Aims and Objectives: We studied the short- and long-term outcomes and quality of life (QOL) in patients undergoing a two-staged modified Duhamel’s procedure for Hirschsprung’s disease.

Materials and Methods: Patients who had undergone this modified procedure, with initial Hartmann’s procedure based on contrast enema, followed by bowel preparation and low colo-anal anastomosis below the dentate line were included. The patient who underwent this procedure over 10 years with a minimum 2-year follow-up were analysed based on an interview-based questionnaire.

Results: Of the 152 patients, 69 responded. Mean age at the time of interview was 7.72 ± 3.04 years with mean follow-up of 4.9 years (2–11 years). Perineal excoriation and soiling was present in 60.9% and 36.2% of patients initially which reduced to 0 and 4.3% by the end of 2 years. In the first 6 months, postoperative period, 15.9% of patients had constipation and 78.2% had altered stool consistency. Good fecal continence score was present in 97.1% of patients in the long term. About 95.4% had good QOL scores. There was no mortality in this series.

Conclusions: Although short-term outcomes showed altered bowel function, soiling, and perineal excoriation, this improved significantly in the long term, with good QOL scores in the majority.

Keywords: Colostomy, contrast enema, hirschsprung’s disease, modified Duhamel procedure, outcome analysis, quality of life
contrast enema (CE) to diagnose HD and identify the transition zone (TZ) provided there is no evidence of perforation, enterocolitis (EC) or massive distension unresponsive to rectal washes [Figure 1]. A Hartmann’s procedure is then performed with a colostomy at least 5 cm above the TZ as defined intraoperatively and/or on the preoperative CE. A frozen section study is performed only in the absence of either. The sigmoid colon is excised irrespective of the level of TZ and the proximal and distal ends of the specimen are marked for histopathological examination (HPE). The excised specimen is studied for ganglion cells using hematoxylin and eosin stains. After adequate weight gain and improvement in the nutritional status, the patients are taken up for modified Duhamel procedure, usually after three to 6 months. Preoperative total gut irrigation with normal saline at 25 ml/kg/h is given through a nasogastric tube till returns are clear. The patient is placed in the lithotomy position. The end colostomy is mobilized and retrorectal tunnel made close to the wall of the rectum. The colo-anal anastomosis is performed between the dentate line and the anocutaneous junction from 3 to 9 o’clock position. Any excess rectal stump is excised so as to have about 2–3 cm of rectum above the peritoneal reflection. A transverse enterotomy is made on the pulled down bowel at the level of the open rectal stump to ensure complete stapler application (Ethicon’s PROXIMATE® GIA linear cutter), followed by anastomosis of the anterior rectal wall to the colon. The rent in the mesentery is closed. Patients are kept nil per oral for 5 days and then discharged over the

![Management algorithm of patients with Hirschsprung](image-url)

**Figure 1:** Management algorithm of patients with Hirschsprung
next 2–3 days when oral intake is adequate. They are followed up routinely a week later and every month for 3 months. They are seen once in 6 months and then yearly at least till the age of five.

Short-term outcomes were defined as the outcomes noted in the first 6 months post operatively: wound infection, pelvic collection, and anastomotic leak in the immediate postoperative period and later perineal excoriation, EC, stool frequency, bowel function, and anastomotic stricture.

Long-term outcomes, i.e., (>6 months after surgery), studied were measures of fecal continence (constipation and incontinence), intestinal obstruction, outlet obstruction, late onset EC, and QOL.

Perineal excoriation was defined as significant when patient required medications in the form of sitz bath and emollients. Wound infection was categorised according to standard definitions. EC was assessed using HD-associated EC score. Constipation was defined as <1 bowel movement in 2 days. Faecal incontinence was assessed using the Clinical Bowel Function Scoring System [Table 1]. QOL was measured using scoring criteria for children with fecal incontinence [Table 2]. Scoring system was graded as good (9–13), fair (5–8), and poor (0–4) depending on the number denoted to bowel control, diet and lifestyle modification, school absence, and peer rejection.

Institutional Ethics Committee approval was taken vide reference no. NK/42/MCh/1391 dated 4.4.2012 Statistical analysis was carried out using Chi-square and Fischer’s exact test.

**RESULTS**

One hundred and fifty-two, patients underwent modified Duhamel procedure over 10 years. However, only 69 parents responded to telephone calls and letters. This included 60 boys and 9 girls with a male/female ratio of 6.6:1. Patients and parents were interviewed for the study. The mean age of the children at the time of interview was 7.72 years ± 3.04 years (range 3–15 years). The mean follow-up period was 4.9 ± 2.4 years (range 2–11 years). The age at initial presentation was neonatal (29%), infancy (23.3%), 1–5 years (34.8%), and >6-year-old (12.9%). Fifty-seven, (82.6%) patients had history of delayed passage of meconium and > 98.6% presented with constipation (98.6%).

CE was available for review only in 58 patients. In 91.3% of CE the TZ could be easily made out. Of the positive CE the TZ was at the recto sigmoid junction in (79.2%) followed by descending colon-sigmoid junction (13.2%) and the rest had TZ lower than rectosigmoid (7.5%). Intraoperatively, during the first surgery, TZ was seen at recto-sigmoid in 60.9%, and descending-sigmoid junction in 13% of patients. In 24.6% of patients, there was dilatation of large bowel up to the peritoneal reflection.

On histopathology, (available for 61 patients) absence of ganglion cells was reported in the distal resected segment in 54 (88.5%) patients. In 7 (11.4%) patients, ganglion cells were reported to be present in the distal segment in spite of strong clinical and radiographic features suggestive of HD. Of these, 3 had TZ at rectosigmoid, 1 at descending colon, whereas in 3 patients no TZ was

**Table 1: Clinical bowel function score**

| Item               | Criteria | Score |
|--------------------|----------|-------|
| Frequency of defecation | Every 2d or 1-2 /d | 2     |
|                    | 3-5/d or 2-3/d | 1     |
|                    | <1/wk or >5/d | 0     |
| Soiling            | Absent    | 4     |
|                    | Accidental | 3     |
|                    | Frequent   | 2     |
| Incontinence       | Accidental | 1     |
|                    | Frequent   | 0     |
| Fecal sensation    | Normal    | 2     |
|                    | Defective  | 1     |
|                    | Absent     | 0     |
| Pain or difficulty with defecation | Never | 2     |
| Defecation         | Accidental | 1     |
|                    | Frequent   | 0     |
| Fecal consistency  | Normal    | 2     |
|                    | Loose      | 1     |
|                    | Liquid     | 0     |

Total score: Good 9-13, Fair 5-8, Poor 0-4

**Table 2: Quality of life scores (QOL)**

| Item               | Criteria | Points |
|--------------------|----------|--------|
| Soiling            | Absent   | 4      |
|                    | Accidental | 3     |
|                    | Frequent  | 2     |
| Incontinence       | Accidental | 1     |
|                    | Frequent  | 0     |
| School absence     | Never    | 2      |
| Unhappy/anxious    | Never    | 2      |
|                    | Accidental | 1     |
|                    | Frequent  | 0     |
| Food restriction   | No       | 2      |
|                    | Somewhat  | 1     |
|                    | Much     | 0     |
| Peer rejection     | Never    | 2      |
|                    | Accidental | 1     |
|                    | Frequent  | 0     |

Total score: Good 13-9, Fair 8-5, Poor 4-0
seen on CE. In the last group, full-thickness rectal biopsy, performed before the definitive procedure showed absent ganglion cells in all seven patients.

**Short term outcomes**

Wound infection occurred in 2 (2.9%) patients in the immediate postoperative period. Anastomotic leak occurred in 2 (2.9%) patients. Single stitch at Martin’s anastomosis had given way, which was managed by laparotomy and primary repair. Perineal excoriation was seen in 42 (60.9%) patients in the first 6 weeks. At 12 weeks, this had completely resolved in all patients. EC was seen in 10 (14.5%) patients aged 1–3 years, 3–6 months after surgery. Twenty-nine (42%) patients did not have any soiling after the surgery. During the first 6 months, soiling ceased at variable time periods in 25 (36.2%) patients [Table 3].

**Long-term outcomes**

EC occurred in 4 (5.8%) after surgery, which resolved with medical management. None of them had a history of preoperative EC.

Of the 15 patients with persistent soiling at 6 months, it stopped in 8 (11.6%) over a period of 2 years, in 4 (5.7%) by 5 years, while 3 (4.3%) continue to have soiling.

One patient, a case of neurocristopathy complex developed an anastomotic stricture, and underwent a second Duhamel pull through procedure. At 4 years follow-up, he has had no further complaints.

**Bowel function**

After 6 months, 53 (76.8%) patients had up to 3 bowel movements per day, which was significant improvement as compared to short term results 38 (55.1%) \( (P = 0.0213) \). Ten (14.5%) patients had >3 bowel movements per day and were successfully managed with dietary modifications. Six patients (8.6%) had persistent constipation, for which rectal biopsies were done in 3 and myectomy in one. All four specimens had ganglion cells on HPE and improved postprocedure. The other two patients (patients with global developmental delay) (2.8%) continue to have constipation and are being managed with rectal washes and enemas [Table 4].

**Fecal continence scores**

Good fecal continence score was present in 67 (97.1%) patients as compared to fair and poor scores in one (1.4%) patient each. The mean fecal continence score was 12.04 ± 1.31.

**Quality of life scores**

The score is validated for children between the ages of 8–16 years. However, children as young as 5 years could easily respond to the questionnaire. Fifty-five (79.7%) children could be assessed for QOL. Fifty-two (95.4%) patients had good, 1 (1.4%) fair, and 2 (2.9%) patients had poor QOL scores. The mean QOL score was 11.85 ± 1.15 (range 7–13). Nearly all children with good scores (9–13) had no soiling or incontinence. Neither was there school absence nor peer rejection. One patient in the fair group had significant peer rejection and displayed significant school absence. The two patients with a poor score had delayed developmental milestones. They had soiling, were on food restriction, and faced peer rejection. They were both studying in normal schools but were behind their contemporaries.

There was no mortality in our series.

**DISCUSSION**

Several procedures for HD are well-defined anatomically; however, outcomes are variable. Studies on QOL should choose the best possible definitive procedure; however, these are limited in number. In addition, studies with a comprehensive assessment of QOL club together different surgical procedures.

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**Table 3: Outcomes (short and long-term)**

| Surgical outcomes       | Short-term (%) | Long-term (%) |
|-------------------------|----------------|---------------|
| Wound infection         | 2 (2.9)        | Nil           |
| Anastomotic leak        | 2 (2.9)        | Nil           |
| Perineal excoriation    | 42 (60.9)      | 3 (4.3)**     |
| Soiling                 | 40 (57.9)      | 1 (1.4)       |
| Intestinal obstruction  | 10 (14.5)      | 4 (5.7)       |
| Enterocolitis           | 10 (14.5)      | 1 (1.4)*      |

*Patient with deaf mutism and neurocristopathy complex, **2 patients with global developmental delay

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**Table 4: Bowel functions**

| Bowel functions          | Short-term (%) | Long-term (%) | \( P \)  |
|--------------------------|----------------|---------------|---------|
| Up to 3 bowel movements/day | 38 (55.1)   | 53 (76.8)      | 0.0213  |
| More than 3 bowel movements/day | 20 (29)     | 10 (14.5)      |         |
| Constipation (<1 bowel movement in 2 days) | 11 (15.9)   | 6 (8.6)*       | 0.016   |

*Three patients underwent rectal biopsy and 1 underwent myectomy and improved over the next 3 months, 2 remain constipated (global developmental delay)
Many centers advocate a single-stage procedure to avoid the complications and social implications of a colostomy. Diagnosis and level of aganglionosis are usually based on frozen section biopsy. At our center, we have been performing a Hartmann’s procedure based on CE and intra-operative findings. We believe that a properly functioning stoma is an excellent marker of ganglionosis. When the “functioning” stoma is brought down, we ensure adequately ganglionated bowel is anastomosed hence almost eliminating major complications. This significantly improves the overall QOL, as shown in our study. In addition, more than two-thirds of our patients presented beyond neonatal age, with massive dilatation of proximal bowel. The colostomy helped to normalize this bowel, before definitive surgery and also gave time to improve nutritional status.

After the definitive procedure, we had an infection rate of 4.3% as compared to other series reporting infectious complications up to 14%.7 Bowel preparation leads to minimal fecal contamination and a lower wound infection rate in spite of the presence of a colostomy. Our anastomotic leak rate was 2.9%, lower in comparison to most series in the literature (upto 5.9%).8 We attribute this to pulling down the ganglionic bowel, meticulous surgery, well prepared bowel, and improved nutritional status.

Compared to other operative procedures for HD, patients undergoing modified Duhamel procedure have lower rates of EC. A low incidence of outlet obstruction, supported with low constipation rates as in our series, may play an important role. In our study, early EC was seen in 14.5% of patients, which progressively decreased with time and remained in only 4.5% in the late follow-up period. Other studies with standard Duhamel have shown an incidence of EC ranging from 7% to 21%.4,9 We feel that removal of the entire sigmoid colon, which is usually very dilated in these patients, has given us better results. There have been other reports of near total colectomy in patients with adult HD and refractory constipation with good outcomes.10,11 In addition, the coloanal anastomosis is performed between the anal verge and dentate line. The low anastomosis completely divides the fibers of the internal sphincter and aids in removing the inherent spasm in these patients. These lower most fibers heal within 6–8 weeks, reverting the problems of excoriation and soiling as shown in our results. Small enterotomy in both native rectum and “pull down” bowel: “Martin’s anastomosis” ensures that the stapler is completely applied and there is no residual spur, which further helps in managing constipation.

Bourdelat et al. reviewed 2,430 patients who underwent a Duhamel operation.12 At 15–30 years follow-up, the majority of patients achieved good or excellent anal function. Only 8.07% of patients had constipation, and 5.3% had soiling. However, Baillie et al. reported residual postoperative problems in 10%–80% including soiling, constipation, EC, and anastomotic strictures.3 Others have reported 32% to 35% incidence of soiling and perineal excoriations.13–16 Long-term perineal excoriation is not only an indicator of persistent soiling and incontinence but also may be an indicator of anastomotic stenosis.17 In our series, soiling and perineal excoriation was seen in 30.6% and 60.9% patients, respectively, in the first 6 months which reduced to 4.3% and 0% at the end of the study.

Frequent bowel movements and loose consistency of stools in the early postoperative period is common which progressively improves with time. We report a similar experience. Incontinence ranging from 4% to 80% has been reported following Duhamel.7,13,15,16 Most studies show mild-to-moderate incontinence which significantly improves from 27% to 4% over a

### Table 5: Studies on outcomes of Duhamel’s procedure

| Report                      | Number of patients | Outcome (%) | Follow up period | Percentage responders |
|-----------------------------|--------------------|-------------|------------------|-----------------------|
| Duhamel, 196419| 270                | 3.7 constipation, 100 continence | Not mentioned | 100                     |
| Foster, 199021| 43                 | 100 asymptomatic | 7 years | 100                     |
| Hung, 199122| 198                | 100 good outcome | <9 years | 100                     |
| Rescola et al., 199223| 103               | 94 normal at 15 years | Mean 6 years | 56                     |
| Marty et al., 199524| 91                 | 12.1 fecal soiling | Not mentioned | 78                     |
| Heji et al., 199525| 63                 | 20.4 good 20.4 at >4 years | Mean >6 years | 84                     |
| Kittur and Jhadav, 200126| 40             | Good | 8 years | 100                     |
| Sharma et al., 200427| 48                 | Satisfactory, Failure rate 14 | 1-2 years | 92                     |
| Current study | 152                | Good fecal continence scores 97.1, Good QOL 94.5 | Mean 4.9 years (2-11 years) | 45.3                     |

QOL: Quality of life
follow-up period of 8–12 years, especially as the patient approaches adolescence.[5,6,18,19] Livaditis demonstrated an improvement of incontinence from 12% to 2% over a period of 13 years.[20] Yanchar and Soucy noted that problems with fecal continence were common among those <15 years old. This improved significantly, with only 8% having fair or poor continence compared with 57.5% and 48% between 5 and 15 years of age and those <5 yearold.[4] We have, however, seen excellent fecal continence scores in a much shorter time frame.

Various series of Duhamel procedures have shown good clinical outcome scores between 20.4% and 100% [Table 5]. In this study, we used the fecal continence scoring system similar to that used by Bai et al. [Tables 1 and 2]. In their series, following a Swenson’s procedure, as many as 51.1% of patients had long-term bowel dysfunction.[1] Good fecal continence score was seen in only 46.7%. However, in our series, using the same criteria, a good fecal continence score was present in 67 (97.1%) patients. Fair and poor scores were seen in one (1.4%) patient each.

Faecal soiling is physically, emotionally, and psychologically disabling. It has a significant impact on patients’ social and emotional development and QOL of the child and their parents.[4,13] Fecal incontinence also leads to dietary modifications and sometimes peer rejection and abstinance from school and play. QOL in HD has been measured by some authors with scores common with anorectal malformations which may give an objective analysis of continence issues alone. Others have used PedQL and their subscores in various age groups.[12,26,27] In general, although most patients had good (40%) or fair (46.7%) QOL after surgical correction for HD, the long-term outcome and QOL were not as good as surgeons had expected (13.3% with poor QOL).[10] In this study, of the 60 children between the ages of 5–15 years, 55 (79.7%) could be assessed for QOL score. Of this subset, 52 (95.4%) patients had good, 1 (1.4%) patient had fair and 2 (3.8%) patients had poor QOL scores. Two of the latter had global developmental delay. Abstaining from school and peer rejection was noted in 4 (5.7%) patients. This was considerably less as compared to other series where poor peer interaction and school abstinence were noted in 15.6% and 13.3% respectively.[11] The rest had appropriately adjusted to society. Bai et al. noted that the greatest negative impact on the QOL was not related to constipation, but to faecal soiling.[1] In their series, 87% of patients who had undergone Swenson’s procedure were graded as having a good-to-faith QOL. This lower QOL may be due to a very high rate of fecal soiling (37%). Low QOL scores have been seen in 32%–80% of patients in other studies with high faecal incontinence rates.[11] In our series following modified two staged Duhamel there is early resolution of soiling and excoriation almost completely by 6 months and significantly improved clinical bowel functional score. We attribute higher QOL scores as compared to literature to excellent faecal continence in all except 3 (4.3%) patients.

Our study is one of the few which has assessed QOL after a single standardized procedure. We have been able to show that a functioning stoma ensures good bowel function after the definitive procedure. Based on the responses in our study of almost 45% of patients, we believe that the two-staged Duhamel procedure with our modifications appears to be an feasible option for the surgical management of HD giving excellent long-term outcome and QOL.

**Conclusions**

Protocized approach to patients with HD is required for the optimization of results. Our modifications of an initial colostomy, excision of the sigmoid colon, bowel preparation, and a low colo-anal anastomosis between the dentate line and mucocutaneous junction are essential for good results. After a brief period of soiling, perineal excoriation and altered bowel function, there is resolution in most patients by 12 weeks. The late postoperative outcome, bowel function, and QOL scores were found to be excellent in >95% of patients who responded.

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**Conflicts of interest**

There are no conflicts of interest.

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