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Intestinal lengthening and nutritional outcomes in children with short bowel syndrome

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Abstract

BACKGROUND: Intestinal lengthening remains a treatment option in pediatric short bowel syndrome. However, clinical indications and nutritional outcomes from bowel lengthening are not well defined.

METHODS: A retrospective review of a prospective database was conducted of patients who underwent bowel lengthening using serial transverse enteroplasty (STEP) at a single center.

RESULTS: Sixteen children who underwent 19 STEP procedures were identified. STEP increased median small bowel length from 84 cm (range, 19 to 295 cm) to 103 cm (range, 24 to 375 cm) (P = .0001). Caloric provisions from parenteral nutrition were decreased after STEP (69% vs 0%, P < .01). Of 15 subjects in the nutritional analysis, 12 (80%) had improved enteral tolerance, and 9 (60%) achieved enteral autonomy after STEP. Six STEP procedures were not associated with improvements in enteral nutrition, and 5 (83%) were performed in children with gastrochisis.

CONCLUSIONS: A majority of children with short bowel syndrome were weaned off parenteral nutrition after STEP. Gastrochisis may portend a less optimal outcome from the procedure.

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The medical and surgical management of short bowel syndrome, the most common etiology of intestinal failure in children, remains a challenge. These patients require complex clinical care that incorporates advanced nutritional support, prevention of sepsis, treatment of liver dysfunction, and surgical care for ostomies, gastrostomies, central venous access, and intestinal reconstruction. Despite these clinical challenges, a distinct improvement in the long-term outcome of children with short bowel syndrome has been reported within the past decade.1-4

Operative bowel lengthening, a form of autologous intestinal reconstruction, represents an innovative surgical strategy to improve the enteral feeding tolerance of children with short bowel syndrome. The goals of bowel lengthening in pediatric short bowel syndrome are two-fold: (1) the small bowel remnant is lengthened to potentially increase the mucosal surface area, slow luminal transit time, and allow improved nutrient absorption; and (2) the dilated small bowel is tapered so that bowel motility is optimized and the risk for bacterial overgrowth is reduced. Various surgical techniques for bowel lengthening have been described in the pediatric population, including the classic longitudinal intestinal lengthening and tapering procedure, as described by Bianchi,5 and the more recent serial transverse enteroplasty (STEP).5,6

Introduced in 2003, the STEP technique for bowel lengthening has quickly become a widespread tool for the surgical management of pediatric intestinal failure.6-8 The operation involves elongating the remnant small bowel by
applying sequential intestinal stapling devices from alternating sides in a plane perpendicular to the long axis of the bowel. The technique results in a lengthened small bowel in a zig-zag configuration and, in turn, also tapers the bowel. The procedure has been described as a safe technique in that the stapler applications are placed parallel to the mesentry, and therefore the blood supply to the intestine is not put at risk. To date, there are limited data regarding contemporary outcomes from intestinal lengthening for pediatric short bowel syndrome. Perhaps more important, there is still no consensus as to which subsets of children with intestinal failure will benefit from operative bowel lengthening. The purpose of this study was to review our institution’s experience with bowel lengthening for short bowel syndrome. We specifically sought to analyze nutritional outcomes after the STEP procedure and whether subgroups of children with short bowel syndrome may benefit differently from the operation.

Methods

Since 2005, the Seattle Children’s Hospital Intestinal Failure Program has served as a regional referral center for children of all ages with pediatric intestinal failure and short bowel syndrome. Our program prioritizes intestinal rehabilitation and reserves solid organ transplantation for patients with refractory short bowel syndrome and the failure to advance on enteral nutrition over time. Outcomes in our program have compared favorably with those at established intestinal failure treatment centers. Our indications for operative bowel lengthening include children with short bowel syndrome who have developed sufficiently dilated small bowel remnants and cannot advance further on enteral nutrition or have developed frequent complications from bacterial overgrowth in the dilated intestinal segment.

The Seattle Children’s Hospital Intestinal Failure Program maintains a prospective database that includes baseline patient information at the time of referral and subsequent data from inpatient and outpatient encounters. After approval from the Seattle Children’s Hospital Institutional Review Board (IRB #12646), a retrospective review of patient data was performed from this prospectively collected database. Additional variables for the purpose of this study were collected in a retrospective fashion using the hospital’s electronic medical record. All children who underwent bowel-lengthening procedures at our institution from 2004 (at the inception of our program) to 2011 were identified from this prospective database and included in the data analysis. For portions of the nutritional analysis, each STEP operation was analyzed as an individual occurrence, so as to best describe outcomes from the procedure. Data were collected from each subject’s initial encounter with the Intestinal Failure Program (either inpatient or outpatient) and until most recent clinical follow-up.

Intraoperative small bowel length measurements were performed using a sterile silk suture placed along the antimesenteric border of the bowel. For each STEP procedure, the remnant small bowel length and diameter were measured immediately before and after the STEP bowel-lengthening technique.

Each patient was evaluated by a dedicated intestinal failure nutritionist during all inpatient and outpatient encounters. The percentage of calories from parenteral nutrition (PN) was used to describe patients’ nutritional provisions and was calculated as the percentage of total caloric intake provided by PN and its lipid component. We defined enteral autonomy as full enteral nutritional provision with permanent discontinuation of PN support. For evaluation of anthropometric data, patients who underwent intestinal transplantation or died were removed from the analysis.

Descriptive measures were used where appropriate to summarize patient demographics and characteristics. Continuous variables are described as median (range). The nonparametric Wilcoxon’s signed-rank test was incorporated to compare outcomes before and after bowel lengthening. Stata version 11.0 (StataCorp LP, College Station, TX) was used for data analysis.

Results

Of 104 patients enrolled in the Intestinal Failure Program, 16 children underwent a total of 19 bowel-lengthening procedures. All patients underwent the STEP bowel-lengthening technique, including 3 patients who underwent 2 STEP operations each. The primary gastrointestinal diagnoses of patients who underwent bowel lengthening with STEP are outlined in Table 1. STEP was performed at 1 year of age (range, 0.1 to 18 years), and subjects were followed clinically for 26 months (range, 1 to 55 months) postoperatively. Eight of the 16 patients (50%) were referred from outside our institution.

STEP increased the remnant small bowel length from 84 cm (range, 19 to 295 cm) to 103 cm (range, 24 to 375 cm) (P < .001). On average, the small bowel remnant was lengthened by 38% (range, 4% to 72%). The procedure resulted in significant tapering of the small bowel diameter from 4.75 cm (range, 4 to 10 cm) to 2 cm (range, 1.5 to 4 cm) after STEP (P < .01), with an average luminal

<table>
<thead>
<tr>
<th>Primary Diagnosis</th>
<th>Frequency</th>
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<tbody>
<tr>
<td>Gastroschisis</td>
<td>5</td>
</tr>
<tr>
<td>Necrotizing enterocolitis</td>
<td>5</td>
</tr>
<tr>
<td>Gastroschisis with atresia</td>
<td>3</td>
</tr>
<tr>
<td>Jejunoileal atresia</td>
<td>2</td>
</tr>
<tr>
<td>Meconium disease</td>
<td>1</td>
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</tbody>
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tapering of 57% (range, 44% to 67%). Nine (range, 4 to 31) stapler loads were used to lengthen the bowel.

One subject was enterally autonomous before STEP and was excluded from the nutritional analysis. After STEP, 12 of the remaining 15 children (80%) had improved enteral feeding, and weight-for-age anthropometric data were improved (Table 2). Caloric provisions from PN were decreased from a baseline value of 69% (range, 26% to 100%) to 0% (range, 0% to 100%) after STEP \( (P < .01; \text{Fig. 1}) \). Nine patients (60%) achieved complete enteral feeding autonomy at 13 months (range, 0.5 to 30 months) postoperatively. Patients who were weaned completely from PN had lower baseline caloric requirements from PN before bowel lengthening (52% vs 100%, \( P < .05 \)).

Six of the 19 STEP procedures were not associated with improvements in enteral tolerance. Five of these procedures were performed in children with primary diagnoses of gastroschisis or gastroschisis with atresia. Fig. 2 presents the caloric requirement from PN after STEP as stratified by the remnant bowel length before STEP. Only 1 child with a remnant small bowel length <50 cm achieved enteral autonomy after STEP in intermediate follow-up. Three patients (19%) underwent second STEP procedures for recurrent small bowel dilation with continued PN dependence, and 1 of these patients ultimately achieved enteral autonomy.

Two subjects (13%) required intestinal transplantation for failure to wean from PN. Two subjects (13%) died from late complications of intestinal failure, including progressive liver failure from PN-associated cholestasis and infection after intestinal transplantation. One child developed gastrointestinal hemorrhage from a staple-line ulcer and required a segmental bowel resection 2 years after STEP. No child currently has biochemical evidence of PN-associated liver disease.

### Comments

Short bowel syndrome in the pediatric population requires chronic and complex care. Because of advances in medical, surgical, and nutritional care in the past decade, overall survival from intestinal failure in children has dramatically improved.\(^1\)\(^-\)\(^4\) In addition, recent data demonstrate that a multidisciplinary approach to the treatment of children with short bowel syndrome may impart a survival benefit.\(^11\)\(^,\)\(^12\) Operative bowel lengthening is one of many tools used to increase enteral tolerance in these patients. For this study, we sought to review our institution’s experience with bowel lengthening within the framework of an intestinal rehabilitation program.

Our findings reflect the limited published outcomes data for contemporary bowel lengthening in children and demonstrate that the STEP procedure imparts a nutritional benefit in certain patients. In this series, 60% of patients achieved complete enteral autonomy and 80% tolerated increasing amounts of enteral nutrition after STEP. Recent studies have demonstrated that bowel lengthening is associated with a 38% to 68% incidence of weaning from PN.\(^8\)\(^,\)\(^13\)\(^-\)\(^15\) The sole publication from the International STEP Data Registry reported a 48% incidence of enteral autonomy after the procedure.\(^7\) The incidence of mortality and the need for intestinal transplantation after bowel lengthening range from 0% to 22% and 12% to 18%, respectively, in the recent literature. Our experience corroborates these data and reinforces the fact that bowel lengthening may not lead to improved outcomes in certain subgroups of patients with short bowel syndrome.

<table>
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<tr>
<th>Variable</th>
<th>Weight-for-age Z score</th>
<th>Height-for-age Z score</th>
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<tr>
<td>Before bowel lengthening</td>
<td>-1.67 (-4.55 to .64)</td>
<td>-1.64 (-5.78 to .44)</td>
</tr>
<tr>
<td>Most recent follow-up</td>
<td>-.60 (-4.31 to 1.31)</td>
<td>-1.34 (-2.06 to .27)</td>
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<td>( P )</td>
<td>.02</td>
<td>NS</td>
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**Table 2** Median (range) anthropometric data immediately before bowel lengthening and at most recent clinical evaluation.
enterocolitis who developed significant dilation of his remnant small bowel and refractory bacterial overgrowth. Resecting the entire dilated segment would have significantly limited his bowel length and jeopardized his enteral tolerance, and therefore STEP was performed primarily for its tapering effect.

In addition, our findings demonstrate that young children with gastroschisis as their primary diagnosis may not share completely in the nutritional benefits from the STEP procedure. In total, our series included 8 patients with gastroschisis who underwent bowel lengthening; of these children, 4 remain partially dependent on PN, 3 required repeat STEP procedures, and 2 were never able to advance on enteral nutrition despite small bowel lengths of 45 and 77 cm, respectively, after STEP. The reason for a less robust effect from the STEP in our children with gastroschisis is unclear. It has been hypothesized that children with gastroschisis and intestinal atresia may have inherent problems with severe intestinal dysmotility. Although previous animal data demonstrate that STEP preserves intestinal motility, it is possible that dysmotility unique to gastroschisis plays some undefined role in intestinal function after bowel lengthening. It is interesting to note that the 2 patients in our series who remained completely dependent on PN after bowel lengthening had both gastroschisis and jejunoileal atresia.

Another unresolved issue in short bowel management is how best to define the ideal candidate for bowel lengthening. To date, the ability to identify patients who should bypass these extensive operations and proceed directly to intestinal transplantation remains a challenge. In the pediatric population, patients who have insufficient bowel length to effectively advance on enteral nutrition and who have developed progressive dilation of their small bowel remnants are routinely considered for operative bowel lengthening. However, there are limited data available to delineate whether bowel lengthening is effective in patients with the shortest bowel lengths. For example, autologous intestinal reconstruction may double the bowel length in a patient with 10 cm of small bowel. But whether this additional length affords a valuable nutritional benefit is not known. In our series, the children who benefited the most from the STEP procedure by weaning off PN were those patients with the longest bowel lengths (Fig. 2) and children who were tolerating a reasonable percentage of enteral nutrition before bowel lengthening. An alternative explanation is that intestinal adaption in patients with the shortest bowel lengths may require more time to achieve enteral advancement. In this way, longer term follow-up would be needed to fully define the benefits of the STEP procedure in this cohort. Regardless, consideration should be given to stratifying clinical and nutritional outcomes by patients’ initial bowel lengths in future studies of bowel lengthening.

This study was limited by the small number of patients who underwent STEP and the fact that there was no valid cohort to compare these data. Although the data in this series are intriguing, the possibility that individual patient factors may have skewed our clinical results cannot be excluded. This is particularly relevant because the population of children with intestinal failure is heterogenous. It is also possible that patients with short bowel syndrome would show similar nutritional benefit from a longer time course of medical management alone. To date, a randomized controlled trial comparing surgical bowel lengthening and medical management in pediatric short bowel syndrome has never been attempted and is likely impractical.

References