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SWENSON'S OPERATION FOR TREATMENT OF HIRSCHSPRUNG'S DISEASE, SHORT AND LONG TERM OUTCOME.

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ABSTRACT

Background / Aim : Many definitive operations exist for Hirschsprung's disease, each of these operations is highly effective in the relief of functional obstruction, on the other hand each operation has some complications. The aim of this study is to evaluate Swenson's operation as a method for treatment of Hirschsprung's disease and assessment of short and long term outcome.

Methodology :

Forty patients underwent Swenson's operation over 4 years, they were 25 males and 15 females with mean age 6.64 years (range 6 months- 12 years). Preoperative history and clinical assessment, laboratory, radiologically and manome-

try were done in all cases. Post-operative evaluation of outcome was studied.

Results :

No mortality in our cases. Soiling occurred in 2 cases, minor incontinence in 7 cases (17.5 %) and major incontinence in 5 cases (12.5 %), enterocolitis in 5 cases (12.5 %) .constipation in 4 cases (10%).

Leakage associated with wound and pelvic collection in 2 cases (5 %) and was treated with antibiotics and leaving the drains for 6 to 8 days. One case developed rectovaginal fistula, one case of anal stricture, one case of recurrent Hirschsprung's disease. On follow up these symptoms improved through along the first and second year post-operatively.

Conclusion:

Swenson's operation is the original curative pull-through procedure especially if precisely performed. However, it has some complications. Anal stricture may complicate ischaemia of the pulled through segment. Enterocolitis remains a serious complication. Incontinence has a relatively high incidence but with significant improvement with time.

Key words: Hirschsprung's disease, Swenson's operation.

INTRODUCTION

Hirschsprung's disease is a congenital disorder characterized by distal aganglionosis, usually involving the rectosigmoid, which results in functional obstruction of the colon (1). Nitric oxide which is a neurotransmitter in the noncholinergic inhibitory nerves, is absent in the ganglionic region and the internal anal sphincter causing spasticity of both (2). The transitional zone lies in the rectum or sigmoid colon in 75%-80%, and reaching the splenic flexure in 13.5%-15%, while total colonic aganglionosis in 0.16%-0.4% of the cases (3).

Many definitive operations exist

for Hirschsprung's disease those include Swenson, Duhamel, Soave, transanal pull-through which is the latest development of treatment (4). Each of these operations is highly effective in the relief of functional obstruction, on the other hand each operation has some complications (5) and so, the long-term outcome and quality of life are not as good as surgeons expected (6).

Soiling is common after Swenson's operation possibly due to much excision of the internal anal sphincter (7). Soave's operation entails stripping of the rectal mucosa and endrectal pull-through of the proximal ganglionic colon after resection of the aganglionic segment (5). This technique leaves the aganglionic muscle cuff intact and allows for preservation of the spastic internal anal sphincter that predispose to postoperative constipation and enterocolitis (8). In Duhamel's procedure, the rectum is retained and the normal ganglionic colon is brought down behind the rectum avoiding manipulation of the rectum anteriorly (9). So the new rectum is composed of normal colon posteriorly and inert part anteriorly,

and peristalsis does not close the lumen and there is no forward movement of the bolus (9).

PATIENTS AND METHODS

Our study included 40 patients between 2002-2006. They were 25 males and 15 females with mean age 6.64 ± 4.4 years (6months- 12 years). All patients had been submitted to thorough history and clinical examination as regard delayed passage of meconium, enterocolitis, abdominal distension, loss of weight, and empty rectum. Routine laboratory investigations, barium enema, and rectal muscle biopsy one cm. above the dentate line. Swenson's operation was done as one stage or associated with colostomy. Preoperative colostomy was done due marked abdominal distension or to correct the general condition in 5 patients. Operative colostomy was done if there was colonic distension in 4 patients. Frozen section was done in all patients from the edge of colonic segment anastomosed to the anal canal to confirm the presence of ganglion cells.

Postoperative assessment and follow up for 1-3 years for early complications as leakage, enterocoli-

tis, urine retention, incontinence, fistula and constipation. Anal digital examination was done after 3 weeks for anal canal resting and maximum squeeze tone. Barium enema was done if there was stricture or suspicious of recurrence of the disease. Anorectal manometry was done when necessary in the presence of incontinence or constipation to measure the resting and maximum squeeze pressures.

RESULTS

This study included 40 patients in the period from 2002-2006 , all patients were treated by Swenson's operation, they were 25 males and 15 females with mean age 6.64 ± 4.4 years (6 months- 12 years). Constipation (table 1) was the commonest presentation in all cases(100 %). Abdominal distension in 25 cases (62.5%) with recurrent attacks of intestinal obstruction in 10 cases (25 %). Delayed passage of meconium in 15 cases (35 %) and enterocolitis in 2 cases (5%). Barium enema was diagnostic in all cases. Rectal muscle biopsy was aganglionic in all cases (100%)(table1). The mean operative time was 125.3 ± 44.22 minutes (90-140 minutes). Oral feeding started 2-4 days.Hospital stay was

5.1 ± 1.9 days (4-10 days).

Early postoperative complications (Table 2) was wound infection in 2 cases (5%) and controlled by antibiotics and wound care, urine retention in one case (2.5%) and managed by a catheter for one week. Minor incontinence in 5 cases (12.5%), soiling in 2 cases (5%) and major incontinence in 8 cases (20%). Enterocolitis in 5 cases (12.5%) and respond to metronidazole in pediatric dose for one week. Constipation in 4 cases (10%) that responded to the transient use of laxatives, anastomotic leakage in 2 cases (5%) and treated by diverting colostomy for 10 days in the hospital and completed the treatment in the outpatient clinic for another 5 to 10 days as the leakage was minor. These fistulae stopped within 2 and 3 weeks. One case developed rectovaginal fistula that was treated by colostomy followed by perineal repair after 4 months, then closure of the colostomy 3 weeks later.

late complications were anastomotic stricture in one case (2.5%) which was treated with repeated anal dilatation, one case (2.5%) developed recurrence of the disease and treated by re-Swenson's operation with great response, minor incontinence in 4 cases (10%) and major incontinence in 2 cases (5%) (table 3).

Follow up of the incontinent children for one year, the number decreased from 6 cases (4 minor and 2 major) to 4 cases (3 minor and one major) which remain the same through 2 and 3 years. Anorectal manometry was done for 6 cases of incontinence, (table 5). The mean resting pressure was 58.1 ± 15.1 mmHg in normal cases while it was significantly lower in the incontinent cases with mean 42.9 ± 16.2 mmHg. Also the mean maximum squeeze pressure was 87.6 ± 28.7 in the incontinent cases which was significantly lower than normal cases with mean 107.7 ± 28.5 (table 4, table 5).

Table(1) Preoperative data (no. 40)

Findings	No	%
Delayed passage of meconium	15	37.5
Constipation	40	100
Recurrent intestinal obstruction	10	25
Enterocolitis	2	5
Abdominal distension	25	62.5
Diagnostic barium enema	40	100
Rectal muscle biopsy(aganglionic segment)	40	100

Table (2) Early postoperative complications :

<u>Complication</u>	No.	%
Wound infection	2	5
Urine retention	1	2.5
Minor incontinence	7	17.5
Major incontinence	5	12.5
Enterocolitis	5	12.5
Constipation	4	10
Leakage	2	5
Fistula	1	2.5

Table(3) Patients morbidity

<u>Causes</u>	NO.	%
Fistula (rectovaginal)	1	2.5
Enterocolitis	5	12.5
Minor incontinence	4	10
Major incontinence	2	5
Recurrent constipation	4	10
Recurrence of the disease	1	2.5
Stricture	1	2.5

Table (4) Long term follow up of morbid cases :

	1 Years		2Years		3Years	
	NO.	%	NO.	%	NO.	%
Enterocolitis	5	12.5	4	10	3	7.5
Incontinence	6	15	4	10	4	10
Minor incontinence	4	10	3	7.5	3	7.5
Major incontinence	2	5	1	2.5	1	2.5
Constipation	4	10	3	7.5	2	5

Table (5) Anorectal manometry in continent and incontinent cases

	Continent	incontinent	P-Value
	(No= 6)	(No= 6)	
Resting pressure	58.1 ± 15.1	42.9 ± 16.2	0.01
Maximum squeeze pressure	107.7± 28.5	87.6 ± 28.7	0.05

DISCUSSION

Surgery for Hirschsprung 's disease must satisfy to criteria : The first is eradication of the aganglionic segment and the second is the preservation of fecal incontinence (17).The clinical outcome of affected patients with Hirschsprung's disease has improved deramatically with the development of effective operative procedures.The commonly used procedures include the Soave pull-through, the Duhamel procedure, the Swenson's procedure, and transanal endorectal pull-through. It is generally thought that there are no significant differences as regard the outcome and long term function of these patients treated by these different procedures. These include a reported increase in the incidence of enterocolitis after Swenson's operation , diarrhea and incontinence after Soave endorectal pull-through and constipation after Dohamal procedure (19,20).

In our study we had no mortality. Mortality rate of 2.4% (24), 2.5 % (20), 7.6% (21) had been reported.Infection as a cause of death had been reported by many authers.The mortality of enterocolitis reaches as high as 30% (23) and pneumonia or

peritonitis was responsible for 18% of causes of death (15). An increased mortality rate has been noted among those with total colonic aganglionosis or Down syndrome or associated congenital heart disease (13). The incidence of soiling after Swenson's operation was 46.4 % (19) and 50% (26) due to excessive excision of the internal anal sphincter .

Early surgical complications as wound infection and pelvic abscess occurred in 2 cases (5%) in our series and associated with anastomotic leakage which is similar to that of Sherman(24) who has leakage in 5.6% and 8.1% (26). Anastomotic leakage and pelvic abscess nessecitated a diverting colostomy until infection subsides and leakage was controlled. Rectovaginal fistula occurred in one case(2.5%) in our series, while it was 1.4 % (18) and 6.2%(20), in other studies.

Anal stenosis occurred in one case (2.5%) in our cases which is better than those of Sherman (24) 7.6% ,Kleinhaus (20) 9.5%, and Nixon (21) 14.1 % . This complication is probably related to ischaemia in the pull-through segment, and if the operation in complicated by local infec-

tion due to anastomotic leakage⁽¹⁶⁾. The surgical options include either an anorectal myotomy or a redo procedure (25).

Enterocolitis is a serious complication of Hirshsprung's disease even after the corrective pull-through procedure. In one series its incidence was 33%, the mean time between the operation and the attack of enterocolitis was 29 months and the longest period was 176 months (12). In our series, the incidence of enterocolitis was 12.5 % in the early postoperative period and 15% in the first year . Other reported incidences were 22.5 % (24), 15.6 % (20) and 11.1% (21). The most prevalent explanation for persistent risk of enterocolitis after corrective procedures is related to the persistently hypertonic anal sphincter associated with the disease causing functional obstruction and inflammation of the colon. Other theories include an alteration in the intestinal mucosa, a deficiency in the intestinal secretory immune system and infection with *clostridium difficile* (14) and consideration should be given to the use of Vancomycin and metronidazole until culture results are available (11). The role of coexistent intestinal neuronal dysplasia in

the proximal colon may be relevant (27). Anorectal manometry revealed a high mean resting pressure among those with postoperative enterocolitis, anal dilatation or even sphincterotomy may prevent recurrence of enterocolitis (23).

Although some reports suggested a good patient satisfaction after surgery, other investigators have found increased incidence of complications and incontinence which resolve with time (23).

Incontinence may be related to decreased neural sensation in the area resulting from the pull-through procedure or to a weakened anal sphincter, making it difficult to expel flatus without passage of liquid stool (22). The number of incontinent cases in our series was 6 cases (15%) which decreased to 4 cases (10%) in the second and third year postoperatively. Marty (18) had an incidence of 12.6 % and Sherman (24) had an incidence of 4.6%. Postoperative anorectal manometry of incontinent cases in our series revealed a relatively low mean resting pressure (42.9 + 16.2 mmHg) and maximum squeeze pressure (87+ 28.7 mmHg) suggesting that incontinence

is related to a weakened internal anal sphincter.

Chronic constipation in patients with corrected Hirshsprung's disease is usually related to postoperative anal stenosis or residual aganglionosis . In our series we had 4 cases (10%) of constipation postoperatively , one of them was due to aganglionosis and treated by reoperation , and one case had anal stricture which was treated by repeated dilatation.

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