Pediatr Surg Int (2012) 28:773–779
DOI 10.1007/s00383-012-3117-3

ORIGINAL ARTICLE

Total colonic aganglionosis: a systematic review and meta-analysis of long-term clinical outcome

Danielle Mc Laughlin · Florian Friedmacher · Prem Puri

Published online: 28 July 2012

Abstract
Purpose Total colonic aganglionosis (TCA) is a severe form of Hirschsprung's disease (HD), occurring in less than 10 % of the cases. It is a challenging surgical condition and outcomes of pull-through (PT) surgery are reported to be inferior to those in patients with recto-sigmoid HD. As even large centres only see a few patients with TCA, there is little information on the long-term outcome of patients after PT operation for TCA. The aim of this meta-analysis was to investigate the long-term clinical outcome in patients with TCA.
Methods MEDLINE® and EMBASE® databases were searched for relevant articles that reported the outcomes of patients with TCA published between 1980 and 2011. The search terms were "Hirschsprung's disease", "Total colonic aganglionosis" AND "Outcome". All published studies containing adequate clinical data for a mean follow-up period of not less than 4 years were included. Reference lists of retrieved articles were reviewed for additional cases. Detailed records of morbidity and mortality were extracted and analysed.
Results This search yielded 225 articles reporting on outcomes in TCA. Of these, 189 were excluded for having too short a follow-up period, small or single case series, inadequate clinical data and duplicated patient groups. Ultimately, 36 articles from 37 centres containing useful clinical information on the outcomes of TCA in 969 patients were identified. There were 152 early deaths prior to PT (15.7 %). Of 817 survivors, 739 underwent PT. The mortality rate for TCA post-PT was 5.7 %. The most frequently reported post-operative complication was enterocolitis in 42 % of the cases. 17.5 % of patients underwent subsequent major surgery including redo PT, stoma reformation or other laparotomy. Long-term follow-up data were available in 396 patients. Satisfactory or normal bowel control was reported in 60 % of the patients. Soiling, faecal incontinence or other poor outcome was reported in 33.5 % of the cases and 6.5 % of the patients had undergone conversion to a permanent ileostomy for post-operative complications.
Conclusion This meta-analysis reveals that a large number of patients with TCA have long-term problems with bowel control.

Keywords Total colonic aganglionosis · Total colonic Hirschsprung's disease · Long-term outcome · Systematic review · Meta-analysis

Introduction

Total colonic aganglionosis (TCA) is a severe form of Hirschsprung's disease (HD) characterised by the absence of ganglion cells in the entire colon extending into the terminal ileum. This relatively rare condition occurs in less than 10 % of the HD cases [1, 2]. Despite recent advances in surgical techniques and meticulous pre- and post-operative management, TCA remains a challenging surgical condition. For a long time, it has been thought that the length of aganglionosis in HD negatively impacts the long-term functional outcome [3] and that results of pull-through (PT) surgery for TCA are inferior to those in patients with

D. M. Laughlin · F. Friedmacher · P. Puri (✉)
National Children's Research Centre, Our Lady's Children's Hospital, Crumlin, Dublin 12, Ireland
e-mail: prem.puri@ucd.ie

D. M. Laughlin
Children's University Hospital, Temple Street, Dublin 1, Ireland

recto-sigmoid HD [4, 5]. Few studies have evaluated the long-term clinical outcome and bowel function in patients with TCA. Even in large centres, only a few patients with TCA may be treated over the course of a year, and therefore, only studies with limited numbers of patients are reported in the literature. The lack of large multi-centre studies for TCA has resulted in conflicting reports regarding the long-term results of surgery. The aim of this study was to investigate the long-term clinical outcome of patients with TCA based on the meta-analysis of the published literature.

Methods

A systematic literature search was performed to identify articles that reported the outcomes of patients with TCA. MEDLINE®(1966–present) and EMBASE®(1980–present) electronic databases were searched in April 2012 using a combination of the following terms: "Hirschsprung's disease", "Total colonic aganglionosis" AND "Outcome". References lists of identified articles were screened for additional publications of interest. All identified articles were independently assessed by each of the three authors. Only retrospective or prospective studies published between 1980 and 2011 with a minimum mean follow-up of 4 years from the time of PT surgery were eligible for inclusion. Series referring to results of already selected cohorts were excluded. Reports on patients selected for specific complications or anomalies were excluded to avoid selection bias and confounding towards a poorer outcome.

Detailed data regarding study design, patient characteristics, procedure type and outcomes were extracted into an electronic datasheet in a standardised manner. Outcome parameters of patients with TCA included mortality rates, post-operative morbidities (immediate and late complications) and long-term outcomes. Patients were considered to have a good long-term outcome when their bowel function was described as "normal", "satisfactory" or "continent" without soiling or need for washouts/dilations to achieve stooling. Conversely, patients with soiling, incontinence or permanent stoma were considered to have a poor functional outcome along with patients who required long-term medications, washouts/dilations or had a high frequency of bowel motions (>5/day). Data for each outcome category were not available in every paper. Outcome incidence is given per total number of patients reported on in that category.

Statistical analysis of contingency tables comparing outcomes between two groups formed on the basis of publication date, and comparing outcomes for three groups with different follow-up lengths, was performed with Fisher's exact test using GraphPad Prism 5.0 (GraphPad Software Inc., La Jolla, USA). A p value <0.05 was considered as statistically significant.

Results

The literature search produced 225 articles, of which 189 were excluded because they did not fulfil the inclusion criteria (Fig. 1). Ultimately 36 articles, published between 1982 and 2011, were included in this analysis [3, 6–41]. Data from 969 individual cases of TCA from 37 different centres worldwide was collected. In 27 articles, 804 cases of TCA were reported from a total of 8,818 patients with HD; therefore, the incidence of HD in which TCA was present was 9.1 %. Gender was reported in 859 cases, 559 were male and 300 females, determining a male-to-female ratio of 1.86:1. The mean follow-up period for all studies was 9.6 years. The earliest study commenced in 1954 and the longest study period was 35 years. The three largest cohorts all originated from Japan and employed national surveys of surgeons [8, 26, 32]. All other studies used retrospective methodology with some employing postal questionnaires, clinical review or interview to generate follow-up data. The largest study reporting long-term functional outcome involved 58 patients with TCA from Ireland and Italy [9]. A comprehensive list of the articles is given in Table 1. Details of the type of PT surgery performed were available in 722 patients. The most common operation for TCA was Martins' procedure in 230 cases closely followed by Duhamel procedure in 229 cases. Straight ileo-anal anastomosis was performed in 197 cases (Soave in 113, Swenson in 41 and not defined in 43). In 66 patients, other procedures performed included Rehbein, Kimura and various patch graft procedures.

The overall mortality rate was 193/969 (19.9 %). The pre-PT mortality rate was 152/969 (15.7 %), and of those who had PT, 42/739 (5.7 %) died postoperatively (excluding 3 unrelated deaths). A total of 543 complications occurred after PT, the most frequent being enterocolitis arising in 42 % of the patients (248/592). Table 2 presents a breakdown of morbidity rates post-PT. Data on long-term functional outcome were reported in a total of 396 patients post-definitive surgery for TCA. A patient distribution flowchart is given in Fig. 2 showing long-term outcomes. Between two publication periods, there was a significant decrease in overall mortality and the number of patients undergoing PT increased, but long-term functional outcome was not significantly different (Table 3). However, a significant improvement in the long-term functional outcome was detected between groups followed-up at 4–7.9 and 8–11.9 years (Fig. 3).

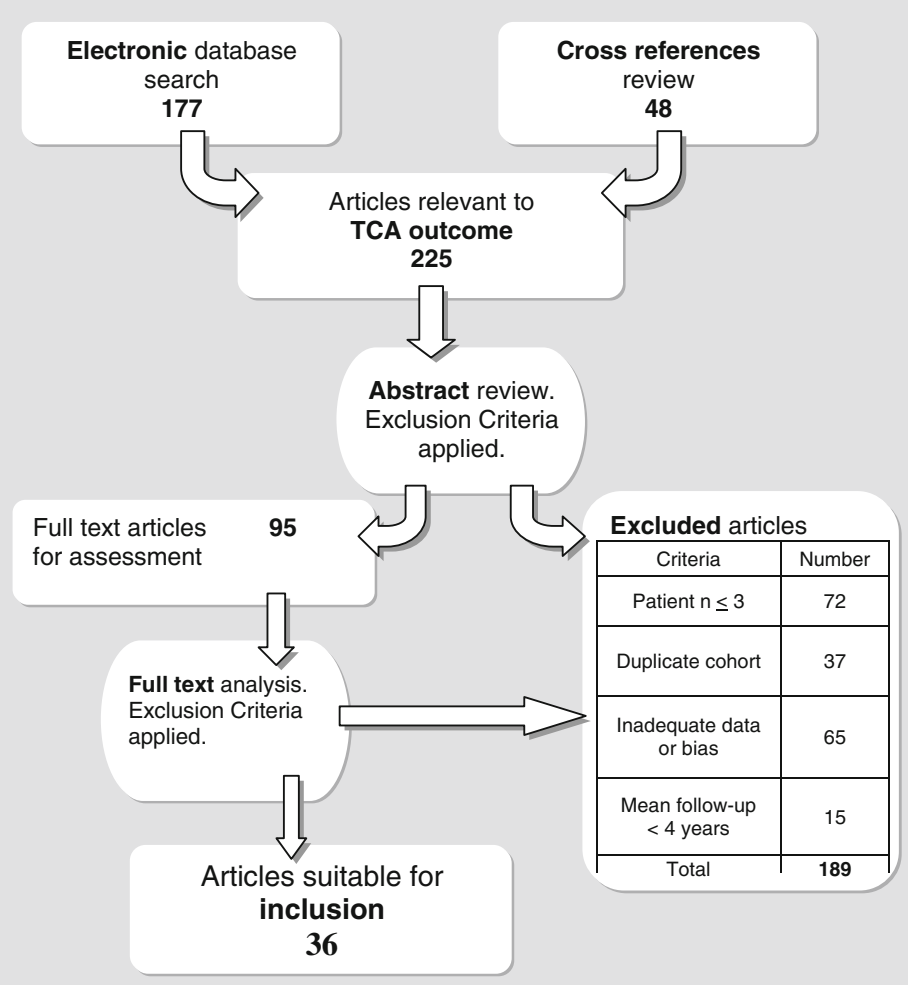


Fig. 1 Literature search flow chart

Discussion

In the present study, we found a TCA incidence of 9.1 % of the total HD cases, which is comparable to previous studies [2]. For the first time, the gender distribution in a large group of TCA patients is shown. HD has a male preponderance of approximately 4:1 [42]; however, our results confirm that TCA differs from recto-sigmoid HD for gender ratio supporting previous evidence that in TCA the male-to-female ratio approaches 2:1 [32].

Mortality rates for TCA are high both pre- and post-definitive PT surgery. In a 40-year multi-centre follow-up of 2,430 HD patients post-Duhamel surgery, an overall mortality rate of 1.6 % was found [43]. The post-PT mortality rate in TCA in the present study was higher at 5.7 %. The overall mortality rates have improved significantly for TCA between an early and a more recent group of cases. Several factors are likely to influence this trend. In the early group, many patients were treated prior to the era of intravenous nutritional support, when PT surgery was in its infancy [11, 15, 44]. The first successful surgical procedure for TCA was described by Sandegard in 1953 [45]. The earliest cases included herein date from this time period. Total parenteral nutrition came into use during the 1970s and, with improvements in intensive care for neonates, has had a diminishing effect on mortality in TCA.

Good functional outcome in recto-sigmoid HD varies depending on the procedure, but can be expected in approximately 90 % of the cases [46, 47]. Long-term bowel function following PT operation in patients with TCA only achieves a good outcome in 60 % of the cases. Faecal incontinence and soiling severely impact the quality of life [48, 49], and are seen in 25 % of the patients at long-term follow-up. This rate did not show significant improvement over time. Faecal continence rates are more satisfactory for HD as compared to TCA. In a large survey of the members of the surgical section of the American Academy of Paediatrics examining the outcomes of 1,196 operated HD patients, a long-term soiling rate of 7.5 % was reported [37]. The incidence of TCA patients suffering from long-term soiling or incontinence reported in the articles included in the present study varied from 6 to 86 %

Table 1 Articles in order of year of publication

Reference	Years	Method	Period	Mean follow-up	Patients	Overall mortality		PT done		Good outcome	
				Years	*n*	*n*	%	*n*	%	*n*/total[a]	%
Travassos et al. [7]	2011	R	1988–2010	11.5	15	2	13.3	15	100	6/12	50
Bischoff et al. [10]	2011	R	n/a	9	27	0	0	27	100	15/19	79
Ieiri et al. [8]	2008	S	1998–2002	n/a	101	16	15.8	83	82.2	n/a	n/a
Menezes et al. [9]	2008	R, I	1975–2005	12.5	58	4	6.9	56	96.5	22/45	49
Barrena et al. [12]	2008	R, I	1972–2007	18	35	4	11.4	33	80.5	21/31	42
Choe et al. [16]	2007	R, I	1985–2004	6.2	17	6	35.3	11	64.1	1/7	14
Mirshemirani et al. [29]	2007	R	1990 –2005	7.5	5	1	20	4	80	4/4	100
Escobar et al. [20]	2005	R	1972–2004	11	36	7	19.4	29	80.5	17/23	74
Wildhaber et al. [34]	2005	R, Q	1974–2002	17.5	25	3	12	20	88	9/18	50
Fouquet et al. [22]	2002	R	1980–1999	8.5	26	2	15.4	18	69.2	11/15	73
Dodero et al. [17]	2001	R	1974–2000	8.5	24	0	0	24	100	21/24	88
Tsuji et al. [36]	1999	R	1980–1996	15	48	3	6.3	41	85.4	13/26	50
Baillie et al. [35]	1999	R, I, Q	1980–1991	8.4	11	0	0	11	100	6/10	60
Hoehner et al. [25]	1998	R	1969–1996	6.6	29	1	3.4	27	93.1	9/23	39
Nishijima et al. [31]	1998	R	1979–1997	12	11	1	9.1	10	90.9	5/9	56
Suita et al. [32]	1997	S	1988–1992	n/a	107	20	18.7	71	71	n/a	n/a
Marty et al. [38]	1997	R, Q	1971–1993	7.9	16	1	6.3	13	81.3	5/12	42
Emslie et al. [18]	1997	R, I, Q	1985–1994	7.8	5	0	0	5	100	3/5	60
Azzis et al. [11]	1996	R	1971–1994	12	16	6	37.5	10	62.5	4/7	57
Hengster et al. [24]	1996	R, I, C	1965–1993	23	12	7	58.3	6	50	3/5	60
Fortuna et al. [21]	1996	R. Q	1975–1994	9.4	5	0	0	5	100	3/5	60
Heij et al. [3]	1995	R, Q	1977–1991	5.5	9	2	22.2	9	100	1/7	14
Endo et al. [19]	1994	R	n/a	11	9	0	0	9	100	4/7	57
Klein et al. [27]	1993	R	1960–1991	4.25	29	13	44.8	19	65.5	n/a	n/a
Bickler et al. [39]	1992	R, C, I	1960–1991	10.4	21	1	4.8	16	76.2	5/6	83
Foster et al. [6]	1990	R	1955–1980	8	9	2	22.2	9	100	n/a	n/a
Bergmeijer et al. [13]	1989	R	1979–1985	4	9	3	33.3	6	66.7	3/5	60
Carcassonne et al. [14]	1989	R	1977–1986	6	7	0	0	7	100	7/7	100
Ross et al. [40]	1988	R	1976–1988	4	16	2	12.5	14	87.5	2/11	17
Festen et al. [41]	1988	R	1974–1987	6.7	11	2	18.2	10	90.9	7/9	78
Cass et al. [15]	1987	R	1954–1983	9.2	32	15	46.9	17	53.1	7/12	50
Galifer et al. [23]	1987	R	1971–1984	8.8	6	1	16.7	6	100	4/5	80
Ikeda et al. [26]	1986	S	1978–1982	n/a	137	56	40.9	67	48.9	n/a	n/a
Nfékété et al. [30]	1986	R	1960–1984	6	27	11	40.7	14	51.8	7/12	58
Weitzman [33]	1986	R, I	1965–1985	14.4	5	0	0	5	100	3/5	60
Martin [28]	1982	R	1960–1981	5	13	1	7.7	12	92.3	8/10	80
			All articles	9.6	969	193	20	739	76	236/396	60

R Retrospective, *S* Survey, *Q* Patient Questionnaire, *I* Interview, *C* Clinical assessment, *n/a* not available

[a] Total is number of patients in whom long-term outcome is determined

[3, 7, 12]. The overall rate of poor bowel function in operated TCA cases is 40 % when other outcomes such as conversion to permanent stoma, high stool frequency and dependence on medications, rectal washouts or dilations are factored in. Sherman et al. [50] reported that 10 (1.3 %) of 797 patients with HD had undergone conversion to a permanent stoma for post-operative complications. In the present study, the incidence of patients with TCA who opted for a permanent stoma for complications after PT operation is much higher at 6.5 %. This figure excludes 22 surviving patients who did not undergo PT and were given a long-term stoma as management and an additional 11 patients who were reported to have a temporary or diverting stoma at long-term review. Several investigators

Table 2 Post-PT complications

Morbidity	*n*/total	%
Enterocolitis	248/592	41.9
Major re-operation (redo PT, unplanned stoma, other laparotomy)	89/509	17.5
Other re-operation (sphincterectomy, myectomy, anal dilation under GA, spur resection)	74/509	14.5
Anastomotic breakdown (leak, abscess, fistula, peritonitis)	29/509	5.5
Other (wound infection, peri-anal excoriation, hernia, sepsis, stricture)	103/509	20.2
Total	543	

have shown that continence post-PT improves over time [5, 50–52]. We have found that outcome is significantly better in patients at a mean follow-up of 8–11.9 years when compared to a group at 4–7.9 years. This trend did not persist in the group followed-up for longer than 12 years and was in fact reversed. The earliest cases included in this study were managed in the 1950s and 1960s, during a time of rapid development in the field of HD surgery, we therefore feel that the older group of patients reported here may not have had the advantages of modern surgical techniques and bowel management programs.

Many patients underwent Martins' modification of the Duhamel operation for TCA, however 85% of these cases dated pre-1996. This procedure was designed to increase

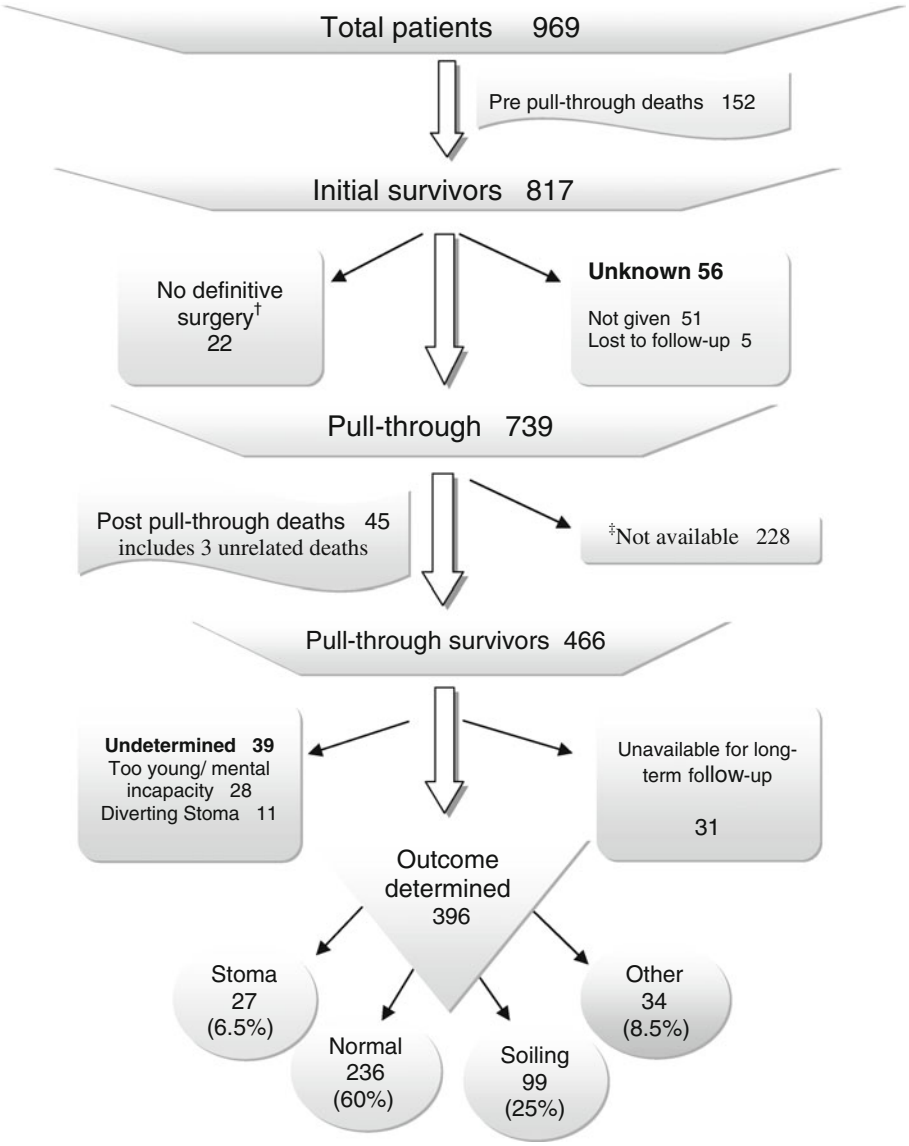


Fig. 2 Patient distribution flowchart showing long-term outcomes

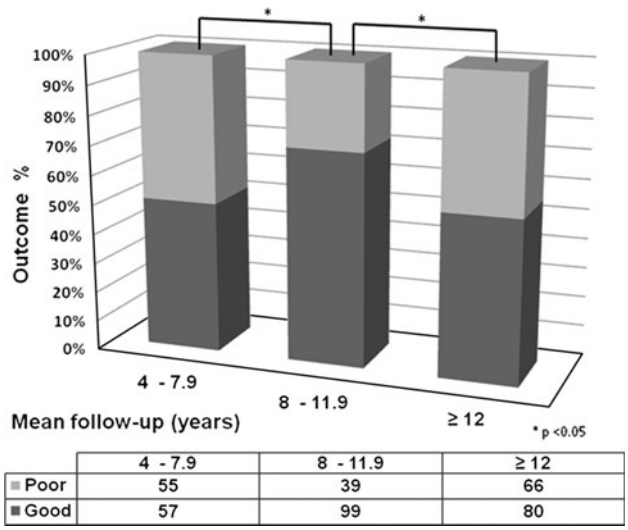
† Surviving patients in whom PT was not done

‡ This figure represents the number of PT survivors in articles which did not report on long-term clinical outcome

Table 3 Comparison of two patient groups based on the publication period

Published between	1982–1996	1996–2011
Number of studies	18	18
Patients	373	596
Overall mortality	122/373 (32.7 %)	71/596 (11.9 %)*
Pull-through done	241/373 (64.6 %)	498/596 (83.5 %)*
Good long-term outcome	170/290 (58.6 %)	66/106 (62.26 %)

* $p < 0.0001$



	4 - 7.9	8 - 11.9	≥ 12
Poor	55	39	66
Good	57	99	80

Fig. 3 Comparison of long-term functional outcome by follow-up period

absorption using a long side-to-side anastomosis of the ganglionic ileum to aganglionic colon. Most surgeons have had less satisfactory outcomes with the Martin long Duhamel operation and have abandoned this procedure in recent years in favour of the standard modified Duhamel operation [20].

Enterocolitis after PT procedure was the most common post-operative complication occurring in 42 % of the patients. Many authors have reported that an increasing length of aganglionosis is associated with higher risk of enterocolitis [53, 54] and our results would concur with this. In a literature review, Little and Synder [55] found an incidence of enterocolitis of 10.6 % in 10,381 patients with HD and TCA outcomes compare poorly with this.

In conclusion, this meta-analysis reveals that a large number of patients with TCA have long-term problems with bowel control. Although some improvements have been seen at a longer duration of follow-up, continence rates have not improved over the last 3 decades and many patients with TCA have poor long-term functional results following PT.

References

1. Ryan ET, Ecker JL, Christakis NA, Folkman J (1992) Hirschsprung's disease: associated abnormalities and demography. J Pediatr Surg 27(1):76–81
2. Singh SJ, Croaker GD, Manglick P, Wong CL, Athanasakos H, Elliott E, Cass D (2003) Hirschsprung's disease: the Australian Paediatric Surveillance Unit's experience. Pediatr Surg Int 19(4):247–250
3. Heij HA, Devries X, Bremer I, Ekkelkamp S, Vos A (1995) Long-term anorectal function after Duhamel operation for Hirschsprung's disease. J Pediatr Surg 30(3):430–432
4. Ludman L, Spitz L, Tsuji H, Pierro A (2002) Hirschsprung's disease: functional and psychological follow up comparing total colonic and rectosigmoid aganglionosis. Arch Dis Child 86(5):348–351
5. Rescorla FJ, Morrison AM, Engles D, West KW, Grosfeld JL (1992) Hirschsprung's disease. Evaluation of mortality and long-term function in 260 cases. Arch Surg 127(8):934–941 discussion 941-932
6. Foster P, Cowan G, Wrenn EL Jr (1990) Twenty-five years' experience with Hirschsprung's disease. J Pediatr Surg 25(5):531–534
7. Travassos DV, van der Zee DC (2011) Is complete resection of the aganglionic bowel in extensive total aganglionosis up to the middle ileum always necessary? J Pediatr Surg 46(11):2054–2059
8. Ieiri S, Suita S, Nakatsuji T, Akiyoshi J, Taguchi T (2008) Total colonic aganglionosis with or without small bowel involvement: a 30-year retrospective nationwide survey in Japan. J Pediatr Surg 43(12):2226–2230
9. Menezes M, Prato AP, Jasonni V, Puri P (2008) Long-term clinical outcome in patients with total colonic aganglionosis: a 31-year review. J Pediatr Surg 43(9):1696–1699
10. Bischoff A, Levitt MA, Pena A (2011) Total colonic aganglionosis: a surgical challenge. How to avoid complications? Pediatr Surg Int 27(10):1047–1052
11. Azzis O, Fremond B, Dabadie A, Jouan J, Bracq H, Babut JM (1996) Total colonic form of Hirschsprung disease. Treatment and long-term follow-up in 16 cases. Chirurgie 121(3):225–229 discussion 230
12. Barrena S, Andres AM, Burgos L, Luis AL, Hernandez F, Martinez L, Lopez-Santamaria M, Lassaletta L, Tovar JA (2008) Long-term results of the treatment of total colonic aganglionosis with two different techniques. Eur J Pediatr Surg 18(6):375–379
13. Bergmeijer JH, Tibboel D, Molenaar JC (1989) Total colectomy and ileorectal anastomosis in the treatment of total colonic aganglionosis - a long-term follow-up-study of 6 patients. J Pediatr Surg 24(3):282–285
14. Carcassonne M, Guys JM, Morrison-Lacombe G, Kreitmann B (1989) Management of Hirschsprung's disease: curative surgery before 3 months of age. J Pediatr Surg 24(10):1032–1034
15. Cass DT, Myers N (1987) Total colonic aganglionosis: 30 years' experience. Pediatr Surg Int 2:68–75
16. Choe EK, Moon SB, Kim HY, Lee SC, Park KW, Jung SE (2008) Outcomes of surgical management of total colonic aganglionosis. World J Surg 32(1):62–68
17. Dodero P, Magillo P, Scarsi RL (2001) Total colectomy and straight ileo-anal soave endorectal pull-through: personal experience with 42 cases. Eur J Pediatr Surg 11(5):319–323
18. Emslie J, Krishnamoorthy M, Applebaum H (1997) Long-term follow-up of patients treated with ileoendorectal pull-through and right colon onlay patch for total colonic aganglionosis. J Pediatr Surg 32(11):1542–1544

19. Endo M, Watanabe K, Fuchimoto Y, Ikawa H, Yokoyama J (1994) Long-term results of surgical-treatment in infants with total colonic aganglionosis. J Pediatr Surg 29(10):1310–1314
20. Escobar MA, Grosfeld JL, West KW, Scherer LR, Rouse TM, Engum SA, Rescorla FJ (2005) Long-term outcomes in total colonic aganglionosis: a 32-year experience. J Pediatr Surg 40(6):955–961
21. Fortuna RS, Weber TR, Tracy TF, Silen ML, Cradock TV (1996) Critical analysis of the operative treatment of Hirschsprung's disease. Arch Surg 131(5):520–524
22. Fouquet V, DeLagausie P, Faure C, Bloch J, Malbezin S, Ferkhadji L, Bauman C, Aigrain Y (2002) Do prognostic factors exist for total colonic aganglionosis with heal involvement? J Pediatr Surg 37(1):71–75
23. Galifer RB, Frapier JM, Bosc O, Massip A, Rieu D, Couture A, Veyrac C, Barneon G (1987) Total colonic aganglionosis without ileal involvement: an analysis of 6 cases. Chirurgie Pediatrique 28(3):137–144
24. Hengster P, Pernthaler H, Gassner I, Menardi G (1996) Twenty-three years of follow-up in patients with total colonic aganglionosis. Klin Padiatr 208(1):3–7
25. Hoehner JC, Ein SH, Shandling B, Kim PCW (1998) Long-term morbidity in total colonic aganglionosis. J Pediatr Surg 33(7):961–965
26. Ikeda K, Goto S (1986) Total colonic aganglionosis with or without small-bowel involvement: an analysis of 137 patients. J Pediatr Surg 21(4):319–322
27. Klein MD, Philippart AI (1993) Hirschsprung's disease: 3 decades experience at a single institution. J Pediatr Surg 28(10):1291–1294
28. Martin LW (1982) Total colonic aganglionosis preservation and utilization of entire colon. J Pediatr Surg 17(5):635–637
29. Mirshemirani A, Sadeghian N, Kouranloo J (2009) Long-segment aganglionosis: 15-year experience. Acta Medica Iranica 47(1):71–74
30. Nfekete C, Ricour C, Martelli H, Jacob SL, Pellerin D (1986) Total colonic aganglionosis (with or without ileal involvement): a review of 27 cases. J Pediatr Surg 21(3):251–254
31. Nishijima E, Kimura K, Tsugawa C, Muraji T (1998) The colon patch graft procedure for extensive aganglionosis: long-term follow-up. J Pediatr Surg 33(2):215–218
32. Suita S, Taguchi T, Kamimura T, Yanai K (1997) Total colonic aganglionosis with or without small bowel involvement: a changing profile. J Pediatr Surg 32(11):1537–1541
33. Weitzman JJ (1986) Management of Hirschsprung's disease with the Swenson procedure with emphasis on long-term follow-up. Pediatr Surg Int 1:100–104
34. Wildhaber BE, Teitelbaum DH, Coran AG (2005) Total colonic Hirschsprung's disease: a 28-year experience. J Pediatr Surg 40(1):203–206
35. Baillie CT, Kenny SE, Rintala RJ, Booth JM, Lloyd DA (1999) Long-term outcome and colonic motility after the Duhamel procedure for Hirschsprung's disease. J Pediatr Surg 34(2):325–329
36. Tsuji H, Spitz L, Kiely EM, Drake DP, Pierro A (1999) Management and long-term follow-up of infants with total colonic aganglionosis. J Pediatr Surg 34(1):158–161
37. Kleinhaus S, Boley SJ, Sheran M, Sieber WK (1979) Hirschsprung's disease—a survey of the members of the Surgical Section of the American Academy of Pediatrics. J Pediatr Surg 14(5):588–597
38. Marty TL, Seo T, Matlak ME, Sullivan JJ, Black RE, Johnson DG (1995) Gastrointestinal function after surgical-correction of Hirschsprung's disease: long-term follow-up in 135 patients. J Pediatr Surg 30(5):655–658
39. Bickler SW, Harrison MW, Campbell TJ, Campbell JR (1992) Long-segment Hirschsprung's disease. Arch Surg 127(9):1047–1050 discussion 1050-1041
40. Ross MN, Chang JHT, Burrington JD, Janik JS, Wayne ER, Clevenger P (1988) Complications of the martin procedure for total colonic aganglionosis. J Pediatr Surg 23(8):725–727
41. Festen C, Severijnen R, Vanderstaak F, Rieu P (1989) Total colonic aganglionosis—treatment and follow-up. Z Kinderchir 44(3):153–155
42. Orr JD, Scobie WG (1983) Presentation and incidence of Hirschsprung's disease. Br Med J (Clin Res Ed) 287(6406):1671
43. Bourdelat D, Vrsansky P, Pages R, Duhamel B (1997) Duhamel operation 40 years after: a multicentric study. Eur J Pediatr Surg 7(2):70–76
44. Casasa JM (1992) Total colonic aganglionosis. Cir Pediatr 5(2):75–77
45. Sandegard E (1953) Ovanliga Varianter Av Morbus Hirschsprung. Nordisk Med 50(32):1115
46. Erdek MA, Wilt E (1994) Hirschsprung's disease: one institution's ten year experience and long-term follow-up. Am Surg 60(8):625–628
47. Menezes M, Corbally M, Puri P (2006) Long-term results of bowel function after treatment for Hirschsprung's disease: a 29-year review. Pediatr Surg Int 22(12):987–990
48. Diseth TH, Bjornland K, Novik TS, Emblem R (1997) Bowel function, mental health, and psychosocial function in adolescents with Hirschsprung's disease. Arch Dis Child 76(2):100–106
49. Bai Y, Chen H, Hao J, Huang Y, Wang W (2002) Long-term outcome and quality of life after the Swenson procedure for Hirschsprung's disease. J Pediatr Surg 37(4):639–642
50. Sherman JO, Snyder ME, Weitzman JJ, Jona JZ, Gillis DA, Odonnell B, Carcassonne M, Swenson O (1989) A 40-year multinational retrospective study of 880 Swenson procedures. J Pediatr Surg 24(8):833–838
51. Heikkinen M, Rintala RJ, Louhimo I (1995) Bowel function and quality-of-life in adult patients with operated Hirschsprung's disease. Pediatr Surg Int 10(5–6):342–344
52. Yanchar NL, Soucy P (1999) Long-term outcome after Hirschsprung's disease: patients' perspectives. J Pediatr Surg 34(7):1152–1160
53. Elhalaby EA, Teitelbaum DH, Coran AG, Heidelberger KP (1995) Enterocolitis associated with Hirschsprung's disease: a clinical histopathological correlative study. J Pediatr Surg 30(7):1023–1026 discussion 1026–1027
54. Surana R, Quinn FM, Puri P (1994) Evaluating the risk factors in the development of enterocolitis complicating Hirschsprung's disease. Pediatr Surg Int 9:234–236
55. Little DC, Synder CL (2008) Early and late complications following operative repair of Hirschsprung's disease. In: Holschneider A, Puri P (eds) Hirschsprung's disease and allied disorders, 3rd edn. Springer, New York, pp 375–385