Seminars in Pediatric Surgery 23 (2014) 291–297

Contents lists available at ScienceDirect

Seminars in Pediatric Surgery

journal homepage: www.elsevier.com/locate/sempedsurg

Short bowel syndrome in children: Surgical and medical perspectives

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ARTICLE INFO

Keywords:
Short bowel syndrome
Intestinal failure
Autologous gastrointestinal reconstructive surgery
Children
LILT
STEP

ABSTRACT

The main cause of intestinal failure in children is due to short bowel syndrome (SBS) resulting from congenital or acquired intestinal lesions. From the first lengthening procedure introduced by Bianchi, the last three decades have seen lengthening procedures established as fundamental components of multidisciplinary intestinal rehabilitation programs. Debate on indications and timing of the procedures is still open leaving SBS surgical treatment a great challenge. However, enteral autonomy is possible only with an individualized approach remembering that each SBS patient is unique. Current literature on autologous gastrointestinal reconstruction technique was reviewed aiming to assess a comprehensive pathway in SBS non-transplant management.

Introduction

The purpose of this review is to provide current state-of-the-art review of the non-transplant management of short bowel syndrome (SBS). Although optimal therapy for SBS remains unknown, a variety of innovative surgical procedures have been developed in the last few decades in addition to medical therapy. Multidisciplinary management has been recognized as the only effective paradigm leading to successful enteral autonomy. In this review, we will focus on the autologous gastrointestinal reconstructive (AGIR) procedures and on the current pharmacological approaches as integral components of SBS multidisciplinary treatment. We will also provide an update on the protocol-driven strategies to promote intestinal rehabilitation. Recent research into regenerative medicine as applied to SBS will also be discussed.

Short bowel syndrome

Short bowel syndrome is defined as a multisystemic condition caused by suboptimal absorption of nutrients due to inadequate small intestinal length.[1] Data from a large tertiary center in Canada shown that the overall incidence of SBS was 22.1 per 1000 neonatal intensive care unit (NICU) admissions and 24.5 per 100,000 live births. The mortality rate of the condition is high with reported survival rates in pediatric SBS ranging from 73% to 89%.[2]

The actual length of small intestine required for optimal absorption is still controversial; however, bowel length < 100 cm in the first year of life is regarded as abnormal. Less than 40 cm traditionally requires therapy according to the practice of most centers.[3] In addition, the gestational age of the child and the timing of the actual incident that caused short bowel will need to be taken into consideration when defining short bowel syndrome.[1]

The majority of underlying conditions that lead to major loss of intestine in neonates and infants have their origins in intrauterine life. Moreover, massive intestinal resection continues to be associated with significant morbidity and mortality rates. In children, the conditions most commonly leading to extensive small bowel resections are necrotizing enterocolitis (NEC), intestinal atresia, gastroschisis, and extensive aganglionosis in Hirschsprung disease.[4,5]

The consequences of short bowel syndrome are huge. Intestinal failure occurs when intestinal function is insufficient to meet the body's nutrition and hydration needs, and supplementary parenteral nutrition and/or intravenous fluid (PN/IV) support is required.[6] In a study from seven tertiary neonatal units in Italy, intestinal failure was seen in 0.1% (26/30,353) of all live births and 0.5% (26/5088) among those admitted to the NICU.[7] With most children dependent on total parenteral nutrition (TPN) for long periods of time, damage to the liver is common. Several medical therapies have been used to enhance bowel adaptation. TPN is used to enhance growth and nutrition of the child until adaptation occurs. In the event of failed adaptation and in the face of liver failure and reduced venous access, transplantation becomes the

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http://dx.doi.org/10.1053/j.sempedsurg.2014.09.010
1055-8586/

only hope for these children.[8] The current estimated survival on parenteral nutrition for short bowel syndrome at 5 years ranges from 52% to 73%.[9] The concept of establishing intestinal failure centers involving the total care of the patient including optimal enteral nutritional management, parenteral nutrition, and central line care, non-transplant surgery and intestinal transplantation has been accepted widely as of ultimate benefit to the child, but is often difficult to put into practice.[1–10]

Autologous gastrointestinal reconstructive procedures

In all SBS patients, the goal of treatment is to promote and enhance the adaptation process over time to possibly gain enteral autonomy, minimize complications, and offer an acceptable quality of life to both children and carers. Intestinal adaptation has been defined as the small intestine's ability to increase its absorptive capacity to compensate for the reduction of absorptive surface area caused by intestinal resection.[11] Timing of intestinal adaptation is variable,[12] unique to each individual and usually occurs during the first 2 years following massive intestinal resection in adults; longer and perhaps more vigorously in children.[13] Intestinal adaptation in children is influenced by growth and development and thus occurs over a longer period in younger children.[5] Massive resection stimulates modification in thickness and length of the muscle layers and structural modification of crypts to villi. Distention of remain bowel is the most common consequence after massive resection.[14]

The adaptive intestinal process prompted the possibility to design autologous gastrointestinal reconstructive procedures. Indeed, pre-requisite condition for successful non-transplant surgery is evidence of a segment or segments of dilated bowel with a lumen diameter more than twice the normal for age and weight.

Surgical attempts at improving mucosal absorption have been varied and imaginative. In 1980, Bianchi[15] proposed a bowel-lengthening technique called Longitudinal Intestinal Lengthening and Tailoring (LILT). Boeckman and Traylor[16] performed the first clinical application of this procedure in 1981 to a 4-year-old PN-dependent child with a 50-cm jejunum, who was also at risk from precarious venous access. Within 10 weeks, the patient was able to wean off PN and has continued to thrive on enteral nutrition. The pathophysiology behind the LILT is based on the anatomical evidence of mesenteric blood vessels that enter the small bowel loop along its lateral aspects. Accordingly, it is possible to divide the dilated bowel longitudinally in the midline along the mesenteric and antemesenteric borders and to create two fully vascularized isopropulsive hemiloops, which are anastomosed isoperistaltically with the distal loop anastomosed to the remnant colon. The tailored bowel is half the diameter and up to double the length of the original loop without significant loss of absorptive mucosa. Propulsion, although not uniform, is effectively isoperistaltic with a reduction in stasis and sepsis (Figure 1). The benefits of LILT are the combined possibility to double the length in addition to better propulsion—a significant problem in dilated dysmotile bowel. This procedure has been demonstrated clinically to improve fat and carbohydrate absorption and slow transit

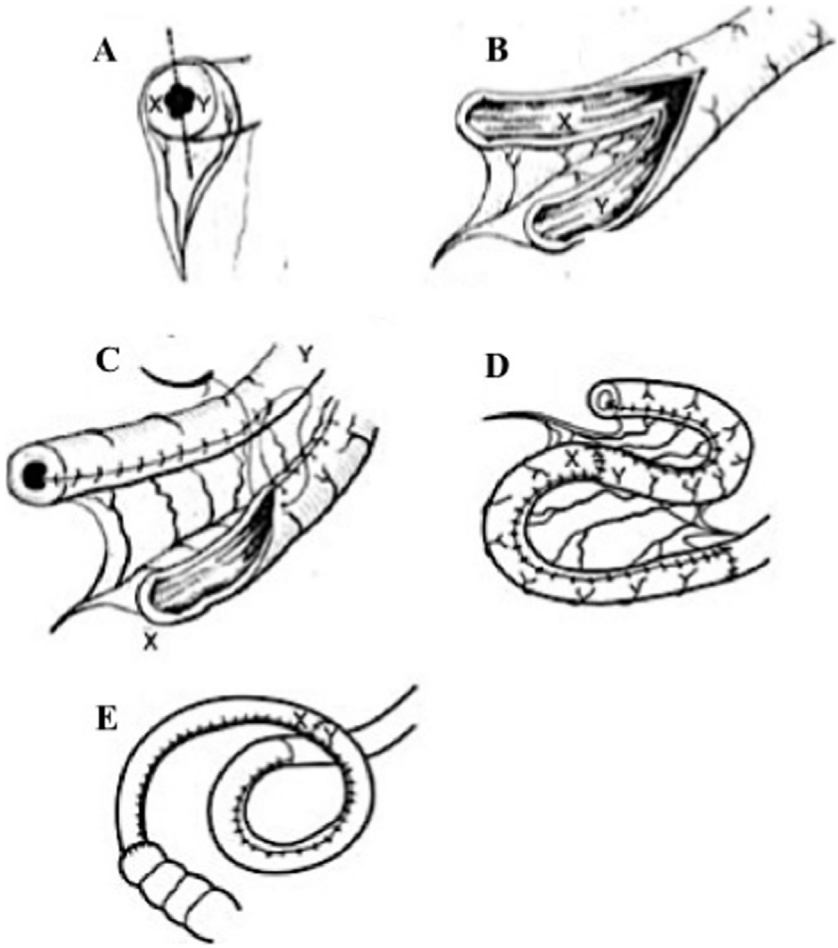

Fig. 1. The LILT procedure. (A) Blunt dissection between the peritoneal leaves of the mesentery, with development of a midline intravascular plane (bowel division depicted by dotted line). (B and C) Formation of hemiloops by manual suturing inverts the bowel edges and preserves all mucosa. (D–E) Isoperistaltic anastomosis between hemiloops in S shape or spiral shape. (Adapted with permission from Bianchi, 1984.)