

A COMPARATIVE STUDY OF THE SURGICAL PROCEDURES TO TREAT HIRSCHSPRUNG DISEASE IN CHILDREN

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Abstract

Purpose: The aim of this study was to compare the clinical outcomes of four surgical procedures to treat Hirschsprung's disease (HD) in children.

Materials and methods: One hundred thirty-eight children with HD were included in this retrospective study. The children underwent four types of pull-through procedures between 2015 and 2020. Sixty-one (44%) children were older than 3 years of age at the time of surgery. In this study, we evaluated the functional outcome by measuring the incidence of postoperative complications.

Results: The surgical procedure consisted of 90 transabdominal (modified Soave – n=72; Swenson procedure – n=8; Swenson-like – n=10) and 48 transanal endorectal pull-through (TEPT) procedures. Early postoperative complications were seen in 13 children and consisted of faecal incontinence (5; 3.6%), anastomotic stricture (4; 2.8%), and cuff abscess (2; 1.4%); one child had dysuria and anastomotic dehiscence (0.7%). Nine of these early postoperative complications were after modified Soave pull-through. Episodes of Hirschsprung-associated enterocolitis occurred more often after transabdominal pull-through procedures (14.4% vs 8.3%; $p=0.04$).

Conclusion: Children who underwent TEPT had a lower rate of postoperative complications, especially episodes of postoperative Hirschsprung-associated enterocolitis. Therefore, in the treatment of HD in children, preference should be given to TEPT, which is less traumatic, has better cosmetic outcomes and has low incidences of postoperative complications.

Key words: Hirschsprung disease, transabdominal pull-through, transanal endorectal pull-through, Hirschsprung-associated enterocolitis.

Introduction.

Hirschsprung's disease (HD), which involve the gastrointestinal tract, is one of the most common life-threatening congenital anomalies during the neonatal period. The estimated prevalence is 1:5000 live births. HD is characterized by the absence of ganglion cells in the submucosal and myenteric plexuses along variable lengths of the intestine [3, 4]. This results in functional intestinal obstruction due to a lack of peristalsis in the aganglionic segment. In developed countries, HD is most often diagnosed and treated during the first year of life. Several surgical procedures have been described to treat the disease [5-10]. Their main purpose is surgical resection of the aganglionic segment of the bowel with pull-through of a normal segment and anastomosis to the anus. However, despite the numerous methods of adequate resection of the aganglionic segment, pre- and postoperative complications are frequent, and overall mortality in low-middle income countries is currently 18% [11]. Postoperative complications such as faecal incontinence, dehiscence, retraction and constipation have an important impact on the quality of life in these children [12, 13]. The incidence of one of the most dangerous complications, enterocolitis, varied from 10% to 30% in different analyses [1, 2, 14, 23, 24]. Studies of HD diagnosed in older children support the hypothesis that there are more postoperative complications when surgery for HD is performed in neonates and infants [15]. Therefore, the **aim** of this study was to compare the clinical outcomes of four surgical procedures to treat Hirschsprung's disease in children.

Patients and methods. We retrospectively analysed the medical charts of 138 children <15 years of age with HD who underwent surgery at the Department of Pediatric Surgery at Tashkent Pediatric Medical Institute Hospital in Tashkent, Uzbekistan from 2015 to 2020. All patients were subsequently evaluated with clinical, laboratory and physical examination studies. Contrast enema was performed to demonstrate the transition zone. The HD diagnosis

was confirmed by intraoperative biopsy. One hundred thirty-eight children with HD underwent surgery during the study period. In this study, 61 (44%) children were older than 3 years of age. For these patients, we started bowel preparation one month prior to the surgical interventions. All surgeries were performed in a single institution by a senior paediatric surgeon.

In this study, we determined the functional outcome by measuring the incidence of postoperative complications, such as faecal incontinence, dehiscence, retraction and Hirschsprung-associated enterocolitis (HAEC). There were no patients with neurological disorders or impairment of neuropsychomotor development that affected sphincter control.

Statistical analyses were performed using SPSS version 23.0 software (SPSS Inc, Chicago, IL). Quantitative variables were expressed as medians and interquartile ranges. Comparisons between quantitative variables were performed using the *t* test. A *p* value below 0.05 was considered statistically significant.

Results. All 138 patients diagnosed with HD who underwent pull-through procedures were included. Ninety patients with HD had a transabdominal approach (modified Soave, Swenson and Swenson-like procedures). Transanal endorectal pull-through (TEPT) was performed in 48 patients with rectosigmoid form (Table 1).

Table 1. Demographic data and characteristics of the 138 patients with HD

	n (%)
Boys	99 (72%)
Girls	39 (28%)
Age	
0-3 years	77 (56%)
4-6 years	43 (31%)
7-10 years	11 (8%)
11 years and older	7 (5%)
Mode of repair, n (%)	
Swenson procedure	8 (6%)
Swenson-like procedure	10 (7%)
Modified Soave procedure	72 (52%)
TEPT	48 (35%)

Modified Soave pull-through was the most common operation and was performed in seventy-two patients. This procedure was performed transabdominally, in addition to the classic procedure after pull-through, the upper intestine was wrapped in the manner of a “cuff”, thereby increasing the density of the tissue connection. Among the patients who underwent modified Soave procedures, 48 (67%) were boys and 24 (33%) were girls, and the mean age at operation was 3.81 years ($SD \pm 0.3$, range 1-12). All children who underwent modified Soave operation had long segments of aganglionosis. In the postoperative period, faecal incontinence occurred in 5 (6.9%) patients, anastomosis strictures occurred in 4 (5, 5%) (fig. 1) patients, and two children with HD after modified Soave pull-through (2.8%) had cuff abscesses. Episodes of HAEC after modified Soave pull-through were observed in 10 (13.8%) children.



Fig. 1 Contrast enema of a patient with a dilated colon after modified Soave pull-through

Eight patients underwent the Swenson procedure. The median age at operation was 3.75 years ($SD \pm 0.65$, range 1-7). All patients who underwent the Swenson procedure had long-segment type disease. Despite observance of the main principles of this procedure, three children developed postoperative complications: 2 cases (25%) of anastomosis stricture, one case (12.5%) of dysuria, and one (12.5%) episode of HAEC.

Ten patients with rectal and rectosigmoid aganglionic zones underwent Swenson-like procedures. Their mean age at surgery was 4.8 years ($SD \pm 1.02$, range 10 months – 7 years). Two children had episodes of Hirschsprung-associated enterocolitis (HAEC) after the Swenson-like pull-through procedure.

Forty-eight patients underwent a TEPT. These children had rectal and rectosigmoid forms of HD. Their mean age at surgery was 3.4 years ($SD \pm 0.35$, range 6 months – 7 years). Nine (18.7%) children in this group had painful defecation in the early postoperative period. Catastrophic anastomotic dehiscence occurred in 1 (2%) patient after TEPT. Four (8.3%) patients with HD after TEPT had episodes of HAEC in the long-term follow-up.

The overall incidence of postoperative complications was higher in children who underwent transabdominal approaches compared to the incidence in those who underwent transanal pull-through procedures. Episodes of HAEC occurred more often after transabdominal pull-through procedures (14.4% vs 8.3%; $p = .04$). The average postoperative time at which the HAEC episodes occurred was not significantly different between the groups; the episodes were diagnosed within the first 3 years after pull-through. All children with episodes of HAEC were treated with irrigations and antibiotics. Patients with faecal incontinence after pull-through procedures were treated with a constipating diet, pectin and loperamide. After this treatment programme, the patient became continent. Six patients with anastomotic strictures were treated with anal bougienage at the outpatient clinic (the duration of which depended on the severity of stenosis). However, in 2 (1.4%) children, anastomotic stenosis remained, and surgical operations were performed to excise the stenosis sites.

Discussion. The incidence of early postoperative complications, such as faecal incontinence, anastomotic stricture, cuff abscess and dysuria, was relatively high ($n=13$; 9.4%) after transabdominal approaches in our study, and nine of them occurred after modified Soave pull-through. After analysing the high incidence of faecal incontinence after the modified Soave procedure, we concluded that it was caused by increased pressure in the anal canal, which was associated with a duplicated serous-muscular layer of the rectum and lower intestine, incomplete synergism of the fibres of the internal and external sphincters of the rectum, which ultimately led to the accumulation and prolonged stagnation of faecal masses, and soiling. Performing definitive conservative management of incontinence with laxatives and dietary modifications depending on the presence of dilated and nondilated colon in children with postoperative faecal incontinence gave us positive treatment results.

There is evidence to suggest that more postoperative complications occur in late-diagnosed HD patients [14, 16]. Anastomotic dehiscence and leakage are more likely to occur in late-diagnosed HD patients and after repeat surgery [17], but in our series, this catastrophic complication occurred after primary TEPT. This unwanted early complication was observed in a boy of 7 years of age and occurred in the era of implementation of this new technique in our surgical practice. We believe that this severe complication could have been avoided if we performed a protective stoma before TEPT.

The incidence of the most serious complication of HD, enterocolitis following pull-through, differs widely from 15 to 45% [18-20]. In our series, postoperative HAEC was observed in 17 (12.3%) children, with a higher frequency after transabdominal approaches. The systematic review and meta-analysis performed by Chen et al., which included 93 studies, compared the clinical outcomes of TEPT and transabdominal approaches [21]. They did not find differences in postoperative HAEC between approaches. According to another study performed by Mao et al., which included 49 studies, the TEPT technique was associated with a greater incidence of postoperative HAEC compared with the Duhamel procedure [22]. Heterogeneity in the frequency of HAEC in children after different pull-through procedures justifies the need for further research on this severe complication.

In conclusion, children who underwent TEPT had a lower rate of postoperative complications, especially episodes of postoperative HAEC. This finding allows us to conclude that in the treatment of HD in children, preference should be given to TEPT, which is less traumatic, has better cosmetic outcomes and has low incidences of postoperative complications.

Compliance with Ethical Standards.

Conflict of Interest: Authors declares that thee have no conflict of interest.

Ethical approval: This article does not contain any studies with human participants or animals performed by any of the authors.

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