Original Article

Duhamel's procedure for Hirschsprung's disease and the functional outcome in a tertiary care center

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ABSTRACT

Introduction: Hirschsprung's disease (HD) is a developmental disorder caused by the migration failure of neural crest cells during intestinal development, resulting in an aganglionic colon and causing a functional obstruction in children. Duhamel's procedure (DP) which involves excision of the aganglionic segment with retrorectal pull through, and anastomosis of the ganglionated bowel above dentate line, is one of the classical surgical procedures for HD. **Materials and Methods**: This retrospective study was conducted in a tertiary care center in South India. The medical records and follow-up details of 17 patients, who underwent DP for HD between January 2014 and December 2018, were obtained and evaluated. Outcomes evaluated included incidence of post-operative constipation, incontinence/soiling, enterocolitis, anastomotic stricture, and leak. **Results:** There were 17 patients with HD who underwent DP during the study period, among which 76.47% of patients had classical HD and 23.52% were long-segment HD. A total of 58.82% of patients presented at birth, with overall 76.47% of male predominance. Fecal soiling (29.41%), perianal excoriation (17.64%), wound infection (11.76%), post-operative enterocolitis (11.76%), stricture formation (11.76%), constipation (11.76%), and hypertension (11.76%), and adhesive bowel obstruction (5.88%) were some complications observed in this study. Most of these complications were successfully managed. **Conclusion:** Staged DP is safe and can address the entire spectrum of HD. Complications are less and comparable to other techniques, and with close follow-up to address the post-operative complications, good quality of life can be achieved for the children with HD.

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

Hirschsprung's disease (HD) is a developmental disorder caused by the migration failure of neural crest cells during intestinal development, resulting in an aganglionic colon and causing a functional obstruction in children. HD is considered a neurocristopathy, a disorder of cells and tissues derived from the neural crest, and may occur as an isolated finding or as part of a multisystem disorder. The incidence ranges from 1 in 4500 to 1 in 7000 live births with a male-to-female ratio of approximately 4:1 [1,2].

According to the length of aganglionosis, HD can be classified as follows: (1) Short segment, (2) long segment, and (3) total colonic aganglionosis. In 80% of individuals, aganglionosis is restricted to the rectosigmoid colon (short-segment disease) [1,2]. Diagnosis is usually made by classical presentation of HD, supported by contrast enema showing a transition zone (TZ). Rectal biopsy showing absent ganglionic cells, nerve hypertrophy, and increased acetylcholinesterase activity on immune histochemical study is confirmatory. Acetylcholinesterase histochemistry is a useful adjunct for the diagnosis of HD and is only performed with frozen sections [3].

Duhamel's procedure (DP) which involves excision of the aganglionic segment with retrorectal pull through, and anastomosis of the ganglionated bowel above dentate line, is one of the classical surgical procedures for HD [4]. The voluntary bowel movement and the absence of soiling or constipation following pull through remain the most important markers of good outcome [5]. This study is an attempt to evaluate the early results of the DP for HD in tertiary care center.

MATERIALS AND METHODS

This retrospective study was conducted in the Department of Pediatric Surgery at Mysore Medical College and Research Institute, Mysore, Karnataka, India. The medical records and follow-up details of 17 patients, who underwent DP for HD between January 2014 and December 2018, were used to obtain

Access this article online	
Received - 22 November 2020 Initial Review - 03 December 2020 Accepted - 06 January 2021	Quick Response code
DOI: 10.32677/IJCH.2021.v08.i01.010	

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data regarding their clinical history, investigation results, details of surgery, and any intraoperative or post-operative complications. Subsequently, patients were called for the assessment of functional outcome and the details of individuals were documented. Diagnosis of HD in our hospital was established accordingly to the clinical manifestation, contrast enema, and histopathology findings.

Important details recorded during the study include age at presentation, detailed history including demographic details, associated anomalies, level of aganglionosis, intraoperative time during DP, length of bowel resected, 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. The diagnosis of HD was confirmed by histopathology either after rectal biopsy or by leveling biopsies when emergency abdominal exploration was done for acute intestinal obstruction or peritonitis.

All the patients included in this study underwent DP by a single surgeon following an initial leveling stoma formation. Definitive procedure was considered after adequate weight gain in infants so that their anal canal could accommodate the stapler used to divide the common wall. DP was deferred in older children till the dilated bowel returned to normal caliber. None of these patients underwent primary DP.

Surgical Technique

During leveling colostomy, for cases with prior proven HD (post-rectal biopsy), rectum was closed such as Hartman's procedure and spastic segment, TZ, including dilated bowel, was resected. End colostomy constructed and doughnut of the stoma was sent for four-quadrant histopathological confirmation of normal innervation. When emergency exploration was done without prior diagnosis, multiple leveling biopsies were taken and loop stoma was constructed proximal to dilated segment and proximal end of stoma biopsied. In this case, bowel resection was planned during definitive procedure. Hence, at definitive procedure, further need for frozen section biopsy could be avoided and we have a biopsy-proven stoma for pull through in either situation. Length of resected bowel varied from 12 cm to 55 cm with a mean length of 26 cm. The reason to perform a longer resection that extends beyond the dilated and thick-walled bowel is to avoid bowel dysfunction due to associated "hypo-" or "dys-" ganglionosis [4].

All the cases had biopsy-proven stoma at definitive procedure, as leveling colostomy was done in every case. During DP, once stoma was mobilized, colon was divided 5–10 cm proximal to the stoma. Colonic tumbledown technique was employed for adequate mobilization in three of the long-segment HD cases. Mobilized colon was routed per retrorectal space to complete the anastomosis 0.5 cm proximal to the dentate line. Common wall was divided by a 55 mm linear stapler fired from either side in the initial six cases followed by proximal colorectal anastomosis. In the remaining 11 cases, single 75 mm linear stapler was applied per rectal, to divide the common wall and rectal stump sutured close to the stapled line (Figs. 1-3).

Delayed passage of meconium was defined as that occurring later than 48 h after birth. Enterocolitis was defined by the presence of distended bowel, fever, and foul-smelling stool along with positivity for serum inflammatory markers. Dilation was defined as a widening of the anastomotic area under general anesthesia. Calibration was defined according to Hegar size of the anal canal assessed in the outpatient clinic [6]. All relevant data were analyzed.

RESULTS

This ranged from neonatal period to 5 years of age. Youngest child was 3 days old, whereas the oldest child was 5 years old. Most common age at presentation was neonatal period, and among them, majority presented within the 1st week, accounting for 11 (64.7%) cases. Four (23.52%) of the patients presented between 1 and 12 months of age. Late presenters accounted for 2 (11.76%) of cases, who presented beyond 1 years of age. In our present study, 13 (76.47%) were male and 4 (23.52%) were female.

Out of 17 cases, 15 (88.23%) of cases had a history of non-delayed passage of meconium. Six cases of the above had non-passage of meconium, which did not respond to bowel wash and underwent emergency leveling stoma formation. Two children of the above had perforation with peritonitis for which emergency abdominal exploration was done in a child with cecal perforation. Of the two cases with no history of delayed passage of meconium, one child had abdominal distension and positive family history of HD for which the child was evaluated. The second child had chronic constipation and proven HD in younger sibling (Table 1).

With respect to age, early neonates had non/delayed passage of meconium. Beyond neonatal period, abdominal distention and constipation were the presentation, which was noted in 6 (35.29%) of cases. Four of these children had a history of delayed passage of meconium, but were not evaluated.

Two children were found to have hypertension in the perioperative period and were on antihypertensive medication for 6 months. Workup for organic causes of hypertension was negative in these cases. One child had Shah–Waardenburg syndrome with sensorineural hearing loss and heterochromia iridis, has been rehabilitated with hearing aids. One child had cerebral palsy with bilateral lower limb paraparesis.

The majority (12, 70.58%) of cases underwent two surgeries, first being leveling colostomy followed by definitive Duhamel pull through. Three (17.64%) of cases underwent three procedures, that is, leveling colostomy followed by DP with covering stoma, and at last stoma closure. The reason for covering stoma was long-segment disease with tumble down in two cases and anticipation of anastomotic healing issues in other case. Among 2 (11.76%) cases that underwent four surgeries, one child developed intestinal obstruction features on post-operative day 14 of DP, for which adhesionolysis was done with covering ileostomy. Ileostomy closure was done at a later date. Another child underwent stricturoplasty with covering ileostomy followed by ileostomy closure later.

Thirteen cases had classical rectosigmoid variety and four cases had long-segment involvement. Intraoperative duration was influenced by many factors including age, prior history of enterocolitis, bowel adhesions, long-segment disease with need for tumble down procedure, and intraoperative bleeding. Operating time varied between 150 and 240 min with a mean of 180 min. Level of bowel resected 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.

GI stapling device was used in all the procedures in the present study and Duhamel's clamp was not used in any of the cases. In the first 6 (35.29%) patients, a second 55 mm stapler had to be fired from above to achieve wide anastomosis and to avoid spur. In later 11 (64.70%) cases, single 75 mm stapler was fired per rectal, which gave wide anastomosis and reduced the operative time significantly. There were no incidences of stapler misfiring OR failure. Post-operative enterocolitis, persistent constipation, anastomotic stricture, wound infection, and perianal excoriation were some of the important complications of the procedure (Table 2).

On assessing the functional outcome, we found that the average stool frequency gradually became normal with age and only few patients complained of some degree of fecal incontinence by 12 months post-DP.

DISCUSSION

HD is one of the "classics" of pediatric surgery, and the development of HD management closely portrays the overall development and quality of pediatric surgery. Removal of aganglionic bowel, pulling through of ganglionated bowel, and preserving the anal canal and sphincter mechanism remain the principles to surgical repair regardless of technique [7]. Despite significant developments in the understanding of the pathologic anatomy and physiology of HD, the results of surgical therapy remain far from perfect [8]. 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. A committed histopathology team with expertise in frozen section methods is a pre-requisite for all single-staged procedures [3,4].

Staged procedures for HD can be performed safely with acceptable functional outcomes even in the absence of frozen section facility. As DP or its modifications require minimal pelvic dissection and can be easily performed with lesser procedure related complications, it is reasonably good option for the management of HD.

Although majority of patients presented as neonates (64.70%) or young infants, there were significant number of late presenters 2 (11.76%) in this study group. Male predominance (76.47%) was evident in present study, like most other series [9]. 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. In this study, 88.23% of children had a history of delayed or non-passage of meconium. In 4331 index cases of HD, there were 330 cases in which a family history of HD was recorded, giving an overall rate of 7.6% for familial recurrence. In our series, 2 (11.76%) children had history of HD in their siblings [10]. Classical rectosigmoid disease was seen in 76.47% of patients, whereas 23.52% of patients had long-segment disease.

Post-operative complications observed in our study included wound infection (11.76%), perianal excoriation (17.64%), enterocolitis (11.76%), stricture formation (11.76%), constipation (11.76%), fecal soiling (29.41%), and adhesive bowel obstruction (5.89%). These complications have been variedly reported in several large series [5,11-13].

Hirschsprung-associated enterocolitis (HAEC) represents the primary cause of high morbidity and mortality in HD patients. There are several hypotheses for the cause of HAEC involving dysbiosis of the intestinal microbiome, compromised mucosal barrier function, changed innate immune responses, and translocation of bacteria [14].

Table 1: Mode of presentation

Presenting complaints	Delayed passage of meconium as primary mode	Perforation with peritonitis	Chronic constipation with abdominal distension (retrospective history of delayed meconium passage)	Chronic constipation with abdominal distension as primary mode
Non-delayed passage of meconium	9	2	4	–
Normal passage of meconium	–	–	–	2

Table 2: Complications

Complication	Number (%)	Management
Soiling	5 (29.41)	Four had Grade 1 soiling: Dietary management One had Grade 2 soiling: On BMP
Perianal excoriation	3 (17.64)	Barrier skin protective agents
Wound infection	2 (11.76)	Wound drainage, intravenous antibiotics
Postoperative enterocolitis	2 (11.76)	Bowel rest, intravenous antibiotics, fluids, rectal washes, and flatus tube insertion
Stricture formation	2 (11.76)	One case improved on serial dilatation and the second underwent laparotomy and stricturoplasty
Constipation	2 (11.76)	Managed with dietary modification, toilet training, judicious use of oral laxatives, and rectal washes
Hypertension	2 (11.76)	No organic cause identified, antihypertensive drugs withdrawn in 6 months
Adhesive bowel obstruction	1 (5.88)	Adhesiolysis was done

BMP: Bowel management program

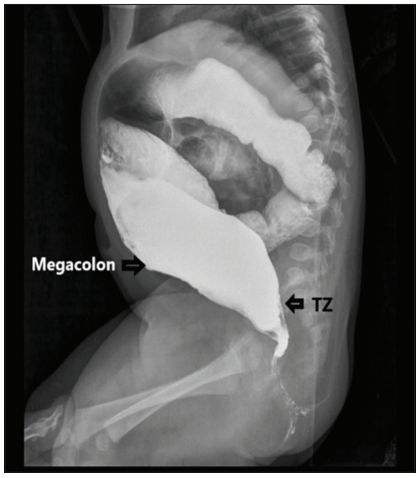

Figure 1: Arrow showing transition zone with proximal megacolon in a child with rectosigmoid Hirschsprung's disease

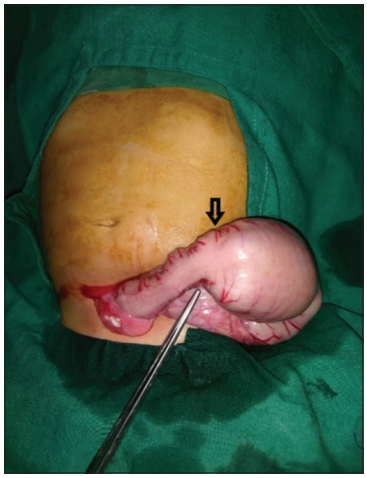

Figure 2: Arrow showing transition zone in sigmoid colon, with distal spastic segment and proximal megacolon during leveling colostomy

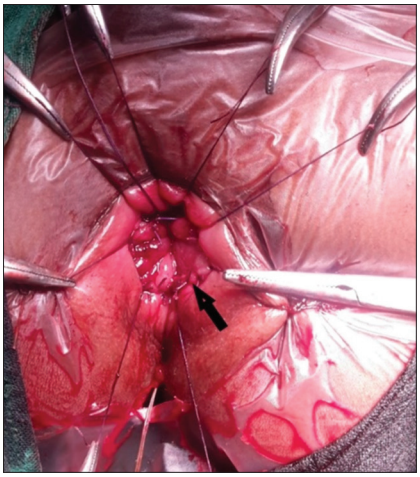

Figure 3: Arrow showing retrorectal pull through during Duhamel's procedure and anastomosis 0.5 cm above dentate line

The incidence of enterocolitis in the post-operative period has been found to range from 5% to 26% in different studies [5,12,13]. HAEC is more common in patients with Trisomy 21 or long-segment disease and is less frequent among late presenters [11]. We encountered enterocolitis in 11.76% of our patients.

The success of a procedure to correct HD is judged by the continence achieved by the patient. Continence is the ability to control the passage of feces; to hold on if necessary; and to pass completely when the individual wants at a socially convenient time [15]. Assessment of functional outcome was done in patients using Krickenbeck classification in the present study [16]. Voluntary bowel movements were established in 15 (88.23%) children. By voluntary bowel movement, we mean feeling of urge, capacity to verbalize, and hold the bowel movement.

Constipation was observed in 2 (11.76%) patients. Constipation has been reported to occur in 5–8% of patients following DP in varies series [5,11-13]. Children with constipation and soiling underwent digital rectal examination and contrast enema X-ray. There was no evidence of anastomotic stricture, megacolon, or residual spur in both the cases. One child had Grade 2 constipation and was managed with dietary change, toilet training, judicious use oral laxatives, and rectal irrigation as necessary. The second child had Grade 3 constipation and needed bowel management program (BMP) with daily rectal irrigations.

In a review of 2430 long-term follow-up of post-operative HD patients, 5.3% of patients showed fecal soiling [17]. According to available evidence, there are no significant differences in the incidence of incontinence between different operative techniques in the long term [8]. Most of these patients have been found to improve with time and do well with dietary modifications and bulking agents before any surgical intervention [8,18,19]. In the present study, 5 (29.41%) children had a history of soiling on early assessment. On further follow-up beyond 1 year post-DP, soiling resolved in four children with Grade 1 soiling. Soiling persisted in 1 (5.89%) child, who had long-segment disease and Grade 3 soiling, hence needed long-term BMP.

Average stool frequency was found to improve from 4/day at 8 weeks after surgery to 2.4/day at 6 months after surgery and 1.4/day at 1 year after surgery. Lower urinary tract symptoms were seen in two patients in the present study. Symptoms were transient and improved within 2 weeks post-DP. No mortality occurred during this study. Most of the series on post-operative complications in DP have reported a low mortality rate. HAEC represents the primary cause of high morbidity and mortality in HD patients [14,20]. Parents were educated in the present series, regarding need for early reporting for bowel decompression, antibiotics, and fluid resuscitation, to avoid morbidity and mortality in case features of enterocolitis appear.

CONCLUSION

DP is safe, has better learning curve for young surgeons, and can address the entire spectrum of HD. Staging of DP with initial leveling colostomy helps to overcome short comings of not having frozen section facility. Stapled anastomosis helps to overcome variability in hand sewn anastomosis to some extent. Complications are comparable to other series of DP and other techniques. With adequate follow-up, good quality of life can be achieved for the children with HD following DP.

ACKNOWLEDGMENT

We sincerely thank Dr. Usha Kini and team, for their support in evaluation of rectal and leveling biopsy samples.

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Funding: None; Conflicts of Interest: None Stated.

How to cite this article: Aravind KL, Nisha N, Sushmitha R, Madiwal C. Duhamel's procedure for Hirschsprung's disease and the functional outcome in a tertiary care center. Indian J Child Health. 2021; 8(1):51-55.