

ORIGINAL ARTICLE

Modified Duhamel Retrorectal Pull-Through for Hirschsprung's DiseaseSHAFIQ UR REHMAN¹, MUHAMMAD ANWAR², ZARLISH FAZAL³¹Assistant Professor of Paediatric Surgery, Sahiwal Medical College Sahiwal²Associate Professor of Paediatrics, Rashid Latif Medical College, Lahore³House Officer, Fatima Jinnah Medical University, Sir Ganga Ram Hospital, LahoreCorrespondence to: Shafiq-ur-Rehman, E-mail: shafiqlangriyal@gmail.com, Cell: 0300-9695031**ABSTRACT****Objectives:** To evaluate the post-operative complications and short-term outcomes of modified Duhamel retrorectal pull-through procedure for Hirschsprung's disease.**Design:** Prospective descriptive study**Place and Duration of Study:** Department of Paediatric Surgery Sahiwal Teaching Hospital Sahiwal from 1st January 2018 to 31st December 2020.**Methodology:** Thirty seven histologically confirmed cases of Hirschsprung's disease having previous decompressing colostomy/stoma age between 1 to 12 years of age were included. Children under one year of age, with severe comorbidities like Down syndrome and cardiac abnormalities, and those who require re-do pull through procedure were excluded. Modified Duhamel retrorectal pull-through procedure was performed in all cases. The demographic information included age, sex, proximal level of aganglionosis, complications of definite procedure, length of hospitalization and mortality. Other information recorded included long term complications like obstructive symptoms, enterocolitis, incontinence and soiling at follow up. Krickenbeck classification was used to evaluate faecal incontinence and constipation. Kelly's clinical score was used to assess the anal sphincter. Bowel habits were assessed only in children above three years of age.**Results:** Twenty six (70.27%) were males and 11 (29.72%) females with male to female ratio 2.3:1 and mean age at operation was 2.89±1.9 years. Twenty nine (78.37%) children were ≤3 years of age and 8 (21.62%) were >3 years. Mean weight was 12.91 kgs, operation time was 126.81 time, fasting time was 6.67 days and hospital stay was 11.91 days. Length of aganglionic segment was short segment 27 (72.97%), long segment 9 (24.32%) and total colonic 1 (2.7%). Hirschsprung associated enterocolitis 7 (18.91%) and wound infection 6 (16.21%) were most common reported complications. Constipation in 5 (13.51) and soiling with retentive constipation was present in 3 (8.10%) patients. Out of total 31 patients who reached toilet training age, 28 (90.32%) developed satisfactory voluntary bowel habits.**Conclusion:** Modified Duhamel pull-through procedure was found to be safe, applicable and with lower associated complications and satisfactory short term functional outcomes in our settings.**Key words:** Hirschsprung's disease, Modified Duhamel pull-through procedure, Complications, Outcomes**INTRODUCTION**

Hirschsprung's disease (HD) is a congenital condition characterized by absence of ganglion cells in submucosal (Meissner's) and myenteric (Auerbach's) plexuses in distal bowel extending proximally for variable lengths. It is anomaly of enteric nervous system of neural crest origin, so also referred as neurocristopathy. Histopathology by frozen or paraffin section is the gold standard for diagnosis. In 1948, Swenson and Bill described the first reconstructive procedure for surgical management of this disease.¹ With the passage of time various pull through techniques, Swenson, Soave, Duhamel and trans anal endorectal pull through, have been developed for the management of Hirschsprung's disease.²⁻⁴

Till date the superiority of any procedure is not established. It is not clear if one of these procedures gives significantly better outcomes. Regardless of the technique that is used, majority of Hirschsprung disease patients experience bowel function problems during follow-up.⁵ There are various modifications of pull through techniques for Hirschsprung's disease including Duhamel and Soave procedures.⁶ The surgical management of HD has moved from multistage open procedures to single-stage trans anal surgical techniques.⁷ The three most popular pull through procedures Swenson, Duhamel and Soave can be

performed primarily with laparoscopy. Diagnosis and status of transitional zone can be confirmed with frozen sections biopsy. However adequate experience is necessary for the success of procedure and outcomes.^{8,9}

Modified Duhamel retro rectal pull through is one of the most frequently used technique and has gained popularity due to technical ease, minimal anal stretching and better visibility.^{5,10-12} There is lot of variation in the diagnosis and treatment of Hirschsprung's disease across the centers. The most aspects of management of Hirschsprung's disease lack consensus.¹³ The surgeon's experience, expertise and available technology, are key factors in determining the choice of procedure. In our settings, our choice is modified Duhamel rhetorical pull through technique. This study is aimed to evaluate the post-operative complications and short-term outcomes of this procedure.

MATERIALS AND METHODS

This prospective study was conducted at Department of Paediatric Surgery, Sahiwal Teaching Hospital Sahiwal from 1st January 2018 to 31st December 2020. A total of 37 Duhamel retrorectal pull through procedures were performed. The confirmed cases of Hirschsprung's disease having previous decompressing colostomy/stoma, who

presented between 1 to 12 years of age were included. Children under one year of age, with sever comorbidities like Down syndrome and cardiac abnormalities, and those who require re-do pull through procedure were excluded. Hirschsprung's disease diagnosis was confirmed by clinical presentation, contrast study and histological findings. The neonates that presented with intestinal obstruction/peritonitis, underwent laparotomy, serial biopsies and stoma formation. The children that presented at older age, HD was diagnosed with barium enema and rectal biopsy. Then they underwent surgery for stoma formation and serial biopsies. Modified Duhamel pull through procedure was performed at age range between 1 to 12 years. Patients were admitted through outdoor patient department.

Pre-op assessment was carried out with clinical history, standard clinical examination and laboratory tests. Associated anomalies were assessed by abdominal sonography. All patients were prepared for bowel evaluation. 0.09% N saline enema, 20ml per kg of body weight, 08 hourly, was initiated 48 hrs prior to surgery. Fasting started one day preoperatively. Ceftriaxone and metronidazole were started 24 hours before operation and continued postoperatively. The ganglionic segment was resected and ganglionic segment was used for pull through. A full thickness histological specimen was obtained from the distal most part of the gut used for pull through in order to make sure that a bowel segment with ganglion was treated. An incision was given through the posterior wall of rectum just a cm proximal to dentate line. The ganglionic bowel was pulled through this incision behind the rectum and side to side anastomosis was carried out to the remaining aganglionic segment with the help of surgical stapler.

Data from each patient was noted in the study specific proforma. The study variables included age, gender and clinical presentation, age at definite pull through, proximal level of aganglionosis, complications of definite procedure, duration of hospitalization, mortality and requirement for subsequent surgery. Other information recorded included long-term complications like obstructive complaints (caused by anal achalasia, stricture, residual aganglionosis, transitional zone, residual fibrosis and twisted distal colon), enterocolitis, incontinence and soiling at follow up. Krickenbeck classification was used to evaluate constipation and faecal incontinence. Kelly's clinical score was used to assess the anal sphincter. Children more than three years of age were evaluated for bowel complaints (sensation of urge, ability to report urge, capability to control). All patients were followed in outdoor patient department. Follow up was continued till the child became toilet trained. Minimum followed up period was six months or till death whichever was earlier. Concepts of enterocolitis, soiling, fecal incontinence and constipation were clearly defined. Hirschsprung associated enterocolitis was defined by fever, lethargy, abdominal distension, explosive diarrhea, leukocytosis, shift to left, dilated gut loops and cut-off sign in rectosigmoid with absence of distal air. Soiling was defined as symptoms caused by pseudo incontinence due to retentive functional constipation. Fecal incontinence was defined as "uncontrolled loss of feces into places inappropriate to the social context, with no evidence

of fecal retention".¹⁴ Statistical analysis was performed using SPSS. Analysis was based on Student t test and significance was considered at $p < 0.05$ with confidence rate of 95%.

RESULTS

There were 26 (70.27%) males and 11 (29.72%) were female respectively with male to female ratio 2.3:1. Neonatal intestinal obstruction and peritonitis was most common mode of presentation and present in 23 (62.16%) patients whereas 14 (37.83%) patients presented at more than one month of age. All patients underwent colostomy and serial full thickness biopsies as a first stage procedure. Modified Duhamel retro rectal pull through procedure was performed as definite procedure in all 37 patients. Twenty nine (78.37%) children were ≤ 3 years of age and 8 (21.62%) were > 3 years at the time of operation. Length of aganglionic segment was, short segment in 27 (72.97%), long segment in 9 (24.32%) and total colonic in 1 (2.7%) [Table 1]. Mean and range of parameters of patients like age, weight, duration of operation, post-operative fasting time and hospital stay are shown in table 2. Hirschsprung associated enterocolitis 7 (18.91%) and wound infection 6 (16.21%) were most common reported complications (Table 3). Voluntary bowel habits were assessed in 31 patients. Eight patients were more than 3 years of age at the time of definite procedure whereas 24 patients reached toilet training age during follow up period. Twenty eight (90.32%) developed satisfactory voluntary bowel habits. Anal sphincter was assessed in all patients according to Kelly scoring system. Strong anal sphincter squeeze was seen in all patients. True fecal incontinence was not present in any patient. Soiling with retentive constipation was present in 3 (8.10%) patients (Table 4). No mortality was noted.

Table 1: Demographic information of the patients (n=37)

Characteristics	No.	%
Gender		
Male	26	70.27
Female	11	29.72
Age at the time of operation		
≤ 3 years	29	78.37
> 3 years	8	21.62
Aganglionosis		
Short segment	27	72.97
Long segment	9	24.32
Total colonic	1	2.70

Table 2: Descriptive statistics of the patients

Parameter	Mean (Range)
Age at operation (Y)	2.89(1-9)
Weight at operation (Kg)	12.91(10-37)
Operation time (min)	126.81(112-154)
Fasting time after operation (d)	6.67(5-11)
Hospital stay (d)	11.91(9-17)

Table 3: Frequency of post-operative complications

Post-operative complications	No.	%
Wound infection	6	16.21
Ileus	2	5.40
Anastomotic leak	1	2.70
Enterocolitis	7	18.91
Soiling	3	8.10
Constipation	5	13.51

Table 4: Functional outcomes according to Krickenbeck classification

Functional outcome	No.	%
Voluntary bowel movements	28	90.32
Soiling	3	8.10
Grade I	1	
Grade II	1	
Grade III	-	
Constipation	5	13.51
Grade I	1	
Grade II	3	
Grade III	1	

DISCUSSION

Post-operative complications after pull through procedures for HD are variable which occur in significant number of patients. The reported incidence varied between 0 and 80%.¹⁴ New surgical techniques have improved the results of surgery by decreasing the duration of operative procedure, controlling the haemorrhagic loss, adequate analgesia and shortened hospital stay.^{15,17,18} Post-operative complications including constipation. Fecal incontinence and enterocolitis among others continue to affect the quality of life of children with Hirschsprung's disease.^{19,20}

The incidence of wound infection, 6 (16.21%), in this study is higher than reported in the literature.^{21,22} Similar high incidence of wound infection 20 (23%) is reported in one study.²³ Presence of previous colostomy is common factor in both studies.

Anastomotic leakage is one of the most serious complications after pull through procedures and the reported incidence varies from 1.3% to 8%.²⁴⁻²⁶ In our study it was 1(2.7%) and managed successfully with diverting ileostomy.

Hirschsprung disease associated enterocolitis is the dreadful complication. It may develop pre or post pull through surgery. It is the most common complication, 7 (18.91%) reported in this series. The reported incidence of enterocolitis varies among studies.²⁷⁻³⁰ Our incidence is in accordance with published literature. However, the variation in the incidence of enterocolitis in the literature can be due to the difference in the definitions of enterocolitis.

The constipation frequency varied from 6-34%.³¹ Reported incidence of constipation after definite surgery in different series is, Gunadi et al³² 15%, Dai et al³³ 14% and Stensrud et al³⁴ 25%. The higher constipation rate in patients who under Soave than Duhamel procedure has been reported.³⁵ Reported incidence of constipation in our study was 5 (13.51%). Four patients with grade I and II constipation were treated with dietary changes and purgatives. One patient developed grade III constipation and was diagnosed a case of rectal spur. Rectal spur was divided with GI stapler and constipation settled. Reported incidence of soiling in this series was 3(8.10%). Soiling was seen in early follow up period and in patients with constipation. True fecal incontinence was not reported in this study. Similar to our finding, less soiling has been reported in Hirschsprung disease patients in whom Duhamel procedure was adopted.³⁶ The soiling after the Duhamel procedure could be a result of the "overflow" incontinence subsequent to constipation, because the Duhamel technique has a lesser chance of the injury to anal canal.^{36,37} Nevertheless, high incidence of long-term

fecal incontinence, 15%, has been reported in one study.³³

To avoid such variability, it is necessary to create consensus on objective and clear definitions of primary long-term outcome, (voluntary bowel functions, constipation, soiling, fecal incontinence) preferably including quality of life.

CONCLUSION

Modified Duhamel pull through procedure was found to be safe, applicable and with lower associated complications and satisfactory short term functional outcomes in our settings. Therefore, it is recommended to be continued, especially where the facility and expertise for frozen section biopsy and laparoscopy are not available.

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