ORIGINAL ARTICLE

Serial transverse enteroplasty for short bowel syndrome: Hong Kong experience

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Objective To report our experience with the first series of serial transverse enteroplasty operations (a novel bowel-lengthening procedure for patients with short bowel syndrome).

Design Case series.

Setting A tertiary paediatric surgery referral centre in Hong Kong.

Patients Four patients with short bowel syndrome aged 11 months to 14 years underwent serial transverse enteroplasty between November 2007 and June 2010.

Results A total of six such serial procedures were performed; two patients had repeated operations. Median pre-serial transverse enteroplasty small bowel length was 17.5 cm. The median increase in small bowel length was 90%. One patient experienced transient postoperative intestinal obstruction that resolved after conservative management. The median postoperative follow-up period was 31 months. The median enteral nutrition tolerance increased from 24% to 47%. The median weight-for-age z score increased by 0.55, and the median weight-for-height z score increased by 0.98. One patient had successfully weaned off parenteral nutrition.

Conclusion Serial transverse enteroplasty is a feasible and safe treatment for short bowel syndrome patients, which helps to improve enteral nutrition and promote growth. Repeated serial transverse enteroplasty can be performed in patients with ultra-short bowel lengths. Follow-up is necessary to assess the long-term outcomes.

New knowledge added by this study

- Serial transverse enteroplasty (STEP) is a safe and feasible bowel-lengthening procedure for patients with short bowel syndrome (SBS).
- STEP improves the enteral nutrition tolerance of SBS patients.

Implications for clinical practice or policy

- A multidisciplinary approach is essential for management of these patients.
- STEP should be considered an adjuvant therapy for selected SBS patients.

Key words
Intestinal obstruction; Intestine, small; Reconstructive surgical procedures; Short bowel syndrome

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Introduction

Short bowel syndrome (SBS) is a condition of inadequate bowel length or malfunction leading to malnutrition and growth retardation.[1] In the paediatric age-group, common causes of SBS include necrotising enterocolitis, gastroschisis, midgut volvulus, intestinal atresia and long-segment Hirschsprung's disease.[2] Owing to intestinal failure, most SBS patients have to depend on total or partial parenteral nutrition (PN). Multiple morbidities include cholestatic jaundice, liver failure, catheter-related sepsis, and small bowel bacterial overgrowth (SBBO).[3]

Management of SBS requires a multidisciplinary approach.[4] Surgical management of SBS consists of small bowel transplantation and non-transplant surgery. The long-term graft survival rate of small bowel transplantation is still unsatisfactory, despite immunosuppressive therapy.[5] Non-transplant surgery for SBS aims to increase small bowel absorptive surface area and intestinal transit time. Bowel lengthening procedures had been described by Bianchi, and Kimura and Soper.[6,7] However these entailed long anastomotic bowel segments that were technically demanding to construct. In 2003, Kim et al[8,9] in Boston described a novel form of serial transverse enteroplasty (STEP). Lengthening of dilated small bowel was achieved by serial transverse applications of a stapling device from alternate directions, so as to create a zigzag channel. In this article, we review our

治療短腸綜合症的橫切小腸成形術：香港經驗回顧

目的 報告首批短腸綜合症病人接受橫切小腸成形術來增加腸管長度的經驗。

設計 病例系列研究。

安排 香港一所小兒外科轉介中心。

患者 2007年11月至2010年6月期間4名年齡介乎11個月至14歲進行橫切小腸成形術的短腸綜合症病人 。

結果 病人中兩人重覆進行手術，共進行手術6次。術前小腸長度的中位數為17.5 cm，術後長度增加比率的中位數為90%。其中一名患者術後有短暫性的腸梗阻，但經保守治療後已康復。術後平均跟進期為31個月。腸內營養耐受性由24%增加至47%。病人年齡別體重Z值的中位數增加了0.55，而身高別體重Z值的中位數亦增加了0.98。其中一名患者更於術後不需要再使用靜脈營養。

結論 橫切小腸成形術可改善腸內營養及促進病人成長，是治療短腸綜合症有效及安全的方法。超短腸綜合症的患者可重覆接受手術，但必須跟進術後的病人來評估此手術長遠的治療結果。

first series of STEP operations performed on four patients with SBS.

Methods

From November 2007 to June 2010, four children with SBS had six STEP operations performed in our centre. Their ages ranged from 11 months to 14 years. Three patients had a primary diagnosis of midgut volvulus and one had a retroperitoneal yolk sac tumour resulting in massive gut resection. All of them were PN-dependent, and two suffered from cholestatic jaundice and failure to thrive. Two patients endured repeated catheter-related sepsis and were running out of venous access for long-term PN. The pre-STEP small bowel lengths ranged from 10 cm to 40 cm. In all the patients, the ileocaecal valve was lost at the initial operation before STEP. In patients A and B, a repeated STEP operation was performed after intervals of 31 months and 13 months (Table 1). In all patients, a preoperative fluoroscopic small bowel transit study was performed to ensure a dilated small bowel width of at least 4 cm necessary to facilitate the procedure.

After laparotomy and adhesion lysis, the STEP procedure was performed as described by Kim et al.[8,9] The anti-mesenteric longitudinal border of the small bowel was marked (Fig a). The first intended stapler application started from most distal part of small bowel. A small mesenteric window was created in which a 16F Foley catheter could pass through (Fig b). The Foley catheter was used to guide an endo-GIA stapler to pass through the mesenteric defect. The bowel was kept flattened with the mesenteric border in the middle on firing of the stapler, leaving an uncut end of approximately 2 to 2.5 cm, depending on age and size of patients (Fig c). A reinforced figure-of-eight polyglactin stitch was applied at end of staple line to prevent leakage. The next stapling was approximately 2 to 2.5 cm proximal to the previous site, applied from the opposite direction. Firing of the stapler was repeated up to the proximal jejunum near duodenojejunal junction, with all firings perpendicular to the long axis of small bowel (Fig d).[9] The number of staplings ranged from 2 to 10 (median, 5).

A gastrostomy was fashioned in every patient to facilitate enteral feeding. On postoperative day 7, a fluoroscopic small bowel transit study was repeated to exclude any leakage from staple line.

Results

The post-STEP small bowel length was increased to 15 to 72 cm, with a median increment of 90% (range, 50-140%). There was no intraoperative staple line leakage. All patients resumed oral or gastrostomy feeding 1 week after the operation. Patient B had intestinal obstruction postoperatively, which resolved on conservative treatment. Patient C had one episode of gastrointestinal bleeding 13 months after the operation and received a blood transfusion. The patients were followed up from 23 to 40 (median, 31) months. A significantly increased weight-for-age *z* score was observed in patients A and D, who had the STEP performed aged younger than 3 years,

TABLE 1. Patients with serial transverse enteroplasty (STEP) operations performed

Patients	Sex/age at STEP	Primary diagnosis	Indications for STEP	Pre-STEP bowel length (cm)	No. of staple firings	Post-STEP bowel length (cm)
A*	M/11 m	Midgut volvulus	Cholestatic jaundice and failure to thrive	15	5 + 6	36
B*	F/14 yr	Retroperitoneal yolk sac tumour	TPN† dependent with difficult venous access	20	5 + 4	40
C	M/11 yr	Midgut volvulus	Cholestatic jaundice and failure to thrive	10	2	15
D	M/26 m	Midgut volvulus	TPN dependent with difficult venous access	40	10	72

* Repeated STEP operation done

† TPN denotes total parenteral nutrition

suggesting a catch-up growth. The weight-for-height *z* score was increased in all patients irrespective of age, suggesting improved nutrition after STEP. The median weight-for-age *z* score increased by 0.55 (at a rate of 0.023 units per month), and median weight-for-height z score increased by 0.98 (at a rate of 0.039 units per month) [Table 2]. Enteral feeding with amino acid–based formula was facilitated by nocturnal gastrostomy feeding via a milk drip chamber. The extent of PN was titrated according to the growth and weight of each patient. The median enteral nutrition tolerance increased from 24 to 47% after STEP, at the rate of 0.82% increments per month. Patient B was weaned off PN 16 months after her second STEP operation. In patients A and C who had cholestatic jaundice before STEP, fish-oil emulsion lipid in a dose of 1 g/kg/day was added to the PN. The bilirubin level improved to normal postoperatively. Loperamide was given before and after STEP to control diarrhoea, with doses adjusted to avoid vomiting and abdominal distension (Table 3).

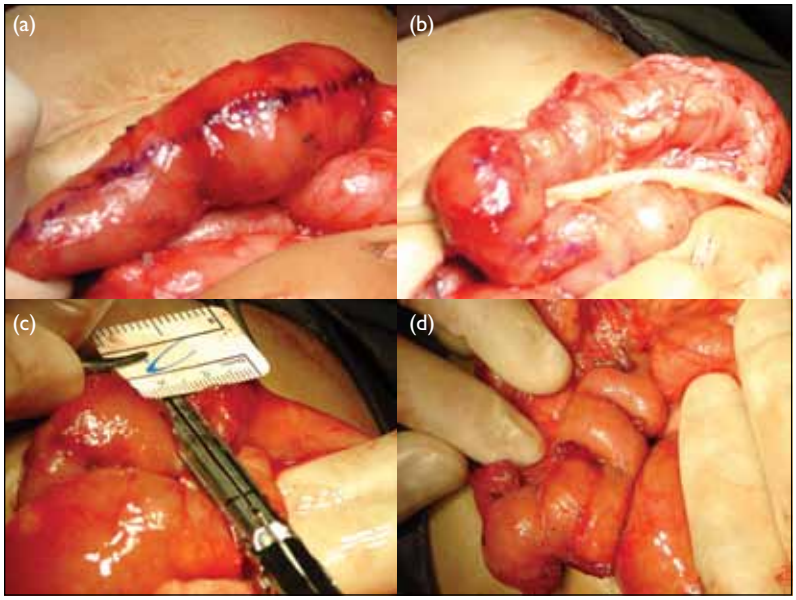

FIG. The serial transverse enteroplasty procedure
(a) The anti-mesenteric longitudinal border of the small bowel was marked. (b) A small mesenteric window was created into which a 16F Foley catheter could be passed through. (c) The bowel was kept flattened with the mesenteric border in the middle on firing of the stapler. (d) Firing of the stapler was repeated up to the proximal jejunum near the duodenojejunal junction

Discussion

Short bowel syndrome is a condition of intestinal insufficiency, causing malnutrition and failure to thrive. Residual small bowel length is the most critical factor influencing prognosis.[2,10] It is still not clear whether the presence of the ileocaecal valve and the length of colon play a significant role in the long-term outcome.[10,11] The management of SBS requires a multidisciplinary approach. Advances in amino acid–based enteral formula and introduction of fish-oil emulsion in PN have improved nutritional absorption in SBS patients.[12,13] In our series, amino acid–based

TABLE 2. Results of serial transverse enteroplasty (STEP) operations: weight-for-age and weight-for-height

Patients	Post-STEP follow-up period (months)	Weight-for-age z score‡		Weight-for-height z score‡	
		Pre-STEP	Post-STEP	Pre-STEP	Post-STEP
A*†	40/9	-3.35	0.74	-3.00	0
B*	32/19	-1.41	-1.41	0	0.67
C†	30	-3.42	-3.27	-0.67	0
D	23	-0.16	0.78	0	1.28

* Repeated STEP operation done, follow-up period after first/second STEP respectively; pre-STEP parameters measured before first STEP procedure
† Fish-oil emulsion parenteral nutrition was given
‡ From "Child growth standard, World Health Organization website": http://www.who.int/childgrowth/standards/en/

TABLE 3. Results of serial transverse enteroplasty (STEP) operations: nutrition, bilirubin level, and loperamide (anti-diarrhoeal) dosage

Patients	Parenteral nutrition given (Kcal/kg/day)		Estimated enteral nutrition tolerance (%)		Total bilirubin (μmol/L)		Loperamide (mg/kg/day)	
	Pre-STEP	Post-STEP	Pre-STEP	Post-STEP	Pre-STEP	Post-STEP	Pre-STEP	Post-STEP
A*†	66.6	57.0	28	47	206	30	0.28	0.70
B*	33.6	0	40	100	17	11	0.18	0.18
C†	54.0	51.3	10	19	79	15	0.25	0.20
D	79.7	47.7	20	47	5	4	0.30	0.66

* Repeated STEP operation done; pre-STEP parameters measured before first STEP procedure
† Fish-oil emulsion parenteral nutrition given

enteral formulae were given to all patients before and after STEP. Enhancement of adaptation after gut resection, pharmacological agents for intestinal motility, and aggressive treatment of SBBO also play important roles in SBS management.[3,14]

Short-term survival after small bowel transplant had improved remarkably over the past 20 years, but the 5-year survival was still only 50 to 60%.[5] After Kim et al[9] described the first operation in 2003, STEP has become a popular bowel-lengthening procedure for patients with SBS. Compared to the Bianchi procedure,[6] STEP is technically less complicated, and requires no bowel anastomosis. The procedure also works well in unevenly dilated bowel, which is a common feature in SBS patients with SBBO.[9] More importantly, STEP can be performed in patients who have undergone a prior bowel-lengthening procedure,[15] which was consistent with the repeated STEP operations performed in two of our patients.

There was no intra-operative staple line leakage in our series, though one patient had early postoperative small bowel obstruction and symptoms that resolved on conservative management. Modi et al[16] reported early postoperative complications as follows: staple line leakage (5.3%), bowel obstruction (5.3%), intra-abdominal abscess (2.6%), and intra-abdominal haematoma (2.6%). We believe the application of a figure-of-eight reinforcement stitch at end of staple line can prevent leakage. Also, accurate intra-operative measurement to create a new bowel channel of at least 2 cm wide can prevent bowel obstruction. One patient in our series had gastrointestinal bleeding 13 months after STEP, which could have been due to staple ulceration or ischaemic enterocolitis. Ching et al[17] reported gastrointestinal bleeding in 19% of patients after STEP after a median follow-up period of 23 months.

The preoperative small bowel length in our patient cohort was ultra-short, ranging from 10 to 40 cm (median, 17.5 cm). In all patients, the ileocaecal valve was also resected in the initial procedure before STEP. All patients depended on long-term PN, with enteral nutrition tolerance of 10 to 40%. In patients A and C, the nutrition was inadequate before the STEP operation as evident by the low weight-for-age and weight-for height *z* scores. Nutrition by the parenteral route was complicated by the poor liver function.

In short gut syndrome, supplementary enteral feeding is given orally and via gastrostomy. However, the amount of enteral feeding given is not the same as that absorbed. The actual amount of enteral nutrition absorbed by the gut is difficult to assess. In our series, the estimated enteral calorie uptake was calculated from the difference between expected total calorie requirement and the calories given via the PN. This estimate could be inaccurate, especially in patients A and C as before STEP they had severe failure to thrive with malnutrition. Also, the increased enteral tolerance could be the result of increases in bowel length after STEP and facilitation of nocturnal gastrostomy feeding.[18] Titration of loperamide as an anti-diarrhoeal agent was also an important factor that could affect enteral absorption. Thus, assessing the effect on enteral nutrition due to STEP alone was difficult. Nevertheless, SBS requires a multidisciplinary input as an adjunct to STEP management.

The post-STEP small bowel length in our series was 15 to 72 cm, with a median increase of 90%. Also the width of the bowel was narrowed to 2 to 2.5 cm after STEP, which preserved bowel motility and prevented SBBO.[19] Kaji et al[20] suggested that after the STEP operation there was an increase in jejunal villus height and decreased rate of crypt cell apoptosis. In our series, the median increment of enteral tolerance after STEP was 113%. One of our patients had received PN for 17 years (after massive gut resection from treatment of retroperitoneal yolk sac tumour). She was weaned from PN and had her central venous catheter removed 16 months after her second STEP operation. In the first report of the international STEP data registry on 38 patients, the mean small bowel length increased from 68 to 115 cm, representing a 69% relative increase. The overall postoperative improvement in enteral tolerance was 116%.[16] In a recent report from the Boston group on 16 patients with long-term follow-up after STEP, six (38%) were weaned from PN after a median of 248 days.[17]

In patients A and C, lipid PN was given at a dose of 1 g/kg/day both before and after STEP, and soybean-oil emulsion PN was given before STEP. They had deranged liver function and hyperbilirubinaemia pre-STEP, and after the operation the total bilirubin normalised. We believe that the improvement could also be attributed by the introduction of fish-oil emulsion to the PN at post-STEP 6 and 3 months. Sudan et al[21] described liver disease being reversed in 80% of patients after bowel-lengthening procedures.

In patients B and C, STEP was performed at the age of 14 and 11 years, respectively. The weight-for-age *z* score did not improve significantly after STEP, whereas the weight-for-height *z* score increased after the operation. Patient B had Down's syndrome and a short stature. In patient C, we fired two staples during STEP, increasing the small bowel length from 10 cm to 15 cm only. In other patients, STEP was performed below the age of 3 years, and both the weight-for-age and weight-for-height *z* scores of patients increased thereafter. We suggest that STEP should be considered in selected SBS patients before puberty, to enable catch-up growth. After STEP, our patients had median increases in weight-for-age *z* scorer of 0.023 units per month and weight-for-height *z* scores of 0.039 units

per month. The median enteral tolerance improved 0.82% per month. The increase in growth parameters and enteral nutrition tolerance was comparable to results reported in other literature.[17,21,22]

Patients with a residual small bowel length of less than 10% of the expected length were associated with high mortality and decreased chance of weaning off PN.[2] Patients A and B in our series with preoperative small bowel lengths of 15 cm and 20 cm, respectively, had repeated STEP procedures. Patient B was weaned from PN after the repeated STEP. Repeated STEP was first described in pig models in 2006.[23] The feasibility for repeated STEPs allows SBS patients to gain additional bowel length, which can improve long-term prognosis. Re-dilatation of bowel can occur after STEP, leading to bowel dysmotility, SBBO and worsening enteral tolerance.[24] Technically, the surgeon should be more careful in performing second STEP, as there may be more severe intra-peritoneal adhesions and the bowel may remain in zigzag position after first procedure, making further stapling more difficult.[15,25] However, we should not hesitate performing a second STEP operation if enteral nutrition tolerance begins to plateau or bowel dysmotility recurs after the first operation, especially in patients with ultra-short small bowel lengths.

Management of SBS is challenging and requires a multidisciplinary approach. Together with advances of nutritional regimens and pharmacological agents, STEP helps to improve the enteral nutrition tolerance and growth of SBS patients. Repeated STEP can be performed in patients with ultra-short bowel lengths. Follow-up is important to assess the long-term prognosis of these patients.

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