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Original Article

Surgical outcomes of total colonic aganglionosis in children: A 26-year experience in a single institute

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Abstract

Background: There is a lack of consensus regarding the treatment of total colonic aganglionosis (TCA) with respect to perioperative morbidity, mortality, complications, and functional outcomes. The aim of this study was to review the results of surgical TCA treatment over a 26-year period and characterize the outcomes.
Methods: We retrospectively reviewed the clinical characteristics, surgical courses, and outcomes of TCA patients who underwent definitive pull-through operations from 1986 to 2012. Follow-up data were collected by chart reviews and telephone interviews using a standardized questionnaire.
Results: We identified nine infants with TCA (8.6%) from among 105 infants with Hirschsprung's disease treated during the 26-year period. Neither sex predominated (male/female ratio = 4:5). All infants underwent laparotomies and simultaneous enterostomies. All patients eventually underwent modified Duhamel pull-through procedures at a mean age of 179 days (range, 47–352 days). Two infants died of complications after surgery including heart failure and sepsis. The remaining infants recovered smoothly with antilaxative medications, and all but one was weaned off these medications. Although the surviving patients did not catch up on growth, they and their families were satisfied with the surgical results.
Conclusion: Infants with TCA had satisfactory outcomes after the modified Duhamel pull-through operation. Based on our experience, we suggest that the pull-through operation could be performed earlier, even when there are loose stools from the enterostomy.

Keywords: child; fecal incontinence; Hirschsprung disease; retrospective study; total colonic aganglionosis

1. Introduction

Hirschsprung's disease is an embryonic developmental disorder characterized by the absence of ganglion cells in the lower enteric complex. The disease may associate with chromosomal anomalies, monogenic syndromes, or other congenital anomalies, such as trisomy 21, congenital central hypoventilation syndrome,[1] congenital heart disease, or urinary tract anomalies. Sporadic disease is sometimes associated with *RET* gene mutations.[2] Total colonic aganglionosis (TCA) is a rare and severe phenotype of Hirschsprung's disease occurring in about 2–13% of cases.[3] This form of disease is characterized by the absence of ganglion cells in the entire colon extending into the terminal ileum, and carries higher morbidity and mortality rates than does the short-segment

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form of Hirschsprung's disease.[4] Various procedures are proposed for the surgical management of TCA. However, there is no consensus as to which method is the best, with respect to perioperative morbidity, mortality, complications, and functional outcomes, as described in a recent systematic review.[5−7] The aim of our study was to review the TCA cases treated in our institution over the past 26 years, with emphasis on the experience of early pull-through and analysis of functional outcome.

2. Methods

We retrospectively analyzed the medical records of nine consecutive patients with TCA who were treated in a tertiary referral center from 1986 to 2012. There were 105 patients with surgically treated Hirschsprung's disease in our hospital during this period. Data were analyzed for sex, clinical presentation, family history of Hirschsprung's disease and associated anomalies, weight at the definitive operation, surgical management, and pre- and postoperative complications.

Informed consent was obtained from the patient or their parents. Follow-up data were collected by telephone interviews using a standardized questionnaire previously described in the literature[8] regarding growth development, stool frequency, stool consistency, fecal soiling, incontinence, and enterocolitis. A scoring system was used for the assessment of objective functional outcomes (Table 1).[8] Enterocolitis was defined as the presence of abdominal distention, diarrhea, vomiting, and fever. Additionally, the most common complication of the pull-through operation was diarrhea, with or without perineal excoriation and enterocolitis.[9,10]

3. Results

Among the nine patients with TCA, there were four males and five females (Table 2). Four patients presented with symptoms in the neonatal period (within 28 days after birth) with features of intestinal obstruction. Four of the five remaining patients presented later in infancy (<1 year) and had previous surgeries performed at other hospitals (2 ileostomies, 1 ileostomy with a Kimura patch, and 1 misplaced sigmoid colostomy). The initial presentations of the five referred patients were intestinal obstruction in four patients and toxic megacolon with perforation in the other patient.

Table 1
Telephone questionnaire regarding functional outcome of patients with TCA.[a]

Follow-up questionnaire	Score = 2	Score = 1	Score = 0
Recurrent abdominal distension	None	Mild	Severe
Frequency of defecation	1−2/d	3−5/d	>5/d
Stool consistency	Normal	Loose	Liquid
Soiling	None	Occasionally	Permanently
Urgency period[b]	Normal	Short	Absent
Diapers required[b]	None	Occasionally	Permanently
Long-term use of medication[b]	None	Antibiotics	Antidiarrheal
Diet	Normal	Restricted	TPN
Range of scores	11−16	6−10	0−5
Objective functional outcome	Good	Fair	Poor

TCA = total colonic aganglionosis; TPN = total parenteral nutrition.
[a] See Wildhaber et al.[8]
[b] If age >3 years.

Two girls had a family history (a sibling and a father) of Hirschsprung's disease, and another boy was born of consanguineous parents. Associated anomalies were hypoplastic right heart syndrome in one patient, imperforate anus in one patient, and a complex case of urogenital sinus, vaginal atresia, and Pallister−Hall syndrome in another patient.

Eight patients (i.e., all except the girl with the misplaced sigmoid colostomy) had leveling enterostomy performed with a view to future definitive pull-through operations. All patients eventually underwent the modified Duhamel pull-through operation. We delivered the ganglionated bowel through an incision in the posterior aspect of the native aganglionated rectum. The common wall between the ganglionated pull-through colon and the aganglionated native rectum was then divided using an endo-GIA linear stapler.

The mean patient age at the definitive pull-through operation was 179 days (range, 47−352 days). The mean body weight at the definite operation was 5137 g (range, 2810−7600 g). The average interval between initial ileostomy and definitive pull-through surgery was 5.1 months (range, 1−10 months).

3.1. Preoperative period

Four of the nine patients in this series experienced preoperative complications. Three of the nine patients had one or more attacks of enterocolitis before the definitive Duhamel pull-through surgery. One patient developed toxic megacolon with perforation, another (who had a previous ileostomy with Kimura patch at another hospital) developed pouchitis with intractable diarrhea, and the third patient (who had a misplaced sigmoid colostomy at another hospital) had recurrent intestinal obstruction with enterocolitis. Wound infection with sepsis complicated the preoperative course in one other patient.

3.2. Postoperative complications

The most common post−pull-through complication was enterocolitis, which occurred in five patients. Other complications included wound infections in two patients and perineal skin excoriation in three patients. One patient with coexisting hypoplastic right heart syndrome died of heart failure, and another patient with urogenital sinus died of a presumed episode of sepsis outside the hospital. Other postoperative conditions included a herniated incision ($n = 1$), intestinal adhesions requiring enterolysis ($n = 1$), toxic megacolon with bowel perforation ($n = 1$), anal stricture requiring anoplasty ($n = 1$), and chronic lower gastrointestinal bleeding with iron deficiency anemia ($n = 1$).

3.3. Long-term follow-up

At a median follow-up age of 9 years (range, 1−28 years), one patient had died of congenital heart disease, and another was unavailable for follow-up. Two patients <3 years of age were too young to be evaluated for voluntary bowel