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Duhamel Versus Swenson Pull-Through for Total Colonic Aganglionosis: A Multi-Institutional Study

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ABSTRACT

Background: Total colonic aganglionosis (TCA) is a rare variant of Hirschsprung disease (HD) where the colon and portion of distal ileum lack ganglion cells. Most pediatric use either a straight ileoanal (Swenson or Yancey-Soave) or a short Duhamel pull-through for TCA. There are no large studies comparing these techniques. We aimed to compare short-and medium-term outcomes between these approaches.

Method: A retrospective review was performed among children with TCA from 2001 to 2019 undergoing a primary Duhamel or Swenson pull-through across three large children's hospitals. Patients undergoing redo and patients with greater than 30 % small bowel aganglionosis were excluded. We gathered data on demographics, operative approach, and outcomes at one, two, and three years. Continuous variables were analyzed with t-tests and categorical variables with Chi square or Fisher's tests.

Results: There were 54 patients, with 26 (48 %) undergoing Duhamel and 28 (52 %) undergoing Swenson pull-through. There were no differences in sex, age, medical comorbidities, or operative details, including age at pull-through, laparoscopic vs open, length of involved small bowel, and operative time. Length of stay and post-operative complications were not different. Three years after pull-through, patients undergoing Duhamel had fewer stools per day (1-3 stools 69.6 % vs 14.3 %, $p = 0.003$) and were less likely to be prescribed fiber supplementation (4.2 % vs 43.8 %, $p = 0.003$). There were no differences in irrigations, botulinum toxin administration, loperamide, or HD admissions.

Conclusion: Both Duhamel and straight pull-throughs are safe for treatment of TCA, with acceptable short- and medium-term outcomes. Further studies on patient-reported outcomes are necessary to examine long-term differences.

Level of Evidence: III.

1. Background

Total colonic aganglionosis (TCA) is an uncommon form of Hirschsprung disease (HD), affecting 5–10 % of those diagnosed with HD, and occurring in 1/50,000 births. In TCA, the entire colon (and usually a segment of distal ileum) lacks ganglion cells. These children usually present in the neonatal period with intestinal obstruction and require an urgent operation for diagnosis and decompression with an ileostomy. Definitive reconstruction involves removal of the aganglionic colon and ileum, with pull-through procedure consisting of connection of the normally innervated small bowel just above the anal sphincter. Patients with TCA experience more morbidity and mortality than short segment HD and require more complex operative management. Although a variety of surgical techniques have been described for management of TCA, most pediatric surgeons in North America and Western Europe use either a straight ileoanal pull-through (i.e. Swenson or Yancey-Soave) in which the small bowel is directly connected to the anus or a Duhamel pull-through in which a short segment of aganglionic rectum is retained and the small bowel is anastomosed posteriorly, resulting in a small pouch [1,2].

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Children with TCA are known to have multiple issues following pull-through surgery, which include continued obstructive symptoms, frequent stools with dehydration and/or perineal excoriation, poor growth, soiling, and enterocolitis [3–8]. Although there are theoretical advantages of each of the two common operations, to date there have been few studies examining outcomes. Our study aimed to combine the experience of three high volume centers to specifically examine the effect of Duhamel vs straight pull-through on outcomes in patients with TCA.

2. Methods

2.1. Patient selection

We performed a retrospective cohort study across three large pediatric hospitals from 2001 to 2019. Patients were included if they had a diagnosis of TCA with a primary pull-through at one of the three institutions. Patients were excluded if they had their primary operation elsewhere and were transferred for management, if their pull-through at one of the three institutions was a redo, or if they had >30 % aganglionosis of the small intestine. The cohort was divided into subgroups by type of pull-through: Duhamel vs Swenson. The medical record at each institution was reviewed for each patient after appropriate institutional IRB approval was obtained. De-identified data were aggregated and analyzed.

2.2. Demographics, medical history, and operative course

Data were gathered on patient demographics and medical history, including the location of their primary operation, sex, gestational age, family history, and comorbidities. Information on operative history was also obtained. This included the age at first operation, age at definitive pull-through, and any additional operations prior to definitive pull-through. Information was collected regarding operative course, including the approach (laparotomy, laparoscopy, or open through the site of the prior stoma). Additional data points included the length of involved small bowel and the total operative time. We defined postoperative complications as occurring within 30 days of operation and included surgical site infection, prolonged ileus, and post-operative Hirschsprung-associated enterocolitis (HAEC). Length of stay was also captured.

2.3. Long term follow up

Data were recorded at the one-, two-, and three-year follow up time points as available. Number of stools were assessed, categorized into 1–3, 4–6, and 7+ per day. If the patient was documented as having a range of number of stools outside of those predetermined categories, the higher number was used. Additional data points included need for irrigations, botulinum toxin injection, or disease-related admission. Medication use was analyzed, including loperamide, fiber, and metronidazole. All postoperative long-term factors were captured over the preceding year.

2.4. Statistics

Descriptive statistics were computed for all categorical variables, and medians and interquartile ranges for continuous variables. Categorical variables were compared with Chi-square or Fisher's exact tests where appropriate. Continuous variables were compared with Wilcoxson rank-sum tests and described by medians and interquartile ranges. A p-value of <0.05 was considered significant. Statistics were computed with R Statistical Software.

3. Results

3.1. Population characteristics

There were roughly equal numbers of Duhamel (n = 26, 48.1 %) and Swenson (n = 28, 51.9 %) in the cohort (Table 1). Most of the cohort was male (n = 40, 74.0 %). Patients infrequently had a family history of HD (n = 17, 31.5 %), medical comorbidities (n = 14, 25.9 %), or a diagnosis of trisomy 21 (n = 4, 5.6 %). The median gestational age was 40.0 weeks.

Patients underwent initial ileostomy placement at a median of 8.5 (3.25–29.25) days (Table 2). Definitive pull-through was performed at a median age of 15.0 (10.0–28.5) months, and the majority were via an open approach (n = 39, 72.2 %), with a median small bowel involvement of 10.0 cm and a median operative time of 225 min. Within the 30 days following pull-through, there were low rates of postoperative complications, including surgical site infection (n = 7, 13.0 %), prolonged ileus (n = 4, 7.4 %), and only one patient with HAEC (1.9 %). The median length of stay was 8.5 days.

3.2. Comparison of demographics and outcomes

All Duhamel procedures were performed at one institution; the other two institutions performed the Swenson procedure (Table 1). There were no differences in demographics or medical history between the two subgroups. All patients underwent initial ileostomy placement prior to definitive pull-through. There were no differences between subgroups in age at operation, age at pull-through, operative approach, length of involved small bowel, operative time, early postoperative complications or length of stay (Table 2).

3.3. Long-term follow up

Outcomes were then compared over three years with interval analysis at each year (Table 3). At the one-year time point, the Swenson patients were prescribed fiber more frequently (43.0 % vs 8.0 %, $p = 0.006$). There were otherwise no differences in outcomes at the one-year time point. At two years, there was interestingly no longer a difference between subgroups in the use of fiber. However, the Duhamel group had a higher utilization of metronidazole (64.0 % vs 24.0 %, $p = 0.01$). At the three-year time point, there was higher fiber utilization in the Swenson group compared to the Duhamel group (44.0 % vs 4.0 %, $p = 0.003$). However, the difference in metronidazole usage did not persist. At three years post pull-through, there was a difference in reported stool frequency, with more Duhamel patients experiencing only 1-3 stools per day compared to higher numbers experienced by Swenson patients (70 % vs 14 %, $p = 0.003$). At no time point (one-, two-, or three-year) was there a difference in use of irrigations, botulinum toxin injection, HD-related admission, or loperamide usage. Within the 3 year follow up period, one patient who underwent a Swenson pull-through required an ileostomy due to obstructive symptoms secondary to transition zone pull-through and later underwent a redo pull-through.

4. Discussion

The choice of technique in pull-through surgery for TCA has been an area of debate since Sandegard first described the successful treatment of long segment Hirschsprung disease by a pull-through procedure in 1953 [9]. Multiple adaptations of various techniques have been described to minimize postoperative morbidity related to enterocolitis, electrolyte abnormalities, stooling frequency, and incontinence. One such modification, described by Martin et al., involved the formation of a stapled side-to-side

Table 1
Patient demographics and medical history.

	Cohort n = 54	Duhamel Pull-Through (DPT) n = 26 (48 %)	Straight Pull-Through (SPT) n = 28 (52 %)	p-value
Sex				0.28
Male	40 (74.0 %)	17 (63.4 %)	23 (82.1 %)	
Female	14 (26.0 %)	9 (34.6 %)	5 (17.9 %)	
Gestational age, weeks	40.0 (38.0–40.0)	40.0 (38.3–39.0)	39.0 (38.0–40.0)	0.26
Family history of HD	17 (31.5 %)	9 (34.6 %)	8 (28.6 %)	0.85
Medical comorbidities	14 (25.9 %)	6 (23.1 %)	8 (28.6 %)	0.88
Trisomy 21 diagnosis	3 (5.6 %)	2 (7.7 %)	1 (3.6 %)	0.95

Table 2
Operative Outcomes. Continuous variables represented as median [IQR].

	Cohort n = 54	DPT n = 26 (48 %)	SPT n = 28 (52 %)	p-value
Age at first operation, days	8.5 (3.3–29.3)	9.5 (4.3–49.5)	8.5 (3.5–22.8)	0.07
Age at pull-through, months	15.0 (10.0–28.5)	10.0 (9.0–18.8)	18.0 (12.0–31.5)	0.09
Operative approach				0.06
Open	39 (72.2 %)	15 (57.7 %)	24 (85.7 %)	
Laparoscopic	5 (9.3 %)	3 (11.5 %)	2 (7.1 %)	
Via stoma site	10 (18.5 %)	8 (30.8 %)	2 (7.1 %)	
Length of involved small bowel, cm	10.0 (0.0–20.0)	8.5 (0.0–20.0)	10.0 (5.0–20.0)	0.06
Operative time, minutes	225 (215–308)	225 (218–344)	266 (213–300)	0.49
Early post-operative complications				0.11
Surgical site infection	7 (13.0 %)	3 (11.5 %)	4 (14.3 %)	
Prolonged ileus	4 (7.4 %)	4 (15.4 %)	0 (0.0 %)	
Post-operative enterocolitis	1 (1.9 %)	1 (3.8 %)	0 (0.0 %)	
Post-operative length of stay, days	8.5 (6.0–12.0)	7.5 (5.0–11.8)	9.0 (7.0–11.3)	0.22

ileocolostomy with the retained aganglionic descending and sigmoid colon, and later with the entire colon [10–12]. Many of these procedures have since been abandoned with many surgeons in North America and Western Europe favoring either a straight ileoanal pull-through (i.e. Swenson or Yancey-Soave) in which the small bowel is directly connected to the anus) or the Duhamel pull-through in which a short segment of aganglionic rectum is retained and the small bowel is anastomosed posteriorly, resulting in a small pouch. Large studies comparing both techniques have been limited due to the rarity of the disease and lack of enough volume of TCA at single institutions.

Our multi-institutional study examining Duhamel vs Swenson for TCA shows both techniques to be safe with favorable long-term outcomes. Post-operative complications were minimal and not different between the two techniques and there were no differences in irrigation requirements, botulinum toxin administration, loperamide usage, or admissions related to HD. Patients undergoing the Duhamel procedure had fewer stools per day and patients undergoing the Swenson procedure were more likely to be prescribed fiber supplementation. The low incidence of major early complications requiring surgical intervention and the low need for repeat reconstruction support the benefits and appropriateness of managing TCA at tertiary centers.

There was a low incidence of early enterocolitis (within 30 days postoperatively) across both approaches. However, the incidence of HD-related admission in the first year, which included admission for enterocolitis, was high in both procedures (50 % DPT, 43 % SPT) and decreased over the 3 year follow up (33 % DPT, 13 % SPT), still indicating a high level of morbidity in patients with TCA.

Loss of the entire colon leads to reduced fluid absorption and faster transit of intestinal contents. The theoretical advantage of the Duhamel procedure is the small rectal pouch which can act as a stool reservoir. This concept was supported by our data, as there was a significant decrease in stooling frequency with the Duhamel procedure. This has also been well described in the literature and has been shown to improve over time as the intestine adapts by increasing water and sodium reabsorption in the terminal ileum [2,6,8,13]. There was no difference in loperamide use between the two groups but patients undergoing the Swenson procedure had higher fiber use.

Table 3
Follow up outcomes.

	1 year follow up			2 year follow up			3 year follow up		
	DPT 26 (48)	SPT 28 (52)	p-value	DPT 26 (48)	SPT 28 (52)	p-value	DPT 26 (48)	SPT 28 (52)	p-value
Number of stools per day			0.07			0.58			0.003
1-3	11 (44 %)	2 (11 %)		8 (33 %)	4 (29 %)		16 (70 %)	2 (14 %)	
4-6	9 (36 %)	11 (61 %)		12 (50 %)	6 (43 %)		5 (21 %)	9 (64 %)	
7+	5 (20 %)	5 (28 %)		3 (13 %)	4 (29 %)		2 (9 %)	3 (21 %)	
Irrigations	16 (62 %)	12 (57 %)	0.25	8 (32 %)	6 (35 %)	1.0	5 (21 %)	5 (33 %)	0.46
Botulinum toxin injection	5 (19 %)	8 (36 %)	0.21	5 (20 %)	4 (25 %)	0.72	3 (13 %)	2 (13 %)	1.0
HD related admission	13 (50 %)	9 (43 %)	0.77	9 (36 %)	5 (29 %)	0.75	8 (33 %)	2 (13 %)	0.26
Loperamide	13 (50 %)	11 (52 %)	1.0	13 (52 %)	8 (47 %)	1.0	11 (46 %)	8 (50 %)	1.0
Fiber	2 (8 %)	9 (43 %)	0.006	3 (12 %)	5 (29 %)	0.23	1 (4 %)	7 (44 %)	0.003
Metronidazole	15 (58 %)	6 (29 %)	0.08	16 (64 %)	4 (24 %)	0.01	13 (54 %)	5 (31 %)	0.20

After surgery for HD, children may still develop obstructive symptoms and or enterocolitis due to a functional obstruction caused by an inability of the internal anal sphincter to relax. In the first year, 60 % of patients in both groups needed rectal irrigations and by 3 years only 20 % of Duhamel and 33 % of Swenson patients required irrigations. Botulinum toxin injection was used in both groups to treat non-relaxing internal anal sphincter with no difference between the two groups. Botulinum toxin utilization varied from 19 % in the Duhamel and 36 % in the Swenson group during the first year to 13 % in both groups at the 3 year follow up.

This study is the first to examine the outcomes from Duhamel vs Swenson pull-through in children with TCA. However, it is vulnerable to certain limitations. The study is it is retrospective and comes from multiple institutions; some of the differences identified may therefore reflect institutional practice rather than differences related to the operation performed. However, the finding of lower stool frequency in the Duhamel group is not related to institutional practice, and fits with the existing literature. As discussed, outcomes may be more favorable at our institutions given high referral pattern and surgeon and institutional experience with managing TCA. Additionally, we limited our analysis to the first three years post-operatively, which limits analysis of functional outcomes related to toilet training or quality of life due to patient age. Finally, given its retrospective nature, the study is vulnerable to selection bias and misinformation bias.

5. Conclusion

In conclusion, our results show that in high-volume centers, both Duhamel and Swenson pull-through is safe with comparable outcomes in the short-term. Future studies will focus on long-term follow-up with a focus on functional outcomes and quality of life.

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