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ORIGINAL ARTICLE

Autologous intestinal reconstruction: a single institution study of the serial transverse enteroplasty (STEP) and the longitudinal intestinal lengthening and tailoring (LILT)

Adil A. Shah[1,2] · Mikael Petrosyan[1] · Ashanti L. Franklin[1] · Alfred A. Chahine[1] · Clarivet Torres[1,3] · Anthony D. Sandler[1]

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Abstract

Purpose To review the effectiveness of the longitudinal intestinal lengthening and tailoring (LILT) and serial transverse enteroplasty (STEP) operations in a cohort of patients with short bowel syndrome (SBS).

Methods We conducted a retrospective analysis of children with SBS treated at our institution from 2004 until 2014. Children aged 0 days to 18 years with SBS who underwent autologous intestinal reconstruction were included in the study.

Results Twenty-two SBS patients underwent 31 different lengthening procedures (LP). Seventeen patients underwent their primary lengthening procedures at our institution: 9 (53%) patients underwent a LILT, 7 (41%) underwent a STEP and 1 (6%) had a simultaneous LILT and STEP procedure. 12/22 patients had a second STEP, two had a third STEP and one patient had an intestinal transplantation after the LP. Median intestinal length at the time of surgery was 25 cm (range 12–90 cm). There was no difference in gain of intestinal length after LILT vs. STEP ($p = 0.74$). Length of stay and initiation of feeds were similar. Serum albumin increased after autologous bowel lengthening ($p < 0.001$). 50% were weaned off parenteral nutrition (PN) (5/9 of the LILT, 1/7 of the STEP, 1/1 of the combined LILT/STEP). There were no surgical complications or deaths.

Conclusion In patients with SBS, LILT and STEP procedures are effective for autologous intestinal reconstruction and enable intestinal rehabilitation.

Keywords Short bowel syndrome · Intestinal rehabilitation · Autologous intestinal reconstruction

Introduction

Short bowel syndrome (SBS) is a multi-systemic disease that is caused by an anatomical and/or functional loss of a significant portion of the small intestine due to an antenatal and/or acquired neonatal disorder. Antenatal insults are most commonly due to bowel atresia, volvulus, gastroschisis, or in utero intestinal obstruction [1]. It is associated with a significant reduction in the quality of life, with an increased morbidity and mortality [2–4]. The degree of malnutrition induced by SBS is not only dependent upon residual intestinal length, but also location of resected intestine, presence of a competent ileocecal valve (ICV), presence of the colon, and the function of the remaining intestine [5, 6].

SBS is associated with inadequate nutrient absorptive capacity and abnormal peristalsis with intraluminal stasis, resulting in portal and systemic translocation of bacteria that can induce sepsis [7]. Goals of therapy include early medical management coordinated in an intestinal rehabilitation program that encompasses adequate nutrition, preservation of hepatic function, and management of central access [8, 9]. Pro-adaptive surgery, such as stoma closure, ostomy in continuity, stricturoplasty, enteroplasty, and autologous gut reconstruction, with the longitudinal intestinal lengthening and tailoring (LILT) and serial transverse enteroplasty (STEP) procedures, are adjunctive surgical procedures that complement medical management in patients with short bowel syndrome. The decision to perform a bowel lengthening procedure is usually reached when there is failure to progress to enteral feedings and

✉ Mikael Petrosyan
MPetrosy@childrensnational.org

1 Department of General and Thoracic Surgery, Children's National Health System, Washington, DC, USA

2 Department of Surgery, Howard University Hospital and College of Medicine, Washington, DC, USA

3 Department of Gastroenterology, Hepatology, and Nutrition, Children's National Health System, Washington, DC, USA

when life-threatening complications, such as liver disease and recurrent central line sepsis, become entrenched. The choice of operation in short bowel syndrome is influenced by the caliber and length of the remnant intestine, blood supply, and intestinal function [5, 6, 10]. This study evaluates the LILT and STEP surgical procedures in a single cohort of patients from a single surgical center.

Methods

A retrospective chart review was conducted at our institution from January 2004 until February 2014 at Children's National Health System, Washington, DC. The institutional database was queried for children ages 0 days to 18 years with a diagnosis of short bowel syndrome who underwent an autologous intestinal reconstruction procedure. LILT was performed as a modification of the original Bianchi procedure with a single anastomosis as first described by Pokorny and reported by Chahine and Ricketts [11]. LILT was the preferred initial procedure and performed more frequently at our institution. The STEP was reserved for a second procedure or selected initially for lengthening when the LILT was avoided due to the mesenteric vascular configuration of the dilated bowel. In patients with refractory SBS who remain PN dependent, and develop dilated hypoperistaltic intestines despite optimal medical management and autologous intestinal reconstruction, an ostomy in continuity (Bishop–Koop or Santulli type) is offered as an adjunct measure. The ostomy in continuity is offered as a salvage procedure that allows decompression of the dysfunctional dilated small bowel, but still maintains partial continuity of the remnant colon enabling both limited absorption and intestinal adaptation [12]. Institutional review board (IRB) exemption was obtained for this retrospective chart review.

At CNHS, patients with short bowel syndrome are entered into the intestinal rehabilitation program if they fulfill the following criteria [10]:

1. Infants with no liver disease or mild cholestasis who have extreme short bowel as defined by at least 10 cm of small bowel and > 50% of their colon.
2. Children with more than 35 cm of small bowel with an increased bilirubin level, but a normal international normalized ratio (INR).
3. Children with advanced liver disease with portal hypertension-associated hypersplenism or thrombocytopenia but normal INR, who had at least 50 cm of bowel and abnormal but potentially repairable intestinal anatomy, or history of tolerance of at least 30% of caloric needs by the enteral route

Experience with an intestinal rehabilitation program in the management of short bowel syndrome has previously been described by our group [10, 12, 13]. It briefly consists of a complete evaluation of the patient including biochemical, physical, radiological, and histological evaluation as well as identifying the anatomic variations. Continual parent education and support is a key component since their contribution is vital for improved outcomes. Patients are followed at least weekly by a pediatric gastroenterologist, a dietitian, a nurse coordinator, and a nurse practitioner. The medical management of patients consists of an aggressive dietetic regimen with a focus on control of the metabolic balance as well as prompt and effective treatment of patient complications. Enteral tube feedings and parenteral nutrition management/weaning is managed in a multidisciplinary manner and allows for timely identification and management of complications of intestinal failure such as sepsis, catheter-related, and bacterial overgrowth. Surgical input is obtained to identify options to restore intestinal continuity, repair enterocutaneous fistulas, and resect strictured/obstructed bowel, including intestinal lengthening (Bianchi procedure) and serial transverse enteroplasty.

Outcome measures included (1) survival (immediate and long-term), (2) length of stay, (3) return of bowel function, (4) pre- and post-operative intestinal length as well as the increase in intestinal length after a LILT or STEP, (5) surgical complications (immediate and delayed), (6) post-operative weight gain, (7) transition from parenteral to enteral nutrition, (8) improved hepatic function, and (9) re-operations.

Descriptive analysis was used to describe the study population. Patient demographics and symptoms of SBS were analyzed and surgical outcomes of LILT and STEP were also compared using Pearson's Chi square or Fisher's exact test for categorical variables and Student's *t* test for continuous data. A p value of < 0.05 was considered statistically significant.

Results

There were 92 patients with short bowel syndrome enrolled in our institution's multidisciplinary intestinal rehabilitation program (IRP) during the study period. Of those, 22 (24%) underwent 31 different lengthening procedures. Sixty-four percent were male (Table 1). Median gestational age (GA) was 34 (IQR 24–40) weeks (Table 1). The median age at initial procedure was 28 months (range 3–129). Median intestinal length at the time of surgery was 25 (IQR 10–90) cm. The most common etiology of SBS in the cohort was intestinal atresia (32%). The mean height and weight *Z* score upon enrollment into our IRP was -2.72 (± 1.74) and -1.49 (± 0.64), respectively. At the time of entry into the program,

Table 1 Demographics

	Overall ($n=22$)		LILT ($n=10$)		STEP ($n=11$)		LILT + STEP ($n=1$)
	Median	IQR	Median	IQR	Median	IQR	
GA (weeks)	34	24–40	35.5	27–28.5	32.5	32–34	27
Age at surgery (months)	28	3–129	26	3–131	29	4–126	3
Initial intestinal length (cm)	25	10–90	25	8–86	29	11–92	18
Follow-up (months)	43	4–99	45	2–80	39	6–45	32
	n	%	*n*	%	*n*	%	*n*
Sex							
Males	14	64	6	60	8	78	1
Females	8	36	4	40	3	22	–
Etiology							
Intestinal atresia	7	32	1	10	6	54	–
NEC	4	18	2	20	1	18	1
Midgut volvulus	4	18	3	30	1	9	–
Gastroschisis	2	9	1	10	1	9	–
Gastroschisis + intestinal atresia	4	18	2	20	2	18	–
Hirschsprung disease	1	5	1	10	0	–	–

Table 2 Operative characteristics

$n=22$	LILT	STEP	LILT + STEP
	n	*n*	*n*
Initial procedure at CNHS	9	7	1
Initial procedure at OSH	1	4 (18%)	–
Second procedure	0	12	0
Transplant	0	1	0
Acute surgical complications	0	0	0
Mortality	0	0	0

all patients were dependent on parenteral nutrition (PN) with a mean caloric energy requirement from PN of 94% and with an average albumin of 2.76 (±0.60) g/dl. At the time of enrollment into our IRP, 59% had an elevated total bilirubin with an average bilirubin of 8.80 (±4.80) mg/dl. Median follow-up was 43 (IQR 4–99) months.

Seventeen patients underwent their primary lengthening procedures at our institution: 9 (53%) patients underwent a LILT, 7 (41%) underwent a STEP, and 1 (6%) had a simultaneous LILT and STEP procedure (Table 2). The remaining five patients had their initial procedure performed at an outside facility. Twelve patients had a secondary lengthening procedure, all a STEP, and two patients subsequently had a third STEP (Table 2). The choice of procedure was dependent on bowel length and blood supply. A total of ten patients had an ostomy in continuity, of which five had an ostomy in continuity (four Bishop–Koop and one Santulli) procedure performed at the time of the second or third lengthening STEP procedure to decompress the dilated small bowel, while still using the remnant colon as a salvage procedure for patients with severe intestinal dysfunction. There was no difference in gain of intestinal length after the initial LILT [mean 29.5 (±16.1) cm] vs. STEP [mean 25.57 (±17.6) cm] $p=0.74$, performed at our institution (Table 3). In addition, there was no difference in length of stay or initiation of enteral feeds in patients who underwent a LILT vs. a STEP procedure. Three (14%) patients had an ileocecal valve (ICV) (1 in LILT and 2 in STEP group). The presence of an ICV was not associated with PN independence, $p=1.00$. To date, only 1 of 22 patients has undergone intestinal transplantation after autologous intestinal lengthening due to progressive liver disease.

At the completion of this study, 11 (50%) of the patients were weaned off PN. Of those who only underwent one

Table 3 Comparison between initial LILT vs. STEP

	LILT	STEP	*p* value
Increase intestinal length (cm), mean ± SD	29.6 ± 16.1	25 ± 17.6	0.74
Time to initial feed (days), mean ± SD	9.3 ± 1.4	8 ± 2.8	0.30
LOS (days), mean ± SD	21.1 ± 16	25.9 ± 17.4	0.54

lengthening procedure, seven patients were weaned off PN (5/9 of the LILT, 1/7 of the STEP, 1/1 of the combined LILT/STEP), ($p = 1.00$). Figure 1a, b demonstrates the time to weaning for STEP and LILT subgroups, respectively. Four received multiple procedures prior to being weaned off PN (one had a LILT/ STEP, two had a second STEP, one had a third STEP). Of these, two had difficulty weaning off PN after multiple lengthening procedures due to significant intestinal dysmotility and required an ostomy in continuity to improve their enteral tolerance. For those who remained on PN, their PN caloric requirement decreased from an average of 93–40%, $p = < 0.001$. There was an overall improvement in albumin ($p = < 0.001$) and total bilirubin ($p = < 0.001$) after lengthening (Table 4). The mean height and weight *Z* scores also significantly improved after surgery, −0.67 (± 1.09, $p = 0.007$) and 0.54 (± 2.08, $p = < 0.001$), respectively. There were no acute surgical complications including post-operative bleeding, clinical anastomotic leak, stenosis or death in either group. While there were no surgical complications, 12 patients failed to progress to complete enteral nutrition and underwent a secondary STEP procedure. Two of the 12 patients underwent a third STEP procedure. The median time to a second lengthening procedure was 13.5 (IQR 6–45) months. 65% of the study population had recurrent central line sepsis requiring multiple central line removal and replacement. The overall 10-year survival rate was 100%.

Discussion

In this relatively large recent retrospective experience from a single center, the outcomes of the LILT and the STEP procedures as an initial lengthening procedure were evaluated. The LILT remains the preferred procedure at our institution and was most frequently the procedure of choice when feasible. The STEP was reserved for a second procedure or selected initially for lengthening when the LILT was avoided due to the mesenteric vascular configuration of the dilated bowel. Despite this partiality to LILT, there

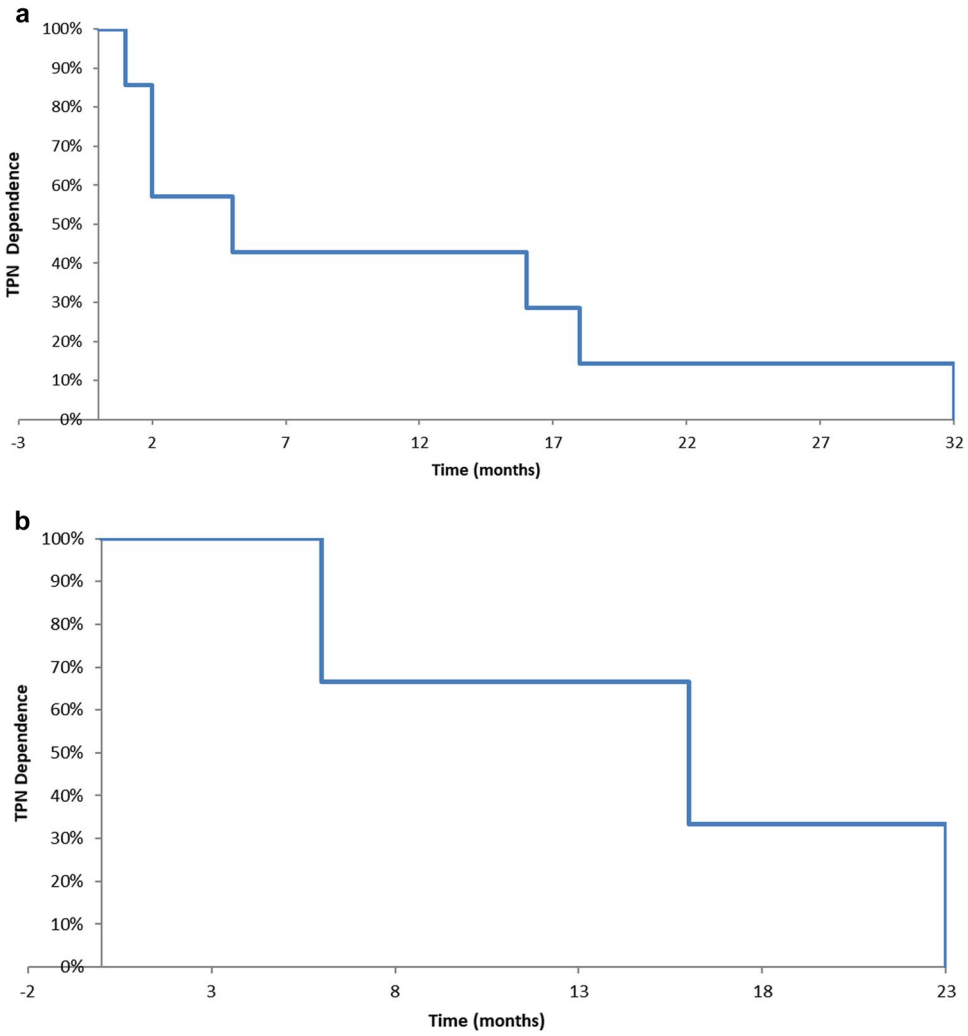

Fig. 1 **a** Time to weaning in patients weaned off parenteral nutrition after undergoing serial transverse enteroplasty (STEP). **b** Time to weaning in patients weaned off parenteral nutrition after undergoing longitudinal intestinal lengthening and tailoring (LILT)

Table 4 Comparing pre-operative and post-operative characteristics

	Enrollment in intestinal rehab (pre-operative)	Post-operative	*p* value
Overall			
Mean *Z* score weight (kg)	−1.49 (±0.64)	0.54 (±2.08)	<0.001
Mean *Z* score height (cm)	−2.72 (±1.74)	−0.67 (±1.09)	0.007
Median albumin (g/dl)	2.8 (IQR 1.5–3.8)	3.5 (IQR 2.9–4.2)	<0.001
Median total bilirubin (mg/dl)	8.75 (IQR 2.4–19.3)	0.3 (IQR 0.1−1)	<0.001
LILT			
Mean *Z* score weight (kg)	−1.61 (±2.33)	0.34 (±0.95)	<0.001
Mean *Z* score height (cm)	−2.78 (±2.4)	−0.77 (±0.69)	0.007
Median albumin (g/dl)	2.7 (IQR 2.4–2.8)	3.7 (IQR 3.2–4.0)	<0.001
Median total bilirubin (mg/dl)	11.9 (IQR 8.5–14.8)	0.3 (IQR 0.2–0.3)	<0.001
STEP			
Mean *Z* score weight (kg)	−1.26 (±1.17)	−0.41 (±1.36)	<0.001
Mean *Z* score height (cm)	−1.87 (±1.53)	−1.31 (±1.64)	0.007
Median albumin (g/dl)	2.7 (IQR 2.6–3.2)	3.5 (IQR 3.1–3.9)	<0.001
Median total bilirubin (mg/dl)	5.0 (IQR 2.8–9.0)	0.3 (IQR 0.3–0.3)	<0.001

was no difference in surgical outcomes between the LILT and STEP procedures. Current literature suggests some controversy regarding which procedure provides the greatest intestinal length, time to wean off PN, and adequate growth and development of the child. In a 20 year study, Walker et al. [14], found a 75% survival rate in patients after a LILT procedure and a 44% success in weaning off PN. This study also found that survival is more likely when the LILT procedure is performed after 6 months of age [14]. Khalil et al. [15] found that the LILT had an improved survival rate and had an overall increased intestinal length of 90 cm. In the literature, 44–91% of children were weaned off PN after the LILT [14–16].

The overall survival rate for the STEP procedure was 89% in one study [17]. Similar to the LILT procedure, 47–88% were weaned off TPN after the STEP [7, 17–21]. While there appears to be no difference between the LILT and STEP, intestinal vascularity may preclude the performance of the LILT. In certain circumstances, the STEP may be more technically feasible in the duodenum and proximal jejunum [22–24]. Furthermore, the LILT procedure may also compromise vasculature with manipulation and dissection of the mesentery [8, 25]. Despite these reservations, we found no benefit of one procedure over the other for absolute bowel lengthening nor were there any cases in which the vascularity of the bowel was compromised during the LILT procedure.

Frequently, children with SBS require more than one lengthening operation to optimize nutritional status. Bianchi suggested that a STEP procedure should be completed after a LILT if there remains concern of bowel length after the initial LILT procedure [7]. Other studies have performed a second STEP after an initial STEP procedure [26, 27]. We have found both approaches to be useful. Our personal preference for lengthening is generally the LILT procedure, as we observe a smoother configuration of bowel relative to the STEP. With time, the STEP can result in "diverticular out-pouching" of the intestine, but in this series, this appears to make no difference in outcome. Of note, five out of nine patients after a single LILT were weaned off PN, while one out of seven were weaned off PN following a single STEP. Although of no statistical consequence, this finding might suggest a trend of improved outcome following LILT. Again, selection bias is evident as in general those patients that could not undergo LILT underwent the STEP procedure, which may indicate less than optimal intestinal blood supply.

There were five patients who underwent an ostomy in continuity at their second or third STEP procedure for intestinal bacterial overgrowth and diverticular out-pouching observed with the STEP procedure. Only one of the patients needing an ostomy in continuity was weaned off PN. Fourteen percent of our study population had an ileocecal valve (ICV) post-operatively. We did not find any association between the presence of ICV and PN dependence; however, this finding may be underpowered due to sample size. Our data and the literature suggest that the presence of the ICV does not predict surgical outcome [16, 28–30]; however Walker et al. [14] found that patients with an ICV had improved surgical outcome.

Although we did not have any acute surgical complications, multiple complications are reported in the literature. Both the LILT and STEP are associated with post-operative bleeding, stricture formation, leaks, and less commonly abscesses [24]. In addition, LILT has been associated with intestinal necrosis, perforation, and fistula formation [8, 24]. Most common causes of mortality after an autologous

intestinal reconstruction are liver failure and sepsis [28–30]. The post-operative mortality rate of autologous intestinal reconstruction is not well known, as the cause of death in this patient population is multi-factorial. In a systemic review, the average mortality rate was 30.2% and 14.3% for LILT and STEP, respectively [24].

This current study is limited by its small sample size, but is relatively large for a single center over a 10-year period. A prospective randomized-controlled analysis would be required to determine the superiority of either the LILT or STEP procedure. While ideal for scientific rigor, a randomized prospective study may not be feasible because of the relative infrequent performance of autologous intestinal reconstruction, individual intestinal anatomy and surgical nuance. Furthermore, no direct measurements of intestinal absorption were obtained. There also remains the possibility that the data failed to account for patient factors that may skew the results, given the heterogeneous nature of patients with SBS. It is also difficult to ascertain if patients undergoing lengthening procedures would have demonstrated similar improvements with a longer course of medical management. Needless to say, despite the retrospective nature of this review, both procedures are effective and useful for autologous intestinal lengthening.

In patients with SBS, LILT and STEP procedures enable intestinal rehabilitation. We advocate for a systematic multidisciplinary intestinal rehabilitation team to manage the highly complex patients with SBS. With meticulous medical and surgical management, patients with autologous intestinal lengthening show significant improvement in enteral tolerance and nutritional parameters and can frequently avoid the need for intestinal and/or liver transplant.

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Compliance with ethical standards

Conflict of interest The authors have no conflicts of interest to report.

Research involving human participants and/or animals All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed consent Not applicable. Institutional review board (IRB) exemption was obtained for this retrospective chart review.

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