

Long-term results of total colonic aganglionsis patients treated by preservation of the aganglionic right hemicolon and the ileo-cecal valve

Eva E. Amerstorfer¹ · Günter Fasching² · Holger Till¹ · Andrea Huber-Zeyringer¹ · Michael E. Höllwarth¹

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Abstract

Purpose In 1989, Sauer introduced a technique to treat patients with total colonic aganglionsis (TCA) where he preserved the ileo-cecal valve and the right hemicolon to retain water and electrolyte absorption. This report examines the long-term outcome of patients who underwent this technique.

Methods All TCA patients treated between 1981 and 2005 according to Sauer were invited in 2013 to participate in a questionnaire survey to assess their long-term outcome focusing on bowel function and quality of life (QoL), using standardized scores.

Results Of eight TCA patients (2 females, 6 males), seven participated in the follow-up survey (median follow-up time 13.5 years (range 8–31.6)). Early postoperative complications involving bouts of enterocolitis in 3 and anal strictures in 5 patients ceased with age. Bowel movements ranged from median 3–4 times a day (range 1–8). Bowel-function score (BFS) was reduced in 6 patients (median 16 points, range 8–19, max 20), who also reported soiling incidences with the need for night-time protective aids in two. QoL assessment, however, documented a good outcome with a median score of 10 points (range 7–13, max 13).

Conclusion This long-term investigation of TCA patients treated according to Sauer's technique documented a good QoL despite a reduced BFS.

Keywords Total colonic aganglionsis · Sauer's procedure · Long-term study · Ileo-cecal valve · Bowel function · Quality of life

Introduction

Total colonic aganglionsis (TCA) represents a severe, rare disorder of the intestine, accounting for up to 12 % of patients suffering from Hirschsprung's disease (HD) [1]. It manifests clinically in most cases in the newborn period or early infancy by constipation and ileus or episodes of enterocolitis [1]. In contrast to classic rectosigmoid HD, in TCA the aganglionsis affects the whole colon and may extend to the lower ileum or even to the mid or proximal small bowel [2, 3]. Surgical treatment involves resection of the aganglionic colon and affected ileum. Total colectomy results in a highly increased stool frequency with fluid and electrolyte disturbances which are associated with considerable morbidity [2, 3].

To minimize the loss of fluids and electrolytes, various surgical techniques ranging from Martin's modification of Duhamel's retrorectal pull-through to Kimura's and Boley's colonic patch graft procedures have been proposed to partially retain the absorptive and reservoir functions of a part of the aganglionic colon [2, 4–8]. Furthermore, preservation of the aganglionic ileo-cecal valve (ICV) has been reported to be beneficial in the treatment of patients with TCA, as it acts as a natural barrier against bacterial backwash which could result in malabsorption of bile acids, vitamin B12 and fatty acids [9, 10]. In 1989, Sauer has introduced a technique which combines the advantages of preserving parts of the aganglionic colon and the aganglionic ICV to achieve better fluid and electrolyte absorption on the one hand and avoid bacterial backwash

✉ Eva E. Amerstorfer
ee.amerstorfer@gmail.com

¹ Department of Pediatric and Adolescent Surgery, Medical University of Graz, Auenbruggerplatz 34, 8036 Graz, Austria

² Department of Pediatric and Adolescent Surgery, Clinical Center Klagenfurt, Feschnigstrasse 11, 9020 Klagenfurt, Austria

on the other hand [9, 11]. Due to the limited numbers of patients, large experiences with the different surgical strategies are missing and long-term follow-up investigations are rare. Thus, the aim of this study was to investigate the long-term outcome of TCA patients treated by Sauer's technique.

Patients and methods

The hospital database research revealed that in total eight patients with TCA were treated from 1981 to 2005 at two tertiary care centers utilizing Sauer's method [11]. Basically, the technique represents a one-stage pull-through procedure performed after an ileostomy has been placed previously in a ganglionic area. Briefly, after appendectomy, a resection of the large bowel from the rectum to the ascending colon is performed and the aganglionic ileum is resected from the pertained ICV up to the ganglionic ileum. The latter is anastomosed to the ICV. An isolated ganglionic segment of the ileum is thereafter anastomosed side-to-side in an isoperistaltic manner to the 180° downward-rotated aganglionic right colon and finally, anastomosed to the anus (either transabdominally or after a transanal mucosectomy) (Fig. 1).

The hospital data of the identified patients were retrospectively reviewed for early and late postoperative complications, as well as relaparotomies. In 2013, the patients were invited either by telephone call, written or electronic means to participate in a long-term follow-up survey. Anthropometric details such as body weight and length, stool frequency and consistency, fecal incontinence, and information about their nutritional and subjective well-being were inquired. The study included a questionnaire to evaluate bowel function, using the "Bowel-function score" (BFS) published by Rintala and Lindahl [12] and a questionnaire to assess the quality of life, using the "Quality-of-Life Score" (QoLS), which has originally been established for children with fecal incontinence aged 8–16 years, published by Bai and colleagues [13], and has been adapted for our adult patients. The data were descriptively statistically evaluated using Microsoft Office Excel 2010®. Ethical approval was obtained from the local ethical committee (EK 25-189 ex 12/13).

Results

Eight patients (two females, six males) with TCA were operated according to Sauer's technique. The definite operation was conducted at various ages with a median of 9 months, ranging from the age of 1 to 65 months (Table 1).

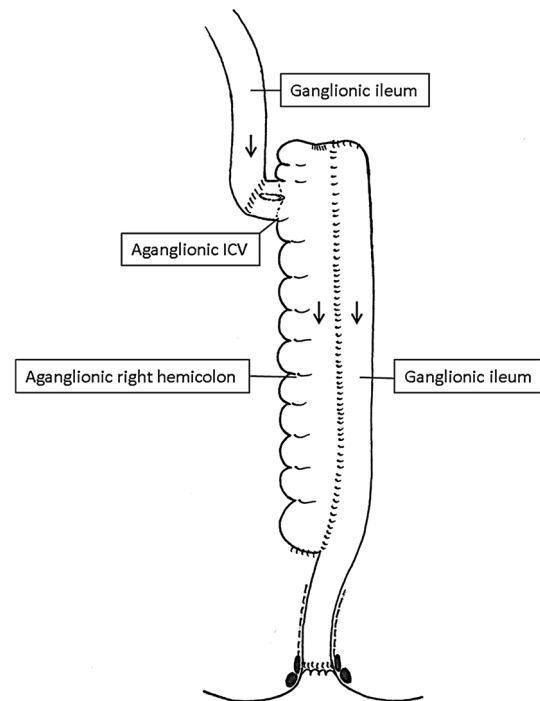


Fig. 1 After appendectomy and resection of the aganglionic ileum and the aganglionic rectum up to the *right* hemicolon, an isolated ganglionic segment of the ileum is anastomosed side-to-side isoperistaltically to the 180° downward-rotated aganglionic right hemicolon and then anastomosed to the anus. The distal end of the ganglionic ileum is anastomosed end-to-back to the ileo-cecal valve (ICV). In contrast to the original technique [11], a myotomy of the ICV had to be performed postoperatively in 2 patients and was therefore integrated in the drawing

Seven patients received primarily a diverting ileostomy, four of them in the neonatal period. One patient underwent a single-stage procedure at the age of 1 month without a prior ostomy. The first patient was operated in three stages and finally underwent a retrorectal pull-through of the ganglionic ileum according to Duhamel. All other patients underwent a single-stage procedure including the ileal pull-through and closure of the ileostomy. The pull-through was conducted according to Duhamel in two patients, whereas in six patients the ileum was anastomosed to the anal mucosa via transabdominal approach according to Soave ($n = 3$) or after transanal mucosectomy according to De la Torre ($n = 3$) (Table 1).

Two patients were primarily misdiagnosed for TCA. One of them was treated by laparoscopic-assisted transanal endorectal pull-through of the transverse colon according to De la Torre at the age of 2 months. The second patient underwent several surgical procedures including an ascendo-rectostomy pull-through according to Soave. Several relaparotomies were needed in both patients before the definite surgical procedure according to Sauer was conducted at the age of 25 months in one and at the age of 5 years in the other patient.

Table 1 General information of the TCA patients treated according to Sauer's technique

ID	Sex (male/female)	Age at pull-through (months)	Pull-through	Age at follow-up (years)	Relaparotomy (n)	Sphincter dilatation (n)	Enterocolitis (n)	Bowel movements min	Bowel movements max	Stool constancy	Soiling	Food intolerance	BMI (kg/m ²)	Bowel function score (max = 20)	Quality of life Score (max = 13)
1	M	16	Duhamel	33	0	9	3	3	4	Watery	Accidental	1	20	16	9
2	M	4	Soave	Lost to follow-up	1	2	0								
3	F	3	Soave	17/9/12	0	1	0	2	3	Semisolid	Accidental	1	22	14	10
4	M	4	Soave	14/8/12	4	0	1	4	5	Semisolid	Daily night-time	1	19.5	10	9
5	F	14	De la Torre	10/8/12	0	1	1	5	8	Watery/semisolid	Daily night-time	1	17	8	7
6	M	1	De la Torre	8/2/12	0	0	0	2	3	Semisolid	Accidental	0	13	17	12
7	M	25	De la Torre	11/1/12	1	2	Recurrent pouchitis	5	8	Watery/semisolid	Frequent	1	15	17	10
8	M	65	Duhamel	19	0	0	0	1	2	Watery/semisolid	Never	0	20	19	13

Early postoperative complications included stenosis of the aganglionic ICV in two patients requiring myotomy of the ICV. A re-laparotomy for ileus had to be performed in total in three patients (Table 1). An anal stricture occurred in five patients, who required median 2 dilatations (range 1–9 dilatations; Table 1). In one patient, the anastomotic stricture was resected transanally. A mild enterocolitis, including a recurrent intestinal candidiasis documented in one girl, occurred in three patients and was successfully managed conservatively (Table 1). One patient suffered from a recurrent pouchitis.

Follow-up (Tables 2, 3)

In 2013, seven patients agreed on participating in our long-term survey. One patient was lost to follow-up. The follow-up time ranged from 8 to 31 years (median 13.5 years). All investigated patients assured a good subjective well-being. The median calculated body mass index (BMI, kg body weight/body length m²) was 19.5, ranging from 13 to 22. While the youngest patients correlated with the lowest BMI values, patients above the age of 14 years (n = 4) presented with a normal age-adapted BMI value. All patients were on normal age-adapted oral feeds and had no special dietary requirements. Five patients, however, reported on selective food intolerances with avoidance of foods such as flatulent vegetables, legumes, and mushrooms. One of these patients was intolerant to lactose and fructose.

All patients assured the ability to defecate voluntarily. Bowel movements occurred from 1 to 8 times a day with a watery to semisolid consistency. Soiling never occurred in only one patient. Three patients reported from rare soiling incidences less than once a week, while one patient referred to soil frequently. Two patients aged 10 and 14 years at follow-up documented on the need for protective night aids for regular night-time soiling. The youngest TCA patient, aged 8 years at follow-up, was trained to use transanal irrigation every second day and thus, achieved day-and-night continence.

The BFS was reduced in six patients with a median of 16, ranging from 8 to 19 points (Table 2). However, correlating with the positive life style feeling, the reported QoLS showed good results with a median value of 10 points, ranging from 7 to 13 points (Table 3).

Discussion

The involvement of the entire colon and variable length of ileum is the hallmark of TCA. Simple resection of the aganglionic segment including the ICV and performance of a straight ileoanal pull-through result in loss of the entire absorptive surface of the colon with the potential

Table 2 Long-term evaluation of bowel function using the bowel-function score by Rintala and Lindahl [12]

Bowel function score	ID 1	ID 3	ID 4	ID 5	ID 6	ID 7	ID 8	Points
<i>Ability to hold back defecation</i>								
Always	3	3		3	3	3	3	3
Problems less than 1/week			2					2
Weekly problems								1
No voluntary control								0
<i>Feels/reports the urge to defecate</i>								
Always	3	3	3		3	3	3	3
Most of the time								2
Uncertain				1				1
Absent								0
<i>Frequency of defecation</i>								
Every other day to twice a day								2
More often	1	1	1	1	1	1	1	1
Less often								1
<i>Soiling</i>								
Never							3	3
Staining less than 1/week, no change of underwear required	2	2			2			2
Frequently staining, change of underwear often required						1		1
Daily soiling, requires protective aids			0	0				0
<i>Accidents</i>								
Never						3	3	3
Fewer than 1/week	2	2			2			2
Weekly accidents; often requires protective aids								1
Daily requires protective aids during day and night			0	0				0
<i>Constipation</i>								
No constipation	3		3	3	3	3	3	3
Manageable with diet		2						2
Manageable with laxatives								1
Manageable with enemas								0
<i>Social problems</i>								
No social problems					3	3	3	3
Sometimes (foul odors)	2							2
Problems causing restriction in social life		1	1					1
Severe social and/or psychic problems				0				0
Maximum score	16	14	10	8	17	17	19	20

consequences of fluid loss as well as electrolyte imbalances. A variety of surgical procedures has been advocated to minimize fluid and electrolyte losses of these patients [2, 4–8, 10, 14]. One attempt to overcome these problems was to create a reservoir by forming an ileoanal pouch such as a J-pouch or S-pouch, which represents a well-established technique for the treatment of children with familial adenomatous polyposis and ulcerative colitis and is associated with an acceptable long-term functional outcome in terms of continence and bowel frequency habits [15–17]. Another

strategy aimed at utilizing the reservoir as well as water and electrolyte absorptive function of the colon. Martin modified the Duhamel procedure by extending the ileo-retrorectal side-to-side anastomosis up to the splenic flexure [2], which he later modified by performing a total ileo-colonic side-to-side anastomosis up to the caecum [7]. Shandling combined the total ileo-colonic side-to-side anastomosis with an endorectal pull-through procedure according to Soave [8]. However, utilization of the total aganglionic colon does not only bear a higher surgical

Table 3 Long-term evaluation of QoL using the adapted QoL Score by Bai and colleagues for patients with fecal incontinence [13]

Item and criteria	ID 1	ID 3	ID 4	ID 5	ID 6	ID 7	ID 8	Points
<i>Soiling</i>								
Absent							4	4
Accidental	3	3			3			3
Frequent			2	2		2		2
<i>Incontinence</i>								
Accidental	1	1	1		1	1	1 (never)	1
Frequent				0				0
<i>School or work absence</i>								
Never				2	2	2	2	2
Accidental	1	1	1					1
Frequent								0
<i>Unhappy or anxious</i>								
Never			2		2	2	2	2
Accidental	1	1						1
Frequent				0				0
<i>Food restriction</i>								
No		2	2	2	2		2	2
Somewhat	1					1		
Much								0
<i>Peer rejection</i>								
Never	2	2			2	2	2	2
Accidental			1	1				1
Frequent								0
Maximum score	9	10	9	7	12	10	13	13

complication risk but is also associated with a higher incidence of enterocolitis and bowel obstruction, wherefore this technique has been abandoned in several institutions [18–20].

Sophisticated research investigations in animal studies and human subjects have shown that the right colon has superior efficiency compared to the transverse and left colon in terms of water and electrolyte absorptive capacities, also leading to a reduced transit time [21–30]. Thus, the right colonic patch has achieved popularity in TCA surgical management [4–6, 31]. Kimura has implemented a complex procedure, originally designed to treat intractable ileostomy diarrhea, where he patched the right colon to the ileum, leaving the patch vascularized by the mesenteric pedicle in the first stage [6]. At the time of definite pull-through, a few months after the side-to-side ileocolostomy, the graft had gained enough blood supply via intramural collaterals enabling severance of the mesocolon for adequate bowel mobilization. Kottmeier and his colleagues developed a similar procedure for extensive TCA in which the distal aganglionic ileum was patched to the ganglionic ileum [14]. Boley was the first to perform a primary ileoendorectal pull-through with a right colonic patch anastomosed to the distal ganglionic ileum in an antiperistaltic direction, covered by a temporary loop ileostomy. [5].

Sauer’s strategy for TCA, which retains the right colon and the ICV, has two potential advantages: (a) the preservation of the right colon as an isoperistaltic onlay with its water and electrolyte absorptive capacity which further enables the possibility of a reservoir between the right colon and ileum, and (b) the preservation of the ICV as a natural barrier to prevent backwash, thus, reducing the incidence of enteritis and associated malabsorption (Fig. 1) [9, 11].

This follow-up study shows that preservation of the right colon as an isoperistaltic onlay led to an acceptable stooling pattern of watery to semisolid stools of median 3–4 times per day, which is comparable to long-term results from other studies. Five patients treated according to Boley were reported on normal growth, continence and acceptable bowel function with a stooling pattern of average 2–4 stools a day after a follow-up period of 2–11 years [32]. Long-term results with a follow-up time of 5–17 years from 10 TCA patients treated according to Kimura’s technique also documented a comparable bowel movement frequency of 1–3 times a day in older patients and a normal body weight achieved in children between 2 and 4 years after the definite procedure [20]. In comparison, long-term results with a follow-up time of 4–129 months available from 11 TCA patients treated by a

long segment side-to-side anastomosis of ganglionic with aganglionic bowel according to Martin's modification showed a slightly higher bowel movement frequency of 4–5 watery stools a day associated with night-time soiling [33]. Thus, the long-term absorptive benefit of an aganglionic patch or longer bowel segment is questionable, especially, as long-term results with a follow-up time of 1–5 years of 10 HD patients (6 of them with TCA and 4 undergoing a redo operation after a failed primary pull-through of long segment aganglionosis) treated by an ileoanal anastomosis with a J-pouch showed a similar bowel movement frequency of 2–5 times a day (median 3), as documented by Rintala and Lindahl [16]. These results are further supported by Coran who has also reported on a stooling frequency of 3.5 times a day 3 years after performing a straight endorectal ileoanal pull-through procedure in 10 TCA patients [34], which indicates that the bowel movements frequency declines with age and becomes similar regardless of the type of initial procedure performed.

Fecal soiling and incontinence is another area of concern in TCA patients that have undergone surgical procedures, which has been reported to vary from 6 to 86 % [35–39]. This survey documented that only one patient does not soil at all. Three of our patients reported on rare accidental soiling occurring less than once a week with one of them achieving day-and-night continence using regularly transanal irrigation. Only one patient suffers from frequent soiling accidents and two patients suffer from night-time soiling needing protective aids. These two patients have the lowest BF and QoL scores. Related to a recent bowel-function assessment of classic rectosigmoid HD patients compared with a healthy control group by Jarvi and colleagues [40], the BFS of most of our TCA patients was comparably reduced as observed in rectosigmoid HD patients who were reported on a mean BFS of 17.1 ± 2.8 . Nevertheless, one of our TCA patients achieved a normal BFS of 19 points when compared to the mean BFS of 19 ± 1.2 obtained from healthy individuals by Jarvi and colleagues [40]. Thus, our results do not confirm earlier findings from a meta-analysis by Laughlin and colleagues, which documented that the overall bowel-function rate of TCA patients is poor in about 40 % [37].

It is furthermore encouraging that all patients confirmed a positive lifestyle and none of our patients required a permanent stoma to manage incontinence, which has been recently reported to be required in about 6.5 % of patients with TCA [37] and was also required in two of nine TCA patients as documented in a recent retrospective study from a single center [41].

Apart from soiling problems to some extent, the QoL questionnaire, however, confirmed an overall good lifestyle of our TCA patients, indicated by a median QoL score of

10 points, which is comparable with the QoL of healthy individuals (mean 11.6 ± 0.7), who have been assessed with the same questionnaire by Bai and colleagues [42]. Only one of our TCA patients with a QoL score of 7 was considered to have a fair QoL (5–8 QoLS), which correlated with the lowest documented BFS of 8 points in this case. This finding suggests that daily soiling and frequent fecal accidents may have a severe impact on the patients' psychosocial life and development. This has been also documented in a study comparing TCA with rectosigmoid HD patients regarding their long-term functional and psychosocial outcomes, revealing that most TCA patients were considered incontinent from the psychological interview and affected with more emotional and behavioral problems and a lower self-esteem than their pairs [43]. It is therefore noteworthy, that our youngest TCA patient with occasional soiling incidences achieved day-and-night continence by transanal irrigation and thus, scored high in the BF questionnaire and in the QoL assessment. Our generally good results of the QoL questionnaire are also consistent with long-term QoL results obtained from patients treated by a modified Martin's procedure or straight ileoanal pull-through, who were also reported on a good QoL except one patient with a fair result [35].

Even though all our patients are on normal oral feeds, selective food intolerance has been found to be a common feature in these patients, which, however, does not seem to impair their QoL.

Enterocolitis has recently been determined as the most common postoperative complication after pull-through procedure in TCA patients, occurring with an incidence of 42 % [37]. In our patients, enterocolitis occurred only in a mild form and early after the surgical procedure in three patients (37 %). Thus, the incidence of enterocolitis has been found to be equal or overall lower than those reported in TCA patients treated with other techniques: Duhamel (37–43 %), Martin modification (33–56 %), Kimura (50 %), or Boley (20 %), respectively [20, 32, 35, 44, 45]. The incidence of enterocolitis has been attributed to the preservation of an aganglionic bowel segment [20, 46, 47], which has therefore already been suggested to be only used as a 10 cm segment for absorptive purposes [48]. Techniques without utilization of aganglionic bowel such as the ileoanal pull-through with a J-pouch construction also resulted in a 30 % enterocolitis rate [16]. A straight ileoanal pull-through has, however, been reported with contradictory enterocolitis rates of 0 % [34], 57 % [35] or 100 % [45], which rises the impression that in terms of postoperative enterocolitis incidences, none of the techniques is superior than the other. Although one might speculate that the preservation of the ICV represents an active barrier against bacterial ascent [49, 50], it has to be mentioned that the preservation of the aganglionic ICV

also might imply the risk of a functional stenosis, which happened in two of our patients requiring a secondary longitudinal myotomy.

Despite the ideal operative therapeutic technique of patients with TCA remains controversial, our long-term results indicate that Sauer's technique, preserving the right hemicolon and the ICV, might be a favorable method for these rare cohort of patients, as typical postoperative complications of this disease such as enterocolitis, malabsorption or failure to thrive are mild or even absent. Although the overall bowel function is reduced and there is an incidence of soiling, these problems seem not to impact the patients' QoL, which has been reported as good in the majority, enabling the patients to lead a fairly normal lifestyle.

Conflict of interest This study has no conflict of interest and has received no funding.

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