



Functional outcomes in Hirschsprung disease: A single institution's 12-year experience



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ABSTRACT

Aims: Hirschsprung disease (HD) is a chronic condition associated with long-term morbidity. We assessed the short and long-term functional outcomes of operated patients in a single institution over a 12-year period.

Materials and methods: We conducted a retrospective review of all children operated for HD between 2002 and 2014. Postoperative functional outcomes were assessed using the Rintala Bowel Function Score (BFS, 0–20, 20 = best score). We assessed hospital admissions, complications including Hirschsprung associated enterocolitis (HAEC) and the need for further surgical procedures.

Results: 72 (52 male) patients were studied, of whom, 6 (8%) had a positive family history, 5 (7%) had Trisomy 21 and 5 (7%) had total colonic HD. The median age at diagnosis was 6.5 days (2 days–6.7 years) and median follow-up was 6 years (1–12 years). All patients except two underwent a Duhamel pull-through procedure. The median age at surgery was 4 months (6 days–90 months), 37 (51%) procedures were performed single-stage and 7 (10%) were laparoscopically assisted. Our early complication rate was 15%; 11 (15%) patients were treated for HAEC and 43 (60%) did not require any further surgery. 12 (17%) underwent injection of botulinum toxin, 7 (10%) needed residual spur division and 4 (5%) required an unplanned, post pull-through enterostomy for obstructive defecation symptoms and HAEC. Two (3%) patients underwent an Antegrade Colonic Enema (ACE) stoma. The median BFS was 17 (5–20). There were two deaths both out of hospital.

Conclusions: Long-term functional outcomes following Duhamel Pull-Through surgery are satisfactory although 40% of patients needed some form of further surgical intervention. The management of anal sphincter achalasia has improved with the use of botulinum toxin and we advocate aggressive and early management of this condition for symptoms of obstructive defecation and HAEC.

Level of evidence: III.

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Hirschsprung disease (HD) is a chronic condition with long-term effects on the physical and psychosocial health of a patient [1]. The outcomes of surgery have improved over the last few decades and there is now an increased recognition in the literature focusing on the functional outcomes of pediatric patients as they mature into adults. Long-term symptoms include constipation, bloating, soiling and fecal incontinence. Whereas the majority of these symptoms can be managed either medically or through limited surgical intervention, some patients have had an enterostomy fashioned to improve their quality of life. Hirschsprung associated enterocolitis (HAEC) also has a well-recognized morbidity and mortality that can continue to affect patients postoperatively.

Several surgical pull-through procedures have been described with varying outcomes. Our preference has been to use the Duhamel

technique, aiming for a single-stage procedure provided satisfactory decompression could be attained preoperatively.

We sought to assess the short and long-term outcomes of all operated patients with HD in our institution over a 12-year period.

1. Materials and methods

A retrospective case note review was performed for all histopathologically confirmed cases of HD undergoing pull-through surgery at our institution between January 2002 and January 2014 inclusive. Any patients with a pull-through procedure performed external to our institution were excluded. Patient demographics, comorbidities, timing of diagnosis and surgery, need for a preoperative enterostomy and details of the surgery were all collected.

Short-term outcomes included postoperative complications such as bowel obstruction, sepsis or an anastomotic leak. HAEC was clinically suspected on the basis of toxemia, offensive/bloody stools, abdominal distension and raised inflammatory markers.

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Long-term functional outcomes were objectively assessed using the Rintala Bowel Function Score (BFS) [2]; a validated tool for use in patients with anorectal malformations and HD. Seven domains focus on bowel control, symptoms of soiling and constipation as well as the social impact of the disease. A maximum score of 20 can be attained (0–20). A single investigator contacted parents of all patients at least four years of age by telephone to attain the score. Ethical approval for this was attained from the Hospital (Approval Number #AN5019).

Patient records were also assessed for any surgical interventions including the need for an examination under anesthesia, disimpaction of stool from the rectum/washout, intrasphincteric botulinum toxin injection and rectal spur division. All data are presented as median (range). Data are expressed as median (range). The Mann–Whitney U test was used to compare outcomes between groups with nonparametric data. The Chi-Square test was used to compare outcomes between categorical data. $P < 0.05$ was considered statistically significant.

2. Results

86 patients with histopathologically confirmed HD were identified during the study period. 14 patients were excluded; 10 patients underwent pull-through surgery elsewhere, two sets of case notes were missing, one patient was awaiting surgery and one infant with Smith–Lemli Opitz Syndrome died preoperatively from respiratory failure. 72 data sets were thus included for analysis over the 12-year period. Median follow-up for the series was for 6 (1–12) years.

52 (72%) patients were male, 6 (8%) had a positive family history of HD, 5 (7%) had an associated history of Trisomy 21 and one patient had MEN 2a syndrome who later underwent a prophylactic thyroidectomy at 2 years of age. The median age at diagnosis was 6.5 days (range 2 days–6.7 years). Median patient follow-up was 6 (1–12) years with data included up to December 2015.

70 patients underwent a Duhamel pull-through with two undergoing a Soave (individual surgeon's preference). 7 (10%) cases were laparoscopically assisted including both cases of the Soave pull-through. The median age at surgery was 4 months (6 days–90 months). Overall, 37 (51%) of all cases were performed single-stage. 18 (60%) of cases were performed multistage between 2002 and 2007 with the figure dropping to 17 (41%) between 2008 and 2014. 4 (5%) patients had a covering ileostomy performed at the time of the planned pull-through. Table 1 summarizes the reasons for a pre-pull-through enterostomy ($n = 35$).

The level of the pull-through was at the sigmoid colon in 70% of cases. 7% had total-colonic disease with an ileal pull-through. The histology of the pull-through was ganglionic in 99% of cases.

2.1. Early complications (within 30 days of pull-through)

11 (15%) patients had an early complication. One patient developed adhesive bowel obstruction after multistage surgery requiring laparotomy. There were three cases of suspected obstruction; two necessitated an examination under anesthesia and one was managed on the ward with a rectal tube. One had early problems with sphincter achalasia following primary pull-through and was managed with injection of

botulinum toxin Dysport® (Ipsen Biopharm Limited) into the internal anal sphincter (IAS) and a rectal tube. One patient had misplacement of an epidural catheter into the presacral space, recognized and removed at the time of the pull-through. Two patients (one multistage, one single-stage) developed postoperative sepsis with a pelvic collection, of whom one patient required a defunctioning ileostomy. Four patients required postoperative parenteral nutrition for high-enterostomy output or loose stools until enteral feeding was established. There was one superficial wound dehiscence managed conservatively.

2.2. Longer-term outcomes

2.2.1. Post-pull-through Hirschsprung associated enterocolitis (HAEC)

There were 15 episodes of HAEC in 11 (15%) patients of whom 5 had multistage surgery. The stool specimens were positive for Rotavirus in three cases. The median time at presentation was 18 months post-pull-through (1–90 months). In addition to these cases, 23 (32%) patients also had at least one episode of simple gastroenteritis requiring hospital admission (including one *Norovirus*, four *Rotavirus* and one *Clostridium difficile*).

2.2.2. Further unplanned surgical interventions after pull-through

31 (43%) patients needed at least one unplanned surgical intervention; ranging from zero to six. Table 2 summarizes the most commonly performed surgical interventions with their age at presentation. 12 patients had intrasphincteric injection of botulinum toxin Dysport® (12 U per kg body weight up to maximum dose of 200 U) under endosonographic guidance for obstructive defecation, recurrent abdominal distension and HAEC symptoms. Of these 6 had injection into the internal anal sphincter (IAS) and 6 into the external anal sphincter (EAS) muscles depending on the preference of the surgeon. There was significant improvement of abdominal distension following injection of the toxin into the EAS versus the IAS (5/6 patients improved vs. 1/6 patients respectively, $P < 0.05$). 7 (60%) patients underwent only a single course of botulinum toxin treatment. No patients in the EAS group required formation of an enterostomy compared with two patients in the IAS group.

2.2.3. Functional outcomes: Rintala Bowel Function Score (BFS)

53 patients aged 4 years and above were eligible for the telephone survey of whom 32 (60%) were contactable. The median BFS was 17 (5–20), Fig. 1. The median BFS in the multistage group was 15 compared with 18 for single-stage surgery ($P = 0.08$), which was not statistically significant.

20 (28%) of patients at their most recent follow up were still using laxatives. Two (3%) patients have had an Antegrade Colonic Enema (ACE) stoma procedure for chronic constipation and soiling and became clean postoperatively. Four (5%) patients required an emergency or urgent ileostomy for recurrent obstructive bowel symptoms and/or enterocolitis of which only one patient has an enterostomy in situ to date. Of these four patients, one patient also required excision of a megarectum at the time of his ileostomy.

Table 2

Surgical interventions performed for symptomatic patients following initial surgery.

Intervention	Number of patients (%)	Median (range) age at time of intervention (months)
Disimpaction of stool/retrograde colonic washout	14 (19%)	36 (11–63)
Injection of botulinum toxin	12 (17%)	36 (23–62)
Rectal biopsy	11 (15%)	34 (10–62)
Rectal spur division	7 (10%)	13 (5–145)
Anorectal manometry	6 (8%)	48 (23–144)
Enterostomy formation ^a	4 (5%)	98 (3–126)
Isolated examination under anesthesia	3 (4%)	17 (3–96)
Antegrade Colonic Enema (ACE) stoma	2 (3%)	96 (84–108)

^a One patient had concomitant excision of their megarectum.

Table 1

Causative factors for multistage Duhamel pull through surgery ($n = 35$).

Reason	Frequency (%)
Failed washouts	10 (29%)
Diagnostic uncertainty	8 (22%)
Surgeon Preference	7 (20%)
Late diagnosis	4 (11%)
Presenting with a cecal perforation	3 (9%)
Dilated, thickened bowel	3 (9%)

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