

Clinical Study of Hirschsprung Disease and Long-Term Outcome of Duhamel's Pull-Through

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Abstract: **Background:** Duhamel's Pull Through (DP) is one of the established surgeries described for Hirschsprung's disease (HD). It is the most common procedure for HD at our hospital. **Objective:** Clinical Study and analysis of the results, complications, and follow-up of DP in patients of HD at a tertiary care centre. **Materials and Methods:** 29 patients has been selected. Medical records of 29 patients who had undergone DP for HD at a tertiary care institute were obtained. These patients were followed in the outpatient's department to see any complications. This data were used to critically analyse this operative method and its results. **Results:** Ureter injury (3%), burst abdomen (7%), fecal fistulae (3%), perianal excoriation (3%), parastomal hernia (3%), Soiling (38%), bleeding PR (3%), fecal incontinence (9%), loose motion (35%), urinary incontinence (3%) and constipation (12%) were some complications observed in this study. Most of these complications were successfully managed. **Conclusions:** Staged DP done after an initial colostomy has good results and is being done in all cases of megacolon with constipation. Post-operative complications can usually be managed successfully leading to a significant improvement in the quality of life.

Keywords: Duhamel's procedure, Hirschsprung's disease

1. Introduction

Hirschsprung's disease (HD) is a congenital disorder of the enteric nervous system resulting in congenital aganglionosis of bowel and causes chronic constipation. Operation is the only treatment of this disease and involves the removal of the aganglionic segment and creating intestinal continuity. Duhamel's procedure (DP) which involves excision of the aganglionic segment with retrorectal pull-through, and anastomosis of the ganglionated bowel is one of the traditional surgical procedures for HD. We present the results of this procedure at our institute.

2. Materials and Methods

This was a retrospective study conducted in the Department of Pediatric Surgery, Institute of Post Graduate Medical Education & Research, Kolkata, West Bengal, India. The medical records and follow-up details of 29 patients who underwent DP for HD between April 2021 and April 2023 were used to obtain data regarding their clinical history, investigation results, details of surgery, and any intra- or post-operative complications.

3. Procedure

All the patients included in this study go through DP following an primary stoma formation and leveling biopsy. Neonates and young infants were managed on stoma for a few months to years before their anal canal could accommodate the stapler used to divide the spur. No patients underwent primary DP. Therefore, all these patients had biopsy-proven HD at the time of DP and this allowed biopsy-proven ganglionated segments to be pulled down during final surgery. The stoma which had been proven to be ganglionated at the time of previous surgery, bowel was divided and this part was pulled down retrorectally to complete the anastomosis 1 cm proximal to the dentate line. Spur was divided by a 75 mm linear stapler. In cases of older children when the spur was too long to be taken care of by a single stapler from below, another 75 mm stapler was fired from above to ensure no remnant spur remains, and Martin's modification was done to

anastomose the rectal stump to the pulled down bowel at the same level. Important details included age at presentation, level of aganglionosis, any intraoperative complication and any post-operative complication such as injury to ureter, post operative burst abdomen, faecal fistulae, perianal excoriation, parastomal hernia, bleeding PR, band obstruction, constipation, and incontinence. All relevant data were analysed.

4. Results

Age of Presentation (Table 1)

This ranged from neonatal to 2 years (oldest). Mean age was 11.14 months. 69% of the patients presented at neonatal age, 7% at 1month, 3% at 2months, 7% at 6months, 3% at 10 months, 7% of the patients presented beyond 1 year of age and 3% patient above 2yrs. Delayed presentation increases the morbidity and complications of HD.

Level of Aganglionosis (Table 2)

Classical (Recto-sigmoid) HD was seen in 18 patients (62% cases), mid sigmoid 5 patients (17%), descending colon 1 (3%), transverse colon 2(7%), long segment 5 patients (17%) while 3patients (10% cases) presented with total colonic aganglionosis.

Spastic segment biopsy (Table 3)

Ganglion cells absent in 20 patients (65%), ganglion cells present in 3 (10%), hypertrophied nerve fibre in 2 (6%) while in 6 patients (19%) reports were inconclusive.

Rectal biopsy (Table 4)

Rectal biopsy is done in 9 patients and out of this in 8 patients ganglion cells is absent (88%).

RAIR (Recto Anal Inhibitory Reflex) (Table 5)

RAIR is absent in HD and present in normal children. We did anorectal manometry in 5 patients. Out of these 5 patients, RAIR absent in (60%) and present in (40%) patients. The sensitivity and specificity of RAIR is more in older children.

Table 1

Age of presentation		
Neonate	20	69%
1 month	2	7%
2 months	1	3%
6 months	2	7%
10 months	1	3%
1yr2mnths	2	7%
2 yrs	2	7%

Table 2

Level of aganglionosis		
Rectosigmoid	18	62%
Descending colon	1	4%
Transverse colon	2	7%
Long segment	5	17%
Total colonic aganglionosis	3	10%

Table 3

Spastic segment biopsy		
Ganglion cells (GC) absent	20	65%
Ganglion Cells (GC) present	3	10%
Hypertrophied nerve bundle	2	6%
Inconclusive	6	19%

Table 4

Rectal biopsy	Done in 9 patients	
GC absent	8	88%
GC present	1	12%
Not done	20	

Table 5

RAIR (Recto Anal Inhibitory Reflex)	Done in 5 patients	
Absent	3	60%
Present	2	40%
Not done	24	

Age of Modified Duhamel surgery (Table 6)

Total 29 patients undergone Modified Duhamel surgery out of which 1patient at age of less than 1 year, 8 patients 1-2yrs, 9 patients 2-3yrs, 4 patients 3-4yrs, 2 at 4-5yrs, 2 at 5-6yrs, 1 at 6-7yrs, 1 at 8-9yrs and 1patient at 9-10yrs.

Table 6

Age of Modified Duhamel surgery		
< 1yr	1	3%
1 to 2yrs	8	28%
2 to 3yrs	9	31%
3 to 4yrs	4	14%
4 to 5 yrs	2	7%
5 to 6yrs	2	7%
6 to 7 yrs	1	3%
7 to 8 yrs	0	0%
8 to 9 yrs	1	3%
9 to 10 yrs	1	3%

Perioperative complications (Table 7)

Out of 29 patients 7 patients had perioperative complications.

Peri operative complication		
Ureter injury	1	3%
Ganglion cell positive in resected segment biopsy	1	3%
Post operative burst abdomen	2	7%
Post operative fecal fistulae	1	3%
Perineal excoriation	1	3%
Parastomal hernia	1	3%

Short term outcome showed 13% postoperative complications required redo surgery. Those are ureter injury (n-1), post operative burst abdomen (n-1) and parastomal hernia (n-1).

Early complication (less than 1month) (Table 8)

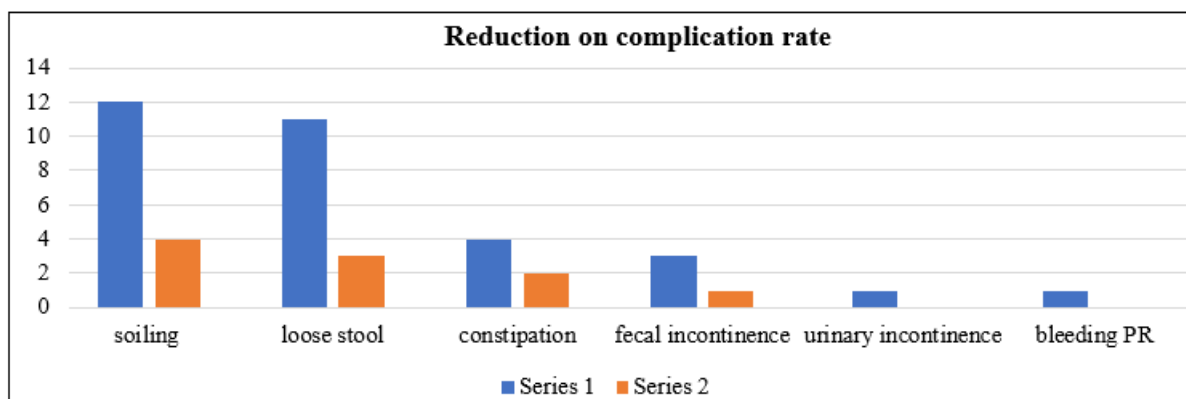
Postoperative loose stool, soiling, constipation, fecal incontinence, urinary incontinence and rectal bleeding.

Early post op complications		
Soiling	12	34%
Loose stool	11	31%
Constipation	4	12%
Fecal incontinence	3	9%
Urinary incontinence	1	3%
Bleeding PR	1	3%
No complaints	3	9%

Late follow up (upto 1yr)

The postoperative complications of early postoperative period reduced significantly in late follow up.

Late follow up		
Soiling	4	14%
Loose stool	3	10%
Constipation	2	7%
Fecal incontinence	1	3%
Urinary incontinence	0	
Bleeding PR	0	
No complaints	19	66%



5. Discussion

HD is the one of the common surgical cause for chronic constipation in children. Three classical Surgical management of HD are Soave, Swenson, and DP. (1)

Patients with HD represent a heterogeneous group, considering the age at which surgical consultation is taken and ways in which these patients present and are managed. Our study also reflects this heterogeneity. While most patients (69%) presented as neonates or young infants, there were significant number of late presenters (13% >1 year of age) whose parents had long been ignorant of their condition. Most of these late presenters were from low socioeconomic group and were denied early attention. Delayed presentation makes children vulnerable for related complications of HD. The mean age at which HD is diagnosed has gone down worldwide and in many developed countries, diagnosis in the neonatal period has increased. This has led to timely surgical intervention resulting in fewer incidences of HD-related complications and post-operative morbidity.

Classical rectosigmoid disease was seen in 62 % of patients, whereas 37 % patients had long-segment disease including three cases of total colonic aganglionosis. Long-segment HD meant more length of bowel to be respected and more intraoperative time. Two cases of total colonic aganglionosis was managed using Martin's modification of DP.

In spastic segment biopsy 10% patients where ganglion cells are present but in rectal biopsy ganglion cells were absent and RAIR was absent in one of the patient and In other two RAIR was not done.

Modified Duhamel surgery was done mostly at 1-2yrs (28%) and 2-3yrs (31%) .

Perioperative complications are very few. Those are ureter injury (3%), burst abdomen (7%), fecal fistulae (3%), perineal excoriation (3%) and parastomal hernia (3%).

Post-operative complications observed in two different timeline first is within one month and second is one year follow up. In first month our study included perianal soiling (38%), loose stool (35%), constipation (12%), fecal incontinence (9%), bleeding per rectum (3%) and urinary incontinence (3%). These complications have been variously reported in several large series. [2] One patient had delayed bleeding after 2 weeks of surgery. On examination, he had polyp formation in the suture line on colonoscopy. This study recorded fewer incidences of perianal soiling and excoriation, in comparison to other studies; these symptoms lessened with time and barrier skin protective agents helped in their healing. Constipation has been reported to occur in 5-8% of patients following DP. [3] In our study, we found constipation in 12% of cases in one month follow up which was reduced to 7% in long term follow up. Assessment of incontinence was done in our patients, 9% of patients were found to be incontinent in 1 month which was reduced to 3% in long term. Fecal soiling was seen in 38% in first month which came down to 14% in 1 year follow up. In a review of 2430 postoperative Duhamel patients, 5.3% of patients showed fecal soiling.[4] Similar observations have been shared by other investigators. [5] Most of these patients have been found to improve with time

and do well with dietary modifications and bulking agents before any surgical intervention.

As many as 3% of patients had lower urinary symptom such as dribbling of urine following DP. This observation was much lower than those of other observers. [6]

The etiology of these symptoms is multifactorial including damage to pelvic splanchnic nerves, hypogastric nerves, or pelvic nerve plexus. Furthermore, a large rectal reservoir may lead to outflow obstruction. Appropriate evaluation using sonogram, voiding cystourethrography, and urodynamic study guides further management in such patients.

No mortality occurred during this study. Most of the series on post-operative complications in DP have reported a low mortality rate and the most common cause has been enterocolitis. [7]

6. Conclusions

Staged DP following levelling colostomy for HD is a solution to the entire spectrum of the disease. It can be done with ease in uncomplicated as well as complicated cases such as those presenting with long-segment disease and massive megacolon. It has less complication and therefore, significantly improves the quality of life in patients suffering from HD

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