Evaluation Of Short And Mid-Term Outcomes In Children With Hirschsprung's Disease Undergoing Different Surgical Procedures

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Abstract

Introduction: The primary aim of surgical procedures for Hirschsprung's disease is to establish proper bowel movement without fecal incontinence, constipation and entrocolitis. In this study we evaluated short and mid-term outcomes in children with Hirschsprung's disease undergoing different surgical procedures.

Materials and Methods: In a prospective single blind study, 50 children with Hirschsprung's disease were selected. Patient eligibility included age under ten years and an elective single stage Duhamel (D group) or trans- anal procedure (T group). Primary end point data collected

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included the development of post-operative constipation and fecal incontinency in groups. Secondary end points of study was intra-abdominal infection, anastomotic leak, wound infection and Hirschsprung associated enterocolitis.

Results: Post-operative constipation persisted in 14 (56%) after 1 month in D group and in 7(28%) in T group (p =0.04). Post –operative constipation persisted in 16 (64%) after 6 months in D group and in 10(40%) in T group (p =.08). There was no significant difference in fecal incontinency rates between groups.

Conclusion: For young children with Hirschsprung's disease who scheduled for operation, we had found no clear difference in mid-term outcome.

Keywords

- Hirschsprung's disease
- Duhamel
- Transanal
- complication

Introduction

Hirschsprung's disease(HSCR), or congenital intestinal aganglionosis is caused by an impairment in cell migration from the neural crest to the gastrointestinal tract, resulting in an absence of neurons in the submucosal ganglion cells(Meissner plexus) or myenteric plexus(Auerbach plexus).[1] The characteristic presentation of HSCR includes delayed passage of meconium beyond the first 24 hours of life, abdominal distention, and bilious vomiting, and a water-soluble contrast enema may demonstrate a transitional zone from ganglionic to aganglionic bowel. [2-3-4]

The only treatment for Hirschsprung's disease is pull-through anastomosis of the ganglionic intestine with perfect function of anal canal sphincter.[5] The first successful treatment was performed about 60 years ago. Currently, various surgical techniques are used for HSCR such as Swenson, Soave, Duhamel and others. [6-7] The goal of treating a child with Hirschsprung's disease is to achieve near-normal anorectal function.

The primary surgical procedure for the disease includes releasing of the distal colon involved in the pelvis through extrarectal submucosal dissection up to 2

cm above the dentate line and creating end-to-end anastomosis. [8-9-10]

The outcome of each technique is determined by the short-term and long-term complications. The final goal of all these methods is to establish proper bowel movement without fecal incontinence, constipation and entrocolitis.[11]

Therefore, we conducted a single institution study to evaluate short and mid-term outcomes in children with HSCR undergoing different surgical procedures. As hypothesis, we investigated whether difference on technique would change functional outcome of patients.

Materials and Methods

This is an approved thesis for obtaining M.D with ethic number of 398320, carried out in the Imam Hossein Children Hospital in Isfahan University of Medical Sciences (IUMS), from May 2013 to October 2019. In a prospective singe blind study, we prospectively included children with HSCR (based on rectal biopsy diagnosis) that referred to our center for operation. Informed consent was obtained from the parents. Patients with past history of enterocolitis episodes, previous abdominal surgery for HSCR, infants with leveling colostomy and patients with chronic fecal impaction were excluded. Chronic fecal impaction was diagnosed by decreased defecation frequency, abdominal distension and filled rectum found on digital rectal examination. Patient eligibility included age under ten years and an elective single stage Duhamel (D group) or trans- anal procedure (T group). Fifty cases of HSCR were selected for study. They were divided into two groups (depending on the type of surgery). Due to being objective of constipation, we have designed the study to be blind so that the registry nurse who collected data would not know the type of technique. The criteria for admission include: clinical diagnosis and the presence of a previous pathology sample confirming hirschsprung, reaffirmation of the pathology diagnosis of Hirschsprung's disease with IHC, undergoing surgery for the first time, and conscious consent from the parents.

Primary end point data collected included the development of post-operative constipation and fecal incontinency. A scoring system called pediatric constipation score **Table 1** was used to measure and compare two surgical groups in terms of the severity of pre-operative constipation.[12-13] Fecal incontinence in

children was defined as involuntary passing of stool in underwear (in children over 4 years old).[14] Any baby who is unable to excrete meconium within 24-48 hours after birth and suffer from an abdominal distension and any child who defecates low frequently with a hard stool as someone who have constipation[15-16].

Secondary end points of study were intra-abdominal infection, anastomotic leak, wound infection and Hirschsprung associated enterocolitis (HAEC). Intra-abdominal infection was diagnosed with history, physical examination and laboratory studies. Further diagnostic imaging such as ultrasound or computed tomography (CT) was used in selected patients. Anastomotic leak was diagnosed clinically (fever, oliguria, ileus, diarrhea, peritonitis) and confirmed by a CT scan. Wound infections were diagnosed according to the following criteria:

1) Infection occurs within 30 days of surgery

2) At least one of the following is present:

a) Purulent discharge from a superficial infection

b) Organisms isolated from aseptically obtained wound culture

3) At least one of the following signs of infection is present:

a) Pain or tenderness

b) localized swelling

c) Redness or heat

d) Fever

Hirschsprung associated enterocolitis (HAEC) was diagnosed clinically (abdominal distension, explosive diarrhea, vomiting, fever, and rectal bleeding.

Assessment of the symptoms in patients was performed by senior author of study (M.M) daily during hospitalization and then, every 2 months in follow up. Patients were followed to 6 months. The statistical package for social science (SPSS 20, Chicago, IL, USA) software was used for data analysis. Categorical variables were compared by chi- square and Fisher`s exact test. Variables were not normally distributed. The Mann- Whitney was used to determine any differences in mean scores of groups. Data were expressed as mean± SD. P value less than 0.05 was considered significant.

Results

In this study, a total of 50 children (25 as D group and 25 as T group) were enrolled. Baseline demographics did not differ between the groups **Table 2.** There were no statistical differences between the D and T

Groups according to gender distribution. Twenty-nine (58%) patients were male and 21 (42%) were female. 21 patients in group D and 5 patients in group T were >= 4 years old which considered for comparison of fecal incontinency in groups.

Table 1: pediatric constipation score

	Pediatric constipation score	Scores		
1	Does your child regularly soil its underclothes by involuntarily passing small amounts of stool?	Yes Always (0)	Sometimes (1)	No (2)
2	Does your child have trouble opening its bowels completely (incomplete emptying)?	Yes Always (0)	Sometimes (1.5)	No (3)
3	How often does your child open its bowels?	Severaltime a day (4)	Ones daily (2)	Less often (0)
4	What does the stool usually look like?	Watery (1)	Variable (0.5)	Thick (0)
5	Does your child have a lot of wind?	Yes Always (0)	Sometimes (1)	Never (2)
6	Can your child tell the difference between stool and air in the bowels?	Yes Always (1)	Sometimes (0.5)	No (0)
7	Does your child feel pain when opening its bowels?	Yes Always (0)	Sometimes (1)	No (2)
8	Does your child have to press hard to empty its bowel?	Yes (0)	Normal (2)	No (4)
9	Does your child suffer from constipation?	Yes Always (0)	Sometimes (2)	Never (4)
10	Does your child have pain in the tummy?	Yes Always (0)	Sometimes (2)	Never (4)
	Total score			

Table 2: patient demographics of patients with hirschsprungs disease.D Group: Duhamel procedure patient;T Group: TERPT patients.

	D Group (n=25)	%	T Group (n=25)	%	Total	%
Male	15	60	14	56	29	**58**
Female	10	40	11	44	21	**42**

Age at operation was 6.37±4.21 years (0.66-14.00) in D group and 1.53±2.43 (0.08-11.69) in T group (p=0.00). Score of pre –operative constipation was5.260 ±4.46 (1.00-18.00) in D group and 7.58 ±6.18 (2.00-24.00) in T group (p=0.09). **Table 3** shows the comparison of groups in regard of postoperative complications.

Entrocolitis after 1 month was seen in 4 of 25 (16%) children in D group. While, one person of 25(4%) has been recognize having entrocolitis after 1 month in T group. There is no significant statistical difference between two group (p =1.000). Entrocolitis after 6 months was seen in 1 of 25 (4%) children in D group. Also, this is recognized in 3 of 25(12%) in trans-anal procedure There is no significant statistical difference between two groups (P =0.6). We have no anastomotic leak in our groups. Five patients (25%) had wound infection in D group. We had no wound infection in T group. Post-operative constipation persisted in 14 (56%) after 1 month in D group and in 7(28%) in T group (p =0.04). Post–operative constipation persisted in16 (64%) after 6 months in D group and in 10(40%) in T group (p =.08). There were no significant differences in fecal incontinency rates between groups.

There was no significant statistical difference between early and mid-evaluation complication rates in groups. **Table 4.**

Table 3: Complication's rate during follow-upfor patients with Hirschsprung's disease.
D Group: Duhamel patients; T Group: Trans anal

	D Group (n /N)	%	T Group (n/N)	%	P value
Enterocolitis after1 month	4/25	16	3/25	12	0.68
Enterocolitis after6 month	1/25	4	3/25	12	0.29
Constipation after1 month	14/25	56	7/25	28	0.04
Constipation after6 month	16/25	64	10/25	40	0.08
Incontinency after1 month	8/21	38	3/5	60	0.37
Incontinency after6 month	6/21	28	1/5	20	0.69

Table 4: Complications' rate during follow-upfor patients with Hirschsprung's disease in regarding to follow up time.
D Group: Duhamel patients; T Group: Transanal

	One month (n /N)	%	Six months (n/N)	%	P value
Enterocolitis(D group)	4/25	16	1/25	4	0.15
Enterocolitis(T group)	3/25	12	3/25	12	1.00
Constipation (D group)	14/25	56	16/25	64	0.56
Constipation (T group)	7/25	28	10/25	40	0.36
Incontinency (D group)	8/21	32	6/21	24	0.52
Incontinency (T group)	3/5	36	1/5	36	0.19

Discussion

Hirschsprung's disease is a heterogeneous genetic disorder, resulting from an anomaly of the enteric nervous system of neural crest cells origin, and characterized by the absence of parasympathetic intrinsic ganglion cells in the submucosal and myentericplexuses.[1] In the newborn, symptoms may appear during the first hours of life with failure to pass meconium, or in the first week with a picture of functional intestinal obstruction.

Enterocolitis, the most common complication, is always severe and is an important cause of mortality in these young patients. In infants and children, the presentation is often less dramatic and may

not mimic acute intestinal obstruction and, in these patients, severe constipation and recurrent fecal impaction are more common.

In spite of the technical advances in the surgical repair of HSCR disease that have occurred over the last 20 years, complications are still common and constipation, incontinence and diarrhea can affect children' lifestyle.[17] Menezes at al.[18] study showed that the long-term soiling and constipation after surgery had 10.3% and 21.7% respectively in post-operative evaluation.

The main goal of this study was to evaluate the post-operative complications based on common techniques of HSCR disease operation in our country.

As could be seen in the results, constipation and fecal incontinence were the most common prevalent postoperative complications, with no statistically differences when accompanied by follow up time. However, percentages differed based on the techniques.

Constipation occurred early after operation in %56 of our cases in first month of operation in D group, however the incidence of this complication was 28% in T group. This result is similar to findings of Ekenze et al. study that showed that the constipation occurred early after operation in %44.1 of their cases whereas it was reported in slightly above a third of patients later throughout their follow-up.[19] Similar studies varied in their percentages, ranging from 8% to 60% of whereas it was reported in slightly above a third of patients later throughout our follow-up. Similar studies varied in their percentages, ranging from 8% to 60% ofcases.[20-21-22] It seems that in trans-anal pull through technique, dilation of sphincter mechanism causes the relaxation of distal part of anal canal temporarily and this can explain the significant differences in constipation rate in first month of operation. However, after six months the mechanism return to pre-operative level and the rate of constipation would be similar in groups. However, anastomosis stricture should be in mind in every case of postoperative constipation.

Likewise, incontinence rates fluctuated widely in different study findings (10-80%).[18-19] Our results are similar to these studies, however trans- anal technique by new devices such as Loane retractors is a recently practice in our clinic, therefore most of our patients in T group are under 4 years and this is the major limitation of our study for evaluation of fecal incontinency. We suggest the more follow up studies in this group to achieve more accurate results.

Conclusion

In this study, we had enterocolitis in five patients overall. However, this complication was shown substantially broad in incidence owing to differences in definitions by authors. Some reported it as high as 26 to 32%[23-24] while Yancharet al.[25] reported the rate of 9.3%.

In conclusion, for young children with HSCR who scheduled for operation, we had found no clear difference in mid-term outcome. However, a multicenter randomized controlled trial is needed to more definitely determine the best approach for children with HSCR.

Ethical Consideration

This study was approved by Ethical committee of Isfahan University of Medical Sciences as thesis for obtaining M.D with ethic number of 398320.

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Conflict of interests

There is no conflict of interest

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