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The Surgical Correction of Hirschsprung's Disease in Adults Using the Modified Duhamel Procedure

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Background: Hirschsprung's disease (HD) is rare in adults, since a majority of cases are corrected in childhood.
Objectives: The authors describe the profile of patients with HD who reached adulthood without having undergone corrective surgery. Also, they describe the outcomes of a modified Duhamel procedure in these patients, in terms of morbidity and mortality.
Methods: This retrospective study, included patients 18 years old and above, diagnosed with HD who reached adulthood without having undergone definitive repair and managed surgically by the Division of Colorectal Surgery, UP-PGH from January 1, 2004 to December 31, 2014. A review from the Department Surgical Database was used and patients' hospital records were used to fill out a Data Collection Form. Descriptive statistics were used to summarize the data.
Results: The 13 patients included in the study were diagnosed at an average age of 16.6 (± 13.16) years. The mean age at the time of definitive surgery was 23.46 (± 6.96) years. The M:F ratio was 5.5:1. The most common presenting symptom was constipation (69.23%). All had a prior proximal bowel diversion, with a transverse loop colostomy (61.54%) being the most common. The transition zone was located in the sigmoid in a third of patients. The mean time from diagnosis to definitive surgery was 6.69 years. Eight (61.54%) have since undergone stoma reversal. There was only one (7.69%) morbidity, a superficial surgical site infection. No mortalities were reported.
Conclusion: The modified Duhamel procedure is a safe definitive surgical procedure for the adult patient with HD.

Keywords: adult Hirschsprung's disease, colostomy, Modified Duhamel procedure

Hirschsprung's disease (HD) in infancy and childhood is well-discussed in literature, including its surgical management and outcomes.[1] There, however, is a paucity of reports with regard to HD in adults due mainly to its rarity. Often these reports are limited to case reports or case series.[2]

Most referrals the authors receive have previously undergone a proximal diversion during infancy. Thus, there are concerns on the function of the ganglionic segment of bowel distal to the diversion that has been subject to disuse. Another area of concern would be how these patients would cope with learning the physiology of defecation after years of emptying themselves through a stoma.[3] At present, no local studies exist describing patients with HD reaching adulthood without having undergone definitive repair.

This study aimed to describe the clinical profile of Hirschsprung disease presenting in the adult, who underwent definitive surgery. This study also aimed to determine the outcomes of performing a modified Duhamel procedure in these patients, in terms of morbidity and mortality.

The authors hope the results from this study may benefit those managing this special subset of patients.

Methods

This is a retrospective, descriptive case series. The study population included patients 18 years old and above, diagnosed with HD who reached adulthood without having undergone definitive repair and managed by the Division of Colorectal Surgery at the Philippine General Hospital (PGH) from January 1, 2004 to December 31, 2014. Approval of the study protocol by the UP Manila

Technical Research Ethics Board (UPMREB) and the PGH Expanded Research Office (EHRO) was obtained. Electronic records of patients were retrieved using the electronic database of the Department of Surgery, the Integrated Surgical Information System (ISIS). Charts of these patients were retrieved from the Medical Records Section for review. A Data Collection Form was filled out. The data gathered included: age, gender, place of origin, presenting symptoms, information regarding the surgical management, morbidity and mortality. All information not found in the records was noted as "unreported." Descriptive statistics were used to summarize the demographic and clinical characteristics of the patients using frequencies and percentages for categorical variables, and mean and standard deviation for continuous variables.

Surgical Technique

The modified Duhamel procedure was performed with the patient in lithotomy position under either general, or combined general and regional anesthesia. A midline laparotomy incision was made (the original description of Martin begins with a left paramedian, rectus-retracting incision).[13] To allow adequate exposure, the caudad extent of the incision was brought to the superior border of the symphysis pubis.

The sigmoid colon was mobilized in a lateral-to-medial fashion with care in identifying and preserving the gonadal vessels, ureter, and hypogastric nerves (Figure 1). The rectum was then mobilized using sharp dissection in its posterior aspect along an avascular plane following the principles of TME surgery for rectal cancer. Care was taken to avoid dissection along the lateral and anterior aspects, not only is this unnecessary to mobilize but this may lead to inadvertent nerve injury. The posterior dissection was carried down until the pelvic diaphragm was reached (Figure 2).

Complete mobilization of the splenic flexure was performed. The mobilization was extended to the root of the transverse colon until the duodenojejunal junction. This maneuver was important since it may be necessary to perform high ligation of the inferior mesenteric artery (IMA) later on, particularly in patients needing a more extensive resection due to findings of a long toneless

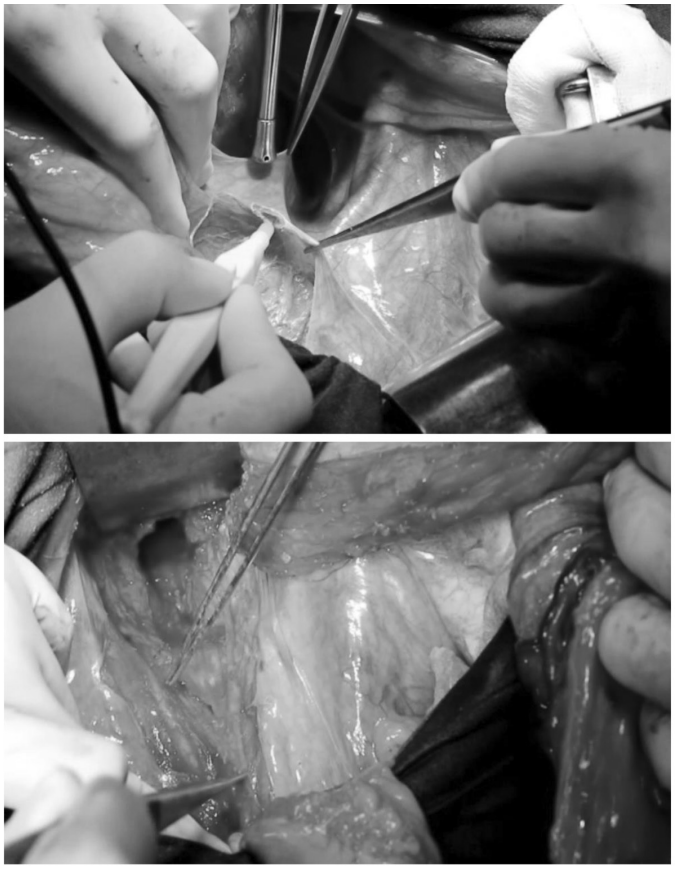

Figure 1. A lateral-to-medial mobilization of the sigmoid is carried out. Care in identifying vital structures (i.e. gonadal vessels, ureter, and hypogastric nerves) is ensured. UP-PGH, 2014.

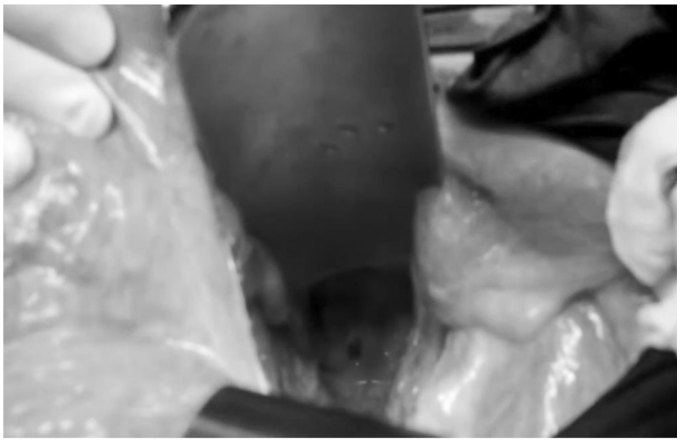

Figure 2. Sharp dissection along the avascular plane along the posterior aspect of the rectum will lead to the level of the pelvic diaphragm. UP-PGH, 2014.

portion of bowel adjoining the megacolic segment. In such cases, perfusion to the colon would be derived from the middle colic artery via the marginal artery.

The rectum was divided just above the anterior peritoneal reflection with staples (Figure 3). Resection of the aganglionic segment was then performed proximally to a level identified by frozen section to have ganglion cells (Figure 4). Due to the chronicity of the disease, adult patients may, at times, require a more extensive resection proximally. The authors preferred removing the pale, toneless, almost rubbery segment adjoining the dilated bowel (Figure 5), even if this had been documented microscopically to have ganglion cells. Long sutures were maintained on the colon so these may be grasped with a clamp as the colon was brought down for the retro-rectal anastomosis.

Figure 5. A grossly toneless segment of colon. The authors routinely include this in the resection as its function is of question, even if ganglion cells are found to be present. UP-PGH, 2014.

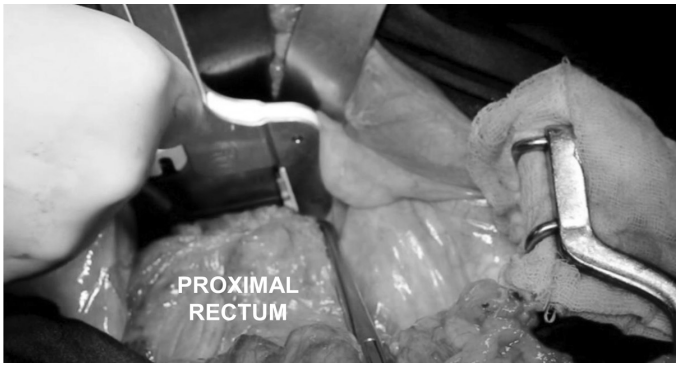

Figure 3. Stapled division of the rectum just above the anterior peritoneal reflection. UP-PGH, 2014.

Figure 4. Resection of the aganglionic colonic segment proximally, with care in preserving the marginal vessels.

The surgeon then transferred for the perineal portion of the surgery. Gentle anal dilatation was done, and a distal rectal wash-out was performed with a mixture of sterile saline solution and an antiseptic. An assist, who remained in the abdomen, inserted a ball of gauze on a long clamp in the retrorectal space and indented this against the posterior aspect of the rectum. A transverse incision was made along the posterior wall of the rectum just above the levator muscles and into the retrorectal space. A full thickness rectal incision became apparent once the ball of gauze was encountered (Figure 6).

A Babcock clamp was then inserted through the incision to grasp the sutures holding the distal end of the colon. The colon was inserted through the lumen of the rectum (Figure 7) and the stapled end resected, revealing its lumen. A retrorectal coloanal anastomosis was performed with full-thickness interrupted suturing using Vicryl 3-0 (Ethicon, Johnson & Johnson) sutures (Figure 8).

Once the retrorectal anastomosis had been completed, sequential linear stapling was performed up to the proximal extent of the rectal stump creating a side-to-side anastomosis between the common wall of the aganglionic rectum and the ganglionic colon (Figure 9).

The authors preferred leaving a proximal diversion in the form of a loop ileostomy. For those with a previous

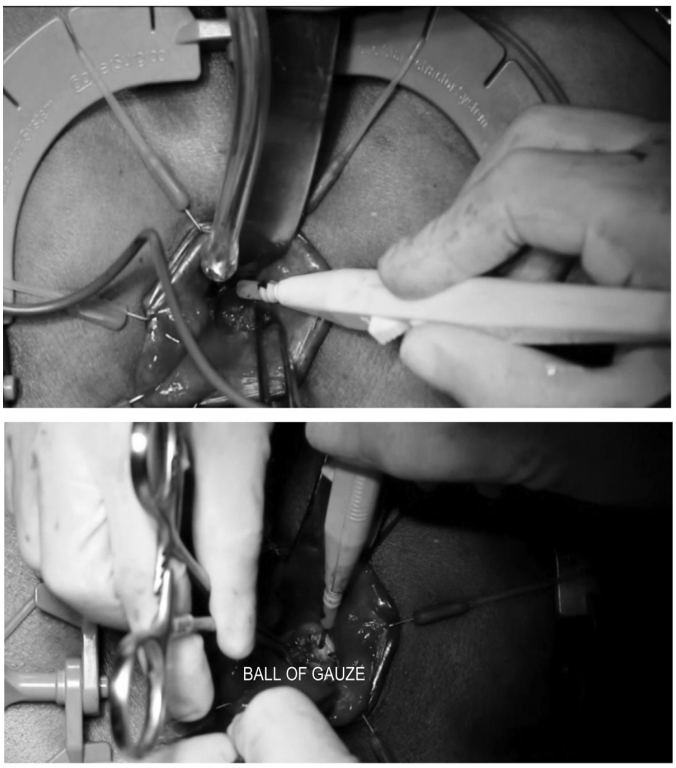

Figure 6. A full-thickness transverse incision along the posterior wall of the rectum just above the levator muscles and into the retrorectal space is made. The retrorectal space is noted to have been entered once the ball of gauze held by the assist is encountered. UP-PGH, 2014.

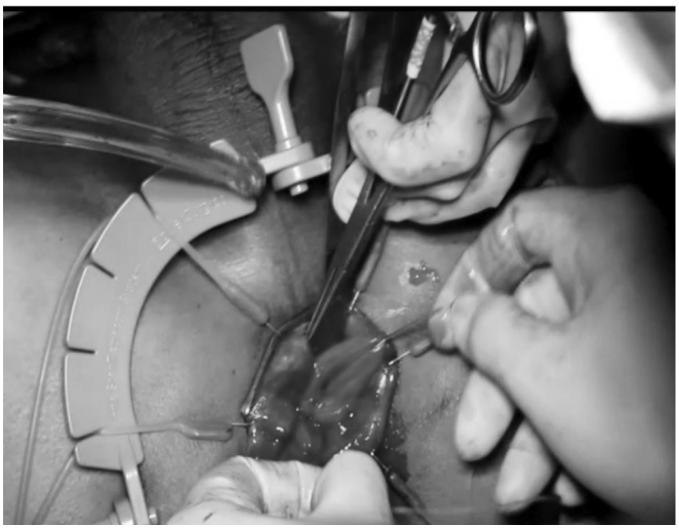

Figure 7. The distal end of the colon being inserted through the transverse incision made in the rectum. UP-PGH, 2014.

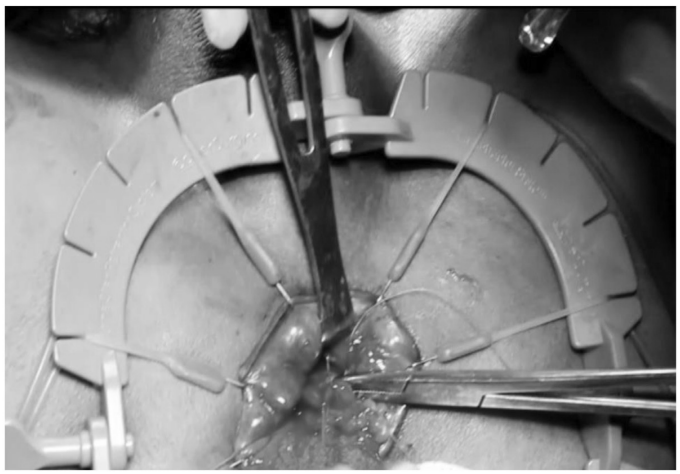

Figure 8. Full thickness retrorectal coloanal anastomosis using simple interrupted Vicryl 3-0 sutures. UP-PGH, 2014.

Figure 9. Sequential linear stapling up to the proximal extent of the rectal stump creating a side-to-side anastomosis between the common wall of the aganglionic rectum and the ganglionic colon. UP-PGH, 2014.

stoma, they often resected up to the level of the distal stoma and brought the proximal stoma down to the pelvis for the retrorectal anastomosis. If the amount of bowel length to allow for a tension-free anastomosis did not suffice, the stoma was taken down and closed, and included in the segment of colon to be anastomosed to the posterior rectum. They have encountered cases where the toneless portion reached the ascending colon. In these cases, they performed rotation of the right colon while maintaining the ileocolic artery (right colonic swing) as described by Deloyers.[16] The defunctioning stoma was reversed after 3 months.

Results

A total of 13 patients were included in the study. There were 11 (84.62%) males and 2 (15.38%) females (M:F ratio 5.5:1). Their ages ranged from 18 to 39 years with a mean age of 23.46 (±6.96; 19.25-27.67 95% CI) years and a median age of 20 years. Nearly all patients (N=12, 92.31%) were from Luzon, with four of these from the National Capital Region (30.77%). Only one patient was from Visayas (N=1, 7.69%), and none from Mindanao. More than half (N=9, 69.23%) presented with symptoms of gut obstruction at an early age. Overall, constipation (N=9, 69.23%) was the most frequent presenting symptom. The other symptoms, in order of decreasing frequency, are: abdominal distention, abdominal pain, vomiting, bloatedness, and weight loss (Table 1).

However, only 5 (38.46%) were diagnosed in infancy or childhood with HD. (Time of diagnosis" was considered as the time that a histopathologic confirmation became available.) The age at the time of diagnosis ranged from 0 to 39 years, with a mean of 16.62 years. With regard to the initial intervention, 69.23 percent

Table 1. Demographic profile of patients with Hirschsprung's disease who underwent a modified Duhamel procedure. UP-PGH, 2005-2014.

Characteristics	Number(N)	Percentage(%)
Age		
Mean (SD)	24.42 (± 6.96)	
Range	18-39	
Median	24	
Gender		
Male	11	84.62%
Female	2	15.38%
Place of Origin		
Luzon	12	92.31%
NCR	4	30.77%
III	5	38.46%
IV-A	3	23.07%
Visayas		
VIII	1	7.69%
Mindanao	0	0.00%
Presenting Symptom		
Constipation	9	69.23%
Abdominal distention	8	61.54%
Abdominal pain	4	30.77%
Vomiting	2	15.38%
Bloatedness	2	15.38%
Weight loss	1	7.69%

(N=9) underwent colostomy for decompression within the year of diagnosis. The longest interval from diagnosis until initial intervention was 14 years. A transverse loop colostomy was the most commonly used form of diversion and was performed on 8 (61.54%) subjects. Eight (61.54%) have since undergone reversal of their protective stoma. There was only one morbidity reported – a superficial surgical site infection. No mortalities were reported. (Table 2). The mean interval in years from diagnosis to definitive surgery was 6.69 (±8.53; 1.53-11.85 95% CI) years (Table 3).

Discussion

Hirschsprung's disease (HD) is characterized by the absence of intrinsic ganglion cells in the submucosal (Meissner's) and the myenteric (Auerbach's) plexuses manifesting as megacolon. This affects one in 2000 to one in 5000 of newborns and carries a male preponderance (4:1).[4-7] In this study, there were 11 (84.62%) males and 2 (15.38%) females, giving a M:F ratio of 5.5:1, which is in concordance with previous studies on adult HD patients, and with data of patients with HD who underwent correction in infancy or childhood.

In this report, most cases of adult HD came from Luzon, one from Visayas and none from Mindanao. (Luzon, Visayas, and Mindanao are the three main island groups in a country that is composed of more than 7,100 islands.) Most specialized hospitals, such as the Philippine General Hospital (PGH), are located in the National Capital Region (NCR, which is located in Luzon). Three of the four accredited training institutions for Colorectal Surgery are in the capital, including the PGH. The majority of consults coming from Luzon is but reflective of the proximity to the PGH of the patient population discussed, and is not representative of the actual distribution of adult patients with uncorrected HD in the country. It cannot be denied, however, that the unique geography of the country, the lack of reliable transport systems, and the concentration of specialized medical care in urban centers are perhaps the underlying

Table 2.Surgical intervention in patients with Hirschsprung's disease who underwent a modified Duhamel procedure. UP-PGH, 2005-2014.

Characteristics	Number(N)	Percentage(%)
Previous Surgery		
Transverse loop colostomy	8	61.54%
Sigmoid loop colostomy	2	15.38
Loop ileostomy	2	15.38
Double barrel ileostomy	1	7.69
Transition Zone:		
Descending colon	4	30.77
Sigmoid colon	4	30.77
Not reported	5	38.46
Modified Duhamel Procedure		
With appendectomy	4	30.77
With frozen section	3	23.08
With takedown of colostomy	5	38.46
With protective ileostomy	5	38.46
Patients who have undergone stoma reversal	8	61.54
Morbidity		
Surgical site infection	1	7.69

Table 3.Timeline of surgical management of patients with Hirschsprung's disease who underwent modified Duhamel procedure. UP-PGH, 2005-2014.

Patient	Age at time of diagnosis (years)	Age at initial surgery (years)	Age at definitive surgery (years)	Follow-up from time of definitive surgery (years)	Time from diagnosis to definitive surgery (years)	Time from definitive surgery to stoma reversal (weeks)
1	20	20	20	5	0	7
2	0	14	21	3	21	14
3	0	0	19	3	19	12
4	5	11	18	9	13	*
5	18	20	20	1	0	*
6	24	24	28	5	4	*
7	2	2	20	0	18	*
8	23	23	23	2	0	*
9	22	22	22	0	0	22
10	20	20	20	4	0	43
11	37	37	37	1	0	14
12	6	6	18	3	12	12
13	39	39	39	0	0	14
Mean	16.62	18.31	23.46	2.77	6.69	17.25
SD	±13.16	±11.83	±6.96	±2.59	±8.53	±11.20
95% CI	8.66-24.58	11.16-25.46	19.25-27.67	1.20-4.34	1.53-11.85	10.48-24.02
Median	20	20	20	3	0	14

reasons for most of these patients to have reached adulthood with undiagnosed, or uncorrected, HD.

A history of long-standing constipation dating back to infancy, or childhood, is the most common presenting symptom of patients with adult HD. Other symptoms were: abdominal discomfort, distention, and pain. In this study, constipation (N=9, 69.23%) was the most frequent presenting symptom supporting previous studies, followed by abdominal distention, abdominal pain, vomiting, bloatedness, and weight loss. Physical examination may reveal a distended abdomen with palpable fecal masses.[4,5] Most of these patients were also observed to have presented with a certain degree of emaciation, likely a result of bowel malabsorption.

In evaluating a patient suspected of having HD, barium enema, anorectal manometry, and rectal biopsy are the primary diagnostic modalities. A barium enema will reveal a transition zone between the aganglionic distal segment, and the dilated innervated colon. This transition zone has been described as being "funnel-shaped" or an "inverted cone."[4] Such narrowing may, however, be absent in as many as 20 percent of patients[7], particularly those with a short or ultra-short aganglionic segment. Retention of barium in delayed films may heighten the suspicion of HD. Barium enema is ubiquitous in medical practice in the Philippines. It appears that the delayed diagnosis of these patients, therefore, is not from unavailability of imaging procedures but from failure of medical practitioners to suspect the diagnosis of HD on the basis of the presenting symptoms.

To confirm the diagnosis of HD, a full-thickness rectal biopsy is performed at the narrowed segment. This is expected to show an absence of ganglion cells, hyperplasia and hypertrophy of nerve fibers, and increased levels of acetylcholinesterase activity. [4-5,8] All of the patients in this study were histologically confirmed Hirschsprung's disease.

There is confusion, however, as to the age that HD may be classified as "adult." Some have arbitrarily classified adult HD as patients over the age of 10 years.[4,9] Others have defined it as those diagnosed beyond 18 or 19 years of age.[4] The patients included in the study were those 18 years and above with histopathologic confirmation of HD.

Patients presenting with HD in adulthood usually have a milder form of the disease, involving only a short segment of aganglionic bowel.[4,11] The expected paucity of symptoms in these patients may also explain the delay in consult, and eventual diagnosis. In this study, age at time of consult ranged from 0 to 39 years old, with an average of 16.62 years. Constipation is often attributed to other pathologic entities, or compensation through hypertrophy of the proximal innervated colon makes the symptoms tolerable. The chronic use of cathartics and enemas[4-5], or even anal digitation, is noted in these patients as a means of assisting defecation. Eventually, with functional decompensation of the colon, rapidly worsening constipation and increasing fecal retention develop with failure of the already dilated colon to propel the feces distally.[4,12]

With regard to the surgical management of HD, a variety of techniques have been described in literature. The more commonly employed procedures are those described by Duhamel[13], Soave[14], and Swenson.[15] In literature, the most common technique employed in adults with HD was the Duhamel procedure.[6,9] The authors perform the modification of the Duhamel procedure as described by Martin and Altemeier.[16] The recognized advantages of the modified Duhamel are: the presence of a fecal reservoir as the native rectum is left in place, the avoidance of incontinence from operative injury as the anastomosis lies above the sphincter complexes, and maintenance of urinary and sexual function as the hypogastric nerves and their branches remain undisturbed during surgical dissection.[16-17]

Vorobyov, et al. performed a modified Duhamel procedure on 82 patients. Of the 82 patients, 60 were assessed regarding functional outcome with 96.7 percent (58/60) claiming good or satisfactory function. A megacolon limited to the sigmoid, in their study, was associated with a good outcome in 89.7 percent of patients. Findings of a more proximal dilatation, however, reduced the acceptable outcomes to 66.7 percent. The criteria they used in assessing postoperative function was based on regularity of bowel movement, presence or absence of incontinence, and a subjective improvement in overall condition compared preoperatively.[5] Miyamoto's review, on the other hand, showed the Duhamel procedure to be associated with

a lower rate of major postoperative complications, and better long-term functional outcome.[8]

The principles of treatment include confirming the location of the transition zone between the ganglionic and the aganglionic segment of the bowel to avoid creating a neorectum with a dysfunctional segment of colon. Based on the data collected, 30.77% (N=4) proximal extent of the resection was up to the level of the sigmoid colon as well as 30% (N=4) up to the descending colon. The rest of the data (N=5, 38.46%) did not mention the portion of the bowel involved. A transition zone at the level of the sigmoid, as noted, is associated with good prognosis and outcomes. But, as mentioned, we often resect more proximal to the histologically-identified transition zone when we find gross evidence of disease chronicity in certain segments of the colon. The authors believe that such intraoperative judgment is vital in ensuring a more satisfactory operative and functional outcome. The creation of a proximal bowel diversion, which most commonly was a transverse loop colostomy, was performed by general surgeons in the local setting.

Admittedly, their measure of ascertaining functional results is lacking in objectivity. However, with all the patients who have had their protective stomas reversed claiming to have regular bowel movement, satisfactory continence, and infrequent bouts of constipation, the authors believe that concerns regarding functional outcome earlier mentioned in this manuscript are baseless and unreal.

The main postoperative complications cited in literature are enterocolitis, constipation and anastomotic stricture. (These, however, are those reported in patients undergoing surgical correction during infancy, or childhood.) In this study, the only morbidity reported after a modified Duhamel procedure was a superficial surgical site infection in one patient. No mortalities were reported. Incidentally, one patient had an anastomotic dehiscence at the time of stoma reversal, and had his bowel exteriorized during re-operation. The same patient has since successfully undergone stoma reversal.

Conclusion

Patients with Hirschsprung's disease in the adult are diagnosed at an average age of 16.6 ± 13.16 years, with constipation as the major presenting symptom (69.23%, N=9). Majority underwent a transverse loop colostomy (61.54%) as the initial operation during childhood. The modified Duhamel procedure as a definitive surgery was considered safe with a 7.69% (1/13) morbidity from a surgical site infection, with no reported mortality. A prospective study to more properly document outcomes related to function is recommended.

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