OVERVIEW OF NON-TRANSPLANT SURGICAL MANAGEMENT OF SHORT BOWEL SYNDROME IN CHILDREN

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The aim of this study was to review the management of Short Bowel Syndrome, focusing on the current trends regarding non-transplant, autologous surgical reconstructive techniques. Management of severe Short Bowel Syndrome is still one of the greatest challenges of medicine. Paediatricians and paediatric surgeons may face this challenge often because of the relatively common occurrence of necrotizing enterocolitis, intestinal malrotation, volvulus, gastrochisis and “apple peel” atresia. Although the short term results of intestinal transplantation are improving, the long term results are still disappointing. On the other hand an increasing number of patients have been saved through modern intestinal bowel rehabilitation programs and autologous intestinal reconstructive surgeries in the last decade. This review summarizes the main medical elements of these programs, such as the control of gastric hypersecretion, inhibition of intestinal motility, elemental enteral feeding, low fat (home) parenteral nutrition and hormonal enhancement of intestinal adaptation. It focuses on non-transplant surgical management of short bowel patients, such as accurate vascular access surgery, controlled bowel expansion program, extracorporeal stool recycling and intestinal lengthening techniques.

Conclusion – Intestinal bowel rehabilitation and autologous intestinal reconstruction should be considered as first line management in short bowel patients. The establishment of national bowel rehabilitation centres should be considered. Intestinal transplantation should be reserved for unsuccessful cases and considered as a last resort.

Introduction

Severe short bowel syndrome is a devastating condition. Epidemiological data suggest that the incidence of severe short bowel syndrome is four in one million, the majority of whom are infants and young children (1). Of all the catastrophic conditions that lead to the subtotal loss of small intestine in early life, the most frequent are necrotizing enterocolitis (NEC) that endangers mainly low-weight new-borns, small intestinal volvulus as a result of intestinal malrotation, gastrochisis and “apple peel” syndrome of the small intestinal atresias. Other rare cases such as incarcerated congenital diaphragmatic hernia have also been reported (2).

After the loss of a certain length of the small bowel, transit time is shortened, the absorption of nutrients becomes ineffective, and malnutrition, dehydration and electro-
lyte deficiency develop. The malabsorption of bile acids escalates diarrhoea and steatorrhoea. The small intestine, in young childhood in particular, is capable of some compensation. The bowel adapts, crypts deepen, and villi become hypertrophic. However this process is slow and usually results in massive dilatation of the bowel. In dilated intestines the muscle constrictions are ineffective in narrowing the lumen, so the peristalsis becomes insufficient; the bowel content becomes stationary, which leads to translocation of bacteria, and sepsis (3). The management of short bowel patients is complex and requires both medical and surgical input from a multidisciplinary team (surgeon, gastroenterologist, dietician, pharmacist, nursing specialists) specialised in intestinal rehabilitation (4).

The aim of this review was to summarise the modern non-transplant surgical management of short bowel syndrome.

**Medical considerations**

**Gastric hypersecretion**

A massive loss of intestine induces increased secretion of gastrin and gastric acid. This can lead to gastric ulcer and massive intestinal haemorrhage. Therefore, the introduction of H2-receptor inhibitors or the more effective proton-pump inhibitors is essential (5). Octreotid has been used with variable success (6).

**Enteral feeding**

In infancy, breastfeeding or hydrolyzed formulas and middle chain triglycerides are introduced. Parenteral supplementation of fat-soluble and B12 vitamins (the latter is absorbed only in the terminal ileum) is always recommended (5). In older children food aversion makes high caloric feeding often difficult. Nasogastric tube (7) or endoscopically inserted percutaneous gastrostomy allows slow, continuous feeding even overnight. Surprisingly, short bowel patients tolerate complex carbohydrates better than simple sugars (8).

**Patients with high output stoma**

High levels of stoma discharge can be returned to the distal stoma (recycling), however microbiological safety should be seriously considered (9). If this is not possible then the stoma-deficit should be reinstated hourly, parenterally. High output stomas may lead to severe loss of sodium, not recognizable in the serum electrolyte level, therefore urinary sodium should be monitored and if it is lower than 20 mM/l oral sodium supplementation is usually recommended (5).

**Reduced intestinal transit time**

Motility inhibitors are routinely used: codeine, morphine and loperamide effectively increase intestinal transit time. Cholestyramine binds free bile acids, reducing their laxative effects. A high fibre diet reduces diarrhoea by absorbing water (5).

**Parenteral nutrition**

Parenteral nutrition (PN) is usually introduced after recovery from the primary surgery when short bowel syndrome is expected. Long term home PN is available in many countries, but requires well-organized health care services and specialist nurses, who visit and teach the parents. It has been observed that with “low fat” PN (less than 1g/kg) liver damage is far less frequent. Recently, omega-3 fatty acid PN has become preferred. However, liver failure is still one of the most feared complications of PN. PN requires long-term, safe central lines, which often become infected, dislodged or blocked. Even today the morbidity and mortality from long term PN is significant. The quality of life of the patients is rather low; their life is bound
to the healthcare provider facility, although home parenteral nutrition, if possible, can improve life quality (10). Sepsis is common. Development and growth are most often delayed (11).

**Hormonal enhancement of bowel adaptation**

Clinical use of hormones such as epidermal growth factor or glucagon-like peptide 2 (GLP-2) is not yet widespread, however promising initial results have been published (5, 12, 13). Jeppesen and Yazbeck (14) reported encouraging results with GLP-2 and with Teduglutide (a protease-resistant analogue of GLP-2 with prolonged biological activity) in adult Short Bowel Syndrome patients. Nutrient absorption was found to increase by 3.5%. A significant increase in protein absorption was found, but no significant increase in carbohydrate absorption, and no significant change in fat absorption. Histological analysis of the small intestine demonstrated an increase in villus height and crypt depth, and more than 20% reduction in PN was observed (13, 14). Oral insulin supplementation was reported to decrease the need for parenteral nutrition in paediatric SBS patients in a pilot study (15).

**Surgical considerations**

**Prevention**

Early and accurate diagnosis of emergencies, such as malrotation, emergency surgery, careful bowel resection, second-look surgery, and avoidance of resection of the ileocaecal valve are the very important common sense factors.

**Vascular access management**

Even tunnelled, good quality, central venous catheters have a limited lifetime and they often become blocked, infected or broken and need replacement. The number of veins available for central catheter placement is limited. Careful indication and technique are mandatory. The percutaneous technique is preferred over surgical exploration (16). Antiseptic non-touch handling of catheters, and a regular 70% ethanol catheter block may prevent catheter-sepsis. Broad-spectrum antibiotics may be effective to treat infected central lines, with urokinase to unblock them. If the removal of the catheter is unavoidable, the new catheter should be placed into the same vein, if possible.

**Small-bowel transplantation**

Intestinal transplantation is only available in certain countries. Although it is true that the short term (1 year) survival, especially with multi-organ (liver and intestine) transplantation, has increased dramatically in the last few years, in some high volume centres by up to 90%, the long term (5 years) survival is significantly lower (50%) and immunosuppressive therapy remains demanding for the patients and may go hand in hand with complications, thus this still remains the last resort of treatment (17).

**Autologous intestinal reconstructive surgery**

The non-transplant management of Short Bowel Syndrome, intestinal rehabilitation with autologous intestinal reconstruction surgery (AIRS) gives us the opportunity to shorten PN and/or restore enteral autonomy, and to avoid intestinal transplantation in an increasing number of patients. The use of AIRS began in the 1980-s, and its objective was to provide a surgical solution for the problems that occur in short bowel syndrome. The success rate of non-transplant management of short bowel patients has increased significantly over the last decade, and
92% overall survival rate has been reported recently and 91% of the surviving patients have been weaned from PN (4, 18, 19). The major goals of AIRS are to increase transit time, improve motility of the dilated bowel, and increase the absorptive surface.

**Increase transit time**

The aim of initial interventions, such as antiperistaltic reverse segment, colon interposition and ileocaecal valve reconstruction valve procedures, was to increase transit time, allowing more time for absorption by creating controlled resistance.

**The Antiperistaltic “reverse” segment procedure**

A bowel segment, 10 cm long in adults and up to 8 cm in infants and children, reversed and reanastomosed antiperistaltically, is believed to slow down transit without causing bowel obstruction. This procedure is used when the absorption surface i.e. bowel length, is thought to be reasonable but transit is rapid, or it could be used in combination with other procedures (Fig. 1).

![Antiperistaltic “reverse” segment procedures](image)

**Colon interposition**

The method was developed by Hutcher et al. (20, 21). The transit time of the colon is longer than that of the small bowel. The benefit of interposing a colonic segment in the small bowel loop is that the chyme passes through the small bowel more slowly, giving the digestive enzymes more time for efficient digestion. Observations suggest that the histological structure of the colon also changes, and it adapts and becomes similar to the small intestine, further improving absorption (22, 23). Different lengths of colonic interposition were used. Glicket et al. (22) used 10-15 cm segments, while Garcia et al. (24) used a 24cm segment. Four of the seven infants with isoperistaltic interposition survived.

**Ileocecal valve reconstruction**

As a result of the original condition (volvulus, NEC) the ileocaecal valve is often removed and an end-to-end ileo-colostomy is performed. The loss of the ileocaecal valve is believed to be well tolerated, however in a survey, one fourth of the non-short bowel patients were found to have persistent diarrhoea as a long-term complication (25). It is easy to understand that this may worsen the symptoms of short bowel syndrome. Some attempts have been made to reconstruct the ileocaecal valve, for example with intussuscepted small bowel nipple valves, but these valves require a very long segment (6-7 cm) of intussusception to become effective in short bowel syndrome (26). Histology analysis revealed that the anatomy of an ileocaecal valve does not resemble the classic sphincter anatomy. It is hypothesized that it could be easily reconstructed with the help of a double layered, 1-2 cm deep, end-to-side invaginated ileo-colostomy. However, this seems to be difficult and risky, and it has not yet been justified in clinical practice (27).

**Improve motility of the dilated bowel**

As a result of the natural bowel adaptation in short bowel syndrome, the intestine becomes dilated. In a large calibre segment the muscle constrictions are ineffective in narrowing the lumen, so peristalsis is insufficient; the bowel content becomes stationary, which leads to
translocation of the bacteria. This is the main source of the sepsis experienced in short bowel patients. Surgical procedures have attempted to improve the motility of the severely dilated intestine. Excision of the “redundant” bowel wall has been associated with further loss of absorptive surface, and plication of the bowel wall is also not effective long term. Bianchi first developed a procedure where the dilated bowel was tailored to half of the calibre, without losing absorptive surface. Bianchi re-fashioned the short and dilated segment with impaired peristalsis to normal calibre but a longer intestine. This is beneficial, because a smaller calibre but longer bowel loop is able to provide adequate peristalsis, to prevent stasis and bacterial translocation, and allows a more sufficient transit time for absorption than a dilated and short segment (3, 4, 28).

**Longitudinal Intestinal Lengthening and Tailoring (LILT)**

This technique is based on the fact that the vasa recta, running in the mesentery, divide before reaching the bowel, and supply one side or the other of the bowel. This way the arteries and veins can be separated from each other just before they reach the bowel wall, so the bowel can be divided into two longitudinal halves, conserving a good blood supply. From the two halves of bowel wall, it is possible to tailor new bowel segments, which have a lumen half the original length, and their joint length is twice as long as the original bowel. However this procedure is surgically challenging and only surgeons with significant experience with this technique are able to execute it safely (18). The overall survival rate is reported to be between 30%-100%, and the ability to wean from PN varied from 28%-100% in different series with LILT (29, 30-33). Complications such as stenosis at the hemiloop anastomosis, inter-loop fistula, loss of the hemi-loop from vascular compromise and recurrent dilatation, have been reported (34, 35). A systematic literature review of the Bianchi and STEP procedures in 2013 reported overall survival of 89%, with no significant difference between the two procedures. The Bianchi procedure has been found to have higher rate of weaned patients, 55% vs. 48%, however the Bianchi procedure has been associated with a higher rate of patients receiving transplantations: 10% vs. 6% (33) (Fig. 2).

![Fig. 2. Longitudinal Intestinal Lengthening and Tailoring.](image)

**Serial Transverse Enteroplasty (STEP)**

This procedure, developed in 2003, is much simpler than the Bianchi operation, there is no need for the challenging disassociation of the mesentery, the extent of the lengthening and the narrowing of the intestine is adjustable within limits (36), but its disadvantage is that the concentric fibres become longitudinal and the longitudinal fibres become concentric, thus making the peristaltic movements uncertain (37). This might be the explanation for the usual bowel dilatation after STEP (38). The STEP procedure has been performed successfully in many centres. Recently, Jones Ba et al. (39) reported the results of 111 consecutive patients enrolled in the international STEP data registry. The overall post STEP mortality was 11%, and 47% of the group attained full enteral nutrition after the procedure. However, patients with longer pre-STEP bowel length were significantly more likely to achieve enteral autonomy (39) (Fig. 3).
Serial Transverse Enteroplasty (STEP)

Spiral Intestinal Lengthening and Tailoring (SILT)

The spiral intestinal lengthening is a new alternative. It barely affects the mesentery, it is simple and does not change the orientation of the fibres dramatically. The heart of the procedure is that the bowel wall is cut along a spiral line, twisted and then lengthened longitudinally to tailor it into a tube again. The mesentery is also incised where it meets the spiral cut-line (37). The procedure was tested in animals (40) and two independent successful human applications have also been reported (41, 42). The SILT technique also seems to have urological application and it seems suitable for creating long Mitrofanoff channels for intermittent catheterisation (43) (Fig. 4).

Increasing the absorptive surface

Mechanical tissue expansion is widely used in plastic surgery for autologous skin transplantation. Mechanical force (controlled pressure or traction) has been proven to increase the absorptive surface and the enterocyte mass of the small intestine (44, 45, 46). This, and the success of the tailoring and lengthening procedure, have led to the idea of controlled bowel expansion (47).

Controlled bowel expansion

In the course of the procedure, a catheter (stoma catheter) is placed into the lumen of the bowel, and a controlled obstruction is used to expand the intestine, obtaining a greater absorbing surface. The expanded dilated bowel loses its motility, but it can be reconstructed using lengthening and tailoring procedures, and refashioning of the dilated short bowel to a normal calibre, but longer segment, without losing absorptive surface. The catheter stoma also gives us the opportunity to recycle the bowel content from the proximal to the distal stoma, reducing the need for PN and atrophy of the distal stoma (47). There are experiments in progress to develop endoluminal devices for intestinal lengthening but these are still far away from safe clinical application (46).

Combination of the procedures

AIRS procedures could be combined and tailored to the individual needs of the patients. Controlled bowel expansion, with tailoring and lengthening procedures and with ileo-caecal valve reconstruction, seems to be most logical sequence in a severe short bowel condition.

Conclusion

With the advancement of PN, enteral feeding and autologous AIRS, the survival of
short bowel patients has improved, and the length of PN needed reduced significantly over the last decade. Intestinal bowel rehabilitation and autologous intestinal reconstruction should be considered as first line management in short bowel patients. The establishment of national bowel rehabilitation centres should be considered.

Conflict of interest: The author declares that he has no conflict of interest.

References


