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Single-stage Modified Duhamel procedure for Hirschsprung's disease: Our experience

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ABSTRACT

Introduction: Primary single-stage pull-through for Hirschsprung's disease (HD) has been reported to give comparable surgical outcomes to staged operations with less morbidity. Herein, we present our experience with single-stage Modified Duhamel procedure for management of HD. **Patients and Methods:** This was a review of 48 cases of HD who underwent single-stage Modified Duhamel procedure without a protective colostomy. **Results:** The age at surgery ranged from 6 months to 10 years (median - 9 months, mean - 2.3 years). The average weight of the child was 7.2 kg (range, 4.9-22 kg). 38 (79.2%) patients had classical rectosigmoid HD, the rest being long segment HD (the proximal most level being the splenic flexure). The average duration of surgery was 175 minutes (range, 130-245 minutes). The average blood loss was 45 ml. The average hospital stay was 7.2 days (range: 6-10 days). The major postoperative complications (*n*=3) included postoperative adhesive intestinal obstruction, anastomotic leak and persistent constipation due to residual aganglionosis. Each required a re-exploration. Minor complications included surgical site infection (*n*=3) and post-operative enterocolitis (*n*=3), which were managed conservatively. Six patients had constipation for a limited period post-operatively. All patients have a satisfactory functional outcome and normal development and growth. **Conclusions:** For HD, we recommend that single-stage Modified Duhamel procedure should be the preferred approach in view of its low morbidity, satisfactory functional outcome and avoidance of stoma, multiple surgeries and economic benefit in view of decreased hospital stay.

Key words: Hirschsprung's disease, Modified Duhamel, single stage

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INTRODUCTION

Hirschsprung's disease (HD) is a congenital disorder characterised by the absence of ganglion cells in the distal bowel resulting into functional obstruction, most commonly affecting newborns.[1] The standard surgical treatment of HD was done in three stages; the creation of a proximal diverting stoma, resection of the aganglionic bowel segment and restoration of bowel continuity.[1-3]

The most popular procedures for definitive treatment of HD include Swenson's procedure, Soave's endorectal pull-through and Modified Duhamel procedure.[1,4] Nowadays, multistage surgery has progressed to open or laparoscopically assisted one-stage repair.[5,6] The rationale of single-stage surgery has been that it provides a cure without multiple operations, decreases surgical scars and hospital stay and avoids a stoma.[1,3,4] Previous reports have mentioned that the results of multi-staged and a single-stage repair is comparable.[2,4,7] The advantages of Modified Duhamel procedure includes ease of performance, reduction of anastomotic leaks and strictures, retention of anal sensory receptors and preservation of nervi erigentes.[8] Herein, we present our experience with single-stage Modified Duhamel procedure for management of HD.

MATERIALS AND METHODS

From January 2008 to October 2010, we diagnosed and treated 52 cases of HD. We retrospectively reviewed the records of these patients. 48 cases of HD, who underwent single-stage Modified Duhamel procedure without a protective colostomy, were included in this study. The diagnosis of all the cases was based on history, clinical examination, radiological investigation (barium enema) and histopathological diagnosis.

The patients who presented in the neonatal age group with delayed passage of meconium, abdominal distension and abdominal radiographs showing dilated bowel loops with paucity of gas in the pelvis were

provisionally diagnosed as HD. They were started on rectal washouts and abdominal distension monitored. Most patients deflated with washouts and were discharged home on twice daily washouts (the parents were taught the same). The remaining four patients who did not respond to washouts were explored and a levelling colostomy was performed with the help of frozen section examination. Out of these four, three presented in the neonatal age group and one at 1 year of age, with severe distension of abdomen and vomiting and did not deflate on rectal washouts. All of them required emergency exploration, in which a levelling colostomy was done because of grossly distended bowel proximal to the transitional zone, in which a primary pull-through procedure could not be considered. These patients were excluded from the study.

All the patients on washouts and other patients who presented at an older age with constipation were investigated with barium enema to know the level of aganglionosis and underwent full thickness rectal biopsy to confirm the diagnosis. The diagnosed patients were kept on washouts and an elective surgery was done at a minimum of 6 months of age and a minimum of 5 kg in weight. No patient had an associated congenital anomaly.

All patients were admitted 2 days prior to surgery and were given a liquid diet on admission. A day prior to surgery, patient was kept nil per orally, started on intravenous fluids, continued on rectal washouts and a complete bowel preparation was done with oral polyethylene glycol solution. The patient was started on intravenous antibiotics on the eve of the surgery and it was continued till fifth day post-operatively. Single-stage Modified Duhamel procedure was done in all the cases by the same surgical team. The extent of the aganglionosis was diagnosed by frozen section examination of multiple seromuscular biopsies taken during surgery. The healthy bowel, 5 cm beyond the distal most level of ganglionosis, was pulled down. The common wall between the native rectum and the pulled-through bowel was resected and stapled using a Proximate linear cutter stapler. As it was difficult to accommodate the GIA staplers through the anus into the pelvis of infants less than 6 months of age and under 5 kg of weight, all patients were operated after they were at least 6 months of age and 5 kg in weight. To temporarily decompress the proximal bowel, a trans-anastomotic rectal tube was kept in every patient, which was removed on postoperative day 4. The patients were kept nil per orally for 5 days. Per-oral feeds were started on day 6 and patients free of complications discharged by day 7. All the patients were kept on regular follow-up with follow-up period ranging from 6 months to 2.5 years. The age, gender, weight of the patient, duration of procedure, blood loss, level of aganglionosis, postoperative complications, duration of hospital stay and functional outcome were studied. Satisfactory functional outcome was defined as normal bowel movements or soiling less than once a week.

RESULTS

A total of 48 patients underwent single-stage Modified Duhamel procedure in our institute, from January 2008 to October 2010. Thirty-three were males (male/female ratio, 2.2:1). The most common age of presentation was neonatal age (62.5%) (Range: Day 2-10 years) [Table 1]. The complaints at presentation included abdominal distension, constipation, vomiting and delayed passage of meconium. The commonest presenting feature in the neonatal age group was abdominal distension, while the patients who first presented beyond the neonatal age most commonly presented with constipation. The age at surgery ranged from 6 months to 10 years (median - 9 months, mean - 2.3 years) [Table 2]. The average weight of the child was 7.2 kg (range, 4.9 – 22 kg). 38 (79.2%) patients had classical rectosigmoid HD, the rest being long segment HD (the proximal most level being the splenic flexure). In the four patients who were excluded from the study, one patient had total colonic aganglionosis, while three patients had long segment HD (two patients had aganglionosis till the splenic flexure and one till the left half of the transverse colon). The average duration of surgery was 175 minutes (range, 130-245 minutes). The average intra-operative blood loss was 45 ml. The average post-operative hospital stay was 7.2 days (range: 6-10 days). Post-operative complications [Table 3] were seen in nine patients (18.75%). Three patients (6.25%) had major postoperative complications, namely, one had post-operative adhesive intestinal obstruction requiring re-exploration and adhesiolysis [Table 3]. The second patient had an anastomotic leak for which a proximal defunctioning right transverse colostomy

Table 1: Detail of 48 patients with Hirschsprung's disease

Sex	%
Male	*N*=33 (69)
Female	*N*=15 (31)
Level of aganglionosis	
Rectosigmoid	*N*=38 (79.2)
Long segment	*N*=10 (20.8)
Age at presentation	
Neonate (<1 month)	*N*=30 (62.5)
1 month-1 year	*N*=12 (25)
>1 year	*N*=6 (12.5)

Table 2: Peri-operative features of 48 children with Hirschsprung's disease

Age at surgery	Weight	Blood loss	Duration of surgery	Hospital stay
6 months-10 years	5-22 kg	35-60 ml	130-245 min	6-10 days
Mean - 2.3 years Median - 9 months	Mean - 7.2 kg	Mean - 45 ml	Mean - 175 min	Mean - 7.2 days

Table 3: Post operative complications following single-stage Duhamel Operation

	Major *N*=3 (6.25%)			Minor *N*=6 (12.5%)	
Complications	Anastomotic leak	Post-op adhesive intestinal obstruction	Residual aganglionosis	Wound infection	Enterocolitis
Number of patients	1 (2.08%)	1 (2.08%)	1 (2.08%)	3 (6.25%)	3 (6.25%)

was done. One patient had persistent constipation after surgery not relieved on conservative measures. The histopathology report in this case was reviewed which revealed aganglionic segment at the proximal excised margin (which was previously reported as ganglionic on frozen section). A re-do pull-through procedure in this patient proved curative. Minor complications (seen in 12.5%) include surgical site infection which was seen in three cases (6.25%) and postoperative enterocolitis which was seen in three (6.25%) patients, which were managed conservatively. There were no intra-operative complications and no deaths in this series. Mean length of follow-up was 17 months. On follow-up, 42 out of 48 (87.5%) patients had a satisfactory functional outcome with at least one bowel movement in the day without laxatives or enemas. There were no complaints of incontinence in any of the patients. Six patients (12.5%) had temporary postoperative constipation which resolved on dietary modification and laxatives within a few months. All six of them have been weaned off medication and have normal bowel movements now. All the patients who had a follow-up of at least 1 year had normal development and growth with the weight and height falling between the 25th and 95th percentiles for that age.

DISCUSSION

The management of HD has come a long way since Swenson first described the pathological basis and came up with a corrective surgery for the same. Previously, the patients used to present with malnutrition, sepsis and colonic distension and colostomy in those cases proved to be life saving.[4] For HD diagnosed in the neonatal period, the conventional approach to the surgical treatment consists of a diverting colostomy in the neonatal period and a definitive pull-through procedure for those at about 1 year of age. However, with understanding of the disease pathology, the management strategies, morbidity and mortality have changed over the years.[1,4]

Though standard surgical education advocated that proximal colostomy should be done to decompress the bowel and allow for weight gain and then carry out the definitive procedure when the child is older. The other approaches include a delayed primary definitive operation without colostomy when the neonate has adequate daily evacuations with the use of suppositories, rectal dilatations and enemas and remains nutritionally well.[2] We, in our study, have tried to bypass the initial colostomy with effective rectal washouts.

With advances in neonatal, anaesthetic and surgical care, a safe one-stage neonatal pull-through has become possible. Since then, a few series reported their one-stage neonatal pull-through (Soave's endorectal pull through) with good results as compared with the conventional approaches.[1,6,9] Previous studies have also reported successful single-stage Modified Duhamel procedure in neonates and infants with similar rates of post-operative complications and outcomes as compared to multi-staged repairs.[2,4,7] We are using the Proximate linear cutter GIA staplers in older infants who can easily accommodate the staplers through their anus in their pelvis. On review of literature, we found few reports of the feasibility of the use of Endo-GIA staplers in neonates and young infants.[2,3,7]

Multi-stage procedures though have traditionally been successful, they are associated with their share of problems, viz., stoma complications, more surgical scars, prolonged hospital stay due to repeated hospital admissions, increased morbidity of multiple procedures and maybe associated with psychological trauma to both patient and his relatives.[1-3,7]

Single-stage transanal endorectal pull-through has gained popularity over the years because it avoids a laparotomy and hence a scar on the abdomen and post-operative intra-abdominal adhesions. It also decreases requirement of analgesia, the time to feeding and hospital

stay. But, it also has its share of complications such as increased rate of anastomotic stricture and perianal excoriations. In addition, long-term outcome following transanal pull-through is still uncertain, especially for continence state that may be affected by the deleterious effect of the anal retraction on sphincter function,[1,6,10] and Srivastava *et al*. found the Modified Duhamel procedure to be better than transanal endorectal pull-through for its better functional outcome.[6]

From our experience, we have adopted single-stage Modified Duhamel procedure as the modality of choice for all patients of HD as almost all patients deflate well with good rectal washouts. There are very few exceptions to this approach which eventually require colostomy as the primary procedure, viz., long segment HD not deflating on washouts, complications of HD such as toxic megacolon, colonic perforation and severe enterocolitis, and in centres where facilities for frozen section are not available.

The post-operative complications (both major and minor) seen in our series are 18.75%. The complication rates in the other studies ranged from 24 to 33%.[4,6,7] One of our patients required a redo-pull-through procedure due to error in the report of the intra-operative frozen section specimen. Error in reporting of intra-operative frozen section is a rare but known complication and may be due to technical issues and pathologist inexperience.[11] The incidence of enterocolitis in our series was 6.25%, which is less than the incidence in other studies (17-29%).[2,4,6,7] The results in our study revealed that the complication rates, minor or major, in our study and the functional outcome are comparable or better than the studies in the past (both multi-staged and single-stage procedures).

In conclusion, cases of HD who deflate well on washouts or enemas are ideal candidates for single-stage Modified Duhamel procedure. This procedure is definitely a feasible option, and we recommend that it should be the preferred approach in view of its low morbidity, satisfactory long-term functional outcome, avoidance of stoma, multiple surgeries and economic benefit in view of decreased hospital stay.

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