



Comprehensive review of procedures for total colonic aganglionosis

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Abstract

Purpose: A variety of procedures have been described for the treatment of total colonic aganglionosis (TCA); however, there is no consensus as to a superior operative method. The objective of this review is to evaluate the operative methods used to treat TCA and their effectiveness with respect to morbidity, mortality, rates of enterocolitis, and functional outcomes.

Methods: We searched Medline (1950–2007) and the Cochrane Library for studies reporting the operative treatment of TCA. Search terms used were “aganglionosis,” “Hirschsprung’s disease,” “total colonic,” and “long-segment.” Studies evaluated were limited to those that solely discussed the operative outcomes for the treatment of TCA. A meta-analysis was then performed for morbidity outcomes in those studies describing a single operative procedure.

Results: The search yielded 271 articles related to TCA. One hundred twenty-two articles discussed operative treatment of Hirschsprung’s disease, and only 26 of these articles discussed operative management of TCA. Nineteen articles were isolated for statistical analysis on the basis of results for perioperative outcomes for a single procedure. These procedures included Martin (4), colonic patch modifications (4), Duhamel (2), endorectal pull-through (1), Kimura (2), Boley (2), Rehbein (2), Soave (1), and J-pouch ileoanal anastomosis (1). Overall mortality was 1.9%. Meta-analysis revealed morbidity rates ranging from 10% to 64% depending on procedure category without significant outliers. Higher morbidity rates were found for the Martin-type procedure using the descending colon but with noted higher within-group variance due to small sample sizes. Enterocolitis was noted in 7 studies with a mean of 22%.

Conclusions: There is no superior operative method for the treatment of TCA with respect to perioperative morbidity, mortality, enterocolitis, and functional outcomes. The operative technique performed should be selected on center and surgeon familiarity and expertise.

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Aganglionosis involving the entire colon (total aganglionosis or TCA) accounts for 5% to 15% of those diagnosed with Hirschsprung’s disease, occurring once in every 50,000 births [1–3]. Total colonic involvement has significantly higher morbidity and mortality than short segment Hirschsprung’s disease and requires more a complex medical and

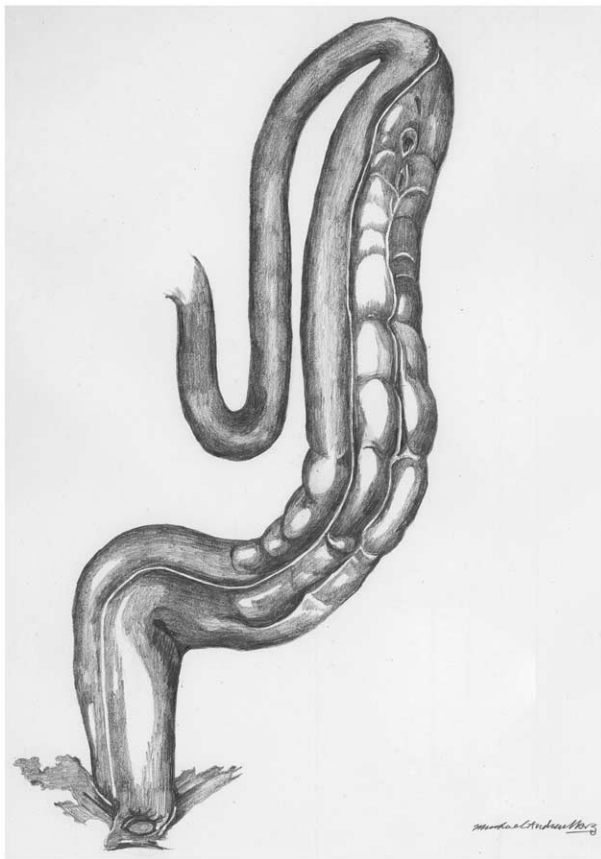


Fig. 1 Martin's ileocolostomy (descending colon)—original work.

operative management [4]. Multiple procedures exist for the treatment of TCA; however, there is no current consensus on a superior operative procedure. We have performed a systematic review of the procedures used to treat TCA to address this question.

Treatment of TCA involves early decompression of the colon by an ostomy using the most distal ganglionated bowel in the neonatal period. This is typically followed by a second reconstructive procedure after the child has had adequate time to grow and nutritional, fluid, and electrolyte abnormalities are corrected. Multiple procedures have been described for the definitive treatment of TCA. Early procedures for the treatment of TCA have evolved from procedures used for rectosigmoid or short segment aganglionosis. These include pull-through procedures initially described by Swenson [5], Duhamel [6], and Soave [7]. Several modifications of these procedures have since been described to decrease resulting morbidity (anastomotic leaks, enterocolitis, poor functional outcomes). One such modification, described by Martin et al [8-9], used the absorptive capacity of the retained aganglionic colon through the formation of a stapled side-to-side ileocolostomy with the descending and sigmoid colon (Fig. 1). This procedure was further modified to use the entire colon [10] (Fig. 2). Resulting morbidity from these Martin-type modifications, in the form of frequent diarrhea,

lead to the development of the ascending colon patch technique described by Boley [11] (Fig. 3). In this procedure, a 15 to 20-cm segment of the ascending colon is anastomosed (2-layer, handsewn) along its antimesenteric border 4 to 6 cm from the distal end of ganglionated ileum. Blood supply to this on-lay patch is provided by either a vascular pedicle supplied by branches of the ileocolic artery or through neovascularization in a staged procedure [12]. The J-pouch ileoanal anastomosis, successfully used in ulcerative colitis patients, has also been applied with success in a limited number of TCA patients [13]. In this procedure, a 7 to 10-cm long stapled J-pouch is formed from the distal ileum with a single layer handsewn ileoanal anastomosis (Fig. 4). A systematic review of the literature was performed to evaluate the various procedures used to treat TCA, and a meta-analysis was performed to compare these procedures.

1. Methods

We searched Medline (1950-2007) and Cochrane Library databases for articles describing the treatment of TCA. Search terms used were "aganglionosis" and "Hirschsprung's disease," and results were subsequently narrowed with the search terms "total colonic" and "long-segment." The search was limited to human subjects and English

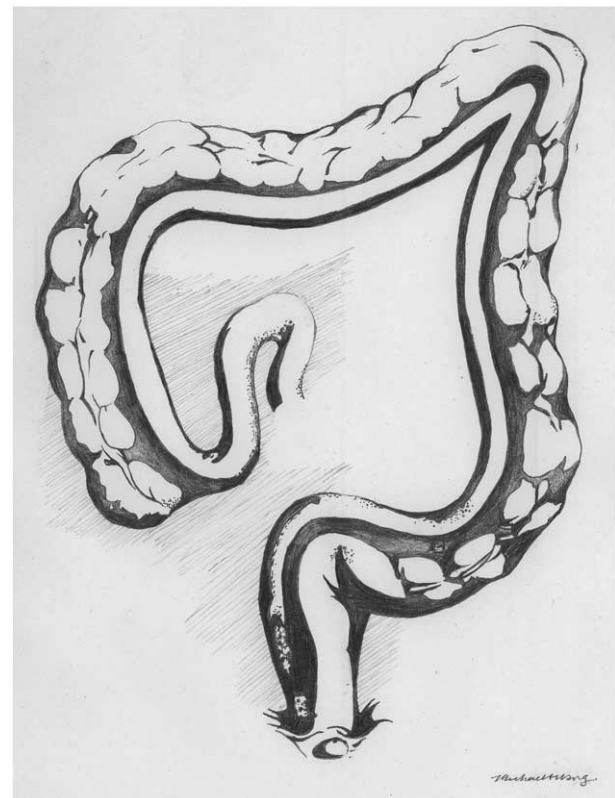


Fig. 2 Martin's ileocolostomy (total colon)—original work.

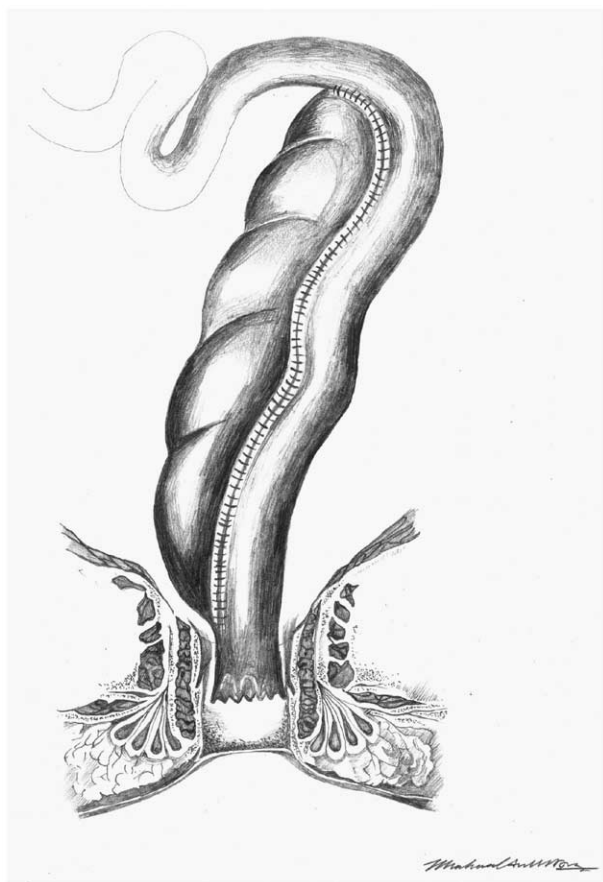


Fig. 3 Ascending colon patch—original work.

language articles. A single reviewer evaluated abstracts for inclusion. Inclusion criteria were (1) studies describing operative management of TCA with a single procedure and (2) studies describing the postoperative outcomes of mortality, morbidity, rates of enterocolitis, and functional outcomes after definitive treatment of TCA. Studies describing TCA with extensive small bowel involvement were excluded, as management of these complex cases is beyond the intended scope of our review. Studies were also excluded if any of the following were noted: (1) nonoperative management was described, (2) there was no discussion of postoperative outcomes, (3) treatment of sub-TCA, and (4) discussion of multiple procedures where the outcomes for total colonic patients could not be extracted. Studies discussing postoperative results of morbidity, mortality, enterocolitis rates, and functional outcomes for a single operative intervention in the treatment of TCA were included for final analysis.

2. Statistical methods

Cumulative morbidity values reported from each individual study, after logit transformation, were combined within

4 of the 5 groups defined by the operative procedures with fixed effect meta-analytic models. One group included only one study and was excluded from the subgroup analyses. Heterogeneity between studies within operative subgroups was assessed by I^2 [14]. Where reported morbidity was either 0 or 1, a statistical adjustment was made to allow calculation of appropriate standard errors of the logit. All calculations were performed with Comprehensive Meta-Analysis Version 2 [15].

3. Results

Our database search initially yielded 271 studies related to TCA (Fig. 5). Of these, 122 articles discussed operative treatment of Hirschsprung's disease of variable lengths of colonic involvement, and 26 discussed operative management of TCA. Seven studies were excluded as they discussed multiple procedures used in the treatment of TCA. Nineteen studies describing a single procedure in the operative treatment of TCA and operative outcomes (mortality, morbidity, postoperative enterocolitis, functional outcomes) were included in

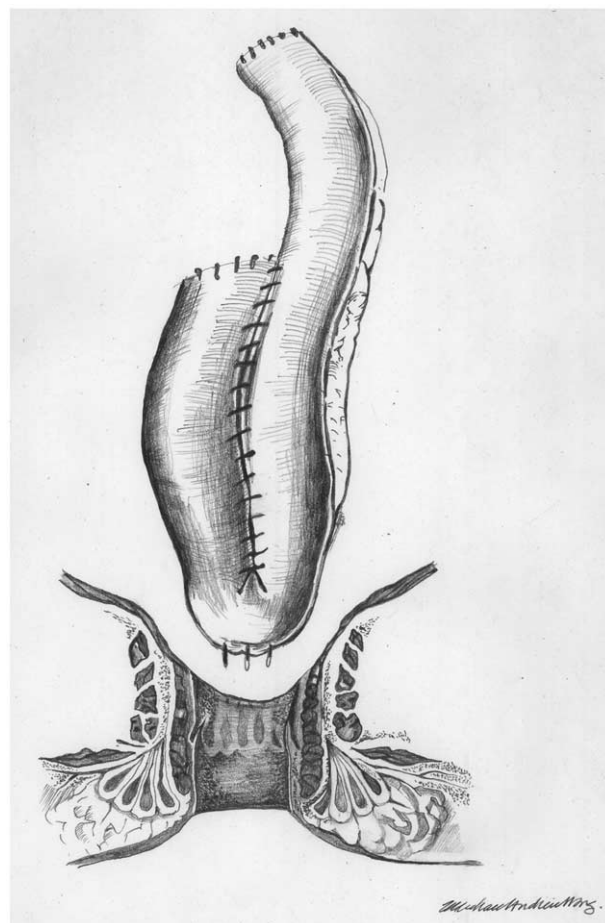


Fig. 4 J-pouch ileoanal anastomosis—original work.

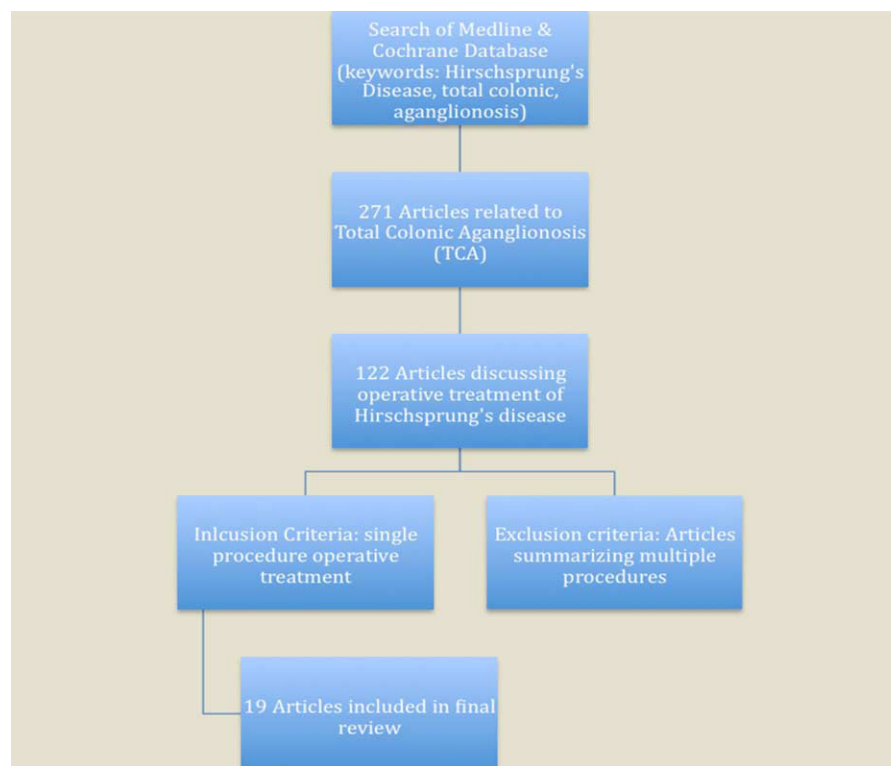


Fig. 5 Literature search flowchart.

the final analysis [8-10,12,13,16-28] (Table 1). All studies reviewed were retrospective series.

A total of 19 operations on 161 patients with TCA were included in this study. A total of 19 articles were included in the analysis that described 5 broad categories of operative techniques. These categories were defined as (1) total colectomy with endorectal pull-through (ERPT) with ileorectal or ileoanal anastomosis ($n = 63$), (2) ERPT with ileorectal anastomosis and ileocolostomy using the entire aganglionic colon ($n = 24$), (3) ERPT with ileorectal anastomosis and ileocolostomy using the left/descending colon ($n = 41$), (4) ERPT with an right ascending colon onlay patch ($n = 23$), and (5) total proctocolectomy with ileoanal pouch anastomosis ($n = 10$). Overall mortality was noted to be 1.9% (3 deaths). The mean age for definitive pull-through procedure when performed was 15.3 ± 8.4 months. Morbidity rates ranged from 0% to 100% with an average morbidity rate of $44.0\% \pm 32.9\%$. Postoperative enterocolitis was reported in 7 studies with a mean of $22.6\% \pm 13.5\%$. These study results are summarized in Table 2. In an overall average follow-up time of 3.9 years after definitive surgery, most children were continent with an average of 3 to 5 stools daily (Table 3). A meta-analysis was performed on the reported morbidity rate for each operative category. An increased morbidity rate (64%) was noted in those patients who received the Martin-type (ileocolostomy using the descending colon) procedure when compared to the other procedure types (Figs. 6 and 7). This group, however, was noted to have significant within-group

variance with high degree of heterogeneity. Heterogeneity within procedure categories are as follows: (1) ERPT only, $I^2 = 73.9$; (2) Martin ileocolostomy (entire colon), $I^2 = 8.2$; (3) Martin ileocolostomy (descending colon), $I^2 = 56.6$; (4) ascending colon patch, $I^2 = 0.0$; and (5) ileoanal pouch anastomosis, $I^2 = 0.0$. The low outcome rate with respect to mortality, the low reporting frequency of enterocolitis, the variable reports of daily stool frequency, as well as the overall small sample sizes made a statistical meta-analysis of these outcomes prohibitive.

4. Discussion

The treatment of TCA has improved with decreased morbidity and mortality [29-36]. This decrease can be attributed to earlier diagnosis of TCA, earlier diagnosis and treatment of enterocolitis (formerly a frequent cause of mortality), and improvements in supportive care [32,37].

Sandegard [38] first described the successful treatment of long segment Hirschsprung's disease by a pull-through procedure in 1953. Swenson [39] subsequently advocated total colectomy with ileoproctostomy in 1955 for the treatment of TCA. Multiple adaptations have since been described in an attempt to minimize postoperative morbidity related to enterocolitis, electrolyte abnormalities, stooling frequency, and incontinence. Variable lengths of the aganglionic colon have been anastomosed to

Table 1 Articles reviewed

Study year	Study author(s)	Study origin	Patients (n)	Procedure description
1968	Martin LW	United States (Cincinnati, Ohio)	5	Ileorectal end/side anastomosis stapled ileocolostomy (up to splenic flexure)
1972	Martin LW	United States (Cincinnati, Ohio)	9	Ileorectal end/side anastomosis stapled ileocolostomy (entire colon)
1976	Burrington JD	United States (Chicago, Ill)	6	Modified Duhamel stapled ileocolostomy (sigmoid/rectum)
1981	Kimura K et al	Kobe, Japan	3	Swenson pull-through ascending colon patch (devascularized patch) (3 stage)
1982	Martin LW	United States (Cincinnati, Ohio)	13	Ileorectal end/side anastomosis stapled ileocolostomy (entire colon)
1984	Shandling B	Toronto, Ontario Canada	2	Soave ERPT ileocolostomy with entire colon
1988	Applebaum H et al	United States (Los Angeles, Calif)	2	ERPT colonic on-lay patch (ascending/transverse colon-Boley)
1988	Kimura K et al	Kobe, Japan	7	Swenson-type pull-through ascending colon patch (devascularized patch)
1989	Shermeta DW	United States (Chicago, Ill)	5	Soave-Boley ascending colon patch
1989	Bergmeijer JH et al	Rotterdam, Netherlands	6	Rehbein (ileorectostomy)
1994	Endo M et al	Tokyo, Japan	9	Modified Duhamel-ERPT ileocolostomy (sigmoid/descending)
1996	Yamagiwa I et al	Yamagata, Japan	1	Modified Duhamel ascending colon patch
1996	Hengster P et al	Innsbruck, Austria	8	Rehbein (ileorectostomy)
1997	Emslie J et al	United States (Los Angeles, Calif)	5	ERPT on-lay patch (ascending/transverse colon)
2000	Coran AG et al	United States (Ann Arbor, Mich)	25	ERPT only
2001	Bonnard A et al	Paris, France	3	Laparoscopic modified Duhamel ileocolostomy (sigmoid/rectum)
2001	Dodero P	Genoa, Italy	24	Soave (ERPT only)
2002	Fouquet V et al	Paris, France	18	Modified Duhamel stapled ileocolostomy (sigmoid/rectum)
2002	Rintala RJ et al	Helsinki, Finland	10	IPAA (J-pouch)

IPAA indicates ileoanal pouch anastomosis. Seven studies reviewed multiple procedures for the management of TCA and therefore excluded in the final review.

ganglionated small bowel in an attempt to use the absorptive capacity of the large colon. The Duhamel procedure, initially described for short segment Hirschsprung's disease, obliterates the septum between the ganglionated colon and the aganglionic rectum with spur-crushing clamps [6]. This procedure was later adapted by Martin [8-10] in the treatment of TCA, first, using the sigmoid and descending colon and subsequently, using the entire aganglionated colon.

Increasing the anastomotic length between the functional small bowel and aganglionated colon has not, however, resulted in a linear improvement in morbidity [40]. Lengthy anastomoses with the aganglionated colon have been shown to result in increased long-term morbidity of nighttime incontinence and increased rates of enterocolitis [26,35,41,42]. Kimura, Boley, and others [11,12,18,20,25,43] have described the use of the ascending colon patch graft (given the greater absorptive capacity of the ascending colon compared to the descending colon or rectum) and have shown favorable results. Others still advocate the straight endorectal pull-through techniques, quoting comparable functional results and more favorable morbidity profiles when compared to

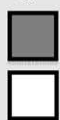
reservoir procedures [44]. More recent procedures have included a restorative proctocolectomy with a J-pouch ileoanal anastomosis and laparoscopic pull-through procedures [13,27]. These procedures have had good functional outcomes with minimal postoperative complications; however, they are limited with respect to short-term follow-up and a small sample of patient experience.

In our review, we encounter no procedure that proves to result in superior outcomes. Overall mortality after treatment of TCA is acceptably low at 1.9%; however, it should be noted that this mortality data encompass only those who survived to receive a definitive operation. Overall mortality for TCA has been as high as 53% and has been improved by appropriate and early diagnosis of TCA and preoperative management of nutrition, fluids, and electrolytes as well as proper treatment of enterocolitis [31,45].

Morbidity rates showed a trend toward increased complication rates in those who received a Martin-type ileocolostomy using the sigmoid and descending colon; however, the degree of heterogeneity of the studies within this category certainly limits the generalizability of this observation ($I^2 = 56.6$). Furthermore, many of the complications noted are minor and are medically managed, whereas very few require

Table 2 Perioperative outcomes

Year	Study	Patients (N)	Mean Age (mo)	Mortality % (n)	Morbidity %	Enterocolitis (%)	Complications noted
1997	Emslie, J. et al.	5	15	0	20	-	Obstruction requiring revision (16 weeks)
1996	Yamagiwa, I. et al.	1	3	0	100	-	Perineal excoriation
1989	Shermeta, D.W.	5	18	0	20	-	Bowel obstruction
1988	Applebaum, H. et al.	2	17.5	0	50	0	Perineal excoriation
1988	Kimura, K. et al.	7	16	14 (1)	43	14	Death: aspiration Enteritis, bowel obstruction
1981	Kimura, K. et al.	3	-	0	0	-	None
2002	Fouquet, V. et al.	18	-	0	100	-	Water/elect abnormalities; septicemia, stoma prolapse; spur formation requiring reoperation (n=2)
2001	Bonnard, A. et al.	3	10	0	33	-	Wound infection
1994	Endo, M. et al.	9	9	0	89	-	Residual obstruction
1976	Burrington, J.D.	6	30	0	50	33	Bowel obstruction x 2 (septum reformation; ileostomy revision)
1968	Martin, LW	5	13.6	0	40	20	Diarrhea; bowel obstruction
1984	Shandling, B.	2	-	0	-	-	None
1982	Martin, LW	13	13	0	25	-	Diarrhea (1)
1972	Martin, LW	9	19	0	50	-	Diarrhea, septum regrowth (n=1), jejeuno-colic fistula
2001	Dodero, P	24	33.8	0	4.2	-	Recurrent diarrhea
2000	Coran, A.G. et al.	25	-	0	25	20	Wound infection
1989	Bergmeijer, J.H. et al.	6	5.5	0	33	-	Anastomotic bleeding; anastomotic leak
1996	Hengster, P. et al.	8	7.4	25 (2)	100	41	Deaths: cardiac arrest; malignant hyperthermia Complications: meningitis, EC, pneumonia; stricture, sepsis,
2002	Rintala, R.J., Lindahl, H.G.	10	18	0	10	30	Wound infection

Legend:

Right colon patch



Ileocolostomy left colon



Ileocolostomy entire colon



ERPT only



IPAA

(-) Values not reported

reoperation. Indications for reoperation included anastomotic bleeding, bowel obstruction, and septum reformation. The most frequent complication in this review was noted to be diarrhea with or without perineal excoriation. This finding is consistent with other observations [46]. Complications were noted in all operative categories and the severity of these complications was evenly distributed throughout all procedure categories.

Enterocolitis is a well-known complication in TCA. Only 7 studies noted postoperative enterocolitis at a mean of 22%. This is slightly lower than previously reported

rates of 30% to 75% [45,46]. The length of the retained aganglionic colon has been correlated with higher rates of enterocolitis [26,46]; however, this was not observed in the series reviewed.

The functional outcome after procedures for TCA reviewed in this series appear to be favorable but with variable lengths of follow-up. In addition, the variability in the reporting of daily stools did not allow us to perform any statistical comparison of these studies with respect to functional outcome. Some believe that anorectal function in Hirschsprung's disease improves throughout childhood

Table 3 Functional outcomes

Year	Study	Functional outcome	Follow-up period
1997	Emslie, J. et al.	All continent; 1-9 stools daily	Mean 7.8 years (Range: 3.5-12 years)
1996	Yamag iwa, I. et al.	All continent; 3-5 stools daily	2.5 years
1989	Shermeta, D.W.	All continent; 3-5 formed stools daily	Mean 1.25 years (Range : 3mo-2.5 years)
1988	Applebaum, H. et al.	All continent; 3-4 soft stools daily	1 month
1988	Kimura,K. et al.	Constipation (n=2); incontinent (n=1); 1-8 stools daily	Mean 5 years (Range: 1-8 years)
1981	Kimura,K. et al.	All continent; formed stools	Mean 2.3 years (Range: 15mo- 39 mo)
2002	Fouquet, V. et al.	Continent <5 years (n=6);continent in 5-9 years (n=5); incontinent at 9 years (n=1)	5-9 years
2001	Bonnard, A. et al.	All continent; 1-9 stools daily	Mean 14.8 months (Range: 1-42 mo)
1994	Endo, M. et al.	All continent; 1-5 loose/semisolid stools daily	Mean 11 years (Range: 2.8-22.4 years)
1976	Burrington, J.D.	All continent; 2-3 semi-formed stools daily	3.2 years (Range: 0.5-5.5 years)
1968	Martin, LW	4/5 assessed; all continent; 2-4 stools daily	Mean 3.9 years (Range: 2.5-8 years)
1984	Shandling, B.	1/2 assessed & continent; 3-4 stools daily	11 months
1982	Martin. LW	Incontinent (n=1); continent (n=12); 1-4 soft stools daily	Mean 2.9 years (Range: 8mo-7 years)
1972	Martin.LW	All continent; 2-3 semi-formed stools daily	Mean 5.7 years (Range: 2.5-12 years)
2001	Dodero, P	3-4 stools daily (3 mo); "only very occasional soiling or loss of stools"	2 years
2000	Coran, A.G. et al.	All continent; 5-10 stools daily	1 year
1989	Bergmeijer, J.H. et al.	All continent; 2-4 semi-formed stools daily	15 months
1996	Hengster, P. et al.	Incontinent (n=1); others continent; 2-10 loose/soft stools daily	11 years
2002	Rintala, R.J., Lindahl, H.G.	All daytime continence; 7% nighttime incontinence and 7% nocturnal BMs; median 4 stools daily	Median 4 years (Range: 6 mo-9 years)

Legend:

Right colon patch



Ileocolostomy left colon



Ileocolostomy entire colon



ERPT only



IPAA

(-) Values not reported

with 100% adult continence regardless of type of pull-through procedure [47,48], whereas others dispute this claim [49,50]. It is widely accepted that short segment Hirschsprung's disease has significantly improved functional outcomes when compared to total colonic involvement [50-52]. Long-term studies of functional outcomes after definitive treatment of TCA patients are minimal [53] and require further investigation.

No operative technique for the treatment of TCA was found to be superior with respect to perioperative outcomes. All procedures were comparable with respect to mortality, morbidity, rate of enterocolitis, and functional outcome. Significant improvement has been made in the overall mortality of TCA, whereas the morbidity is still high. Operative approach to TCA should be based on surgeon familiarity and expertise and overall center experience.

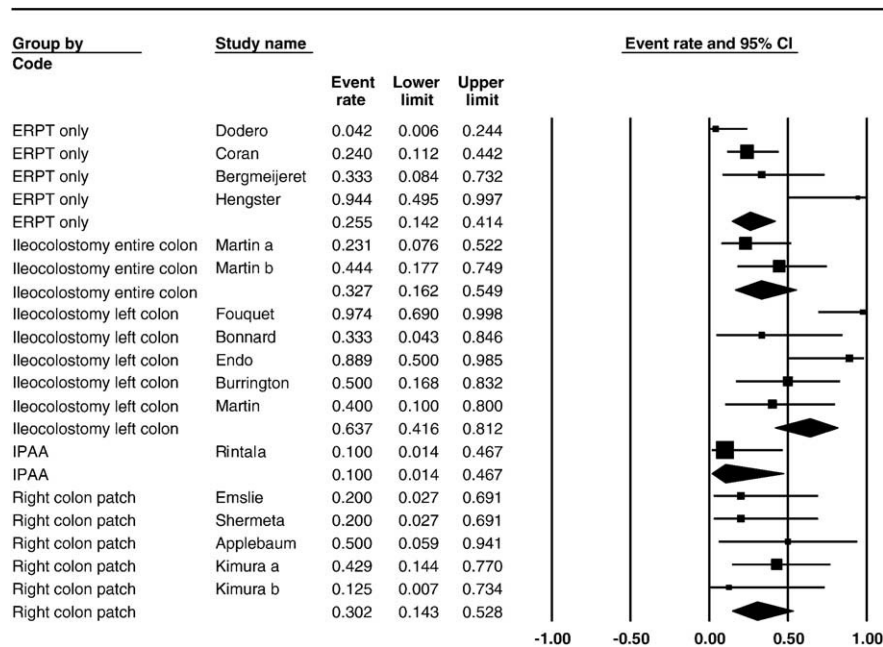


Fig. 6 Forest plot for morbidity (individual procedures).

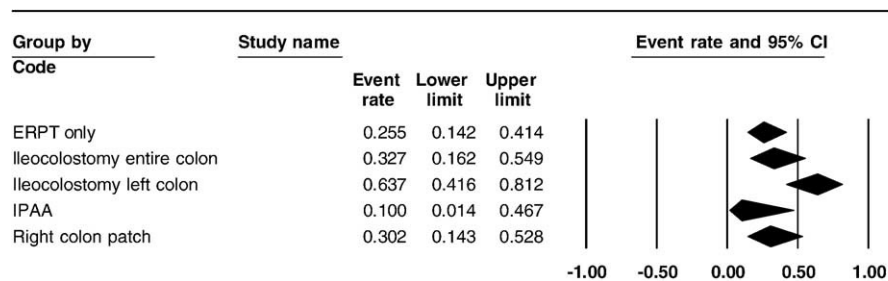


Fig. 7 Forest plot for morbidity (procedure category).

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Discussion

Dr. Patricia Donahoe (Boston, Mass): Very nice paper and certainly a good application of meta-analysis which I think, as Mike LaQuaglia showed yesterday, we need to apply more to our field where we have so many small numbers, so I applaud that approach.

Secondly, just a wish for the future. Do you have any idea what the causes of enterocolitis are in these patients?

Dr. Marquez (response): There are a couple of theories. One is that it is due to partial obstruction even after definitive pull-through, and the second theory is that it is due to internal sphincter spasm following these procedures. At this point, there is no clear answer as to the cause of enterocolitis.

Dr. Jack Langer (Toronto, Canada): I am surprised that the incidence of enterocolitis is that low. In my experience, it is much higher than that. As we are going to present tomorrow, I think evaluating the literature for enterocolitis depends entirely on what the definition of enterocolitis is and that is all over the map, so it is hard to interpret that.

The other thing is you did not mention the standard Duhamel operation which is actually our preferred operation for this. Did you include those in your endorectal pull-through group?

Dr. Marquez (response): Yes, they were included in our endorectal pull-through procedures.