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First STEPs: Serial transverse enteroplasty as a primary procedure in neonates with congenital short bowel

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ABSTRACT

Background: Since its introduction as an alternative intestinal lengthening technique, serial transverse enteroplasty (STEP) has been increasingly used as the surgical treatment of choice for patients with refractory short bowel syndrome (SBS). While primary STEP for the treatment of congenital conditions was proposed in the original description of the procedure, emphasis was placed on a delayed or staged approach to these patients. To date, a comprehensive review of the outcomes from this sub-population has not been reported by the International STEP Data Registry.

Methods: A retrospective review of the International STEP Data Registry was performed to identify all patients who underwent STEP as a primary operative procedure for the treatment of congenital SBS. Changes in pre- and post-STEP values were assessed using paired t-tests with significance set at $p < 0.05$. Data are presented as mean ± standard deviation.

Results: Fifteen patients underwent primary STEP for congenital SBS between September 1, 2004, and April 10, 2012. Thirteen patients had follow-up information available. Causes of congenital SBS included closing gastroschisis, small bowel atresia, and midgut volvulus. Twelve patients had pre- and post-STEP bowel measurements taken. Average pre- and post-STEP bowel lengths were 32 ± 16 cm and 47 ± 22 cm, respectively. Intestinal length was increased by a mean of 15 ± 12 cm for a relative small bowel length increase of 50.4 ± 27.3% ($p < 0.001$). Only one patient required an ostomy at the time of primary STEP. A second patient required a temporary ostomy at 3 months of age that was later closed. There was one death from intestinal failure associated liver disease (IFALD). Another patient experienced IFALD progression and required liver and intestinal transplantation. The most commonly reported complication following primary STEP was obstruction or bowel re-dilatation requiring additional operative interventions. Nine patients underwent second STEP procedures under these circumstances. Eight patients remain dependent on parenteral nutrition, while three patients achieved enteral autonomy.

Conclusions: Primary STEP is a feasible and safe surgical option for the treatment of congenital conditions resulting in SBS. Primary STEP establishes early bowel continuity, creates intestinal length from congenitally dilated bowel, and appears to obviate the need for interval stomas and their associated loss of bowel length in neonates with congenital SBS. However, with recent changes in SBS management emphasizing intestinal rehabilitation, additional studies are needed to assess the long-term impact on intestinal adaptation of STEP performed in the neonatal period prior to adoption of this technique.

Short bowel syndrome (SBS) is owing to the anatomic or functional loss of a large segment of small intestine and results in intestinal malabsorption [1]. Although the worldwide incidence has not been reported, a substantial portion of SBS cases are of congenital etiology and culminate in considerable loss of small intestine length [2,3]. Surgical management of congenital SBS focuses on the principles of bowel conservation and restoration of intestinal continuity [3]. However, conservation principles often give way to practical considerations at the time of the initial surgery resulting in resection of dilated intestinal segments or ostomy creation. Primary serial transverse enteroplasty (STEP) is an alternative approach that offers the potential benefits of promptly establishing intestinal continuity and avoiding further loss of intestinal absorptive area [3–5].

The STEP procedure was first described in 2003 as an alternative intestinal lengthening technique for patients with refractory SBS [6].

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Similar to other bowel lengthening procedures, STEP lengthens the intestine and narrows bowel diameter with the intent of improving peristalsis and preventing small bowel bacterial overgrowth, malabsorption and sepsis [6,7]. In contrast, STEP has gained favorability as its novel approach is intuitive, relatively easy to perform, poses minimal threat to intestinal blood supply, can be utilized for varying degrees of bowel dilatation, and is repeatable should bowel dilatation recur [1,7]. Accepted STEP indications include intestinal failure and bacterial overgrowth refractory to maximal medical management, as well as neonatal intestinal atresias or obstructions with limited small bowel length and a dilated proximal segment [6,7]. Though neonatal intestinal atresias and other causes of congenital SBS have long been considered indications for STEP, the outcomes for this subgroup have not been formally investigated. This study sought to determine the safety and utility of primary STEP by reviewing the short- and long-term outcomes of this subgroup.

1. Methods

Data were obtained from the International STEP Data Registry. The STEP registry was created in 2004 approximately one year after the procedure's introduction at Boston Children's Hospital. Through voluntary reporting, the registry has accrued case data on over 200 patients from more than 45 centers worldwide. Patient variables collected in the registry include patient demographics, primary diagnosis and coexisting medical conditions, operative information, complications, and outcomes.

Study approval was granted by the Kapi'olani Medical Center for Women and Children institutional review board and approval to use the International STEP Data Registry was granted by the registry personnel at Boston Children's Hospital. The registry was searched to identify all patients who underwent primary STEP for congenital short bowel syndrome (SBS). Data reflect patients treated between September 1, 2004 and April 10, 2012. For patients with incomplete registry data, email and telephone requests with referring surgeons were made. The patient variables used for analysis include gestational age, primary diagnosis, pre- and post-STEP bowel measurements (length and diameter), enteral tolerance, complications, ostomy creation, disease progression with development of intestinal failure associated liver disease (IFALD), need for transplantation, and mortality. Statistical analysis was performed using SPSS software (version 19.0, SPSS Inc./IBM, Chicago, IL). Mean change in bowel length as well as percent change was assessed using paired t-tests with significance set at $p < 0.05$. Data are presented as mean ± standard deviation.

2. Results

Fifteen patients from 13 medical centers who had undergone primary STEP for congenital SBS were identified. Thirteen patients had follow-up information available. Mean gestational age was 35 weeks. Causes of congenital SBS include intestinal atresias ($n = 9$), closing gastroschisis ($n = 5$), and midgut volvulus ($n = 1$) (Fig. 1). All patients with operative measurements ($n = 12$) experienced intestinal tapering and gained intestinal length with the STEP procedure. Mean intestinal width was tapered from 4.4 ± 1.0 cm to 1.7 ± 0.3 cm ($p < 0.001$). Pre-STEP bowel length ranged from 10 to 66 cm (mean = 32 ± 16 cm). Post-STEP bowel length varied between 15 and 89 cm (mean = 47 ± 22 cm). Intestinal length was increased by a mean of 15 ± 12 cm translating to a relative increase in small bowel length of 50.4 ± 27.3% ($p < 0.001$; Fig. 2).

Few short-term complications and challenges to restoring or maintaining bowel continuity were described. Only 1 patient required ostomy creation at the time of primary STEP. A second patient required a temporary ostomy at 3 months of age that was later closed. No intra-operative complications were reported.

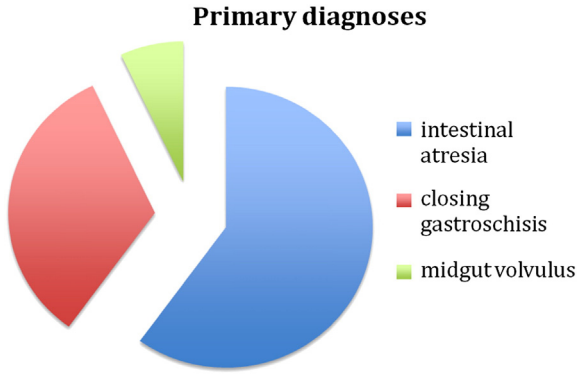

Fig. 1. Primary diagnosis in 15 patients undergoing primary serial transverse enteroplasty.

Over time, the most commonly reported long-term complication following primary STEP was obstruction or bowel re-dilatation requiring additional operative interventions. Nine patients underwent second STEP procedures under these circumstances. Late complications were related to the development and progression of intestinal failure associated liver disease (IFALD). One death resulted from IFALD while another patient required liver and intestinal transplantation (Fig. 3).

As all study subjects were neonates, pre-STEP enteral tolerance percentages were not applicable. Eleven patients had post-STEP follow-up data regarding enteral tolerance. Eight patients still require parenteral nutrition (PN) with 5 meeting half their daily caloric needs enterally. Full enteral autonomy was achieved in 3 patients.

3. Discussion

Primary STEP has been described in case reports as a useful tactic for managing the dilated proximal bowel segment in neonatal intestinal atresias. Few intra-operative or technical complications occurred, and the primary intent of preserving bowel length while providing the necessary intestinal tapering for a primary anastomosis was accomplished [4,5,8]. Using data from the International STEP registry, this current report is the first to review the short and long-term outcomes of a cohort of patients undergoing primary STEP.

Short-term outcomes resemble those reported in all-inclusive STEP studies. The STEP procedure was rapidly adopted as early investigations demonstrated its value as a simple technique with easily reproducible results, namely increased bowel length, and few technical drawbacks [7–11]. We found that primary STEP can similarly be performed in neonates with congenital short bowel with few complications in the peri-operative period. While staple

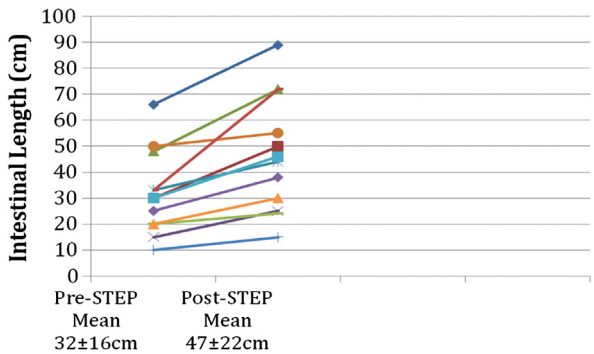

Fig. 2. Graphic representation of the intestinal lengthening achieved in 12 of 15 patients undergoing primary serial transverse enteroplasty (STEP). Mean intestinal length increased by 15 ± 12 cm for a 50.4 ± 27.3% relative increase in small intestine length ($p < 0.001$).