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Review Article

Lengthening and Tailoring the Bowel: Surgical Approaches in the Treatment of Paediatric Short Bowel Syndrome

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Abstract

Short Bowel Syndrome (SBS) is a multi-system disorder caused by mal-absorption of nutrients due to an inadequate length of small bowel. Due to the mal-absorptive state SBS patients need Parenteral Nutrition (PN) to support enteral state but the long-term use of PN could lead to liver disease. To increase the intestinal length aiming to enhance absorption of nutrients, various surgical procedures have been developed. The two commonly used procedures for bowel lengthening are: Longitudinal Intestinal Lengthening and Tapering (LILT), and Serial Transverse Enteroplasty (STEP). Spiral Intestinal Lengthening and Tailoring (SILT) has recently been developed after successful trials on animals. Several SILT procedures have now been performed in children since the first clinical application in 2012 with positive results. The presences of these three lengthening procedures allow the surgeon to employ the right technique to the right patient at the right time. We firmly believe that it is important to tailor the procedure to either the patients' needs and the clinical findings to deliver a clinical approach centred to patients with short bowel.

Keywords: Short bowel syndrome; Paediatric; Intestine; Lengthening, Surgery

Introduction

Short bowel syndrome has been reported to have a congenital condition only in very limited cases [1], but in the most classical scenario there is usually a precipitating cause. The commonest causes of SBS in the paediatric population are Necrotizing Enterocolitis (NEC), intestinal atresia, malrotation and volvulus, and gastroschisis [2]. According to the experience of a leader centre on SBS treatment, this condition is considered to be a multi-system disorder caused by mal-absorption of nutrients due to an inadequate length of small bowel [3].

In patients with SBS, a significant level of natural intestinal adaptation occurs after intestinal resection. This happens relatively soon after surgery, where a change in structure, motility and physiology occur in the remnant bowel [4]. There is proliferation of the villi, lengthening of the crypt cells, increase in microvilli along epithelial surface, and an increase in mucosal weight. Hyperplasia causes the thickness and length of the muscle layers to increase. In terms of motility, there is an increased intestinal transit time, and due to the functional adaptation that occurs, there is an increase in the amount absorbed by individual enterocytes [5].

As the patient struggles to absorb the necessary nutrients for survival, all patients are required to start PN. Children, who remain on PN for a long time, have high rates of sepsis, cholestasis, and mortality [6,7]. The prevalence of liver failure in paediatric patients is poorly defined, as it has varied from 7.4% to 84% [8,9].

The aim of autologous intestinal reconstructive procedure is to prolong intestinal transit time and increase functional surface area. As reported by the experience of intestinal rehabilitation units, after lengthening procedure the patient increase the capacity to absorb food, and to be weaned off PN [10-12].

Surgical Techniques

The current surgical management of SBS is based on three main procedures. The oldest and most established procedure is with no doubt the longitudinal Intestinal Lengthening and Tapering (LILT) proposed by Bianchi in 1984 [13]. Subsequently in 2003, Kim described an alternative procedure was called the Serial Transverse Enteroplasty (STEP) [14]. Recently, a new technique called Spiral Intestinal Lengthening and Tailoring (SILT) has been developed and used for the first time in the clinical practice in Manchester [15] following extensive work in large animal model [16]. The common purpose of all these techniques is to increase intestinal length, to tailor the dilated intestinal segment for increasing the nutrients' contact time to the mucosa and finally to improve absorption. Importantly, each technique appears to have specific characteristic and application.

Longitudinal Intestinal Lengthening and Tapering

LILT was first described in 1980 [17] by Bianchi, and then used in the first patient in 1984 [13]. The technique involves dividing the longest segment of dilated bowel along the anti-mesenteric border, with extra care to ensure the highest numbers of blood vessels are preserved. After this, the bowel is then divided into two vascularised hemi-segments along the mesenteric border. The two hemi-segments are than tubularised to form hemi-loops, and sutured together in an isoperistaltic fashion, using a continuous inverting 6-0

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Maxon knotting every fourth row. Bowel continuity is established with a jejuno-colonic anastomosis and a wide oblique hemi-loop anastomosis in the original S-Shape, or the Aigrain spiral [18]. Bowel function is usually resumes between 3 to 5 days [3,10,12].

More recent data shows that survival rates with LILT vary between 30%-100%. The data shows that of these survivors, 28%-100% were weaned off PN [19]. The wide range of the results using LILT procedures suggests that surgical experience in performing this technique and intestinal rehabilitation unit experience may play an important role.

Serial Transverse Enteroplasty

STEP, first proposed by Kim et al in 2003 [20], is a procedure that involves a stapler being applied across dilated bowel in an alternating fashion, such that it leaves the bowel in the shape of a zig-zag [21]. The creation of the zig-zag requires a mesenteric defect at each staple line. The stapler is placed at 90 degrees and 270 degrees - with 0 degrees being the mesenteric border- and incisions are made in the bowel, leaving the bowel approximately 2 cm in diameter [14].

The main advantage of this procedure is that STEP lengthens and tailors the bowel, without damaging the mesenteric blood supply. It is also conceptually relatively simple, and because of the natural re-dilation of bowel, some authors believe that STEP can be used safely after a dilatation due to LILT [22]. Multiple STEP procedures can be performed on the same patient after the process of adaptation and dilation has occurred [23]. The bowel is remarkably lengthened during the STEP procedure, with one study on animals showing an average increase of 64% [14]. Interestingly, a recent article showed that re-STEPs failed to result in significant PN weaning, with no re-STEP patients achieving enteral autonomy during follow-up [24].

The main reported complications are intestinal obstruction, inter-loop fistulae and bleeding from the suture line [22]. Remarkably, the exact physiological implications of STEP procedure are still unknown because this procedure changes the orientation of the muscle fibres in the lengthened segment. It is still not clear whether peristalsis is restored in the STEP segment, or the intestine becomes passive, and whether this procedure has an impact on the function of the intestine. More research studies will be essential to address these open questions.

Spiral Intestinal Lengthening and Tailoring (Silt)

SILT is the most recent lengthening procedure developed and proposed. The procedure was established on the bench [25] and tested in modelled Vietnamese pigs in 2011 [16]. In the original experiment, the animals were kept on water for 24 hours, then liquid food for 48-72 hours post-operatively. Of the 6 pigs that underwent this procedure, 4 had an uneventful post-operative course. Autopsies showed that bowel obstruction did develop in two pigs. In both autopsies, the lumen was shown to have narrowed by 70% to a diameter under 1.5cm. Statistical analysis of the results showed no significant difference in length and width measured straight after SILT and that measured 5 weeks later. All lengthened segments showed peristalsis immediately after the procedure and at terminal laparotomy 5 weeks after, or at autopsy.

The procedure involves drawing spiral incision lines at 45-60 degree angles on the bowel. The mesentery is then incised perpendicularly to the marked points. The incisions are now made on the bowel, and the bowel is then stretched longitudinally over an intraluminal catheter to a larger tube of narrower diameter. Finally, the mesenteric defects are narrowed to prevent intra-abdominal herniation.

The first SILT procedure for a patient with SBS was performed in Manchester in 2012 with good results. The patient was weaned off TPN after 6 months [15]. A 3-year-old patient born at 24 weeks gestation was deemed suitable for the SILT procedure. After natural intestinal adaptation aided by Parenteral Nutrition (PN), gastrostomy feeding, structured controlled bowel expansion, and continuous extracorporeal stool recycling, she underwent Spiral Intestinal Lengthening and Tailoring (SILT). Prior to SILT, the length of the Jejunum was increased from 15cm to 22cm using controlled tissue expansion; a technique that allowed to dilate in controlled was segments of intestine [26]. Using SILT, the 11cm of distended bowel was extended to 20cm. 5 days after SILT, she was started on oral and gastrostomy feeding, and there were no surgical complications. She was weaned off PN, with good liver function 6 months after the procedure. After the first case we have successfully performed 4 more SILT procedures (data not shown) and there also been reported another patient treated with this technique [27].

Discussion

LILT is the oldest and most established surgical procedure to treat patients suffering SBS [13,17,28,29]. In the experience of Manchester Paediatric Autologous Bowel Reconstruction and Rehabilitation Unit (12), we use the LILT in patients with no fibrotic mesentery (as the procedure splits the mesentery in two halves) and bowel diameter of 5-6 cm or greater (Table). We choose this procedure in severe short bowel state (<20cm) when a 100% increase in length is required.

In the STEP procedure the increase in the bowel segment appear to be not as great as the LILT procedure. Although STEP is able to length bowel segment, the reported increase in length using STEP is about 64% [30]. In order to perform the STEP seems to be necessary a dilated segment of bowel with 5-6 cm of diameter but importantly this procedure seems to not be affected by the status of the mesentery. In our experience, we employ STEP in selected patient with remnant bowel of 40 cm and greater with clinically significant dilatation (Table).

The SILT has been shown to increase the length of the segment of bowel by 75% [15]. This procedure can be safely performed in short bowel patients with moderate bowel dilatation (bowel diameter of 2-4 cm).

The status of the mesentery is not a limiting factor as the indentation on the mesentery produced by the procedure is minimal. This is a potential advantage and will make this procedure widely accepted and used. Further studies are needed to fully assess the impact of this technique on the long-term outcome of larger series.

Conclusion

It is really reassuring that intestinal rehabilitative surgeons can employ different surgical techniques to lengthen bowel in paediatric

population. These varieties of technique will allow the surgeon to individualise the choice of procedure according to patients' needs aiming to maximise surgical outcomes and bowel adaptation. The aim of any bowel reconstructive surgeon should be to provide the right operation for the right patient at the right time at the first attempt.

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