underwent definitive operation. In 32 (52.4%) patients a Soave-Boley procedure followed by appendectomy and tube cecostomy and in 22 (36%) patients, Soave-Boley pull-through without appendectomy and tube cecostomy was done. Three patients (4.9%) underwent the State procedure and 3 (4.9%) others, the Duhamel method. The Duhamel-Martin operation was applied to a single case (Table 3).

Considering immediate postoperative complications, 4 cases (6.5%) had wound infection and 2 others (3.2%) suffered intestinal obstruction. Perianal abscess, sepsis and early enterocolitis appeared each in one patient. Fourteen patients (22.9%) had some kind of delayed complications. Four cases had rectal stricture, 4 others, recurrent enterocolitis, and 2 cases who were both mentally retarded, suffered from persistent soiling. Persistent constipation was a matter of concern in three patients and incisional hernia in one case only. In total, 55 patients (90.1%) obtained normal defecation after the operations. Overall, the mortality rate in this study was 4.7%. The reasons for mortality were colonic aganglionosis after ileostomy because of sepsis in one, enterocolitis six months after definitive surgery in the other, and unknown cause 5 days after definitive surgery in the third patient.

Most patients had defecation 2-3 days after surgery; oral feeding was initiated on the 7th postoperative day and almost all patients were discharged from the hospital after 8 to 10 days.

Discussion

Since the reconstructive operation of HD was first described by Swenson and Bill in 1948, patients have been treated soon after diagnosis with a functioning colostomy.[1] When patients were older, a definitive pull-through procedure was performed followed by colostomy closure. This three-stage procedure was extensively advocated because early reports suggested that it was unsafe to perform a definitive operation during the neonatal period.[3] The results of the three-stage Swenson's operation have been well documented through a large multinational study over 4 decades.[4]

Duhamel's procedure was designed to avoid dissecting the anterior rectum and therefore, became a definitive operation in infancy.[5] With Martin's modification, the Duhamel's procedure found wide acceptance for all forms of HD with good results.[6] Surgical stapling devices for intraoperative anastomosis and division of rectal septum are now commonly used and are particularly useful in patients with small bowel transitional zone. Both individual and collected series have revealed low mortality, leak and stenosis rates; hence, improving the results compared to the alternative procedure.[7, 8,9]

The endorectal pull-through as originally described by Soave and then modified by Boley is the third alternative procedure widely utilized for HD .[10,11]

Colostomy or ileostomy is the most frequently applied initial step in the management of HD. But in the past decade, primary pull-through procedure in the neonatal or the first few months of age has been applied in some centers.[12, 13,14]

Recently, the minimally invasive laparoscopic approach has generated considerable interest.[15] Ostomy advantages are numerous; obstruction is relieved, the dilated normal bowel diminishes in caliber which provides less discrepancy and improved safety at the time of reconstruction. Nutrition is also improved, home care is simplified and less traumatic in comparison to colonic lavage. Most importantly, the risk of enterocolitis, particularly its potential mortality, is diminished. The option of a two- or three-stage reconstruction is also important in ostomy site selection. Although some experienced surgeons plan three-stage reconstruction[16], the majority now utilize two stages, the stoma placed at the biopsy proven transitional zone with a low morbidity rate. There are several advantages of transitional zone colostomy or leveling colostomy; the stoma level is selected initially by frozen sections and subsequently confirmed by permanent sections. The interval between stages one and two allows clinical confirmation of normal function, salvaging intestinal length and simplifying the subsequent second stage without a proximal stoma.[17] In such

instances, periostomal skin problems are minimized. Most importantly, transitional zone decompression obliterates previously apparent landmarks at the time of the second stage. A transition zone colostomy is advantageous due to its time consuming nature and low probability of error in frozen section determination. Patients undergoing a modified Duhamel procedure and Boley technique were compared. It was concluded that both procedures were easy to perform, with low morbidity or mortality therefore, proving to be the acceptable treatment for HD.[18] San Fillippo, et al[19] discussing results of definitive procedures in the treatment of 56 patients with HD, concluded that the highest complication rate was in their Duhamel-treated group. The endorectal pull-through procedure was deemed superior because it had fewest complications and the best long-term results. A survey of American surgeons' experience with HD indicated a preference for Swenson's procedure by 23% of the surgeons, Duhamel procedure by 30% and endorectal pull-through by 47%. [20] In this study, we utilized a divided loop end colostomy in LLQ, upper crease curve incision with distal mucous fistula, because of better cosmetic results and ease of distal irrigation and clean pelvic dissection in the second stage.

Although marked improvement in operative mortality and functional results has occurred, no procedure is without complications and the results of three basic current operative procedures are similar. Our endorectal Soave-Boley in second stage is more cost-effective than Duhamel-Martin's procedure because stapler use is eliminated (costing approximately 300-400 US $).

It can be concluded that because of the elimination of one of the stages in the surgical procedure, its good results, diminished complications and since costly staplers are not needed, the Soave-Boley procedure may be rendered as a cost-effective and desirable procedure in the treatment of HD in our country.

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