

ORIGINAL ARTICLE

INTERNAL ANAL SPHINCTER: IS IT PROPERLY MANAGED DURING RESECTIONAL SURGERY FOR HIRSCHSPRUNG'S DISEASE?

By

Kamal Abd Elelah, Abdel Azim Ali, Sabry A. Mohamoud

Department of Surgery, Faculty of Medicine, Mansoura University

Aim: was to compare the technique and the postoperative continence state of Swenson's operation with its Wang's modification.

Methods: 25 cases underwent a resectional surgery over 2.5 years. 13 underwent Wang's modification while Swenson's operation was performed for 12 cases.

Mean age at operation was 16 months (2 months to 5 years).

Results: Wang's operation had superior results than Swenson operation regarding postoperative soiling ($P= 0.027$) that occurred in 6 cases after Swenson operation and in only one case after Wang's operation while anastomotic leak occurred in one case of each group. Further, Wang's operation group had postoperative resting and squeeze pressures of 49.8 ± 7.31 mmHg and 90.8 ± 2.80 mmHg respectively; both are significantly higher than those recorded in Swenson operation group: 40.8 ± 2.30 , 78.0 ± 7.21 respectively ($P= 0.0001$).

Conclusion: Wang's modification of Swenson's operation represents a proper management of the internal anal sphincter during resectional surgery for HD through preservation of the anterior rectal wall and splitting its posterior wall with formation of heart shaped anastomosis.

Keywords: Hirschsprung's disease, Internal anal sphincter, Wang's modification.

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⁾

HD fits the pathological spectrum of neurocristopathies caused by point mutation affecting RET proto-oncogene that is present on the long arm of chromosome 10.⁽²⁾

Nitric oxide which is a neurotransmitter in the non adrenergic non cholinergic inhibitory nerves, is absent in the aganglionic region and the internal anal sphincter causing spasticity of both.⁽³⁾

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 represents 0.16%-0.4% of the cases.⁽⁴⁾

HD have an incidence of 1:5000 live births with a male:female ratio of 3.8:1.⁽⁵⁾

Clinically, HD is considered the most common cause of neonatal intestinal obstruction (17%).⁽⁴⁾ While, 8% of cases have onset of symptoms outside the neonatal period.⁽⁶⁾

Cases having HD can be diagnosed by barium enema, anorectal manometry and rectal suction biopsy.⁽⁷⁾

Traditionally, HD diagnosed in the neonates or early infancy has been treated by diverting colostomy, followed later by the definitive pull-through. Recently, successful one-stage surgical treatment of infants with HD can be safely performed.⁽⁸⁾

A variety of definitive operations exist for HD, including those described by Swenson, Duhamel, Soave, Rehbein and the transanal pull-through which is the latest development in treatment of HD.⁽⁹⁾

Each of these operations is highly effective in the relief of functional obstruction, but, on the other hand, each has its own inherent defects leading to both early and late complications⁽¹⁰⁾ and so, the long-term outcome and quality of life are not as good as surgeons expected.⁽¹¹⁾ Swenson operation entails resection of the aganglionic segment down to within 2 cm. of the dentate line with anastomosis of the normal ganglionic colon with the anal canal.⁽¹²⁾ Soiling is common after this operation which may be due to too much excision of the internal anal sphincter.⁽¹³⁾ Soave's operation entails stripping of the rectal mucosa and endo-rectal pull-through of the proximal ganglionic colon after resection of the aganglionic segment.⁽¹⁰⁾ This technique leaves the aganglionic muscle cuff intact and allows for maximal preservation of the spastic internal anal sphincter that predisposes to postoperative constipation and enterocolitis.⁽¹⁴⁾ In Duhamel's technique, the rectum is retained and the normal ganglionic colon is brought down behind the rectum avoiding manipulations of the rectum anteriorly.⁽¹⁵⁾ So, the new rectum and part of the rectosigmoid are composed of normal colon posteriorly and inert tissue anteriorly, with this anatomic arrangement, peristalsis does not close the lumen and there is no forward movement of the bolus.⁽¹⁶⁾ Wang's et al.,⁽¹⁴⁾ stated that many of these complications occurred because of mismanagement of the internal sphincter and so designed a new operative method in which the rectum is transected 4 cm. above the dentate line.

The aim of our work is to compare the technique and the postoperative continence state of Swenson's operation with its Wang's modification.

PATIENTS AND METHODS

25 cases of HD were admitted at pediatric surgery department of Mansoura University Children's Hospital from Oct. 2002 to Feb. 2005. All cases were diagnosed on the basis of clinical evaluation, barium enemas, anorectal manometry and rectal punch biopsy.

The following criteria were considered suggestive of HD from the clinical point of view:

History of delayed passage of meconium, constipation dating the first days of life, no or very infrequent spontaneous evacuation, absence of soiling, previous attacks of enterocolitis (EC), previous attacks of intestinal obstruction and presence of empty rectum.

On barium enema, HD was diagnosed by the presence of contracted distal bowel, presence of a transition zone, dilated proximal colon and barium retention in the delayed film.

Anorectal manometry was done using POLYGRAM NET system (Medtronic A/S-Denmark) which consists of POLYGRAM NET version 4.01. 525.45, anorectal function testing software, polygraph ID data-acquisition device, COMPAQ compatible computer, Mui scientific water perfusion pump with constant perfusion rate about 0.3 ml/min which connected to four channel anorectal water perfusion catheter with balloon.

Technique of anorectal manometry: The child is placed in lateral position, the pressure catheter is introduced at 5cm from the anal verge and the rectal pressure is recorded, the catheter is slowly withdrawn until a rise in intraluminal pressure is observed which defines the beginning of the anal canal, its distance from the anus is recorded thus determining the length of the anal canal, the recorded zone of high pressure represents the resting anal canal pressure, then the deflated rectal balloon is lubricated and introduced 5cm above mucocutaneous line and held in this position, the pressure catheter is reintroduced till the zone of high pressure is recorded again. Then the catheter is placed against the balloon tubing and both are held in place. Anorectal inhibitory reflex is determined by inflating the rectal balloon intermittently for 10 seconds. A small volume of air is introduced initially and increased gradually until resistance to balloon distention is felt at the syringe. Persistence of the pressure in high pressure zone area with inflation of the rectal balloon indicates absent anorectal inhibitory reflex which strongly suggests the diagnosis of Hirschsprung's disease. Squeeze pressure was measured during infant crying while older children were ordered to squeeze the tubes.

Pathologically, rectal punch biopsy revealed absent ganglia with the presence of hypertrophied nerve bundles.

Colostomised cases, cases having ultrashort segment disease and cases with total colonic aganglionosis were excluded from the study.

After diagnosis, the infant was prepared as follows: Colonic washout with saline twice per day in the three preoperative days, non-residue diet the day before operation, I.V. fluids at the preoperative night with stoppage of oral intake, and I.V. metronidazol and 3rd generation cephalosporin 2 hours before operation.

Swenson's operation was performed for 12 cases while Wang's modification was performed for 13 cases. The consent of the operation was taken from the parents.

Swenson's operation⁽¹²⁾: Left lower paramedian incision is performed. The proximal ganglionic bowel is identified by operative biopsies and then widely mobilized up to the splenic flexure (Fig. 1). Inferior mesenteric artery is ligated and divided at the aortic root. Pelvic dissection is commenced and the rectum is dissected keeping immediately on the muscular wall till the level of dentate line (Fig. 2). Attention is now directed to the perineum where the aganglionic colon is intussuscepted through the anus. The anterior half of the everted rectal wall is divided 1½ cm proximal to the anodermal junction while the posterior wall is cut ½ cm only from this junction. Then coloanal anastomosis is done with 4-0 polygalactin interrupted sutures.

Wang's modification⁽¹⁴⁾: This operation like that of Swenson but differs in the pelvic dissection where posterior dissection of the rectum is completed till the dentate line, laterally, dissection continues till the lateral ligaments only, anteriorly, rectal wall is left undisturbed and so the pelvic autonomic nerves are protected.

At the perineal part of the operation the everted rectum is transected 4 cm above the dentate line (Fig. 3) and its posterior wall is split longitudinally till that line (Fig. 4), the tips of the two halves are trimmed so that the remaining rectal wall, whose anterior aspect is 4cm from the dentate line and the posterior wall is at the dentate line, has the shape of the heart (Fig. 5). Interrupted sutures are placed circumferentially at each quadrant through the seromuscular coats of the proximal bowel and the full-thickness edge of the transected rectum. Each suture is tied and grasped with clamps to prevent retraction; the subsequent sutures are added to complete the anastomosis. Oral intake was started 3-5 days after operation. After discharge the cases were followed up as regards frequency of bowel motions, soiling, attacks of enterocolitis, anal tone and anorectal manometry (ARM) that was performed 6 months after operation.

Statistical analysis: Numerical data are expressed in mean±standard deviation. Student t test and Chi-square tests were used to compare resultant data as appropriate. P value <0.05 was considered significant.

RESULTS

The total number of cases was 25, 17 males and 8 females. Table 1. shows the clinical presentation of the study group. The age at the time of the pull-through ranged from 2 months to 5 years (mean age 16 months). Transition zone was at recto sigmoid junction in 21 cases (84%), the other 4 cases were long segment HD (16%). Swenson's operation

was performed for 12 cases while Wang's modification was performed for 13 cases. Table 2. shows postoperative results of both groups.

Soiling occurred in 6 cases of Swenson's group and in only one case of Wang's group. Urine retention with overflow occurred in one case of Swenson's group that was managed with re-insertion of the catheter for one week. Anastomotic leak occurred in one case of each group on the 4th and 5th day and treated with urgent divided loop colostomy. Residual constipation occurred in a single case of Wang's group, motions occurred every 3-5 days. In this case there was no residual aganglionosis or anastomotic stricture. The case was subjected to daily rectal irrigation and laxatives for one month and then laxatives only for another month. Post operative Hirschsprung's enterocolitis occurred in 3 cases of each group. These cases were re-admitted and received the conservative regimen for management and no colostomy was needed for any of them. No cases of residual aganglionosis or anastomotic stricture have been noted. 6 months after operation, the mean resting and squeeze pressure for Swenson's group were significantly lower when compared to those of Wang's group, Table 3.

DISCUSSION

For an operation to cure Hirschsprung's disease, it must satisfy two criteria: the first is eradication of the aganglionic segment and the second is the preservation of fecal continence.⁽¹⁷⁾

HD is a serious condition but is not a malignant disease in the usual sense of the term, therefore an operation designed to remedy the condition should not be of excessive severity and should carry no risk that one functional anomaly will be substituted for another.⁽¹⁸⁾

The incidence of soiling after Swenson's operation in a series of 45 patients performed by Puri and Nixon⁽¹⁹⁾ was (46.4%). Similarly, it was 50% in our series, which is significantly higher when compared with that of post Wang's operation which was only (7.6%), and the difference is attributed to excessive excision of the internal anal sphincter in the former technique. This in reverse to Wang's modification that preserves the internal anal sphincter so avoiding soiling and at the same time make a posterior incision in the sphincter thus avoiding its spasm.

The incidence of soiling after Wang's et al⁽¹⁴⁾ in a series of 40 patients under went his modification was only 5% and corrected with diet alterations.

Table 1. Shows the clinical presentation of the cases.

Presentation	Swenson (12)	Wang's (13)
Delayed passage of meconium	11 (91.6%)	11 (84.6%)
Constipation	12 (100%)	13 (100%)
Absence of spontaneous evacuation	6 (50%)	8 (61.5%)
Enterocolitis	4 (33.3%)	3 (23.1%)
Soiling*	0 (0%)	0 (0%)
Attacks of intest. Obst.	1 (8.3%)	2 (15.3%)
Associated anomalies:		
Down syndrome	2 (16.6%)	-
Cardiac anomalies	1 (8.3%)	
VSD		
P.S		1 (7.6%)
Other anomalies		
Hypospadias	-	2 (15.3%)
Undescended testis	-	1 (7.6%)
Inguinal hernia	2 (16.6%)	1 (7.6%)

* Soiling: Staining of the underpants with fecal matter either consciously or involuntarily.

Table 2. Postoperative results in both groups.

	Swenson (12)	Wang's (13)	Chi-square
Soiling	6 (50%)	1 (7.6%)	P= 0.027*
Urine retention	1 (8.3%)	-	P= 0.48
Wound infection	1 (8.3%)	2 (15.3%)	P= 0.531
Residual aganglionosis	-	-	NA
Anastomotic leak	1 (8.3%)	1 (7.6%)	P= 0.74
Anastomotic stricture	-	-	NA
Residual constipation	-	1 (7.6%)	P= 0.25
Enterocolitis	3 (25%)	3 (23.1%)	P= 0.637
Death	-	-	NA

NA: Not applicable

* Significant values

Table 3. Comparison of postoperative resting and squeeze pressure of both groups.

	Swenson's group (n= 12)	Wang's group (n= 13)	Student t test
Resting p. (mmHg)	40.8±2.30	49.8±7.31	P= 0.0001*
Squeeze pressure	78.0±7.21	90.8±2.80	P= 0.0001*
RAIR	Absent	Absent	

* Significant value



Fig 1. HD, colon mobilization
Ordinary segment HD

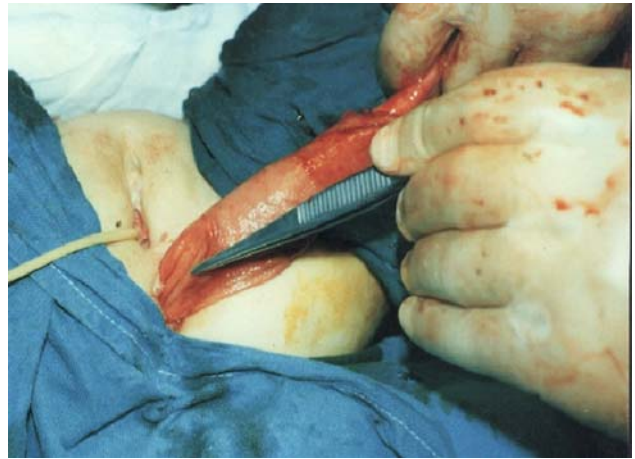
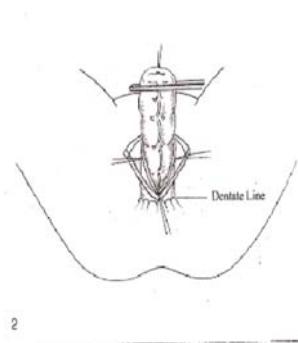


Fig 2. Swenson's operation, rectal mobilization till the dentate line



Fig 3. Wang's operation, rectum transected 4cm above the dentate line

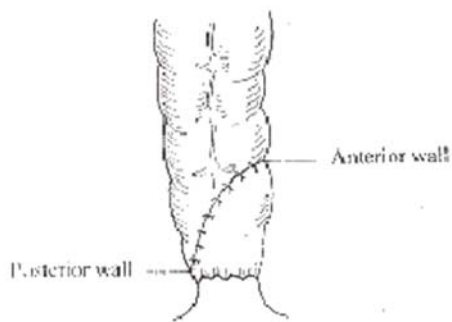


A



B

Fig 4a,b. Wang's operation rectum splitted posteriorly



A



B

Fig 5a,b. Wang's operation, heart-shaped anastomosis completed

One case in our series has suffered from post Wang's constipation (7.6%). Similarly one case after Wang's et al series (2.5%)⁽¹⁴⁾ suffered from intermittent constipation that was treated with enemas and laxatives.

Anastomotic leak occurred in one case of Wang's group of our series for whom diverting colostomy was performed. There was a size discrepancy between the proximal ganglionated colon and the distal heart-shaped, splitted rectum. No cases of anastomotic leak have been reported by Wang's et al.⁽¹⁴⁾

In our series urine retention occurred in one case of post Swenson's group which may be attributed to nerve concussion during anterior dissection of the rectum. Such a complication has not been seen after Wang's operation group due to limited pelvic dissection.

In our series, postoperative EC occurred in 3 cases of post Swenson group (25%) and also, in 3 cases of post Wang's modification group (22.8%). The difference is statistically insignificant. Likely, Coran⁽¹⁹⁾ documented that the incidence of postoperative EC is around 20% regardless of the type of pull-through performed. So, EC may be an intrinsic part of HD and is not an acquired condition after a successful pull through.

In conclusion Wang's modification maximally preserves the internal anal sphincter and so avoiding soiling, and at the same time making a posterior incision in this sphincter, so, avoiding constipation caused by this spastic sphincter. On the other hand, the heart-shaped anastomosis makes it safer as it minimizes the discrepancy between the proximal ganglionic dilated segment and the distal aganglionic, splitted rectum.

Lastly, anterior rectal dissection is avoided in Wang's operation. So, pelvic nerve injury is not reported with this technique.

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