

ANALYZING THE SHORT AND MID-TERM RESULTS IN HIRSCHSPRUNG'S DISEASE CHILDREN UNDERGOING VARIOUS SURGICAL TREATMENTS

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ABSTRACT:

Background: Establishing regular bowel movements free of fecal incontinence, enterocolitis, and constipation is the main goal of surgical operations for Hirschsprung's disease. In this research, we assessed the short- and long-term effects of several surgical techniques on children with Hirschsprung's illness.

Methods: A sample of 50 children with Hirschsprung's illness was chosen for prospective single-blind research. Age under 10 years and an elective single-stage Duhamel (D group) or trans anal operation were requirements for patients (T group). The formation of post-operative constipation and fecal incontinence in groups were the primary endpoints for which data were gathered. Intraabdominal infection, amniotic fluid leak, wound infection, and enterocolitis linked with Hirschsprung was the study's secondary endpoints.

Results: After one month, 14 (56%) in the D group and 7 (28%) in the T group still had post-operative constipation (p-value =0.04). After six months, post-operative constipation remained in 16 (64%) of the D group patients and in 10 (40%) of the T group patients (p-value =.08). Fecal incontinence rates did not significantly vary across the groups.

Conclusion: We discovered no discernible difference in the mid-term result for young infants with Hirschsprung's disease who were scheduled for surgery.

Keywords: Duhamel, Hirschsprung's disease, Trans anal

Introduction: Cerebral intestine aganglionosis, also known as Hirschsprung's disease (HSCR), is brought on by a problem with cellular proliferation from the notochord to the intestinal system, which leaves no neurons in the subcutaneous tissue neural impulses (Conductive axis) or mesenchyme plexus (Auerbach plexus). (1) Delay in passing mucus beyond the initial 24-hour period of life, abdominal discomfort, and bilious nausea are the hallmark symptoms of HSCR. A liquid contrast evacuation may show a zone where the ganglionic and aganglionic bowels are transitioning. (2-4)

Squeeze ligation of the sympathetic ganglia intestine with excellent anal canal sphincter activity is the sole cure for Hirschsprung's disease. (5,6) The first effective procedure was carried out roughly 60 years ago. Currently, HSCR is treated surgically using a variety of methods, including Swenson, Soave, Duhamel, and others. The aim of treatment for a kid with Hirschsprung's disease is to restore anorectal function to a state that is almost normal.

The main surgical treatment for the condition is making an end-to-end anastomosis and releasing the distal colon that is affected in the pelvis by an extra rectal capsular incision up to 2 cm above the edentulous line. (7-10) The short- and long-term consequences of each procedure affect its result. All of these techniques ultimately aim to develop regular bowel movements free from fecal incontinence, constipation, and enterocolitis. (11) As a result, we carried out single-institution research to assess the short- and medium-term results in kids with HSCR who had various surgical treatments. We looked at whether different techniques would affect patients' functional outcomes as a hypothesis.

Methods: The Mayo Hospital in Lahore, Pakistan, where this research was conducted, from December 2022 to December 2023. We recruited into the study children with HSCR (based on colorectal biopsies diagnostic) who were referred to our facility for surgery in prospective single-blind research. The parents gave their informed permission. Patients who had had bouts of enterocolitis in the past, undergone prior abdominal surgery for HSCR, had leveling colostomies as newborns, and had chronic fecal impaction were all disqualified. Reduced frequency of feces, abdominal distension, and a full rectum discovered during a digital rectal examination was used to diagnose chronic fecal impaction. Age under 10 years and an elective single-stage Duhamel (D group) or trans anal operation were requirements for patients (T group). For the research, 50 HSCR instances were chosen. Two groups were created out of them (depending on the type of surgery). We intentionally made the research blind in order to protect the identity of the registry nurse who gathered the data while maintaining the study's objectivity about constipation. The existence of a prior pathology sample that confirmed Hirschsprung's illness as well as a clinical diagnosis is required for admission. Other requirements include having to undergo surgery for the first time and having the parents' informed agreement.

The emergence of post-operative constipation and fecal incontinence was the primary endpoint data that were gathered. The degree of pre-operative constipation in two surgical groups was measured and compared using a scoring system termed the pediatric constipation score (Table 1). (12-13) Stool passing

in underwear without consent is known as fecal incontinence in children (in children over 4 years old). (14) Any infant who has stomach distension and is unable to pass meconium within 24 to 48 hours after birth, as well as any young kid who often passes low, hard stool, is considered to have constipation. (15,16)

The study's secondary endpoints were Hirschsprung-associated enterocolitis, anastomotic leak, intra-abdominal infection, and wound infection (HAEC). With the use of medical history, physical examination, and laboratory tests, an intra-abdominal infection was identified. In certain individuals, further diagnostic imaging such as computed tomography (CT) or ultrasonography was employed. Clinically identified anastomotic leak (fever, oliguria, ileus, diarrhea, peritonitis), and CT scan confirmation was made. The following criteria were used to identify wound infections:

1) At least one of the symptoms of the infection listed below is present.

A fever, heat or redness, regional swelling, pain or tenderness, or all of the above.

2) At least one of the elements listed below is true.

a) Isolated organisms from a wound culture acquired aseptically b) purulent discharge from a superficial infection

3) Within 30 days following surgery, an infection develops

Clinical testing was used to identify Hirschsprung-associated enterocolitis (HAEC) (rectal bleeding, fever, vomiting, explosive diarrhea, and abdominal distension)

Patients' symptoms were evaluated daily while they were in the hospital and then every two months after they were released. Six months of patient follow-up were done. The program SPSS version 26 was used to analyze the data. In order to compare categorical variables, chi-square and Fisher's exact tests were used. Variables weren't distributed regularly. Any discrepancies between the mean scores of the several groups were found using the Mann-Whitney test. Data were presented as mean and SD. Results with a P-value of 0.05 or below were deemed significant.

Results: 50 kids in all (25 in the T group and 25 in the D group) participated in this research. The groups' initial demographics were the same. Table 2. In terms of gender distribution, there were no statistical differences between the D and T Groups. There were 21 female patients and 29 male patients (58%) in total. For the comparison of fecal incontinence in groups, 5 patients in group T and 21 patients in group D were greater than or equal to 4 years old. The mean age at surgery was 6.37, with a standard deviation in the D group of 4.21 (0.66-14.00) and in the T group of 1.53 with a standard deviation of 2.43 (0.08-11.69) (p -value = 0.00). Preoperative constipation score averages were 5.260 with a standard deviation of 4.46 (1.00-18.00) in the D group and 7.58 with a standard deviation of 6.18 (2.00-24.00) in the T group (p =0.09). The comparison of the groups in relation to postoperative problems is shown in Table 3. After one month, enterocolitis was discovered in 4 of the 25 kids (16%) in the D group. While in the T group, after one month, one individual out of 25 (or 4%) was diagnosed with enterocolitis. The p -values indicate

that there is no statistically significant difference between the two groups (p-value = 1.000). After six months, 1 of 25 (4%) children in the D group had enterocolitis. Additionally, 3 of 25 (12%) trans-anal procedures identify this. Between the two groups, there is no discernible statistical difference (P-value = 0.6). Our groups don't have any anastomotic leaks. In the D group, 5 individuals (or 25%) experienced wound infections. In the T group, there was no wound infection. After one month, post-operative constipation remained a problem for 14 patients (56%) in the D group and for 7 patients (28%) in the T group (p-value =0.04). After six months, post-operative constipation remained a problem for 16 patients (64%) in the D group and for 10 patients (40%) in the T group (p-value =.08). Fecal incontinence rates did not vary significantly across groups.

Table 1: Pediatrics Constipation Score

No	Pediatric constipation score	Scores					
1	Does your kid feel abdominal pain?	Never	4	Sometimes	2	Yes Always	0
2	Does your kid have constipation issues?	Never	4	Sometimes	2	Yes Always	0
3	Does your kid have to exert a lot of effort to urinate?	No	4	Normal	2	Yes	0
4	When emptying its bowels, does your child experience any pain?	No	2	Sometimes	1	Yes Always	0
5	Can your child distinguish between feces and intestinal air?	No	0	Sometimes	0.5	Yes Always	1

6	Do you think your kid has a lot of wind?	Never	2	Sometimes	1	Yes Always	0
7	How does the stool typically appear?	Thick	0	Variable	0.5	Watery	1
8	How frequently does your kid go potty?	Less often	0	Once daily	2	Several times a day	4
9	Does your kid struggle to entirely empty its bowels?	No	3	Sometimes	1.5	Yes Always	0
10	Does your kid unintentionally pass a few feces on a frequent basis and stain their underwear?	No	2	Sometimes	1	Yes Always	0

Table 2: Group-T and Group-D Patients' Demographics

	Female	Male
Group-T N (%)	11 (44)	14 (56)
Group-D N (%)	10 (40)	15 (60)
Total (%)	21 (42)	29 (58)

Table 3: Complication rate for patients with Hirschsprung's disease during follow-up.

		Incontinency		Constipation		Enterocolitis	
		after 30 days	after 180 days	after 30 days	after 180 days	after 30 days	after 180 days
Group-T	n	3	1	7	10	3	3
	N	5	5	25	25	25	25
	Percentage	60	20	28	40	12	12

Group-D	n	8	6	14	16	4	1
	N	21	21	25	25	25	25
	Percentage	38	28	56	64	16	4
p-value		0.37	0.69	0.04	0.08	0.68	0.29

The rates of assessment complications in the groups for early and mid-evaluation were statistically indistinguishable.

DISCUSSIONS: Hirschsprung's disease is a heterogeneous genetic disorder characterized by a lack of vagal innate neural impulses in the idiopathic intracranial and myenteric plexuses. It is caused by an anomaly of the enteric nervous system of neural crest cells origin. 1 With the failure to pass meconium in the first few hours of life, or with a picture of a functional intestinal obstruction in the first week, symptoms may appear in newborns.

The most typical consequence, enterocolitis, is invariably severe and a significant factor in death in these young individuals. In infants and young children, the presentation is frequently less dramatic and does not resemble an acute intestinal obstruction; instead, these patients are more likely to suffer from severe constipation and recurrent fecal impaction.

Despite the technical improvements in HSCR surgical repair over the past 20 years, comorbidities are still frequent and constipation, incontinence, and diarrhea can have an impact on children's lifestyles.

(17) In the post-operative evaluation, long-term soiling and constipation had rates of 10.3% and 21.7%, respectively. (18)

This study's primary objective was to assess postoperative complications using the standard methods used to treat HSCR disease in our nation.

Constipation and fecal incontinence were the most frequent postoperative complications, as was evident from the results, and there were no statistically significant differences when follow-up time was taken into account. However, the percentages varied depending on the methods.

In the first month following surgery, 56 of our cases in the D group experienced constipation; however, the incidence of this complication was 28% in the T group. This result is consistent with those of a previous study, which found that constipation occurred early after surgery in 44.1% of cases, whereas it was reported later in follow-up in slightly more than 33% of patients. (19) Comparable studies' percentages ranged from 8% to 60%, whereas during our follow-up, it was reported in slightly more than a third of patients. Comparable studies' numbers varied, ranging from 8% to 60% of instances. (20-22) The distal region of the anal canal seems to momentarily relax as a result of the trans-anal pull-through technique's sphincter dilatation, which may account for the striking disparities in constipation rates during the first month of usage. However, the mechanism would revert to its pre-operative level after six months,

and the incidence of constipation would be comparable across groups. In every instance of postoperative constipation, anastomosis stricture should be taken into consideration.

Similarly, to this, incontinence rates varied greatly among studies (10- 80%).

(23,24) Our findings are consistent with those of these studies; however, because the trans-anal technique using novel devices like Loane retractors is a relatively new procedure in our clinic, the majority of the patients in the T group are infants. This is a major constraint of our research for evaluating fecal incontinence. To get more reliable findings, we advise this group to do further follow-up research.

CONCLUSIONS: There were a total of five individuals with enterocolitis in this investigation. However, since writers' definitions varied, this complication's incidence was determined to be quite wide. Others reported a rate of 9.3%, while others claimed it to be as much as 26% to 32%. (25)

In conclusion, we observed no discernible change in the mid-term prognosis for young kids with HSCR who were scheduled for surgery. To establish the optimum strategy for kids with HSCR, a multicenter randomized controlled study is necessary, nonetheless.

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