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Importance of anorectal manometry after definitive surgery for Hirschsprung's disease in children

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ABSTRACT

Objectives: The purpose of this investigation is to evaluate anorectal function after definitive surgery for Hirschsprung's disease (HD) by anorectal manometry. **Materials and Methods:** We evaluated the anorectal manometric assessment of 18 children who were operated for HD. Functional outcomes were determined by a questionnaire. Rectoanal inhibitory reflex (RAIR) and maximum anal resting pressure (MARP) were monitored. The results were compared between obstructive patients and asymptomatic patients. **Results:** The median age at definitive operation was 19 months (range 12–72 months). Anorectal manometry was performed in 14 male and 4 female patients. All the cases underwent three staged procedure for HD and modified Duhamel procedure was performed as definitive procedure for all the patients. Mean age was 4.3 months (range 25 days to 5 years) at time of diagnosis. Post-operative enterocolitis or severe constipation was observed in seven patients (38.8%). There were no patients with incontinence. Eighteen patients underwent anorectal manometry meanly 2 years after definitive operation. RAIR was absent in 14 (77.7%) patients and abnormal in 4 (22.2%). There were no significant differences in the MARP values between symptomatic and asymptomatic patients. **Conclusion:** The results of our study showed that the majority of the patients have impaired anorectal motility. There were no significant differences in the results of the functional studies for the seven patients with symptoms of obstruction or constipation when compared with asymptomatic patients after surgery for HD.

Key words: Hirschsprung's disease, anorectal manometry, constipation, enterocolitis, complications

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INTRODUCTION

Hirschsprung's disease (HD) is characterised by absence of ganglion cells in the myenteric and submucosal plexus on rectal biopsy. Surgery for HD generally generates satisfactory outcome, unfortunately some patients continue to have persistent bowel dysfunction such as constipation and intestinal motility disturbances.[1-3] The reasons of persistent symptoms are always not clear. Appropriate management of these complications depends on the aetiology of the problem.[3]

Post-operative anorectal manometric evaluation of the patients after surgery gives detailed information for function of anal canal and rectum. Determining the cause of persistent symptoms and treatment of these children can become more rational.[3-12] The purpose of this study is to evaluate the importance of anorectal manometry as a diagnostic tool, for understanding the obstructive symptoms after definitive surgery for HD.

MATERIALS AND METHODS

We reviewed anorectal manometric evaluation of 18 children who were operated for HD between 2000 and 2003. The diagnosis of the HD was based on clinical features, radiology and also confirmed by rectal biopsy. All patients underwent a staged procedure with initial colostomy.

Clinical information about previous treatment was taken from the medical records. The patients and their parents were interviewed by a detailed questionnaire. Questions were asked about stool frequency, consistency, continence, faecal control, faecal soiling, incontinence and other related gastrointestinal symptoms. We classified patients according to results of the questionnaire into two groups: Patients in group A (n = 7) had enterocolitis or constipation. Patients in group B (n = 11) had normal gastrointestinal function. In group A, patients were investigated for

possible anal stenosis, large Duhamel pouch, residual aganglionic segment and intestinal neuronal dysplasia. For that reason physical examination of perineum, anorectal manometry, barium enema, re-evaluating of the pathological samples and rectal biopsy (only one patient) were performed in group A. Anorectal manometry was performed in 14 male and 4 female patients. A computerised manometric technique (Syntic, Enfield, UK) Polygram-lower GI-tract, version 6.4, capillary water pump, 63 kPa) with a four-channel perfusion catheter was applied. During the anorectal manometry, maximal anal resting pressure (MARP) and rectoanal inhibitory reflex (RAIR) were recorded. Manometry was performed while the patient was at rest in the left lateral decubitus position.

The results were compared between the obstructive group and with the asymptomatic patients. Mann–Whitney U test was used for statistical analysis of the data. A "*P*" value of less than 0.05 was considered significant.

RESULTS

There were 18 children in the study (14 males and 4 females). Modified Duhamel procedure was performed as definitive procedure for all patients.

Seven of the patients presented within the neonatal period and, eight presented between 1 and 12 months of age. The three remaining patients presented between 1 and 5 years of age. Mean age was 4.3 months (25 days to 5 years) at time of diagnosis. Presenting symptoms included abdominal distention, constipation and vomiting. One infant presented with intestinal perforation.

The localisation of the aganglionic segment was rectosigmoid in 14, long colonic in 3 and with a colonic atresia in 1 patient. The median age at definitive operation was 19 months (range 12–72 months). Mean length of follow-up was 2 years, with the range of 4–42 months [Table 1].

All of the patients have anal opening with normal calibrer. With barium enema, Duhamel pouch size and configuration were normal. In pathological re-examination, HD associated with Intestinal neuronal dysplasia (IND) was reported in one patient. The diagnosis was confirmed by a new rectal biopsy for this patient.

In the anorectal manometry, four (22.2%) children had abnormal pattern of relaxation, which had confluent changing of RAIR in response to inflation of rectal balloon. RAIR was absent in 14 (77.7%) children. Maximum anal resting pressure (MARP) was 55 (11 cm H_2O in Group A, and 49 (12 cm H_2O in Group B [Table 2]. For the MARP values, there was no significant difference between the groups in (pP > 0.05).

There was no significant difference in faecal continence, sensation, control, soiling between the seven patients with symptoms of obstruction and the asymptomatic patients after surgery for HD ($P > 0.05$).

DISCUSSION

Many children with Hirschsprung's diseaseHD have good surgical results, however, long-term follow up studies expressed several ongoing problems.[1-4] The main problem following surgery for HD, which must be solved is constipation. It is not clear why some children do well after surgery for HD, and others have not. Obstructive symptoms may be related with persisting abnormalities of the enteric plexus, which characterizsed with impaired colonic propulsive forces or anorectal dyssynergia. These anomalies represent an important disturbance of function within the spectrum of obstructive or spastic pelvic floor disorders.[5] It is widely known that the majority of patients, who have severe constipation after operation for HD, have anorectal dyssynergia. Paradoxical contractions,

Table 1: Patient characteristics (*n* = 18)

	Number (%)
Gender	
Male	14 (77.7)
Female	4 (22.2)
Presenting symptoms at diagnosis	
Abdominal distension	11 (61.1)
Constipation	9 (50)
Enterocolitis	3 (16.6)
Intestinal perforation	1 (5.5)
Extent of disease	
Rectosigmoid	14 (77.7)
Long colonic	3 (16.6)
Total colonic	1 (5.5)
Age at diagnosis (mean)	4.3 months
Type of operation	
Duhamel procedure	18 (100)

Table 2: Maximal anal resting pressure (MARP) values of the groups

Group (*n* = 18)	MARP
Group A (*n* = 7)	55 ± 11 cm H_2O
Group B (*n* = 11)	49 ± 12 cm H_2O

impaired colonic propulsion and impaired anal relaxation are diagnostic parameters for anorectal dyssynergia that are shown by anorectal manometry in adult constipated patients.[13,14] However, this study was not performed in children. In some children, the reason of obstructive symptoms may be caused by anal stenosis, large Duhamel pouch, a residual aganglionic segment or intestinal neuronal dysplasia.[4-6,13-14] In our study, we did not't find any anal stenoses, large Duhamel pouch or a residual aganglionic segment. In group A, one patient has IND associated with HD.

The anorectal manometry test helps us to understand the essential pathophysiological process that is responsible for the persistent symptoms.[4-12] This information letsallows us to understand the compliance and motility of rectum and responds of anal sphincter against rectal distension.[4-6,9,15-16] The use of anorectal manometry and early rectal biopsy had decreased to 2.6 months to the diagnosis for HD.[12] In normal individuals, rectal distension inhibits internal anal sphincter pressure by causing a transient pressure decrease in anal canal called the rectoanal inhibitory reflex (RAIR). Specifically, the absence of RAIR is characterizsed for the HD.[5,7,8] There is no agreement with the absence or the presence of RAIR during anorectal manometry after surgical procedures for HD.[2,3,9] It was reported that a normal RAIR could develop in children after a pull-through operation, though the majority continue to have abnormal reflex.[7-10,12] We did not observe normal RAIR in our patients post-operatively.

MARP values did not standardizsed for children. Nagasaki *et al*. reported that the MARP as 13.7 (5.6 cm H_2O was normal in children.[15] In an other study, MARP was reported as 50.7 cm H_2O in normal children.[17]. Keshtgar *et al*. reported the MARP values of 66 mmHg in constipated patients after surgery for HD.[5] In our opinion , the differences in normal MARP values in children may be attributed to some factors such as, age of childrens, differences in analyzing techniques and devices, physchological status of the children and healthcare providers in charge of the procedures. In our study, we measured the MARP values as was 55 (11 cm H_2O in Group A and 49 (12 cmH_2O in Group B. Our MARP values may be accepted in normal range. No clear difference in MARP was noted between patients with symptoms of obstruction, and asymptomatic patients.

Increase of the internal anal sphincter pressure does not explain the obstructive symptoms. Even though the anorectal inhibitory reflex is considered mandatory for continence and defecation, its absence after anal or rectal anastomosis is not always accompanied by incontinence or constipation.[15-21]

In our study, there were no significant differences in the results of the functional studies of the seven patients with symptoms of obstruction and constipation when compared towith the asymptomatic patients after surgery for Hirschsprung's disease HD.

Despite the fact that this study, which had a low number of cases, we conclude that classical manometric evaluation of the anus after surgical procedures for HD does not give enough information for understanding the cause of obstructive symptoms. The studies, which are designed for diagnosis of anorectal dyssynergia and pelvic floor disturbances, may give some important information for understanding of obstructive symptoms.

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