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SHORT COMMUNICATION

Serial transverse enteroplasty (STEP): intermediate outcomes in children with short bowel syndrome

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Abstract Short bowel syndrome is the most common cause of intestinal failure in children. The treatment is based on a multidisciplinary approach involving pediatricians, pediatric surgeons, and nutritionists. Surgical procedures for intestinal lengthening may be decisive, having been revalued after the recent description of serial transverse enteroplasty (STEP). We reviewed the patients who underwent the STEP operation for short bowel syndrome in our hospital in order to evaluate medium-term outcome. Between April 2006 and December 2008, three children were submitted to STEP without postoperative complications directly related to the procedure. In two cases the autonomy for oral/enteric feeding was obtained within 3 and 7 months after surgery with sustained growth, persisting at 5 years of follow-up after STEP. One child remained dependent of parenteral nutrition and was submitted to intestinal transplantation 30 months after STEP. However, since STEP until transplantation, it was possible to increase enteric volume and decrease intestinal dilation and the frequency of occlusive episodes. STEP is an effective and safe technique for intestinal lengthening that may allow increased tolerance to oral/enteric feeding or at least alleviate some complications of short bowel syndrome.

Keywords Serial transverse enteroplasty · Short bowel syndrome · Gastroschisis · Intestinal atresia

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Introduction

Intestinal failure is defined as the inability to absorb enough nutrients to maintain body weight or normal growth and development. Short bowel syndrome is the most common cause of intestinal failure in children [17] and can result from congenital anomalies (e.g., intestinal atresia) or acquired conditions (e.g., necrotizing enterocolitis). It affects 24.5 per 100,000 live births and is higher in preterm infants (353.7 per 100,000) [16]. Treatment is based on a multidisciplinary approach involving the intervention of pediatricians, pediatric surgeons, and nutritionists, trying to optimize the adaptation of the remaining intestine with a conservative attitude in order to obtain full enteral tolerance. The extent and type of the remaining intestine (e.g., presence of ileocecal valve), as well as the underlying disease are factors that significantly influence the achievement of full enteral tolerance.

When weaning of parenteral nutrition is not possible, intestinal lengthening surgery, if indicated, can stop the vicious cycle of infection of central venous catheter (CVC), development of liver disease, and malnutrition. This type of procedure has regained interest with the recent description in 2003 of serial transverse enteroplasty [9, 10], a simpler technique than those previously used (Bianchi, Kimura, and Soper) and with promising results. Serial transverse enteroplasty (STEP) is a surgical technique of autologous bowel reconstruction which consists of applying a linear stapler, perpendicular to the longitudinal axis of the intestine and midway between the

mesenteric border and anti-mesenteric, alternating the side, creating a longer narrower intestinal channel [8–10]. Therefore, dilated intestine is a prerequisite to perform the STEP technique. However, scientific publications with series with long and intermediate outcomes are scarce. In this paper we analyze patients who underwent STEP at a university hospital between April 2006 and December 2008 with particular emphasis on the evaluation of postoperative outcomes (clinical and nutritional).

Case reports

Case 1 is a female child, born at 34 weeks of pregnancy, with prenatal diagnosis of gastroschisis and bowel dilatation. In the neonatal period, she underwent correction of gastroschisis and bowel resection for double intestinal atresia, and proximal jejunostomy was performed. After intestinal transit reconstruction, she presented with intestinal occlusion and was submitted to laparotomy at 3 months of age. Three months later, she had a new occlusion: a new jejunostomy was performed, and she was referred to our hospital because of food intolerance, total dependence of parenteral nutrition, cholestatic jaundice, and malnutrition. Although it was possible to make progress in oral feeding, it was not possible to wean parenteral nutrition. At 11 months a barium meal showed marked distention of jejunal loops (3.9 cm in greatest diameter) 25 cm proximal to the stoma.

At 12 months, it was decided to perform STEP and reconstitution of intestinal transit: Eight enteroplasties were performed with GIA™ (*Covidien*, Fig. 1). After surgery it was possible to progress in enteral intake, with resolution of cholestasis and gradual weaning of parenteral nutrition, achieving total weaning 7 months after surgery. Growth was satisfactory, with the weight evolving from below the 3rd percentile to 50th and length from below the 3rd percentile to the 10th, in a sustained way until the last follow-up assessment performed at 5.3 years post-STEP.

Case 2 is a male child, born at 36 weeks of pregnancy, with prenatal diagnosis of intestinal atresia. In the neonatal period, he was submitted to surgical correction of intestinal atresia type IV, with resection of multiple atretic segments and construction of eight primary anastomosis, leaving 40 cm of jejunum–ileum (without the ileocecal valve, but with preserved colon). He was dependent on parenteral nutrition. The CVC had to be replaced five times due to mechanical and/or infectious complications. At age 4 there was a thrombosis (right subclavian and axillary veins) with superior vena cava syndrome which led to chronic hypocoagulation. At 4 years and 10 months, he had marked duodenal–jejunal dilatation and maintained dependence on parenteral nutrition, so it was decided to perform STEP: enteroplasties were carried out with endo-GIA™ (*Covidien*) resulting in a final length of 60 cm of small intestine (Fig. 2). Postoperatively, the boy had gastroesophageal reflux refractory to medical treatment, requiring antireflux surgery (Nissen fundoplication). Progression to enteral feeding was complicated by very frequent diarrhea due to malabsorption, requiring loperamide, that was suspended 12 months after surgery. It was possible to gain autonomy from parenteral nutrition 3 months after STEP. Currently, 5 years and 7 months post-STEP, he tolerates oral feeding and needs nocturnal continuous enteric intake. Weight and height progress in the 10th–25th percentile.

Case 3 is a male child, born at 33 weeks of gestation, with prenatal diagnosis of gastroschisis. In the first days of life, he was submitted to surgical correction of the malformation and multiple resections of intestinal atresia, leaving 30 cm of jejunum and ileum (with preservation of the ileocecal valve). However, he was dependent on parenteral nutrition and had irregular enteral tolerance. At 4 months old, the patient underwent STEP (intraoperative measurement, 42.5 cm of small intestine and 16 cm of colon), but maintained irregular food intolerance and frequent need for gastric decompression by aspiration. At 6 months a contrast X-ray was performed that showed marked dilatation of the proximal small intestine and intraduodenal and duodeno-gastric reflux. At 10 months old he had intestinal obstruction and underwent laparotomy: a new STEP was performed. However, he continued to be TPN dependent with irregular enteral tolerance, vomiting, diarrhea, and recurrent abdominal distension. At 2 years and 4 months, he was referred to our hospital. Given the limitation in oral and enteral feeding, he was subjected to a new STEP with 19

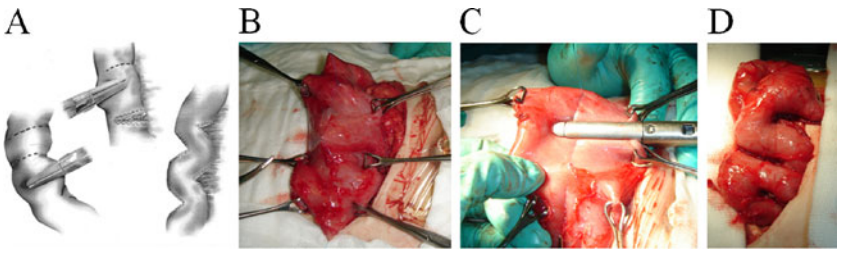

Fig. 1 Schematic drawing of STEP and perioperative photos of STEP performed in case 1. **a** Serial application of linear stapler. **b** Intestinal distention. **c** Applying linear staple perpendicular to the intestinal long axis and parallel to the mesenteric blood supply. **d** Zigzag-shaped channel of lengthened small bowel

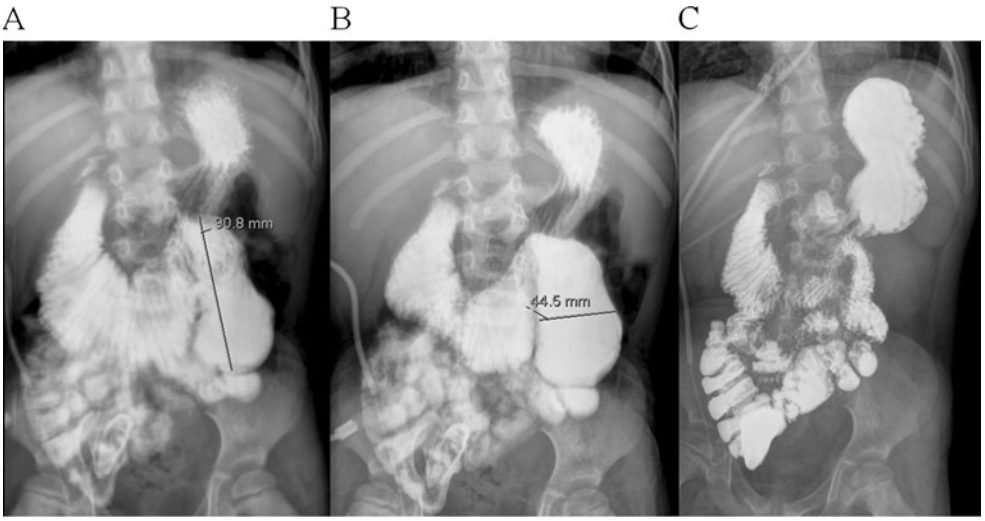

Fig. 2 Bowel contrast study: **a**, **b** Pre-STEP—substantial dilation of the duodenum and jejunum. **c** Twelve days post-STEP—reduction in the diameter of the distal segments of duodenum and proximal jejunum

enteroplasties with endo-GIA™ (*Covidien*) resulting in a final length of 110–115 cm of small intestine. After surgery he presented variable oral/enteric tolerance, with periods of significant increase in oral intake and reduction of parenteral nutrition, alternating with episodes of functional intestinal obstruction. Three months after the last STEP, although able to tolerate oral intake, malabsorption occurred (six to eight semiliquid stools per day), remaining dependent on parenteral nutrition, but with stable growth (height in the 5th percentile and weight in 25th percentile). CVC was replaced 15 times due to mechanical/ infectious episodes. The dependence on parenteral nutrition and the limitations in venous access led to intestinal transplant performed at 5 years old.

Discussion

Intestinal adaptation, as a compensatory response to decreased absorptive surface, evolves with micro- and macroscopic changes, such as proliferation of enterocytes, hypertrophy of the mucosa and smooth muscle, and increased intestinal caliber. However dilatation favors intestinal stasis; it predisposes to bacterial overgrowth [4]. Thus, most patients with short bowel syndrome have insufficient intestinal length associated with excessive intestinal dilation, which was also found in all our patients.

STEP is a way of limiting dilation and acquiring lengthening at the same time, e.g., it is a way of using the dilated bowel. In regard to the other lengthening techniques, it has several advantages: it is a simpler procedure, does not require dilatation over a longer segment; does not imply intestinal anastomoses, does not compromise the blood supply; and the final diameter can be determined by the surgeon, can be performed after a Bianchi procedure if there is redilation, does not preclude transplant [1, 3, 9], and can be repeated [1, 6, 13]. We corroborate some of these advantages in our series. In fact, one of the patients underwent three STEP (two in the hospital of origin and one at our institution), confirming the possibility of repetition of the procedure when redilatation occurs and did not preclude transplant. After the third STEP, it was possible to minimize intestinal dilation, achieving a far better oral tolerance and substantial reduction of occlusive episodes.

This type of surgical procedure has been used in short bowel syndrome [5–7], in intestinal atresia [7, 15], and in severe bacterial overgrowth [12]. The ideal time for surgery is not consensual. It should be considered when progression of enteral feeding is not possible (thus no gain in autonomy from parenteral nutrition), if there is a dilated intestinal segment in the absence of irreversible liver damage, especially if there are complications of parenteral nutrition (cholestasis, recurrent infections of CVC [1, 3]). In our series enteral feeding was tried from the beginning in all patients with variable tolerance. At the time of the decision to perform STEP, the percentage of parenteral nutrition was superior to 60 % in all patients. Ranitidine was used through addition to the parenteral nutrition bag.

With STEP it is possible to increase intestinal length between 52 and 94 % [11, 14] and significant reduction of intestinal dilatation. The weaning of parenteral nutrition is usually achieved in the short term—the average time until suspension is 12 months [3]. Two of our patients achieved complete autonomy of parenteral nutrition at 3 and 7 months post-STEP. In experimental studies in animal models and in clinical studies, there was improvement in nutritional outcomes in the short and medium term, as a result of increased intestinal absorption [2, 5, 11]. In the only published work with midterm results, it was found that patients maintain enteral tolerance and sustain growth 4 years after surgery without increasing mortality [3]. In two of our three patients (one underwent intestinal transplantation), with follow-up time between 5 and 6 years post-STEP, we found improved growth and maintenance of tolerance to oral feeding. The third patient underwent intestinal transplantation 2 years and

7 months after the third STEP. This procedure allowed him a greater tolerance to oral feeding and improved quality of life until transplantation.

The first report of the international registry, which included two of the three patients described here, was published in 2007 [11], involving 19 institutions and 38 patients. This study showed an increase of 31–67 % of oral feeding after a follow-up time (median) of 12.6 months; three patients died and three required intestinal transplantation.

Conclusion

STEP is a promising surgical technique for intestinal lengthening, which may allow complete autonomy from parenteral nutrition or reduce complications by reducing the excessive intestinal dilation and its complications.

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