Anorectal function after total colectomy with ileoanal anastomosis for total colonic aganglionosis

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Received 4 October 2016
Accepted 5 November 2016

The Egyptian Journal of Surgery
2017, 36:119–123

Background/purpose
Total colonic aganglionosis is a rare form of Hirschsprung's disease. Many techniques have been described for its management to improve the outcome as regards anastomotic leakage, recurrent constipation, and incontinence. The aim of this study was to assess the function of anorectum as regards the frequency and continence using the standard scoring system.
Patients and methods
In this retrospective study, 15 patients were evaluated after treatment in Zagazig University Hospitals. All cases underwent transanal endorectal pull-through with ileoanal anastomosis. All patients were evaluated by applying the Wexner score at 1, 3, and 6 months postoperatively.
Results
Fifteen infants were included in the study. None of them reached a high score level at sixth month postoperatively (i.e. totally incontinence). On applying the scoring system, the mean±SD result of all patients at first postoperative month was 15.0667±2.52039; at the second evaluation in the third month postoperatively the mean±SD score was 11.0667±3.28344, and lastly at the sixth month evaluation the mean±SD was 5.8667±2.82506. All results were significant on comparing the third month results with the first month results and on comparing the sixth month score results with either the first or the third month postoperatively.
Conclusion
Although total colonic aganglionosis is a rare form of Hirschsprung's disease, it needs staged surgical intervention. The most important stage is the second definitive one. Despite variable methods described for reconstruction after total colectomy, ileoanal one carries less major surgical complications and acquires accepted anorectal functional results with more improvement with time postoperatively.

Keywords:
endorectal pull through and ileoanal anastomoses, Hirschsprung's, disease, total colonic aganglionosis

Egyptian J Surgery 36:119–123

1110-1121

Introduction

Total colonic aganglionosis (TCA) with or without the involvement of a part of the small intestine is an infrequent form of Hirschsprung's disease (HD) and its incidence is about 3–12% of all infants with HD [1,2]. Many techniques have been described for the treatment of TCA. There is no superiority for one technique over others [3]. The longer the length of the remaining colon is adversely affecting the children's ability to defecate, the greater the incidence of postoperative enterocolitis [4].

Many different surgical techniques have been used for TCA [5–7], with outcomes mostly related to the type of surgical technique performed [8]. The techniques used include the Soave and Swenson techniques and the 'long' Duhamel procedure as modified by Martin *et al.* [9,10]. In certain parts of the world, the Kimura colonic patches had been a popular method for very long ganglionic segments [7,11].

Although a comparison of TCA patients managed with the Soave procedure showed fewer operative complications compared with those who underwent the extended Duhamel or Martin procedure, patients managed with the Soave procedure took longer time to defecate normally [12].

Different surgical procedures are used for treating TCA [13].

 DOI: 10.4103/1110-1121.204524

Patients and methods

This retrospective study included 15 infants; nine (60%) of them were female and six (40%) were male. These infants had been managed in the Pediatric Surgery Unit, Zagazig University, during the period between January 2008 and December 2015. The research had been approved by the local research committee and the patient's parents have been consented.

All patients underwent preoperative contrast enema and multiple biopsies (rectal, diversion site, and appendix) for diagnosis. All were treated first with proximal leveling ileostomy at the functioning dilated ileum either after exploration for neonatal obstruction or preliminary step in chronically constipated infants.

All patients underwent total colectomy, resection of a part of the involved small intestine, transanal mucosectomy of the rectum, and established continuity with ileoanal anastomoses by suturing the ileum to the anal canal 2 cm above the dentate line (Fig. 1).

Clinical data including age at operation, sex, symptoms at presentation, and diagnostic tools (contrast enema and multiple biopsies) were collected.

All patients were examined using the Wexner score, which is a questionnaire that is administered to parents at 1, 3, and 6 months postoperatively.

The Wexner score

Types of incontinence	Frequency				
	Never	Rarely	Sometimes	Usually	Always
Solid	0	1	2	3	4
Fluid	0	1	2	3	4
Gases	0	1	2	3	4
Wears pad	0	1	2	3	4
Lifestyle alteration	0	1	2	3	4

Never, 0; rarely, <1 month; sometimes, <1/week, 1/month; usually, <1/day, 1/week; always, 1/day. 0=perfect continence; 20=complete incontinence Jorge and Wexner [14].

Results

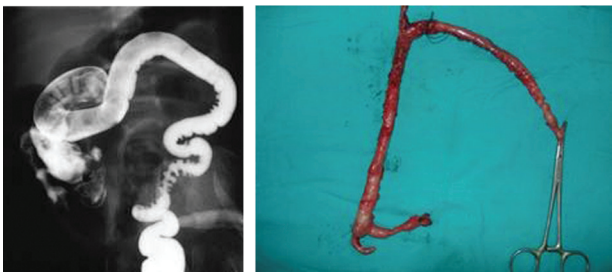
Table 1 shows patients' ages; the mean±SD age was 9.7±3.75.

Table 2 shows the number of female patients was nine (60%), whereas the number of male patients was six (40%).

Table 3 shows that there was a highly statistically significant difference as regards solid state at first, third, and sixth month, with the highest mean (2.5) or highest score at first month and the lowest mean (0.7) or lowest score at sixth month. This indicates that the patient improved progressively over months.

Table 4 shows that there was a highly statistically significant difference as regards fluid state at first,

Figure 1



Contrast enema of TCA and excised whole colon, appendix, and a part of the terminal ileum in TCA. TCA, total colonic aganglionosis.

Table 1 Mean±SD of age (months)

Mean	9.7333
Median	9.0000
SD	3.75056
Minimum	4.00
Maximum	18.00

Table 2 Frequency of male and female

	Frequency [*n* (%)]
Sex	
Female	9 (60.0)
Male	6 (40.0)
Total	15 (100.0)

Table 3 Mean±SD of solid status of incontinence

Types	Mean±SD	*P*
Solid first month	2.5333±0.99043	0.00**
Solid third month	1.6000±0.63246	
Solid first month	2.5333±0.99043	0.00**
Solid sixth month	0.7333±0.59362	
Solid third month	1.6000±0.63246	0.00**
Solid sixth month	0.7333±0.59362	

*Highly significant.

Table 4 Mean±SD of fluid status of incontinence

Types	Mean±SD	*P*
Fluid first month	3.3333±0.81650	0.00**
Fluid second month	2.4000±0.91026	
Fluid first month	3.3333±0.81650	0.00**
Fluid sixth month	1.2667±0.79881	
Fluid third month	2.4000±0.91026	0.00**
Fluid sixth month	1.2667±0.79881	

*Highly significant.

third, and sixth month, with the highest mean (3.3) or highest score at first month and the lowest mean (1.26) or lowest score at sixth month. This indicates that the patient improved progressively over months.

Table 5 shows that there was a highly statistically significant difference as regards gaseous state at first, third, and sixth month, with the highest mean (2.6) or highest score at first month and the lowest mean (1.4) or lowest score at sixth month. This indicates that the patient improved progressively over months.

Table 6 shows that there was a highly statistically significant difference as regards diaper state at first, third, and sixth month, with the highest mean (3.4) or highest score at first month and the lowest mean (1.4) or lowest score at sixth month. This indicates that the patient improved progressively over months.

Table 7 shows that there was a highly statistically significant difference as regards the score at first, third, and sixth month, with the highest mean [13] or highest score at first month and the lowest mean (5.8) or lowest score at sixth month. This indicates that the patient improved progressively over months (Figs 2 and 3).

Discussion

Total colonic agangliosis is a complex form of HD and requires good surgical strategies to achieve accepted functional results.

The aim of surgery for treating TCA is to excise the entire colon and the variable length of the terminal ileum, and then reconstruct bowel continuity.

The use of pouch or patch reconstructive procedures are associated with more complications (leakage, fistula, intestinal adhesion multiple reoperations, continence, and/or growth disorders).

Total colonic agangliosis remains a serious surgical challenge. Patients suffering from the condition have

Table 5 Mean±SD of gaseous status of incontinence

Types	Mean±SD	*P*
Gases first month	2.6000±0.82808	0.006*
Gases third month	2.0667±0.88372	
Gases first month	2.6000±0.82808	0.00**
Gases sixth month	1.4000±0.63246	
Gases third month	2.0667±0.88372	0.001**
Gases sixth month	1.4000±0.63246	

*Highly significant.

Table 6 Mean±SD of diaper status of incontinence

Types	Mean±SD	*P*
Diaper first month	3.4000±0.73679	0.001**
Diaper third month	2.6000±0.82808	
Diaper first month	3.4000±0.73679	0.00**
Diaper sixth month	1.4000±0.63246	
Diaper third month	2.6000±0.82808	0.00**
Diaper sixth month	1.4000±0.63246	

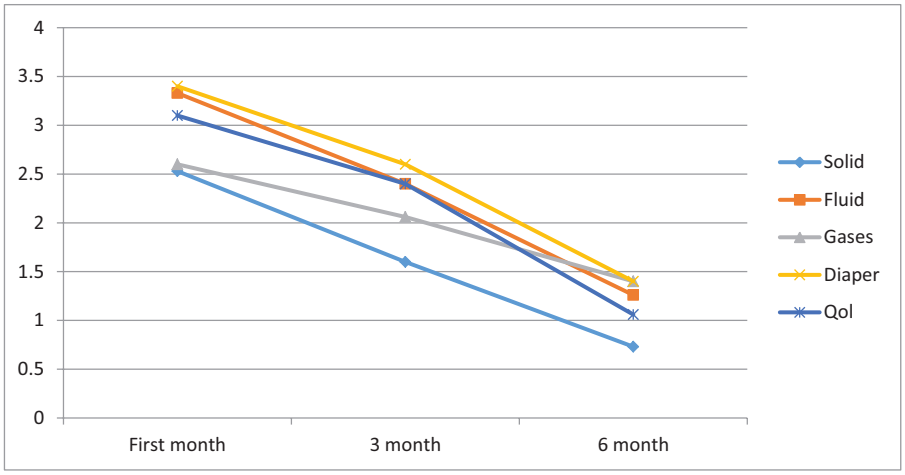
*Highly significant.

Table 7 Mean±SD of score of incontinence

Types	Mean±SD	*P*
First month score	15.0667±2.52039	0.00**
Third month score	11.0667±3.28344	
First month score	15.0667±2.52039	0.00**
Sixth month score	5.8667±2.82506	
Third month score	11.0667±3.28344	0.00**
Sixth month score	5.8667±2.82506	

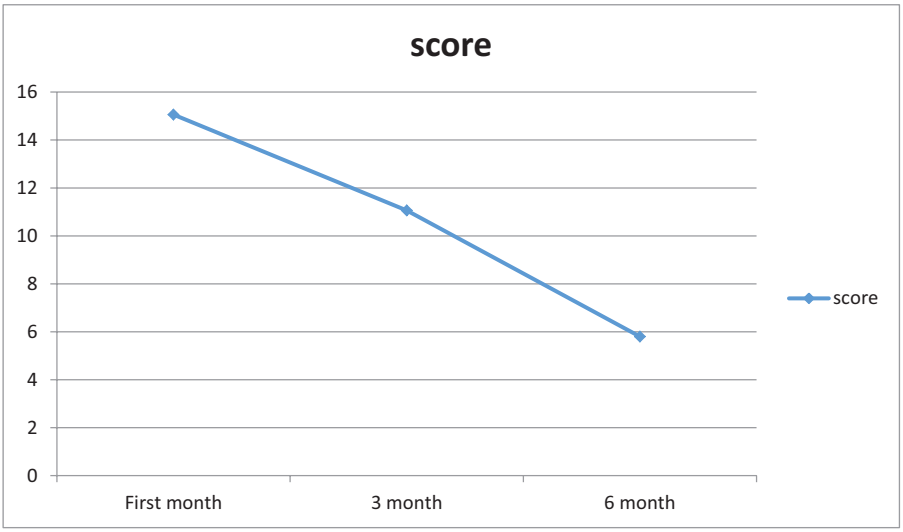
*Highly significant.

Figure 2



The five items of Wexner score with the noticeable changes through the period of 6 months postoperatively.

Figure 3



The mean score of all patients at different time evaluations; the decline of mean results from 14 to 5 indicates the improvement in anorectal functions with time.

multiple complications, sequelae, and often require reoperations. Yeh *et al.* [15] found that it is possible to prevent many of these by properly fixing the stoma, avoiding pouch or patch procedures, delaying ileostomy closure, having pathology expertise, and with meticulous surgical technique by starting the dissection/anastomosis well above the dentate line.

Hoehner *et al.* [16] mentioned that acceptable long-term outcome was most frequent in TCA patients whose definitive repair did not incorporate an extended ganglionic–aganglionic common channel. The use of extensive lengths of aganglionic bowel to maximize fluid absorption is frequently met with substantial morbidity [16].

Ross *et al.* [17] followed up 12 children who underwent Martin's procedure with a 0% mortality rate and an 81.8% morbidity rate. This study would indicate, as do others, that, even though the Martin procedure can safely be performed, the long-term results require close evaluation. A re-evaluation of this procedure and its alternatives is necessary to improve long-term results [17].

In our series, all infants underwent transanal pull-through and ileoanal anastomoses without patch or pouch.

No leakage or fistula was recorded postoperatively, but the expected complications were anorectal function, nutritional, and growth disorders.

We carried out continence evaluation by applying continence score (Wexner score) to all operated cases at 1, 3, and 6 months; the results were encouraging with time. Immediately the continence is disturbed that is clear with high score which gradually decrease through third and sixth month's evaluation.

After 6 months of evaluation of continence status, none of our patients recorded complete continence result (0 score) but the gradual improvement reflects the importance of time factor for infants who underwent total colectomy and ileoanal anastomoses to achieve continence.

On reviewing the literature, we did not find works with functional evaluation of anorectum after total colectomy for TCA using the Wexner score to be compared with our work.

Yeh and colleagues evaluated nine infants who were operated using the modified Duhamel procedure. The follow-up was for 1 year. Three of the five older patients who were available for follow-up reported normal bowel control, and the remaining two soiled occasionally. They had an average of 3–4 loose bowel movements per day. A stool score was applied to patients who were older than 3 years of age. All patients had good overall functional outcomes; the mean functional outcome score was 12.8 ± 0.84 out of a total score of 16 points, in the range indicating 'good' objective functional outcome.

Conclusion

Although ileoanal anastomoses after total colectomy for TCA lacking reservoir colonic stage that modify defecation mechanism and associated with high

frequency rate, soiling or even continence troubles related to excreted ileal contents directly, yet it is believed that with nutritional and electrolyte support no major surgical complications occur and defecation process gaining improvement with time and continence score improves with time.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

References

1 Wildhaber BE, Teitelbaum DH, Coran AG. Total colonic Hirschsprung's disease: a 28-year experience. J Pediatr Surg 2005; 40:203–207.

2 Coran AG, Teitelbaum DH. Recent advances in the management of Hirschsprung's disease. Am J Surg 2000; 180:382–387.

3 Marquez TT, Acton RD, Hess DJ, Duval S, Saltzman DA. Comprehensive review of procedures for total colonic aganglionosis. J Pediatr Surg 2009; 44:257–265.

4 Hoehner C, Ein SH, Shandling B, Kim PC. Long-term morbidity in total colonic aganglionosis. J Pediatr Surg 1998; 33:961–965.

5 Martin LW. Surgical management of total colonic aganglionosis. Ann Surg 1972; 176:343–346.

6 Tsuji H, Spitz L, Kiely EM, Drake DP, Pierro A. Management and long-term follow-up of infants with total colonic agangliosis. J Pediatr Surg 1999; 34:158–161.

7 Goto S, Gunter M, Scherer LR, Bloch T, Grosfeld JL. Surgical treatment of total colonic agangliosis: efficacy of aganglionic patch enteroplasty in the rat. J Pediatr Surg 1986; 21:601–607.

8 Escobar MA, Grosfeld JL, West KW, Scherer LR, Rouse TM, Engum SA, *et al.* Long-term outcomes in total colonic aganglionosis: a 32-year experience. J Pediatr Surg 2005; 40:955–961.

9 Martin LW, Altemeier WA. Clinical experience with a new operation (modified Duhamel procedure) for Hirschsprung's disease. Ann Surg 1962; 156:678–681.

10 Martin LW, Caudill DR. A method for elimination of the blind rectal pouch in the Duhamel operation for Hirschsprung's disease. Surgery 1967; 62:951–953.

11 Emslie J, Krishnamoorthy M, Applebaum H. Long-term follow-up of patients treated with ileoendorectal pull-through and right colon onlay patch for total colonic agangliosis. J Pediatr Surg 1997; 32:1542–1544.

12 Shen C, Song Z, Zheng S, Xiao X. A comparison of the effectiveness of the soave and Martin procedures for the treatment of total colonic agangliosis. J Pediatr Surg 2009; 44:2355–2358.

13 Khaleghnejad A, Foroutan H, Banani A, Mohajerzadeh L, Ruozrokh M, Mirshemirani A. State's pull-through for total colonic agangliosis and GI dismotility. Arch Iran Med 2013; 16:277–280.

14 Jorge JM, Wexner SD. Etiology and management of fecal incontinence. Dis Colon Rectum 1993; 36:77–97.

15 Yeh YT, Tsai HL, Chen CY, Wang JB, Chin TW, Wei CF, *et al.* Surgical outcomes of total colonic agangliosis in children: a 26-year experience in a single institute. J Chin Med Assoc 2014; 77:519–523.

16 Hoehner JC, Ein Barry Shandling SH, Kim PC. Long-term morbidity in total colonic agangliosis. J Pediatr Surg 1998; 33:961–966.

17 Ross MN, Chang JH, Burrington GD. Complications of the martin procedure for total colonic agangliosis. J Pediatr Surg 1988; 23:725–727.