Long-Term Outcome of Children with Short Bowel Syndrome Treated with a Modification of the STEP Technique Avoiding Mesenteric Defect

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Abstract

Background The Serial Transverse Enteroplasty Procedure (STEP) Registry has reported a 47% success to achieve enteral autonomy in pediatric short bowel syndrome (SBS). We have performed the STEP with a technical modification (MSTEP) consisting in stapler application without mesenteric defects that can also be applied to the duodenum. Our experience with this technique is described.

Materials and Methods In this study, 16 children with SBS underwent MSTEP (2005–2019). Indications were nutritional autonomy achievement (n = 11, with duodenal lengthening in 5/11) and bacterial overgrowth treatment (n = 5).

Results With a median follow-up of 5.8 years (0.7–13.7 years), 5 of 11 (45%) patients achieved enteral autonomy, 4 of them with duodenal lengthening. Four of four who preserved >50% colon, while only one of seven with <50% of colon achieved enteral autonomy (p < 0.05). After redo procedures, three of four attained enteral autonomy. Thus, 8 of 11 (73%) progressed to enteral autonomy, including all with duodenal lengthening. One child, already parenteral nutrition free, died due to central line sepsis. All the patients from the bacterial translocation group improved their metabolic/nutritional status, but one required subsequent enterectomy of the lengthened intestine due to multiple ulcers in the staple lines.

Conclusion The effectiveness of MSTEP to achieve enteral autonomy seems similar to the classical STEP. It can be applied to the duodenum. The retained colon length may influence the post-STEP enteral autonomy achievement.

Keywords ► short bowel syndrome ► intestinal lengthening ► serial transverse enteroplasty

Introduction

Serial transverse enteroplasty procedure (STEP) is an ingenious intestinal lengthening technique described in 2003 to treat the subset of patients with short bowel syndrome (SBS) and dilatation of the remnant small bowel.1 Similarly, but in a different way to the Bianchi procedure, it increases the mucosal surface area improving absorption of enteral nutrients and at the same time taper the dilated intestine to reduce the risk for bacterial overgrowth.2 From the technical standpoint, it consists in sequential linear stapler applications alternating from opposite directions in 90 and 270 degrees orientation to the mesenteric axis of the dilated intestine beyond the ligament of Treitz, to create a zig-zag channel.1,3 A mesenteric defect is required to insert the stapler arm at each staple line. Theoretically, since the staplers are not adapted to children size, particularly small...
children, the mesenteric defect diameter required to introduce the stapler jaw might jeopardize the intestinal blood supply. In addition, such defect might be a source of morbidity in the future with the development of internal hernias. For this reason, since 2005, we have invariably performed the STEP with a technical modification (MSTEP) which consists in applying the stapler applications in the same fashion without the creation of the mesenteric defects to insert the stapler arm (Fig. 1). Thus, in cases of associated duodenal dilatation, we were able to apply the same principles to the duodenum gaining an additional length and tapering the duodenal diameter. The aim of this study is to describe the results of children with SBS treated with the MSTEP at our institution, associated or not to duodenal lengthening with at least 1 year follow-up.

Materials and Methods

After approval from the Hospital Institutional Review Board, the medical records of patients with SBS who underwent the MSTEP were reviewed, and data collected retrospectively. From June 2005 to March 2019, 16 children (8 males and 8 females) were surgically treated with MSTEP. All the patients had developed the SBS at the neonatal period. The causes of intestinal failure were necrotizing enterocolitis (n = 6), gastroschisis with intestinal atresia or vanishing (n = 5), Hirschsprung’s disease (n = 2), midgut volvulus (n = 2), and intestinal atresia (n = 1). Median age and weight at MSTEP were 5.2 years (range: 0.25–19 years) and 13.8 kg (range: 3–32 kg), respectively. Preoperatively, as part of our protocol, an upper gastrointestinal radiographic series and barium enema to assess the presence of distal mechanical obstruction or stricture were performed in all children.

Surgical Procedure

Written informed consent was obtained from all parents prior to the procedure. Prophylactic preoperative intravenous (IV) antibiotics and IV fluconazole were administered to all patients. Under general anesthesia, a mid-line incision was made, either anew or through the previous scar. After all the bowel loops had been completely separated, the small intestinal length was measured from the ligament of Treitz down to the retained colon. The stapler firings were performed sequentially starting on the proximal dilated jejunum using the MSTEP technique as described earlier (Fig. 1A–C). In cases of dilated duodenum, a concomitant duodenal lengthening procedure was performed as we have previously described. To minimize gastrointestinal bleeding, the stapler jaws were maintained on the intestinal wall 1 minute closed before firing to act as hemostatic. After our second case, corners of the cutting line were inverted with one or two 4–0 Silk or Vicryl sutures and the vertexes of the cutting line were reinforced with a U-fashion stitch to prevent intestinal leaks. A Jackson–Pratt’s drain was placed to detect postoperative leakages. In the beginning of our experience, the patients underwent a radiologic gastrointestinal contrast study on the seventh postoperative day to rule out stricture or leakage before the initiation of enteral nutrition. The Jackson–Pratt’s drain was removed the same day of the study if no leaks were detected. Antibiotics and IV fluconazole were administered postoperatively for 48 hours.

For the purpose of analyzing our results, our patients were divided in two groups based on STEP indication. Group I (n = 11) named “intestinal lengthening” group with parenteral nutrition (PN) dependence. The main goal in this group was to achieve enteral autonomy defined as full enteral nutrition with permanent discontinuation of PN support. Group II (n = 5) named “intestinal tailoring” group with PN independence. In this group, the patients had dilated bowel resulting in severe metabolic and/or electrolytes disturbances owing to bacterial overgrowth and the MSTEP was applied as a tailoring technique to improve/solve their symptoms.

Intestinal Lengthening Group (n = 11)

Seven children had history of prematurity. The median remnant intestinal length measured at the time of neonatal surgery was 32 cm (range: 0–75 cm) and increased to 50 cm (range: 0–120 cm) at the moment of the MSTEP. The median age and weight at primary MSTEP were 3.8 years (range: 0.2–11 years) and 11.2 kg (range: 3–23.2 kg), respectively. At the time of the procedure, 6 of 11 patients had developed duodenal dilatation and 5 of these 6 underwent duodenal lengthening. Of these five, four were combined with the MSTEP (mean intestinal length preprocedure was 46 cm), and one child underwent only duodenal lengthening because the child just retained the duodenum anastomosed to the ascending
colon. The remaining patient with duodenal dilatation did not undergo the duodenal procedure as the patient’s MSTEP took place prior to the technique development.

Only 1 (9%) of the 11 patients preserved the ileocecal valve and 7 (64%) preserved ≤ 50% of the colon, considering the splenic flexure of colon as the midpoint. In further classifying these 11 patients regarding their retained colons, 1 patient retained the whole colon with the ileocecal valve, 1 the ascending colon, 2 the transverse colon, 2 the left colon, and 3 the sigmoid colon. Concerning two Hirschsprung’s patients with no retained colon, one had undergone a Bianchi procedure at the age of 3 years, and retained 60 cm of ganglionic jejunum. This last patient was the only one from the series who required an ostomy of the lengthened intestine at the time of primary MSTEP. Regarding the central venous access, 4 of 11 preserved only one access.

### Intestinal Tailoring Group (n = 5)

The median age and weight at surgery were 7.7 years (range: 1.1–18 years) and 19.6 kg (range: 7–32 kg), respectively. All patients had history of prematurity and were on full enteral nutrition. Three patients had developed onsets of lactic acidosis requiring high dose of bicarbonate and frequent hospital readmissions. All children showed severe malnutrition and gastrointestinal symptoms including severe abdominal distension and pain. None of the patients had developed duodenal dilatation, one retained the ileocecal valve, and all preserved > 50% of colon. Considering the central venous access, each patient preserved more than two patent accesses. Only one patient had undergone previous alogous gut reconstruction in the form of a reversal antiperistaltic jejunal loop procedure at the age of 1 year.

Additionally, statistical analysis to evaluate the impact of retained colon on PN free-state achievement in Group I was performed with a 2 × 2 contingency table and Fisher’s exact probability test due to the short sample. A p-value of < 0.05 was considered to have statistical significance.

### Results

#### Intestinal Lengthening Group (n = 11)

The mean age at surgery was 3.9 years (range: 1–10 years) and 18.3 kg (range: 7.6–32 kg), respectively. One patient had undergone a Lester Martin procedure at the age of 3 years, retaining 60 cm of ganglionic jejunum. This patient was the only one in the series who required an ostomy of the lengthened intestine at the time of primary MSTEP. Regarding the median follow-up of the lengthened intestine resembled a party streamer serpentine.

#### Intestinal Tailoring Group (n = 5)

The mean stapler firings was 12 (range: 6–22) and the final post MSTEP intestinal length measured was 143 cm (range: 105–180 cm). All patients are alive with a median follow-up of 3.9 years (range: 1.9–9.5 years). Neither duodenal lengthening nor redo procedure was required in this group.

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 Featured Image: Surgical picture of the final intestinal appearance after the modified serial transverse enteroplasty procedure. The lengthened intestine resembles a party streamer. 

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**Fig. 2** Surgical picture of the final intestinal appearance after the modified serial transverse enteroplasty procedure. The lengthened intestine resembles a party streamer serpentine.
Gastrointestinal symptoms improvement, reduction of bouts of lactic acidosis and electrolytes disturbances, and weight gain were observed in all patients after the tapering procedure.

Surgical Complications
After primary MSTEP, three cases (19%) developed surgical-related complications, one in the immediate postoperative course and two in the long term. The immediate postoperative surgical complication occurred in our second case in the form of an intestinal leak from one of the stapler lines. This was resolved surgically. Concerning the long-term complications, one had intermittent intestinal subocclusions secondary to food transgressions (big sunflowers seeds) and in one child from Group II, an ulcer developed in one of the distal staple lines 8 months after MSTEP that leads to chronic anemia from bleeding and required blood transfusion. This case initially was unsuccessfully managed first with oral antibiotics to treat bacterial overgrowth, then with endoscopic argon-beam cauterization, and finally, with wedge surgical resection of the stapler line and subsequent intestinal handsewn suture. However, new ulcers appeared 1 year later in the last three consecutive stapler lines which did not respond to hyperbaric oxygen treatment. The entire lengthened intestine was removed and with 8 months of follow-up, the child is doing well with enteral autonomy. Finally, after redo procedures in the intestinal lengthening group, one patient developed an intestinal leak at the vertex of a 0-degree staple line.

Discussion
In the original description of the classical STEP technique, the choice of applying the 90 to 270 degrees stapler line orientation through a mesenteric defect instead of the 0 to 180 degrees was because the bowel appeared to lay better and less likely to twist or kink. However, the required mesenteric defect with this orientation is at each staple line, double the defects than if the choice would be the 0 to 180 degrees orientation which requires only defects at every other staple line. The main reason that leads us to modify the original STEP in 2005 with the same staple line orientation but without the creation of such defects was to eliminate a potential source of morbidity as previously explained. Although reports of STEP postsurgical complications are scarce, it is fair to remark that our initial fear may probe to be unfounded since to date, after more than 15 years of accumulated experience worldwide with the procedure, there is no description of complications related with the mesenteric defects. Why the need to make mesenteric holes? In the experimental field, Kim et al found that if the mesenteric defects were not created, the staples tend to be placed at less than 180 degrees apart and it could facilitate the development of bowel diverticula, rather than a smooth zig-zag channel. In our experience, initially, the lengthened intestine resembles a party streamer serpentine (Fig. 3), but it straightens with time acquiring a normal appearance (Fig. 4A–C).

As expected, and with the same pre-STEP mean intestinal length (50 cm), our primary success rate (45%) to achieve a free PN state after MSTEP was similar to the 47% reported by the International STEP Data Registry. However, the mean time to achieve a full enteral tolerance state in our series was significantly shorter (11 months) than in the registry (21 months). This fact may be, at least in part, explained by the bias that comes from diverse medical management styles since the registry included cases from 50 centers. However, other anatomical factors which favor the intestinal adaptation, such as the ileocecal valve were retained more frequently in the patients from the registry than in our series (31 vs. 9%). In addition, the length of the retained colon seems to influence the achievement of enteral autonomy after primary MSTEP since the four children who preserved ≥ 50% colon attained it, as opposed to only one of seven of those who preserved < 50% colon. Furthermore, although duodenal dilatation seems a frequent complication in patients with SBS and PN (54% of our population); to date, there are no data provided by the literature regarding this issue. We hypothesized that the attainment of enteral autonomy in a shorter period of time in our cohort could be related with the treatment of the duodenal dilatation which may interfere with the adaptation process by promoting intestinal dysmotility and bacterial overgrowth. In fact, the application of the MSTEP to the duodenum in five patients allowed not only tailoring the dilatation but gaining 21% of additional staple firings, resulting in successful PN withdrawal in all of them, four primarily and one after redo procedure.

Another factor which interferes with the adaptation process is intestinal redilatation, the most frequent post-STEP complication occurring up to 30 to 40% of the cases. Interestingly, although we observed a similar rate with the MSTEP, so far duodenal redilatation following duodenal lengthening did not occur, perhaps owing to the anatomical attachment of the duodenum to the pancreas. This said, there are controversial data regarding the benefits of redo procedures after intestinal redilatation. While a recent study suggests that the redo procedures are not cost-effective,
Wester et al reported a similar chance to be weaned off PN support after a re-STEP compared with the primary STEP.\textsuperscript{10,11} In our experience, the overall success rate to attain enteral autonomy jumped from the initial 45 to 73\% following the re-STEP. Currently, in cases of redo procedure, our choice is to use the 0 to 180 degrees orientation (up and down) stapler application to avoid the cross with the 90 to 270 degrees (lateral) staple lines from the primary MSTEP.

Finally, the reported incidence of post-STEP surgical complications has ranged between 14 and 26\%, and is mainly related with the use of surgical stapling devices.\textsuperscript{6–8} Infrequent but worrisome, the apparition of ulcerations along the staple line months or years after surgery has been described and leads to chronic gastrointestinal bleeding and transfusion requirement.\textsuperscript{7,12–14} Of unknown origin, as is in our case, they appear more in the distal staple lines, closer to the colonic anastomosis, suggesting bacterial overgrowth as one of the possible etiologic factors.\textsuperscript{12,14}

**Conclusion**

In summary, with the main limitation of sample size in this analysis, the MSTEP seems at least as effective to achieve a free PN state as the classical STEP. The main advantage is the possibility to apply this technique to the duodenum in cases of duodenal dilatation. In addition, the length of the retained colon may have an influence in the ability to achieve enteral autonomy after the procedure.

**Conflict of Interest**

None declared.

**References**


