



## Effectiveness of various surgical methods in treatment of Hirschsprung's disease in children

Efikasnost različitih hirurških procedura u lečenju Hiršprungove bolesti kod dece

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### Abstract

**Background/Aim.** Hirschsprung's disease is the most common identifiable developmental disorder of the enteric nervous system, characterized by a failure of its formation in a variable segment of distal bowel. Currently available surgical therapies for Hirschsprung's disease, although lifesaving, are associated with numerous complications. The aim of our study was to evaluate the effectiveness of different surgical methods and the incidence of serious complications after radical surgery of rectosigmoid Hirschsprung's disease. **Methods.** A retrospective analysis, from June 1997 until May 2012 was carried out on 84 patients operated for Hirschsprung's disease of rectosigmoid colon. Transanal endorectal pull-through was performed in 30 (35.7%) patients (group I), while 54 (64.3%) patients were operated by other (Soave, Duhamel or Swenson) procedures (group II). The age at operation, the incidence and severity of postoperative complications, the need for previous colostomy and the number of reoperations are countered in order to evaluate the efficacy of surgical procedures. **Results.** In the group I, the

mean age at operation was  $9.41 \pm 6.37$  months and in the group II the mean age at operation was  $16.8 \pm 13.9$  months which was significantly higher ( $p < 0.01$ ). In the group I there were only 3 (10%) patients with complications, one (3%) of them was prone to only one redo procedure ( $1.00 \pm 0.00$ ) and there was no need for previous colostomy in all patients (100%). In the group II there were 16 (30%) patients with significantly frequent complications ( $p < 0.05$ ), about 2 reoperations on the average ( $1.94 \pm 1.84$ ) in 4 of them (25%) and 22 (41%) redo procedures, which was, in total, significantly higher than in the group I ( $p < 0.01$ ). Only Soave's procedure was performed without previous colostomy in 20 (37%) patients. **Conclusion.** Transanal endorectal pull-through in surgical treatment of patients with Hirschsprung's disease is more effective than other procedures concerning earlier onset, low incidence and less severe complications, which would require further operations, and no scars.

**Key words:** hirschsprung disease; surgical procedures, operative; postoperative complications; reoperation; child.

### Apstrakt

**Uvod/Cilj.** Hiršprungova bolest je jedan od najbolje izučenih urođenih poremećaja crevnog motiliteta kod dece. Različite hirurške metode za lečenje Hiršprungove bolesti razvijene su da bi se sa što manje rizika i više efektivnosti postigao normalan motilitet creva. Značajan napredak učinjen je smanjivanjem opsežnih hirurških procedura, udruženih sa povećanim morbiditetom i mortalitetom, kao i uvođenjem minimalno invazivnih operativnih tehnika u ranom, neonatalnom uzrastu. Cilj naše studije bio je da se proceni efektivnost različitih hirurških metoda i incidencija komplikacija posle opsežnih operacija Hiršprungove bolesti rekto-sigmoidne lokalizacije. **Metode.**

Izvršena je retrospektivna analiza (od juna 1997. do maja 2012. godine) grupe od 84 bolesnika sa Hiršprungovom bolešću. Svi su imali aganglionaran rekto-sigmoidni segment, potvrđen preoperativnom irigografijom i histološki. Operacija metodom endorektalnog provlačenja izvedena je kod 30 (35,7%) bolesnika (grupa I), dok je 54 (64,3%) bolesnika operisano procedurama po Soavi, Duhamelu ili Swensonu (grupa II). Parametri praćenja bili su starost, incidencija i težina postoperativnih komplikacija, neophodnost preoperativne kolostome kao i broj reoperacija. **Rezultati.** U vreme operacije u grupi I prosečna starost dece iznosila je  $9,41 \pm 6,37$  meseci, a u grupi II  $16,8 \pm 13,9$  meseci, što je bilo statistički značajno više u odnosu na grupu I ( $p < 0.01$ ). Postoperativne komplikacije imalo je 3/30

(10%) bolesnika u grupi I kod kojih je urađena operacija endorektalnog provlačenja i 16/54 (29,6%) bolesnika kod kojih su preduzete druge procedure. Pripadnici grupe II imali su značajno češće komplikacije ( $p < 0,05$ ). Bolesnici grupe I imali su samo jednu ( $1,00 \pm 0,00$ ), dok su oni iz grupe II imali oko dve reoperacije u proseku ( $1,94 \pm 1,84$ ). Više od jedne reoperacije urađeno je kod 6/16 (37,5%) bolesnika iz grupe II. Pripadnici grupe II imali su značajno više reoperacija od onih iz grupe I ( $p < 0,05$ ). Operativna procedura kod svih bolesnika (100%) grupe I i samo procedura po Soavi kod 20 (37%) bolesnika

grupe II, preduzete su bez prethodne kolostome. **Zaključak.** Rezultati naše studije pokazuju da je operacija metodom endorektalnog provlačenja u lečenju Hiršprungove bolesti mnogo efikasnija nego ostale procedure, posebno u odnosu na njeno rano preduzimanje, kao i manju incidenciju ozbiljnih komplikacija koje zahtevaju ponovno hirurško lečenje.

#### Ključne reči:

**hiršprungova bolest; hirurgija, operativne procedure; postoperativne komplikacije; reoperacija; deca.**

## Introduction

Hirschsprung's disease (HD) is the most common identifiable developmental disorder of the enteric nervous system (ENS), characterized by failure of its formation in variable segments of distal bowel. Traditionally, surgical therapy for HD considered preliminary proximal defunctioning colostomy, followed months later by the definitive reconstructive pull-through procedure, mostly by Swenson, Duhamel or Soave's technique. These surgical therapies for HD, although lifesaving, were associated with a significant incidence of disturbances of bowel function. The most frequent postoperative complications include enterocolitis after the Swenson's procedure, constipation following Duhamel's repair, and diarrhea and incontinence after the Soave's pull-through procedure. Even reports on long-term outcomes after definitive repair for HD are conflicted highlighting the need for newer curative therapies<sup>1,2</sup>.

In the beginning, the routine use of colostomy have been abandoned in favor of one-stage pull-through, with multiple studies suggesting this approach as safe and efficacious. Over the past few decades, the popularity of minimally invasive surgical techniques has led to a number of modifications to the standard one-stage procedure and solely transanal approach, which has been adopted by many surgeons and associated with the same advantages, without the need for extensive intraabdominal dissection. Several other creative approaches have been described, including a modification of the transanal approach with laparoscopic assistance<sup>3,4</sup>. Current regenerative strategies are under investigation to restore function in aganglionic intestine. Stem cell transplantation to regenerate the ENS is a subject of many recent experimental series. Though auspicious, these discoveries warrant further study to translate cell-based therapies into clinical practice<sup>5</sup>.

The aim of our study was to evaluate the effectiveness of different surgical methods and the incidence of serious complications after radical operation of rectosigmoid HD.

## Methods

We identified cases by retrospective review of HD admissions to the University Children's Hospital in Belgrade, Serbia, over a 15-year-period, from June 1997 to May 2012. During this period there were 84 patients with recto-sigmoid aganglionosis treated surgically. They were divided into two groups: the group I included 30 (35.7%) patients operated on

by single-stage transanal endorectal pull-through (TEPT) and the group II of 54 (64.3%) patients operated on by Soave, Duhamel or Swenson's open technique. They were also observed concerning the presence of colostomy: 50 (59.5%) patients operated on by single-stage TEPT and Soave technique, had no colostomy, while other 34 (40.5%) patients were operated on by three-stage Duhamel and Swenson's procedures. They had previous colostomy, which was subsequently closed, about 4 weeks after the radical procedure.

The study was performed according to the principles of Good Clinical Practice and the Declaration of Helsinki, after the Institutional Ethical Committee approval and signing the form of parents consent.

Prior to surgery, all the patients underwent water-soluble contrast or barium enema that helped to identify a transition zone between a narrowed aganglionic and dilated, otherwise normally innervated segments. Histological findings including the absence of ganglion cells and hypertrophic extrinsic nerve fibers in submucosal and myenteric layers of rectal biopsy specimen, confirmed the diagnosis of HD. Antimicrobial prophylaxis considered monotherapy of ertapenem, in a course of 3 days in the group I, or triple antibiotic treatment of ampicillin, gentamycin and metronidazole, in a course of 5 days in the group II. Colonic lavage, consisting of mechanical irrigation with a large-bore rectal tube and large volumes of irrigant, had been required preoperatively. The surgery was done under general anesthesia.

The age at operation, the incidence and severity of postoperative complications, the need for previous colostomy and the number of reoperations were countered in order to evaluate the efficacy of surgical procedures.

The complication frequency was presented as a whole number and percents.  $\chi^2$  test was performed to establish a statistical significance between the study groups of patients, regarding complication frequency. The average value of redo operations was presented as a mean value ( $\bar{x}$ ) with the standard deviation (SD). Fisher's test was used to evaluate complication frequencies among different surgical procedures in the groups of patients with and without colostomy. Mann-Whitney U-test was used to evaluate statistical significance of mean values of reoperations between the study groups. Statistical significance was set at  $p < 0.05$ .

## Results

From the total number of 84 patients, 67 (79.8%) were males and 17 (20.2%) females, so the ratio was 4 : 1. In the group I,

the mean age at operation was  $9.41 \pm 6.37$  months, which was significantly lower comparing to the group II ( $p < 0.01$ ); there were only 3 (10%) patients with complications, one (3.3%) of them was prone to only one redo procedure ( $1.00 \pm 0.00$ ) (Table 1) and there was no need for previous colostomy in all the patients (100%) (Table 2).

In the group II, the mean age at operation was  $16.8 \pm 13.9$  months. There were 16 (30%) patients with significantly frequent complications ( $p < 0.05$ ), about 2 reoperations on the average ( $1.94 \pm 1.84$ ) in 4 (25%) of them (Table 1) and 22 (41%) redo procedures, which was, in total, significantly higher than in the group I ( $p < 0.01$ ) (Table 3). In this group, only Soave's procedure was performed without previous colostomy in 20 (37%) patients. The patients operated by single-stage TEPT and Soa-

ve's procedure without colostomy had far less complications and reoperations than the others with colostomy (Table 2).

Within these very groups with and without colostomy, there was no significant difference in distribution of complications and reoperations among different surgical procedures ( $p > 0.05$ ) (Table 2).

The group I had significantly lower number of complications ( $p < 0.05$ ). Anastomotic stricture was the only (100%) complication in the group I and the most frequent with Soave's operation (67%), while enterocolitis was the most frequent complication with Duhamel's (45%) and Swenson's (100%) procedures. Almost every third patient with colostomy had stoma complications (38%) with the similar incidence of prolapse (46%) or stoma stenosis (54%) (Table 3).

Table 1

## Evaluated parameters regarding surgical procedures

Evaluated parameters	Group I <sup>†</sup>	Group II <sup>†</sup>	<i>p</i> -values
Number of patients, n (%)	30 (35.7)	54 (64.3)	
Complications, n (%)	3 (10.0)	16 (29.6)	< 0.05*
Reoperation, $\bar{x} \pm SD$	$1.00 \pm 0.00$	$1.94 \pm 1.84$	< 0.05**
Age (months), $\bar{x} \pm SD$	$9.41 \pm 6.37$	$16.81 \pm 13.95$	< 0.01**

Group I – group with transanal endorectal pull-through procedure; Group II – group with Soave, Duhamel and Swenson's procedures.\* $\chi^2$  test; \*\*Mann-Whitney U-test.

Table 2

## Evaluated parameters regarding the presence of colostomy

Evaluated parameters	n (%)	Complications n (%)	<i>p</i> *	Reoperation $\bar{x} \pm SD$	<i>p</i> **
Without colostomy					
TEPT	30 (100)	3 (10)		$1.00 \pm 0.00$	
Soave	20 (37)	3 (15)	> 0.05	$1.00 \pm 0.00$	> 0.05
With colostomy					
Duhamel	29 (54)	11 (38)		$2.03 \pm 1.97$	
Swenson	5 (9)	2 (40)	> 0.05	$1.00 \pm 0.00$	> 0.05
Stoma complications		13 (38)			

\*Fisher test; \*\*Mann-Whitney U test; TEPT – transanal endorectal pull-through procedure.

Table 3

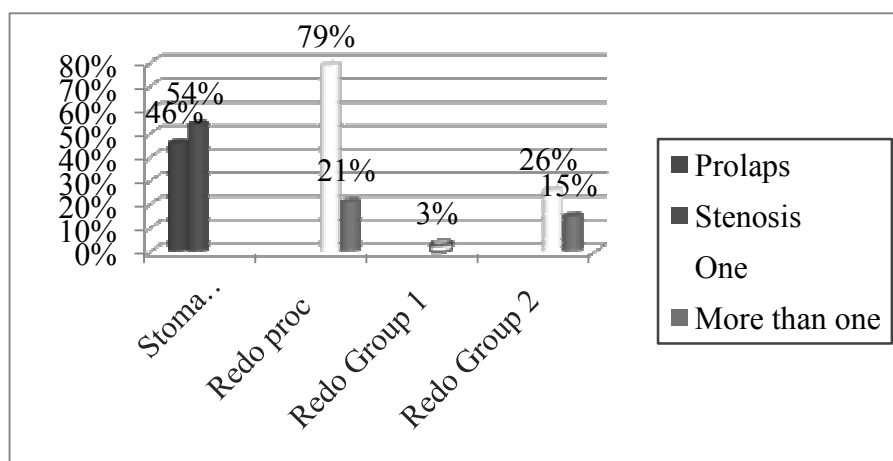
## Distribution of complications and reoperations in evaluated groups

Type of surgical procedure	n (%)	Complications associated with surgical procedures	n (%)	<i>p</i>
TEPT	3 (10)	Anastomotic stricture	3 (100)	
Soave	3 (15)	Anastomotic stricture	2 (67)	
		Anastomotic dehiscence	1 (33)	
Duhamel	11 (38)	Residual aganglionosis	2 (18)	< 0.05*
		Anastomotic dehiscence	2 (18)	
		Obstructed pouch	2 (18)	
		Enterocolitis	5 (45)	
Swenson	2 (40)	Enterocolitis	2 (100)	
Stoma	13 (38)	Prolapse	6 (46)	
		Stenosis	7 (54)	> 0.05*
Reoperations one	19 (23)		15 (79)	
more than one			4 (21)	< 0.01**

\* $\chi^2$ -test; \*\*Fisher test, TEPT – transanal endorectal pull-through procedure.

In the entire group of patients who required reoperation, one redo was performed predominantly in 79% of cases ( $p < 0.01$ ). In the group I, there was only one (3%) redo operation, because of omitting the posterior myectomy of the cuff, by mistake in the first case of TEPT. In the group II, one redo was done in 14 (26%) patients and 2 on the average in each of 4 (15%) patients which was in total significantly higher, 22 (41%), redo procedures ( $p < 0.01$ ) (Figure 1). More than one redo were done because of overlooked residual aganglionosis in 2 cases and residual pouch obstruction with incomplete resection of the common colorectal wall in Duhamel's technique in another 2 cases.

The enterocolitis that is associated with HD in children still presents a significant cause of morbidity and mortality<sup>12</sup>. The occurrence of postoperative enterocolitis in our study is confirmed in almost every tenth patient (8.3%) that is similar with previous reports in the literature<sup>13, 14</sup>. In our study the enterocolitis is predominantly associated with surgical procedures requiring colostomy (Duhamel and Swenson) and also with strictures as complication. The outcomes of TEPT procedure have been similar to open single-stage approaches, and analgesia requirements and hospital stays are decreased. Recent studies also report lower rates of postoperative incontinence and shorter operating times among transanal pull-



**Fig. 1 – Distribution of stoma complications and redo procedures in the evaluated groups.**  
**Group I – group with transanal endorectal pull-through procedure;**  
**Group II – group with Soave, Duhamel and Swenson's procedures.**

## Discussion

There are several surgical techniques that are used for the treatment of patients with HD<sup>6</sup>. However, despite advances in medicine and surgery, there are still children with functional problems even after surgical treatment<sup>7</sup>. It is important to stress out that the main goal of surgery in these patients is to remove aganglionic segment and to enable bowel function management. Previous studies notice that TEPT is better surgical choice, mainly due to infrequent complications and better functional outcome<sup>8, 9</sup>. The main indications that require reoperation refer to the anatomical problems (*eg* strictures, twisted pull-through, obstructions), pathological problems (*eg* residual aganglionosis) and others (*eg* enterocolitis, fistulas)<sup>10</sup>.

Our results clearly demonstrate that TEPT is most effective surgical option in the treatment of patients with HD. The advantage of such method is that TEPT is minimally invasive procedure with no need for colostomy and could be performed in neonatal period. Currently, approximately 90% of patients with Hirschsprung's disease are diagnosed and also could be cured in the newborn period. In spite the fact that TEPT procedure is associated with common complications including constipation, enterocolitis and strictures, we show that the presence of complications and particular need for reoperation are far less frequent than with other techniques<sup>11</sup>.

through procedures<sup>15</sup>. We showed that even Soave's surgical technique did not require colostomy; it had slightly higher frequency of complications comparing to TEPT. It was noticed as well that both Duhamel and Swenson's procedures had highest rates of complications, but the need for more than one reoperation was significantly higher in patients who underwent Duhamel's procedure. It is consistent with previous reports which stated that pouch in Duhamel's procedure is mainly responsible for serious complications like impaction, overflow incontinence and enterocolitis<sup>16</sup>. The overall morbidity and mortality in staged procedures are increased by complications associated with procedures requiring colostomy<sup>17, 18</sup>. Therefore, a single-stage TEPT procedure could be of a great benefit in further surgical practice for patients with HD, since it reduces complications, shorten hospitalization time and hospital costs, possibilities of acquiring infection and finally reduces further necessity for reoperations<sup>18, 19</sup>. Such a technique is more favorable due to less severe postoperative pain and excellent cosmetic result with no scar<sup>19</sup>. The study of Zhang et al.<sup>20</sup> it was noticed that functional outcome (stooling patterns and colonic motility) is satisfactory in patients who undergo TEPT procedure. Although, the limitation of our study refers to the lack of these long-term functional predictors after TEPT, we consider that regarding the surgical procedure, choosing the appropriate time and technique and

including opting for a less invasive approach are the key factors for a good long-term functional outcome.

### Conclusion

The results of our study show that the TEPT procedure in surgical treatment of patients with HD is more effective than other procedures concerning early onset, low incidence

and less severe complications that would require further operations and also, excellent cosmetic result with no scar.

### Conflict of interest

No one of the authors has an affiliation or financial relationship with a commercial entity that has an interest in the subject of this manuscript.

### R E F E R E N C E S

1. Butler TN, Trainor PA. The developmental etiology and pathogenesis of Hirschsprung disease. *Transl Res* 2013; 162(1): 1–15.
2. Teitelbaum DH, Cilley RE, Sherman NJ, Bliss D, Uitvlugt ND, Renaud EJ, et al. A decade of experience with the primary pull-through for hirschsprung disease in the newborn period: a multicenter analysis of outcomes. *Ann Surg* 2000; 232(3): 372–80.
3. Puri P. Hirschsprung's disease. In: Oldham TO, Colombani PM, Foglia RP, editors. *Surgery of infants and children: Scientific principles and practice*. New York: Lippincott-Raven; 1997. p. 1277–99.
4. de la Torre L, Ortega A. Transanal versus open endorectal pull-through for Hirschsprung's disease. *J Pediatr Surg* 2000; 35(11): 1630–2.
5. Goldstein AM, Hofstra RM, Burns AJ. Building a brain in the gut: development of the enteric nervous system. *Clin Genet* 2013; 83(4): 307–16.
6. Peña A, Elicevici M, Levitt MA. Reoperations in Hirschsprung disease. *J Pediatr Surg* 2007; 42(6): 1008–13.
7. Langer JC. Hirschsprung disease. *Curr Opin Pediatr* 2013; 25(3): 368–74.
8. Tannuri AC, Tannuri U, Romão RL. Transanal endorectal pull-through in children with Hirschsprung's disease--technical refinements and comparison of results with the Duhamel procedure. *J Pediatr Surg* 2009; 44(4): 767–72.
9. Aslan MK, Karaman I, Karuman A, Erdoğan D, Cavuşoğlu YH, Cakmak O. Our experience with transanal endorectal pull-through in Hirschsprung's disease. *Eur J Pediatr Surg* 2007; 17(5): 335–9.
10. Sheng Q, Lu Z, Xiao X. Re-operation for Hirschsprung's disease: experience in 24 patients from China. *Pediatr Surg Int* 2012; 28(5): 501–6.
11. Dutta HK. Clinical experience with a new modified transanal endorectal pull-through for Hirschsprung's disease. *Pediatr Surg Int* 2010; 26(7): 747–51.
12. Frykeman PK, Short SS. Hirschsprung-associated enterocolitis: prevention and therapy. *Semin Pediatr Surg* 2012; 21(4): 328–35.
13. Rutenstock E, Puri P. Systematic review and meta-analysis of enterocolitis after one-stage transanal pull-through procedure for Hirschsprung's disease. *Pediatr Surg Int* 2010; 26(11): 1101–5.
14. Luis LA, Encinas JL, Avila LF, Andrés AM, Burgos L, Fernández A, et al. Hirschsprung disease: lessons learned from the last 100 cases. *Cir Pediatr* 2006; 19(3): 177–81.
15. Langer JC, Seifert M, Minkes RK. One-stage Soave pull-through for Hirschsprung's disease: a comparison of the transanal and open approaches. *J Pediatr Surg* 2000; 35(6): 820–2.
16. Chatoorgoon K, Pena A, Laval TA, Levitt M. The problematic Duhamel pouch in Hirschsprung's disease: manifestations and treatment. *Eur J Pediatr Surg* 2011; 21(6): 366–9.
17. Bischoff A, Levitt MA, Laval TA, Peña A. Colostomy closure: how to avoid complications. *Pediatr Surg Int* 2010; 26(11): 1087–92.
18. Pena A, Migotto-Krieger M, Levitt MA. Colostomy in anorectal malformations: a procedure with serious but preventable complications. *J Pediatr Surg* 2006; 41(4): 748–56.
19. Rintala RJ. Transanal coloanal pull-through with a short muscular cuff for classic Hirschsprung's disease. *Eur J Pediatr Surg* 2003; 13(3): 181–6.
20. Zhang S, Wang W, Bai Y, Wang W. Evaluation of anorectal function after transanal one-stage endorectal pull through operation in children with Hirschsprung's disease. *Zhongguo Dang Dai Er Ke Za Zhi* 2007; 9(3): 188–92.

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