STUDY OF OUTCOME AND COMPLICATIONS OF ANORECTAL MYECTOMY IN CHILDREN WITH ULTRASHORT SEGMENT HIRSCHSPRUNG’S DISEASE

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Abstract- 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 treatment with extended sphincteromyectomy. In anal sphincter achalasia, anal sphincter dilatation under general anesthesia may be sufficient to treat the condition; in cases with persistent constipation, sphincteromyectomy is indicated. Some investigators believe that the term ultrashort-segment Hirschsprung’s disease and anorectal achalasia are the same. Our study was performed to define the efficacy of transanal anorectal myectomy and digital dilation under general anesthesia in children with ultrashort-segment Hirschsprung’s disease and internal anal sphincter achalasia. A total of 87 patients were included in our study. Among these, 15 cases (17.24%) were female and 72 (82.76%) were male. In 12 patients (13.79%), the muscle strip had normal ganglion cells in both distal and proximal ends (group A). In 10 patients (11.49%), there was not any ganglion cell in both distal and proximal ends of muscle strip (group B). In 65 patients (74.71%), there were normal ganglion cells in proximal end with no ganglion cell in distal end of the muscle strip (group C). There was no meaningful differences between group A, B and C in their outcome and partially or complete response to anorectal myectomy. We recommend the term “sluggish rectum” for these patients instead of ultrashort-segment Hirschsprung’s disease or internal anal sphincter achalasia that causes ambiguity in diagnosis and treatment of these cases.

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INTRODUCTION

Ultrashort-segment Hirschsprung’s disease is not rare; however, data on the incidence vary considerably because the term ultrashort is not clearly defined (1).

The postulated length of the ultrashort segment varies from 2 to 10 cm (1). Most researchers believe that the term should be restricted to the lower most 2 to 4 cm of the anal channel above the mucocutaneous line which means the a ganglionic segment comprises the lower half of the rectum form the dentate line upward to the third sacral vertebra. Shorter distances restricted to the internal anal
sphincter are referred to as neurogenic anal sphincter achalasia (1).

Detailed reviews of this disorder identify some common characteristics, including a history of chronic constipation, the presence of ganglion cells on suction rectal biopsy at 3 and 5 cm and a contrast enema without a transition zone; and despite these findings but that are not diagnostic for Hirschsprung’s disease they present lack of anorectal reflex relaxation with rectal distention in anorectal manometry (2).

However, there are some children with ganglion cells on rectal biopsy that also present lack of inhibitory reflex and may hence develop obstructive symptoms that resemble those of Hirschsprung's disease (3). This condition has been termed internal anal sphincter achalasia (3).

Anorectal myectomy provides long-term relief of this chronic problem in a subgroup of patients with ultrashort-segment Hirschsprung’s diseases who lack nitrinergic neurons at the internal anal sphincter.

Our study was performed to define the efficacy of transanal anorectal myectomy and digital dilation under general anesthesia in children with ultra short-segment Hirschsprung’s disease and internal anal sphincter achalasia.

**MATERIALS AND METHODS**

This study was designed as longitudinal analytical one. We studied 122 cases which had undergone anorectal myectomy and anal sphincter dilatation under general anesthesia from May 1995 to May 2005 in Pediatric Medical Center of Tehran University, Tehran Pediatric Center, Mehr Hospital and Amirkabir Hospital, Iran. A total of 87 patients were included in our study. These patients had a history of chronic constipation with no response to conservative management, and they did not have anal stenosis or anal malposition or any abnormality in pelvis or perineum. They had a contrast enema without transition zone and a manometry showed failure reflex relaxation with rectal distention.

Transanal anorectal myectomy was done in lithotomy position under general anesthesia. At first, digital dilatation was performed, and two narrow anal retractors were inserted and held by assistants (Fig. 1). A 2 cm transverse incision was made through the mucosa and submucosa, starting 1 cm above the dentate line (Fig. 2). Myectomy was done by sharply incising the full-thickness of the muscle layer followed by the Bovie cautery to excise a 0.5 to 1 cm wide muscle strip in the midline. The initial strip that was removed had to be at least 5 cm in length (Fig. 3). The muscle strip was examined for ganglion cells; if it was ganglionic, the operation was finished and if it was aganglionic the myectomy was continued to maximum of 8 cm. The proximal site was examined for ganglion cells again and the operation was finished (Fig. 4).

**Fig. 1.** Two narrow anal retractors are inserted.

**Fig. 2.** 2 cm transverse incision 1 cm above dentate line.

**Fig. 3.** Initial strip myectomy at least 5 cm.
RESULTS

Out of 122 cases, 35 patients were excluded because of insufficient data and 87 patients were included in our study. 35 cases (17.24%) were female and 72 cases (82.76%) were male. Male to female ratio was 5 to 1. They were from 14 months to 12 years old. Their symptoms were constipation in 87 cases (100%), obstipation in 85 cases (97.7%), abdominal distention in 59 cases (67.8%), soiling in 10 case (11.5%), failure to thrive (FTT) in 7 cases (8%), vomiting in 5 cases (5.7%) and enterocolitis in 3 cases (3.5%) (Fig. 5). Complications after surgery were persistent symptoms in 5 cases (5.74%), rectal bleeding in 2 cases (2.3%), soiling in 2 cases (2.3%), and gas incontinences in 1 case (1.15%). Totally, 79 cases (90.80) got some degree of benefit from anorectal myectomy. 66 cases became symptom free and in 13 cases (14.94%), the symptoms became milder, in the way that the use of laxative was limited to once weekly up to once monthly rather than daily consumption. In 12 patients (13.79%) the muscle strip had normal ganglion cells in both distal and proximal ends (group A). In 10 patients (11.49%), there was not any ganglion cell in both distal and proximal ends of muscle strip (group B).

In 65 patients (47.71%) there was not any ganglion cell in distal end and there were normal ganglion cells in proximal end (group C). 9 cases (75%) had complete and 2 cases (16.6%) had partial response to anorectal myectomy from 12 cases in group A. 8 cases (80%) had complete and 1 case (10%) had partial response to anorectal myectomy from 10 cases in group B. 49 cases (75.38%) had complete and 10 cases (15.38%) had partial response to anorectal myectomy from 65 cases in group C.

In our study there was no meaningful difference in the results of surgery between groups A, B and C (\(P = 0.98\) in Kruskal-Wallis test, \(P = 0.99\) in Fisher's exact test)

DISCUSSION

The term ultrashort-segment Hirschsprung’s disease is not clearly defined. Some authors believe that the postulated length of the ultrashort segment Hirschsprung’s disease varies from 2 cm to 10 cm (1). Some investigators believe that the term of ultrashort Hirschsprung’s disease and anorectal achalasia are the same (2). On the other hand some researchers believe that internal anal sphincter achalasia (IASA) is defined as the inability of the internal anal sphincter to relax (3). Some believe that modification of criteria to allow up to a 4 cm segment of aganglionosis for the diagnosis or ultrashort segment Hirschsprung’s disease would eliminate some of the current ambiguity (4). Some researchers believe that the aganglionic segment has an extension of 1-3 cm (5).

IASA may represent as an isolated disease of the ENS, or it may be an acquired condition of psychological (functional) origin with normal innervation patterns. However, it may also be part of HD or allied disorders (Neurogenic anal sphincter Achalasia) (1). Ultrashort-segment Hirschsprung’s disease is an underlying cause in some diseases for example rectal prolapse (6) and rectal fissure.

In ultrashort-segment HD the limited extent of the disease allows for treatment with extended sphincteromyotomy (1). In internal anal sphincter achalasia, dilatation under general anesthesia may be sufficient to treat the condition, in cases with persistent constipation sphincteromyectomy is indicated (2).

In our study there was no meaningful difference in the results of surgery between groups A, B and C (\(P = 0.98\) in Kruskal-Wallis test, \(P = 0.99\) in Fisher's exact test), and they got benefit from dilatation and
Anorectal myectomy in children

anorectal myectomy. All of these patients had 4 criteria:

1- History of chronic constipation with no response to conservative management.

2- Absence of any anatomical abnormality in pelvis and perineum.

3- Absence of transition zone in contrast enema.

4- Failure of relaxation reflex with rectal distention.

We had no mortality in group A, B and C and the complications were low and were solved by conservative management. Therefore we recommend dilatation under general anesthesia and anorectal myectomy in any child who has these 4 criteria, and we offer the term “sluggish rectum” for these patients instead of ultrashort segment Hirschsprung’s disease and internal anal sphincter achalasia that causes ambiguity in diagnosis and treatment.

Conflicts of interests

The authors declare that they have no competing interests.

REFERENCES


