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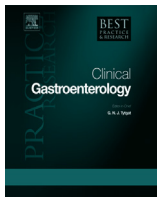

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Surgical management and autologous intestinal reconstruction in short bowel syndrome

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A B S T R A C T

Short bowel syndrome (SBS) is a serious condition with considerable morbidity and mortality. When treatment with parenteral nutrition fails and life-threatening complications occur, autologous intestinal reconstruction (AIR) should be considered before intestinal transplantation (ITx). Single or combined ITx should be reserved for patients with severe liver disease and as last resort in the treatment of SBS. Longitudinal intestinal lengthening and tailoring (LILT) has proven its value in AIR, but its availability depends on the expertise of the surgeons. Serial transverse enteroplasty (STEP) has similar success rates as LILT and fewer patients progress to ITx. STEP is also applicable at small bowel dilatation in ultra-short bowel syndrome. The scope may be widened when duodenal dilatation can be treated as well. Spiral intestinal lengthening and tailoring (SILT) is a promising alternative. More research is needed to confirm these findings. Therefore we suggest an international data registry for all intestinal lengthening procedures.

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Introduction

Parenteral nutrition (PN) is the standard treatment in short bowel syndrome (SBS). After its introduction in the late 1960's [1], several important improvements in the treatment of PN have been introduced. Despite these improvements, PN-dependent patients are still prone to develop severe complications contributing to a high mortality [2–4]. Intestinal transplantation is the standard treatment for patients with intestinal failure who develop severe complications from PN or experience a low quality of life [5]. The accepted indications nowadays for intestinal transplantation (ITx) are recurrent catheter related infections, thrombosis of the central veins, alteration in growth and development of infants, severe dehydration with refractory electrolyte changes and liver failure and/or liver cirrhosis with portal hypertension [5]. The first successful isolated transplantation of the intestine was performed in 1989 [6], two years after the first successful multivisceral transplantation in 1987 [7]. Improvement in surgical techniques, novel immunosuppressive agents and advances in anaesthesia and critical care led to an overall increase of intestinal transplantations worldwide. However, after the initial enthusiasm it became evident that the long-term results were disappointing with a 5-year graft survival of only 50% for patients transplanted after 2000 [8]. As a consequence, the number of intestinal transplantations has decreased from more than 200 procedures in 2008 to only 120 cases in 2012 [8]. Given the severe complications and the disappointing survival of ITx and low quality of life of patients on PN, all efforts should be made to prevent a SBS. The ultimate goal of treating SBS is to achieve enteral autonomy through medical and surgical intestinal rehabilitation. The aim of this review is to discuss the initial surgical management and autologous intestinal reconstruction (AIR) in patients with SBS.

Definition and aetiology of short-bowel syndrome

Definition

Short bowel syndrome (SBS) is a condition in which there is a congenital or an acquired shortage of small bowel length and thus a lack of absorptive small bowel surface. This leads to the incapability of retrieving enough nutrients, water and electrolytes to ensure growth in infants and body maintenance in adults and therefore requiring long-term administration of PN. The broader term 'intestinal failure' includes in addition to SBS, mechanical abnormalities and intrinsic bowel diseases. These patients have a normal bowel length, but experience severe bowel obstructions or functional problems caused by motility disorders or mucosal abnormalities [9,10]. In this chapter, we will limit ourselves to SBS with an anatomical absence or loss of significant bowel length.

The total length of small bowel in a healthy person depends on the gestational age within a natural spread. The younger, the more growth potential there is. Loss of for instance 50 cm is clearly more severe in a premature born child then in a mature neonate [11]. From the second year of life, bowel length does not significantly increase further and stays around 340 cm. However, the diameter of the intestinal lumen still increases with age [12].

There have been made functional and anatomic classifications, both in three types, which enables us to compare treatments of SBS. The classification based on functional outcome was introduced by Shaffer in 2002 [13]. The latest revision has been made recently by Pironi et al. (see Table 1): type I SBS being acute, short-term and usually self-limiting. Type II stands for a prolonged acute condition, often in metabolically unstable patients requiring complex multi-disciplinary care and intravenous supplementation over periods of weeks or months and type III representing chronic intestinal failure in metabolically stable patients, requiring intravenous supplementation over months or years; it may be reversible or irreversible [10,14]. In adults the minimal length needed for functional type I or II SBS is at least 115 cm in patients with an endjejunostomy, 60 cm in case of a jejunocolic anastomosis and 35 cm if a jejunoileal anastomosis with an intact colon is present (Table 1 and Fig. 1) [15]. Although the exact length is not part of the generally accepted functional classification, we thus know that the remaining bowel length is an important prognostic factor to estimate the amount of enteral autonomy that reasonably can be expected. In addition, a detailed description of the remaining length of the duodenum, jejunum, ileum and colon makes a comparison of the effectiveness of AIR possible.