Hirschsprung's Disease:Experience With Boley's Modification Of Soave's Endorectal Pull Through Procedure In Children

M SALEEM N MALIK M TAHIR A HAMEED
Dept. of Paediatric Surgery,The Childeren's Hospital &The Institute of Child Health, Ferozepur Road, Lahore.
Correspondance to:Dr. Mahmood Shaukat.

Boley's modification of Soave's endorectal pullthrough was performed in 33 children with Hirschsprung's disease. A record of age, sex, extent of aganglionosis, complications of definitive operation, and status of faecal continence of the patients after definitive operation was made. There is follow up of 2-6 years. The complications encountered were stenosis of the anastomotic site 7, wound infection 6 and burst abdomen in 2 patients. Twenty-four patients were fully continent, 4 fairly continent and 3 had poor faecal control.
Key Word : Hirschsprung's disease, Aganglionosis, Boley's modification, Soave's endorectal pullthrough.

Hirschsprung's disease was first described in 1691 by Frederick Ruysch, but the morbid anatomy was understood as a result of the work[1] of Danish paediatrician Harald Hirschsprung in 1886[2]. Fifty years passed before any definitive treatment was offered in the form of rectosigmoidectomy performed by Swenson and Bill in 1948[3]. The principle being replacement of the aganglionic colon with normally innervated proximal colon which is brought down to the anus and a direct end to end anastomosis is performed at the anal verge.

Subsequently Duhamel in 1956 described a new procedure which is essentially a retrorectal end to side anastomosis between the normally innervated pulled down colon and the retained rectal pouch[4]. In 1964 Soave described the endorectal pullthrough, in which a seromuscular sleeve of the aganglionic segment is retained[5]. This avoids an extensive pelvic dissection. In its original form, Soave's procedure leaves a small length of the endorectally pulled out ganglionic colon outside the anus. This stump has to be trimmed and anastomosed to anal canal at the dentate line about 2 weeks later. Boley in 1966 described a modification of Soave's procedure, in which a direct primary anastomosis is performed at the dentate line, thus avoiding the 3rd stage of the procedure[6].

Each procedure has its own merits and demerits and modifications have been devised to minimize the immediate or delayed complications. Two methods have stood the test of time, the endorectal pullthrough and the Duhamel operation.

Materials And Methods

A prospective study was conducted at Mayo Hospital Lahore over 4 years to evaluate the results of Soave's endorectal pullthrough (Boley's modification). A total of 33 pullthroughs were performed in this period. In 31 patients endorectal pullthrough was performed with Boley's modification of Soave's procedure. In two cases ileoanal endorectal pullthrough with Boley's modification was performed for total colonic aganglionosis.

These 33 patients were seen for the first time in the out-patient department/emergency of Paediatric Surgery. Age distribution in these patients at the time of presentation is shown in (Table 1). Male to female ratio in these patients was 3.9:1. Extent of disease was determined in all these patients with the help of barium enema, by naked eye appearance at laparaotomy/colostomy and confirmed by serial segmental biopsies. This is shown in (Table 2). Majority of these patients were suffering from rectal or rectosigmoid disease. The sequence of investigations for making a definitive diagnosis was, plain X-Ray abdomen, barium enema,full thickness Rectal biopsy, laparotomy, serial, segmental biopsies and colostomy/ileostomy.

Table - 1: Age At Time Of Presentation.(N = 33)

Age	n=	%age
< 1 months	12	36.36
1-6 months	4	12.12
7m - 1 year	5	15.15
1-2 year	3	9.09
2-3 year	3	9.09
3-5 year	2	6.06%
> 5 years	4	12.12%.

Table - 2 Extent Of Aganglionosis(N = 33)

Extent	n=	%age
Short Segment.	24	72.72
Rectosigmoid	13	39.39
Sigmoid	11	33.33
Long Segment	7	21.21
Descending Colon	3	9.09
Splenic Flexure	3	9.09
Transverse Colon	1	3.03
Total Colonic Aganglionosis		
with ileal involvement	2	6.06

The diagnosis was confirmed by a full thickness rectal biopsy in all these patients. Then a defunctioning ileostomy/colostomy was created. The definitive surgical procedure was performed at a mean age of one year or six months after creation of a defunctioning stoma in older children.

Results

The colostomy/ileostomy was performed in all patients. The complications of colostomy/ileostomy are shown in [Table 3]. Paracolic evisceration was encountered in two patients in whom the colostomy had to be revised. Retraction of colostomy stoma occurred in one patient. Episodes of severe diarrhea were frequently encountered in patients with ileostomy necessitating supplemental fluids for hydration. Recurrent prolapse of stoma[8] and

skin excoriation[7] were common complications directly attributable to the stoma. Skin excoriation was managed by different local bland applications like pertroleum jelly. Recurrent prolapse of colostomy was a troublesome complication. Frequent reductions and anchorage to the anterior abdominal wall with non-absorbable sutures was performed, but did not yield satisfactory results.

Table - 3 complications Of Ileostomy / Colostomy(n = 33)

Complications	n	%age
Paracolic evisceration	2	6.06%
Retraction of stoma	1	3.03%
Diarrhoea	10	30.30%
Recurrent prolapse of colostomy.	8	24.24%
Skin excorriation.	7	21.21%

Table - 4-Complications Of Pull Through(n= 33

Complications	n	Percentage
Early Complications :		
Wound infection	6	18.18
.Mucosal prolapse	1	3.03
Abdominal wall dehiscence.	2	6.06
Perianal skin excoriation	8	24.24
Late Complications :		
1.Adhesive bowel obstruction	1	3.03
2.Anal stenosis and constipation	7	21.21
.3.Enterocolitis and death	2	6.06
4.Incontinence	3	9.09

Table - 5 Status Of Continence(N = 31)

Status	n=	%age
Good	24	77.41
Fair	4	12.90
Poor	3	9.67

Complications of the definitive operation are shown in Table - 4. Wound infection[6] perianal skin excoriation[8] were commonly encountered early complications. Abdominal wound dehiscence was encountered in 2 patients. Mucosal prolapse was seen in one patient. Adhesion obstruction was encountered in one patient three months after pullthrough, responding to conservative management. Temporary anal stenosis and constipation were seen in 7 patients. Six of these patients responded to repeated anal dilatations. In two patients an annular ring developed at the anastomotic site, which needed division.

Post-operative enterocolitis was seen in two patients. One patient developed post-operative enterocolitis three months after pullthrough. He came to the ward with tense abdomen, bleeding P/R, and high fever. He died in emergency and could not be revived. The second patient had developed anal stenosis, because the child's parents did not follow the advice at discharge, of regular anal dilatitation. After three months, he was brought in a moribund condition due to enterocolitis, colostomy was planned, but refused by the parents, the child expired on the second day of admission.

The status of continence was assessed using the protocol of Soper and Figueroa (1971)[6] and Templeton and Diteshein (1984)[7] . Majority of the patients (77.41%) had good results. Four patients had fairly good results and only 3 had poor results. The results were evaluated at least one year after surgery or in younger patients, at the age of 2.5 years. The patients with poor results were managed with dietary modifications, addition of husk of Ispagula in diet and toilet training. .

Discussion

Since Harald Hirschsprung describd the gross anatomy of this disease. , a lot of research work had to be done before the etiology and pathophysiology of this disease could be deciphered. Erenphresis, 1946 was the first to point out that absence of ganglion cells is the basic cause[2] of this disease (cited by Swenson, 1989) . Later on hypertrophy of preganglionic cholinergic fibers was also found to be a regular histological feature alongwith absence of ganglion cells.

The pathophysiology of Hirschsprung's disease is now well established, its etiology however still evades research work[8] . Different theories have been proposed like.Failure of migration of vagal neural crest cells into the affected bowel segment[9] .Harmful microenvironment and destruction of neural elements[10,11] after their migration .Vascular insufficiency[12,13] .Autoimmune damage to ganglia[14,15] .Genetic disorders[16] .Viral illnesses resulting in loss of ganglia .

Still a consensus has not been achieved regarding the eitology.

There has however been a greater triumph as regards the management. It was a non-treatable disease till 1948, when Swenson and Bill described rectosigmoidectomy as a remedy[2] . Swenson's procedure did not give comparable results regarding continence in the hands of other surgeons.More procedures have since been devised. Currently three procedures, Swenson's colo-anal anastomosis, Soave's endorectal pullthrough and Duhamel's retrorectal pullthrough are being performed commonly[17] . Martin's modification of Duhamel's procedure[3] and Boley's modification of Soave's endorectal pullthrough[4] have now been universally incorporated in the standard procedures.

All these procedures are based on the principle of bringing normally innervated bowel down to the anus. The difference is merely in the type of anastomosis and the method of preserving and using the aganglionic rectal pouch. The Swenson's procedure involves an end to end anastomosis after resecting ganaglionic colon[2] between normally ganglionic bowel and anus . Duhamel procedure is an end to side anastomosis between the rectal pouch which is aganglionic and the normal ganglionic bowel brought behind[3] the rectal pouch and anastomosed to its posterior wall . Soave's procedure is a coloanal anastomosis, between the normal ganglionic bowel pulled down through a seromuscular cuff of the[4] aganglionic rectum, prepared by stripping the mucosa . The anastomosis between this normal bowel & anus is performed subsequently at a second stage.

Boley popularized a modification in the endorectal

pullthrough by performing a primary anastomosis at the time of the original pullthrough, thus completing the operation at one sitting [3].

This procedure has been widely practiced with a lesser complication rate, as shown by Kleinhaus et al [18] (1979) and Ikeda and Goto [19] (1984) that 60.4% and 64.9% of the patients treated with primary anastomosis at the time of endorectal pullthrough were free of complications respectively. The incidence of enterocolitis was also less with this procedure (2.1%) while it was 15.1% with Swenson [18] 5.9% in Duhamel and 5% with Soave's pullthrough [10]. In our series it was 6.06%.

This procedure does not require any special surgical gadgets like stapling guns which are expensive and beyond the reach of our patients. There is no pelvic dissection and there is no direct weakness of internal anal sphincter the incontinence rate is less as compared to the two other procedures [3].

According to a survey conducted by Kleinhaus et al in 1979, 47% of the Paediatric surgeons preferred endorectal pullthrough, 30% Duhamel's procedure [18] and 23% Swenson's procedure [10]. Harrison et al in 1985 [20] published a survey which also shows the same tren [20]. An extension of Boley's endorectal pullthrough was described by Boley himself [21] in 1984 for management of total colonic aganglionosis [21].

The results of modified Soave's operation in our series are comparable with the results in literature[18,20], 77.41% of our pateints are fully continent, 3(9.67%) have poor results, which are being managed with dietary modifications and toilet training.

The cause of death in two patients was post-operative enterocolitis. One major contributing factor for high mortality in enterocolitis was delay in reporting for treatment.

This series throws light on many aspects of this not an uncommon condition. It needs long specialized treatment with more than one surgical procedures and multiple hospital admissions. It is very important to think in terms of cost effective management and protocols. With the modified endorectal pullthrough, we achieved consistantly good results and we consider that it is an as equally effective method as Duhamel's procedure. However being less costly it is more suitable for our patients.

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