

Outcome of Duhamel's Pull-through in Hirschsprung's Disease: A Tertiary Center Experience

Vinit K Thakur¹, Sandip K Rahul²

¹Associate Professor, Department of Paediatric Surgery, Indira Gandhi Institute of Medical Sciences, Patna, Bihar, India, ²Assistant Professor, Department of Paediatric Surgery, Indira Gandhi Institute of Medical Sciences, Patna, Bihar, India

Abstract

Background: Duhamel's procedure (DP) is one of the classical surgeries described for Hirschsprung's disease (HD). It is the most common procedure for HD at our center.

Objective: Analysis of the results, complications, and follow-up of DP in patients of HD at a tertiary care center.

Materials and Methods: Medical records of 72 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 their quality of life and any complications. This data were used to critically analyze this operative method and its results.

Results: Wound infection (5.56%), rectal stump leak (1.39%), perianal excoriation (5.56%), enterocolitis (8.33%), stricture formation (1.39%), constipation (6.94%), bleeding per rectum (2.78%), remnant spur and spur-related complication (4.17%), fecal impaction (2.78%), adhesive bowel obstruction (2.78%), incontinence (5.56%), and lower urinary symptoms (9.72%) 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 can be done in complicated cases presenting with perforation, long-segment disease, enterocolitis, and massive megacolon. Post-operative complications can usually be managed successfully leading to a significant improvement in the quality of life.

Key words: Constipation, Duhamel's procedure, Hirschsprung's disease

INTRODUCTION

Hirschsprung's disease (HD) is a developmental disorder of the enteric nervous system resulting in congenital aganglionosis of bowel. This causes chronic constipation. Operative intervention is the only treatment of this entity and involves the removal of the aganglionic segment and establishing 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 classical surgical

procedures for HD. We present the results of this procedure at our institute.

MATERIALS AND METHODS

This was a retrospective study conducted in the Department of Pediatric Surgery, Indira Gandhi Institute of Medical Sciences, Patna, Bihar, India. The medical records and follow-up details of 72 patients who underwent DP for HD between January 2011 and December 2016 were used to obtain data regarding their clinical history, investigation results, details of surgery, and any intra- or post-operative complications.

Procedure Details

Detailed history including age, demographic details, chief symptoms and their duration were noted from the hospital records. All the patients included in this study underwent

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Corresponding Author: Dr. Vinit K Thakur, D4/3, Indira Gandhi Institute of Medical Sciences Campus, Sheikhpura, Patna - 800 013, Bihar, India. Phone: +91-8084044086. E-mail: dr.vinit.igims@gmail.com

DP following an initial stoma formation and leveling biopsy. Neonates and young infants were managed on stoma for a few months before their anal canal could accommodate the stapler used to divide the spur. Older children had to wait for longer duration for the hypertrophied bowel to lessen its caliber. None of these patients underwent primary DP. Hence, all these patients had biopsy-proven HD at the time of DP and this enabled biopsy-proven ganglionated segments to be pulled down during definitive surgery.

At the time of DP, stoma was mobilized and any significantly dilated segment sacrificed. 5 cm proximal to the site of 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 long-segment disease or sometimes in 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, and Martin's modification was done to anastomose the rectal stump to the pulled down bowel at the same level. In cases where stapler misfired or could not divide the spur adequately, we used the Duhamel's clamp and kept it for 5-7 days so as to divide the spur completely.

Important details included age at presentation, associated anomalies, level of aganglionosis, intraoperative time during DP, length of bowel resection, intraoperative blood loss, any intraoperative complication, length of stay in hospital, and any post-operative complication such as anastomotic leak, enterocolitis, stricture formation, perianal excoriation, constipation, and incontinence.

To assess continence in children >4 years of age, Kelly scoring was adopted which included three parameters: (1) Straining or smearing, (2) accidental defecation/soiling, and (3) strength of puborectalis muscle.¹ All these parameters were scored from 0 to 2. Sum total of Kelly score was used to draw inference regarding continence: (1) Good continence - total score 5 or 6, (2) fair - total score 3 or 4, and (3) poor continence - total score 1 or 2.

All relevant data were analyzed.

RESULTS

Age at Presentation (Table 1)

This ranged from 5 days (neonatal) to 7 years (oldest).

Youngest child was 5 days old, whereas the oldest child was 7-year-old. Mean age was 16.24 months. 43% of the patients presented between 1 and 6 months of age. 15% of the patients presented beyond 2 years of age

(late presenters) and most of them came from low socioeconomic group. Delayed presentation is common in developing countries like India and this increases the morbidity and complications of HD.

Sex Distribution (Table 2)

Male to female ratio of HD patients in this study was 5:1.

Associated Anomalies

- Down syndrome: 2 (3.86%)
- Congenital talipes equinovarus (CTEV): 2 (3.86%)
- Cardiac anomalies: Atrial septal defect - 2 (7.6%)
- Ventricular septal defect: 1 (3.86%).

Mode of Presentation (Table 3)

Constipation with abdominal distension was the most common symptom. History of delayed passage of meconium was seen in 52.78% patients while enterocolitis was seen in 16.67% of cases.

Level of Aganglionosis (Table 4)

Classical (Recto-sigmoid) HD was seen in 70.83% cases, while 29.17% cases presented with long segment disease.

Table 1: Age at presentation

Age group	Number of patients (%)
<1 month	08 (11.11)
1-6 months	31 (43.05)
6-12 months	12 (16.67)
1-2 years	10 (13.89)
More than 2 years	11 (15.28)
Total	72

Table 2: Sex distribution

Sex	N (%)
Male	60 (83)
Female	12 (17)

Table 3: Mode of presentation

Presentation	Number of patients (%)
Non/delayed passage of meconium	38 (52.78)
Perforation	3 (4.17)
Constipation and abdominal distension	60 (83.33)
Enterocolitis	12 (16.67)

Table 4: Level of aganglionosis

Level	Number of patients (%)
Rectosigmoid HD (classical HD)	51 (70.83)
Long-segment HD (beyond descending colon)	21 (29.17)

HD: Hirschsprung's disease

Table 5: Complications

Complication	Number of patients (%)	Management
Wound infection	4 (5.56)	Conservative dressings, intravenous antibiotics
Rectal stump leak	1 (1.39)	Proximal diverting stoma followed by anastomotic revision later
Perianal excoriation	4 (5.56)	Barrier skin protective agents
Post-operative enterocolitis	6 (8.33)	Bowel rest, intravenous medications, fluids, and rectal washes
Stricture formation	1 (1.39)	Improved on serial dilatation
Constipation	5 (6.94)	1 managed conservatively; 1 had stricture, managed on dilatation; 2 had remnant spur which needed division by clamp; 1 had remnant dilatation of proximal colon resection and antegrade enema through Malone's procedure
Bleeding per rectum	2 (2.78)	Conservative T/t; antibiotics
Remnant spur and spur-related complications	3 (4.17)	Washes; reapplication of Duhamel's clamp to divide remnant spur
Fecal impaction	2 (2.78)	Rectal washes; laxatives
Adhesive bowel obstruction	2 (2.78)	Conservative management in one; adhesiolysis in other
Mortality	0 (0)	

Intraoperative Time

This was influenced by many factors including age, prior history of enterocolitis, adhesions surrounding the stoma and bowel loops, and intraoperative bleeding.

Operating time varied between 90 and 180 min with a mean of 125 min. In 10 cases, failure of stapler added to the operating time and Duhamel's clamp had to be applied in these cases.

Length of Bowel Resection

This again varied depending on the length of the spastic segment and the length and caliber of the dilated segment. This varied between 20 and 55 cm with a mean of 26 cm.

Intraoperative Blood Loss

This ranged from 15 to 120 ml with a mean of 35 ml.

Patients with significant adhesions, peristomal excoriation, older patients, and those with a history of enterocolitis had more blood loss which necessitated blood transfusion in the post-operative period.

Failure of Stapling Device during Procedure

About 10 (13.89%) of our patients had problems related to the stapling device. In these patients, Duhamel's clamp was applied and kept for 5-7 days in the post-operative period. This decreased child's movement and made him anxious.

In 6 (8.33%) patients, a second 75 mm stapler had to be fired from above to take care of a long spur not adequately taken care of by a single 75 mm stapler.

Length of Stay in Hospital

This ranged between 6 and 15 days with a mean of 8 days.

Table 6: Functional outcome

Average stool frequency (3 weeks)	4.2/day
Average stool frequency (6 months)	2.6/day
Average stool frequency (1 year)	1.8/day
Incontinence	4 (5.56%)
Lower urinary symptoms	7 (9.72%)

Complications (Table 5)

Post-operative enterocolitis, persistent constipation, wound infection and perianal excoriation was some of the important complications of the procedure.

Functional Outcome (Table 6)

Although the average stool frequency gradually became normal with age, a few patients complained of some degree of fecal incontinence and lower urinary symptoms.

Cost Analysis

The average cost of surgical procedure was around Rs. 31,150 which included the cost of operative procedure, stapling device, and post-operative stay.

DISCUSSION

HD is the one of the common surgical cause for chronic constipation in children.^{2,3}

Surgical management of HD has evolved through the three classical surgeries (Soave, Swenson, and DP) and their modifications to single-staged techniques, total endorectal techniques, and laparoscopy-assisted procedures. Developments in histopathological techniques together with frozen section methods have gone hand-in-hand with advancements in surgical management. A committed histopathology team with this facility is a pre-requisite for all single-staged procedures. In a

resource-challenged country like India, we still have places where these facilities are not developed to support single-staged surgeries and often patients present with different complications of HD such as perforation, enterocolitis, or massive megacolon which necessitate staged procedures. DP or its modifications best address the needs of these patients with an acceptable complication rate.

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 some patients presented as neonates or young infants, there were significant number of late presenters (29.17% >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.⁴ This has led to timely surgical intervention resulting in fewer incidences of HD-related complications and post-operative morbidity.

Male:female sex ratio of patients included in this study was 5:1. Male predominance in HD has been like most other series.⁵ However, there was no bias in the selection of the patients. This ratio only represents the way these patients turned up.

Different associations of HD have been described.⁶⁻¹⁰ In our series, we noted Down syndrome, CTEV, and cardiac anomalies as some of the associations. With a reported incidence of 4.5-16% in HD patients, Down syndrome is the most common chromosomal abnormality associated with HD.^{7,11} Patients of Down syndrome had difficulties in toilet training and although their symptoms improved with DP, they still had on and off constipation. Patients of HD with cardiac anomalies in our series did not have any unusual risk for anesthesia or surgery during definitive management. None of our patients had any unusual hereditary syndromes known to be rarely associated with HD such as Shah-Waardenburg syndrome, multiple endocrine neoplasia Type 2 syndrome, and congenital central hypoventilation syndrome.

About 4.17% of patients presented with perforation in the neonatal period. Site of perforation was cecum in one and ascending colon in 2 patients. In these patients, initially, stoma formation at the site of perforation with leveling biopsy was performed before a subsequent DP. It has been observed that 98% of normal full-term infants pass meconium in the first 24 h of life and the remainder by 48 h.¹² In this study, 52.78% of patients gave a history of

delayed or non-passage of meconium in the first 48 h of birth. Many ignorant parents, particularly of late presenters could not recall the time taken to pass meconium after birth. Chronic constipation and abdominal distension were noted in 83.33% of cases. Enterocolitis was seen in 16.67% patients. Reported incidence of enterocolitis has ranged from 12% to 58%.^{4,13}

Classical rectosigmoid disease was seen in 70.83% of patients, whereas 29.17% patients had long-segment disease including one case of total colonic aganglionosis. Long-segment HD meant more length of bowel to be respected and more intraoperative time. One case of total colonic aganglionosis was managed using Martin's modification of DP.

Intraoperative time during definitive DP depended on a large number of factors including the extent of adhesions following previous surgery, length of aganglionic segment, length and caliber of the dilated segment, whether Martin's modification was applied or not to DP, whether some intraoperative difficulty was seen such as failure of the stapling device in dividing the spur. Mean duration of surgical procedure during this study was 125 min.

There was considerable variation in the length of the bowel resected. It varied from 20 to 55 cm with a mean length of 26 cm. It not only depended on the length of the spastic aganglionic segment but also on the length and caliber of the excessively dilated bowel proximal to it which had to be resected to facilitate normal transit. The reason to perform a longer resection that extends beyond the dilated and thick-walled bowel is to avoid bowel dysfunction owing to associated "hypo-" or "dys-" ganglionosis. Since our center does not have facilities for frozen section biopsy to confirm the presence of ganglion cells at the site of anastomosis; we had to be more liberal in resection of the intestinal segment to avoid the inconcordance between the radiographic transition zone and the pathologic extent of aganglionic bowel.

Resected specimens underwent histopathological examination for the confirmation of presence of ganglion cells in the upper segment of the resected bowel. All such specimens sent during this study showed the presence of ganglion cells at proximal end and their absence at distal end.

Intraoperative blood loss also varied depending on whether there was significant adhesion, peristomal excoriation and history of enterocolitis, length of the bowel resected, and age of the patient.

In a significant number of our patients, we had several practical problems such as failure of the stapling device

(10 patients), inadequate division of the spur following application of staples (3 patients), large spurs requiring more than one stapler (6 patients), or non-availability of staplers due to cost constraints.

We had an unacceptably high failure rate of the stapling device. Careful analysis led us to conclude that staplers should be fired in a virgin space so that no suture should interfere in the line of the stapler and if possible, a new gun should be used in each case. Despite these precautions, in many of these patients, we had to resort to the application of Duhamel's clamp for division of the spur. Although these patients had inconvenience in keeping the clamp *in situ* for 5-7 days, there was complete division of the spur following application of Duhamel's clamp. Patients in whom Duhamel's clamp was used had increased post-operative stay in the hospital compared to those patients in whom stapling device was used to divide the spur.

Mean post-operative stay of 8 days in our study is comparable to that reported by other investigators.¹⁴

Post-operative complications observed in our study included wound infection (5.56%), rectal stump leak (1.39%), perianal excoriation (5.56%), enterocolitis (8.33%), stricture formation (1.39%), constipation (6.94%), bleeding per rectum (2.78%), remnant spur and spur related complication (4.17%), fecal impaction (2.78%), and adhesive bowel obstruction (2.78%). These complications have been variously reported in several large series.¹⁵⁻¹⁷

The incidence of enterocolitis in the post-operative period has been found to range from 5% to 26% in different studies.^{15,18,19} We encountered enterocolitis in 8.33% of our patients. These patients improved on conservative management.

Two patients had delayed bleeding after 2 weeks of surgery. On examination, they had granuloma formation in the suture line on examination. They were conservatively managed and did not need any active surgical intervention.

One patient had leakage of the stump. In this patient, a proximal stoma was fashioned to divert the fecal stream.

One patient, who had lost to follow-up for a long time, presented after 3 years with progressive stenosis of the anastomosis and this led to progressive dilatation of the proximal bowel. This patient improved on serial dilatation and washes.

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.^{15,20,21} In our study, we found constipation in 6.94% of cases. In 1 patient, constipation gradually improved on conservative management with rectal washes. 2 patients had remnant spur which led to fecal impaction and this was managed by dividing the spur with Duhamel's clamp. 1 patient had stricture at the anastomotic site which was managed by serial dilatation; in one patient, there was significant dilatation of hypoperistaltic proximal bowel which necessitated its resection and antegrade enema using Malone's procedure.

Assessment of incontinence was done in patients more than 4 years of age using Kelly score. 5.56% of patients were found to be incontinent. In a review of 2430 post-operative Duhamel patients, 5.3% of patients showed fecal soiling.¹⁵ Similar observations have been shared by other investigators.^{22,23} Most of these patients have been found to improve with time and do well with dietary modifications and bulking agents before any surgical intervention.

Average stool frequency was found to improve from 4.2/day at 3 weeks after surgery to 2.6/day at 6 months after surgery and 1.8/day at 1 year after surgery.

As many as 9.72% of patients had lower urinary symptoms such as urinary retention, poor stream, and dribbling of urine following DP. This observation was akin to those of other observers.²⁴

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.¹⁵

CONCLUSIONS

Staged DP following initial colostomy for HD is an answer 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 perforation, long-segment disease, enterocolitis, massive megacolon, and in setups without

frozen section techniques. It has less complication and therefore, significantly improves the quality of life in patients suffering from HD.

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