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

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Abstract*. Children who underwent TEPT had fewer rate of postoperative complications, especially episodes of postoperative HAEC. Therefore, in the treatment of HD in children, preference should be given to TEPT which is less traumatic, had better cosmetic outcome and low incidences of postoperative complications.*

Keywords*: hirschsprung disease, transabdominal pull-through, transanal endorectal pull-through, hirschsprung associated enterocolitis.*

Purpose: aim of this study is to compare clinical outcomes of four surgical procedures to treat Hirschsprung's disease in children.

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

Results: 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 fecal incontinence (5; 3.6%), anastomotic stricture (4; 2.8%), cuff abscess (2; 1.4%), one (0.7%) had dysuria and anastomotic dehiscence (1; 0.7%). Nine of these early postoperative complications were after modified Soave pull-through. The episodes of Hirschsprung associated enterocolitis occurred more often after transabdominal pull-through procedures (14.4% *vs* 8.3%; *P*=.04).

Background. Hirschsprung's disease (HD) is one of the most common life-threatening congenital anomalies during the neonatal period, which involve the gastrointestinal tract. Estimated prevalence is 1:5000 live newborns. HD is characterized by absence of ganglion cells in the submucosal and myenteric plexuses along variable length of the intestine [1, 2]. This results in functional intestinal obstruction due to a lack of peristalsis in the aganglionic segment. Several surgical procedures have been described to treat the disease [3-8]. 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% [9]. Postoperative complications such as fecal incontinence, dehiscence, retraction and constipation have an important impact on the quality of life of these children [10, 11]. The incidence of one of the most dangerous complication – enterocolitis in different analyses varies from 10% to 30% [12, 23].

The aim of this study is to compare clinical outcomes of four surgical procedures to treat Hirschsprung's disease in children.

Patients and methods. We retrospectively analyzed charts of 138 children <15 years of age with HD, operated at the Department of Pediatric Surgery of Tashkent Pediatric Medical Institute

Hospital in Tashkent, Uzbekistan from 2018 to 2023. All patients were subsequently evaluated with clinical, laboratory and physical examination studies. Contrast enema performed to demonstrate transition zone. The HD diagnosis was confirmed by intraoperative biopsy. One hundred and thirty-eight children with HD were operated in the study period. In this study, 61 (44%) children were older than 3 years of age. For such patients, we start bowel preparation one month prior surgical interventions. All surgeries were performed in a single institution and by senior pediatric surgeon.

In this study we measured the functional outcome by the incidence of postoperative complications, such as fecal incontinence, dehiscence, retraction and Hirschsprung associated enterocolitis (HAEC). There was no patient with neurological disorders and impairment of neuropsychomotor development that may affect 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 of below 0.05 was considered statistically significant.

Results. All 138 patients diagnosed with HD and underwent pull-trough procedures were included. Ninety patients with HD had 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 performed in seventy-two patients. This procedure performed transabdominally, and in addition to classic procedure after pull-through the upper intestine wrapped in the manner of a “cuff”, thereby increasing the density of the tissue connection. Among modified Soave procedures 48 (67%) were boys and 24 (33%) were girls, and the mean age at operation was 3.81 years (SD±0.3, range 1-12). All children with modified Soave operation had long segment of aganglionosis. In postoperative period fecal incontinence occurred in 5 (6.9%) cases, anastomosis strictures in 4 (5, 5%) (fig.1) observations and two children with HD after modified Soave pull-through (2.8%) had cuff abscess. Episodes of HAEC after modified Soave pull-through 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 Swenson procedure. The median age at operation was 3.75 years (SD±0.65, range 1-7). All patients with Swenson procedure had long-segment type disease. Despite observance of main principles of this procedure three children developed postoperative complications: anastomosis stricture 2 (25%) cases and one (12.5%) patient had dysuria and one (12.5%) episode of HAEC.

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

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

Overall incidence of postoperative complications was higher in children after transabdominal approaches compared to transanal pull-through procedures. The episodes of HAEC occurred more often after transabdominal pull-through procedures (14.4% *vs* 8.3%; *P*=.04). The average postoperative time when HAEC episodes occurred was not statistically significant in comparing groups, and diagnosed within the first 3 years after pull-through. All children with episodes of HAEC treated with irrigations and antibiotics. Patients with fecal incontinence after pull-through procedures were treated with a constipating diet, pectin and loperamide. After this treatment program patient became continent. Six patients with anastomotic strictures was 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 - excision of stenosis sites.

Discussion. The incidence of early postoperative complications like fecal incontinence, anastomotic stricture, cuff abscess and dysuria in our study was relatively high (n=13; 9.4%) after

transabdominal approaches, and nine of them were after modified Soave pull-through. Analyzing the high incidence of fecal incontinence after modified Soave procedure, we came to the conclusion that it was caused by an increased pressure in the anal canal, which was associated with duplicated serous-muscular layer of the rectum and lower intestine, incomplete synergism of the fibers of the internal and external sphincters of the rectum, which ultimately led to the accumulation and prolonged stagnation of fecal masses, and soiling. Performing definitive conservative management of incontinence with laxatives and dietary modifications depending on presence of dilated and nondilated colon in children with postoperative fecal incontinence and gave us positive treatment results.

There is evidence that supports hypothesis of more postoperative complications in late-diagnosed HD patients [13, 14]. Anastomotic dehiscence and leakage is more likely to occur in late-diagnosed HD patients and after redo surgery [15, 22], 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% [16-18]. In our series postoperative HAEC observed in 17 (12.3%) children, with higher frequency after transabdominal approaches. The systematic review and meta-analysis performed by Chen et al. which included 93 studies, compared clinical outcomes of TEPT and transabdominal approaches [19, 21]. They did not find difference between approaches in postoperative HAEC. According to another study performed by Mao et al. which included 49 studies, TEPT technique was associated with a greater incidence of postoperative HAEC compared with the Duhamel procedure [20]. Heterogeneity in the frequency of HAEC in children after different pull-through procedures justifies the need for further research of this severe complication.

In conclusion children who underwent TEPT had fewer rate of postoperative complications, especially episodes of postoperative HAEC. Thus, it allows us to conclude that in the treatment of HD in children, preference should be given to TEPT which is less traumatic, had better cosmetic outcome and low incidences of postoperative complications.

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