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ORIGINAL ARTICLE

EVALUATION OF ANORECTAL MYECTOMY IN MANAGING ULTRASHORT SEGMENT OF HIRSCHSPRUNG'S DISEASE

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Aim: *The term ultra short is not clearly defined in ultrashort-segment Hirschsprung's disease. The limited extent of the ultrashort-segment Hirschsprung's disease allows for the treatment with extended sphincteromyectomy. This procedure could be diagnostic and therapeutic one. The study was designed to evaluate the role of posterior myectomy in children with refractory constipation in the diagnosis and treatment of ultrashort-segment Hirschsprung's disease.*
Methods: *A descriptive cross sectional study was carried out among 40 pediatric patients (1 – 12 years) with refractory constipation without obvious organic causes. Detailed history, examination, barium enema and anorectal manometry were performed to all children. Posterior anorectal myectomy was performed to the studied patients. The outcome and postoperative complications were assessed for all children.*
Results: *Agangliosis was diagnosed in 24 patients (60%) among whom constipation was improved in 75% while it was improved among (50%) of the ganglionic group. Abdominal distension, soiling and anorectal manometric studies were markedly improved after surgery in both aganglionic and ganglionic groups. Rare postoperative complications were reported among only five patients (12.5%).*
Conclusion: *Anorectal myectomy is an effective procedure in managing refractory constipation in pediatrics and also for the diagnosis of Ultrashort segment Hirschsprung's disease and is curative in such cases.*

Keywords: *Constipation, megacolon, aganglionosis.*

INTRODUCTION

Constipation is a common problem in the pediatric population, which can usually be treated with dietary modifications and laxative therapy. However, some children have refractory symptoms, and when subjected to high-dose laxative therapy, develop abdominal distension, vomiting, cramping, and bloating.[1] There have been many reported operative strategies for the management of severe constipation. Hirschsprung's disease is one of the conditions that require surgical correction.[2]

The disease is considered the most important type of gastrointestinal dysmotility in neonatal pathology. Aberrant craniocaudal migration of neural crest stem cells results in an intestinal aganglionic segment of variable length. In 'classical' Hirschsprung's disease (60-75% of cases), the aganglionic segment spans the rectum and sigmoid colon. Ultrashort Hirschsprung's disease is restricted to the most distal 3-4 cm.[3] It is not a rare disease. Data on its incidence vary considerably because the term ultrashort is not clearly defined.[4] Although this type of the disease was enzyme-histochemically characterized about 40 years ago, its existence is still often ignored by some authors[5] while other authors used to refer to this condition as internal anal sphincter achalasia.[6]

In 1960s, Ultrashort segment Hirschsprung's disease began to be discussed as a cause of refractory constipation in infancy and childhood. The postulated length of the ultrashort segment varies from 2 to l0 cm but most researchers believe that the term should be restricted to the lower most 2 to 4 cm.[7] It is evident that the length of the aganglionic segment does not correlate with the propulsive capacity of the bowel.[8] However, this type of the disease results in poor quality of life and delayed social development.[9]

Once the diagnosis is established, the basic treatment is to remove the poorly functioning aganglionic bowel and create an anastomosis to distal rectum with the healthy innervated bowel (with or without an initial diversion).[10] Many operations have been used to manage Hirschsprung's disease including fecal diversion.[11] and transabdominal resection of the rectum and sigmoid.[12-15] Other less invasive techniques have been described, such as antegrade continence enemas[16,17] botulinum toxin injections [18], and internal sphincter myectomy.[19]

In the ultrashort-segment Hirschsprung's disease, the therapy of choice has proven to be a partial myectomy of the distal internal sphincter. It carries minimal risk and requires little preparation. Generally, it is not difficult to convince parents to agree to it. It has been used for short aganglionosis, both as a definitive and diagnostic procedure.[5,20]

Several techniques have been described for anorectal myectomy[20-23] but all depend on excision of a longitudinal strip from the internal sphincter for a variable distance with variable approaches. This study was designed to evaluate the role of posterior myectomy in children with refractory constipation for the diagnosis and treatment of Ultrashort-segment Hirschsprung's disease.

PATIENTS AND METHODS

This study was carried out as a descriptive cross-sectional study. Over a period of 18 months (August 2007- February 2009) and after approval of our ethics committee, 40 patients with refractory constipation were recruited from pediatric surgery unit of Suez Canal University Hospital in Ismailia, Egypt. Patients of both sexes in age group from 1 to 12 years old suffering from severe refractory constipation without obvious organic cause and not responding to medical treatment with chronic use of laxatives were included in the study.

Exclusion criteria included patients proved to have long segment affected with Hirschsprung's disease, patients with constipation due to other obvious causes and patients with neurological deficits or any congenital anorectal anomaly.

All of the included children were subjected to detailed medical history, focusing on any delayed or difficult passage of meconium. Any diarrhea like illness was recorded, especially if associated with previous constipation, chronic use of laxatives or abdominal distension. Full general examination, abdominal examination and per rectal examination were done to evaluate the general condition of the patient, to detect any abdominal distension and to assess the sphincter tone and faecal loading.

Barium enema and anorectal manometry were an integral part of the study. Barium enemas were done without preparation while anorectal manometry was carried out by a pre-calibrated, water perfusion system using a catheter with 8 holes 0.5 cm apart (Andorfer Inc. Wisconsin, USA). A motility measurement unit was connected to the catheter and infusion system to provide digital readouts and a printed copy of pressures. All children had routine preoperative investigations before subjected to posterior anorectal myectomy.

Anorectal myectomy: All patients underwent posterior anorectal myectomy using the technique described by Scobie and Mackinlay (1977) [22]. No special bowel preparation was carried out. Antibiotics were prescribed before and after the operation. Under general anesthesia, with the patient in lithotomy position, a transanal approach was used to do the operation. Mild digital dilatation was performed and then two stay sutures were held on both sides of the anus (Fig. 1).

A curvilinear transverse incision was made at the mucocutaneous junction on the posterior wall of the anal canal. The internal sphincter is identified. In the posterior midline, a plane is established between mucosa and muscle and the surgeon started his dissection in this submucosal plane (Fig. 2). A muscle strip 0.5-1 cm width and 4 to 7 cm. length was excised. The excised strip is correctly orientated, laid on a foil sheet and sent to the pathology department (Fig. 3). The transverse incision is closed with interrupted polygalactin suture (Fig. 4). Patients were able to go home on the second postoperative day. Follow-up and incidence of postoperative complications was accomplished by regular visits to the out-patient clinic of surgery over 3 months postoperatively.

Gathered data were processed using SPSS version 15 (SPSS Inc., Chicago, IL, USA). Quantitative data were expressed as means ± SD while qualitative data were expressed as numbers and percentages (%). Unpaired t test was used to test significance of difference for quantitative variables while Chi Square test was used to test significance of difference for qualitative variables. A probability value (p-value) < 0.05 was considered statistically significant.

RESULTS

Analysis of the sociodemograhic characteristics of our studied patients has revealed that twenty five patients (62.5%) were males and fifteen (37.5%) were females. The mean age of the studied patients was 5.2 years. Fourteen patients (35%) were preschool aged (< 5 years) while 26 patients (65%) were school aged (5-12 years). The most common symptoms were constipation which was found in all patients (100%), abdominal distention in 24 patients (60%), soiling in 4 patients (10%) and failure to thrive in 1 patient (2.5%).

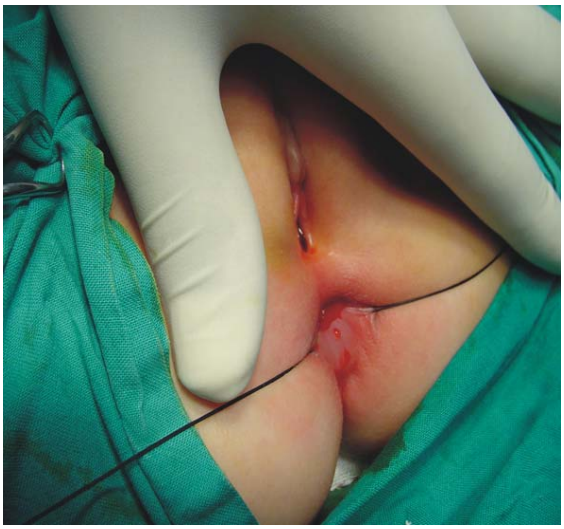

Fig 1. Two stay sutures at both sides of the anus before the operation.

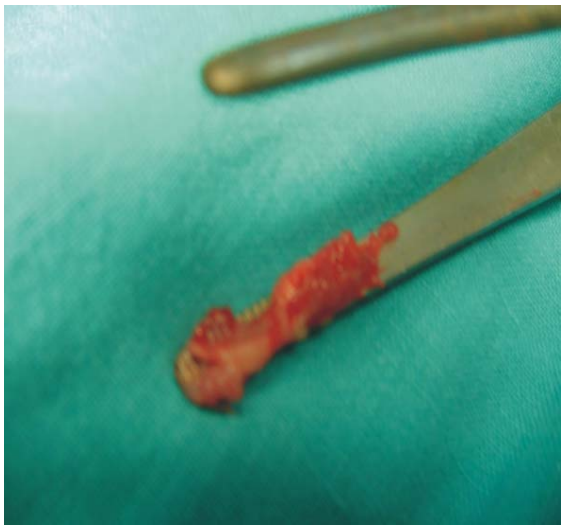

Fig 3. The excised muscle stripe.

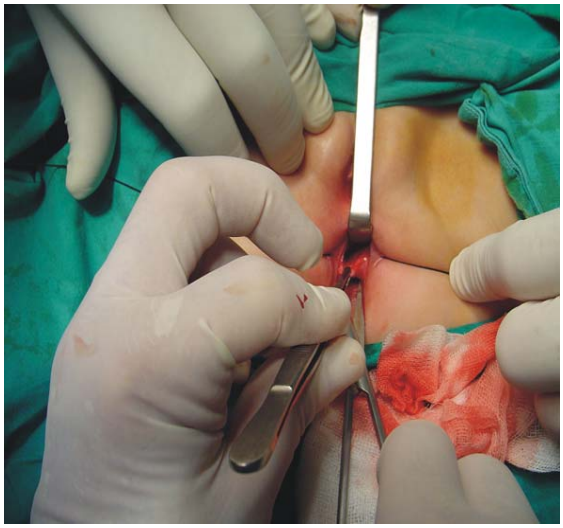

Fig 2. Dissection of the muscle in the sub mucosal plane.

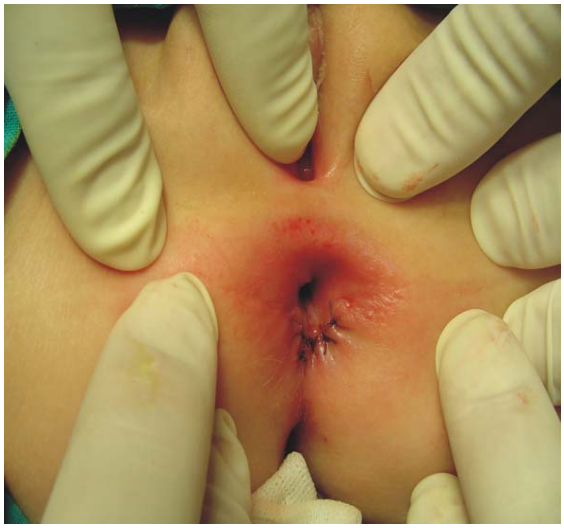

Fig. 4. The anus after closure.

Table 1. Results of rectal biopsy and the outcome after surgery.

		Aganlionic group *n*=24		Ganglionic group *n*=16	
		Preoperative No (%)	**Postoperative No (%)**	**Preoperative No (%)**	**Postoperative No (%)**
Clinical	Constipation	24 (100%)	6 (25%)	16 (100%)	8 (50%)
	Distension	10 (42%)	3 (12.5%)	14 (88%)	8 (50%)
	Soiling	2 (8%)	1 (4%)	2 (13%)	1 (6%)
	Failure to thrive	1 (4%)	1 (4%)	0 (0%)	0 (0%)
Manometric studies	Max. resting anal pressure	77.6 ± 11.3	58.6 ± 8.1	73± 9.3	59.2± 10.5
	Absence of recto-anal inhibitory reflex	22 (92%)	13 (54%)	15 (94%)	6 (38%)

Muscle strip biopsy and histopathological examination revealed aganglionosis and diagnosis of ultrashort-segment Hirschsprung's disease in 24 patients (60%). 16 patients of them (66.6%) were males and 8 patients (33.3%) were females (Male: female ratio of 2: 1). The specimen was totally aganglionic in four patients (16.7%) while it was partially aganglionic in 20 patients (83.3%). The partially aganglionic specimen resemble the total aganglionic one except for the presence of few ganglia in its proximal end, a feature characteristic to ultrashort-segment variety of Hirschsprung's disease.

Anorectal myectomy resulted in improvement of constipation in 18 patients (75%) of the aganglionic group and only 8 patients (50%) in the ganglionic group. In the aganglionic group, distension and soiling were reported in ten patients (42%) and two patients (8%) respectively in the preoperative assessment while they were found in three patients (12.5%) and one patient (4%) respectively in the postoperative assessment. The same occurred in the ganglionic group but in a lesser degree as they were found in fourteen patients (88%) and two patients (13%) preoperatively and decreased to be found in 8 patients (50%) and one patient (6%) postoperatively. There was one child who failed to thrive. The histopathological examination of his biopsy revealed total aganglionosis. He was not improved after the operation Table 1.

The incidence of postoperative complications is low in the study; Only 1 patient (2.5%) developed intersphincteric abscess, three patients (7.5%) reported transient incontinence of flatus after surgery while only one patient (2.5%) showed persistent minor fecal incontinence Table 2.

Table 2. Postoperative complications.

Complication	Number	Percent
Intersphincteric abscess	1	2.5%
Transient flatus incontinence	3	7.5%
Persistent minor fecal incontinence	1	2.5%

The anorectal manometric studies showed significant improvement after surgery. There was significant fall in the maximum resting anal pressure from 77.6 to 58.6 cmH2O in the aganglionic group (p-value < 0.05) and from 73± 9.3 to 59.2± 10.5 in the ganglionic group (p-value < 0.05). However maximum squeeze pressure didn't change significantly after operation in both groups. In the aganglionic group, the recto-anal inhibitory reflex was absent in 22 child (92%) before the operation while, postoperatively, there was marked improvement as it was absent in only 13 child (54%) (p-value < 0.05). In the ganglionic group, the reflex was absent in fifteen patients (94%) preoperatively, and six patients (38%) postoperatively (p-value < 0.05) Table 1.

DISCUSSION

Hirschsprung's disease is a congenital absence of Meissner's and Auerbach's plexuses of the colon. The disease was first described in 1886 and reported in 1888 by Harald Hirschsprung, a Danish pediatrician.[24] Unlike classic Hirschsprung's disease, ultrashort-segment variety of this disorder is usually latent and milder. Children over one year of age, adolescents, and adults who suffer from intractable chronic constipation may have this disease.[25]

Lynn[26] proposed anorectal myectomy for treatment of this entity in 1966. Several terms have been used in the literature for this procedure: anorectal myectomy, anorectal myomectomy, anal myectomy, posterior internal myectomy, posterior internal anal myectomy, posterior rectal myectomy and extended sphincteromyotomy. It has been suggested that the procedure might be most beneficial in patients with short-segment Hirschsprung's disease, the suggestion that is being supported by our data.

Males were found to be more affected by the disease with a male: female ratio of 2:1. This is similar to findings reported by Heikkinen et al[6] and Ahmadi et al[7] who reported male: female ratio of 5: 1.

Constipation is the only symptom that present in all patients which is in agreement with many previous studies.[8,19,23] The second most commonly presenting symptom was abdominal distention (60%). Ahmadi et al[7] reported abdominal distention in 67.8% of his cases while Udassin et al[8] reported it in 29% of his series. There are other less frequent symptoms like soiling and failure to thrive. Other studies reported rare symptoms that are not encountered in our work such as vomiting and enterocolitis.[7]

In our study, total aganglionic specimens represent 10% while partially ganglionic and ganglionic specimens represent 50 % and 40% respectively. Scobie and Mackinlay[22] reported different figures as they record 34.6% total aganglionic, 46.2% partially ganglionic and 19.2 ganglionic specimens.

Improvement of constipation was noted in 75% of aganglionic cases and in 50% of ganglionic cases with no significant difference. Pinho et al[19] had less favorable results as they reported improvement in 50% of aganglionic cases and in 31% of ganglionic cases.

Ahmadi and associates[7] classified patients according to presence of ganglionic cells into 3 groups; group A (normal ganglion cells in proximal and distal ends of muscle stripe), group B (no ganglion cells in both proximal and distal ends) and group C (normal ganglion cells only in proximal end). They reported no meaningful difference in the results of surgery between groups A, B and C, and they got benefit from dilatation and anorectal myectomy.

The reported complications after surgery were rare and mostly transient except for one patient with persistent minor fecal incontinence. The same was observed by Shehata and his colleagues[23] who did not find immediate postoperative complications except for one patient who developed perianal abscess at the myectomy site.

The significant improvement of the anorectal manometric results in the ganglionic groups was similar to that reported by Heikkinen and his team[6] who showed an absence of rectoanal reflex in all his patients before the operation. The same was recorded by Yoshioka and Keighley[27] who evaluated anorectal myectomy in 29 patients (eight with aganglionic specimens and 21 with ganglionic specimens) They found a significant fall in maximum resting anal pressure after operation in 62% of the patients (102.7 ± 16.7 to 79.5 ± 27.8 cm H2O). The operation had no influence on squeeze pressures.

In conclusion we found that anorectal myectomy gives acceptable results when used to treat the ultrashort-segment Hirschsprung's disease. It is a satisfactory procedure with minor transient complications.

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