

# Autologous gastrointestinal reconstruction for short bowel syndrome

**Parenteral nutrition-dependent patients with short bowel can realistically hope for a return to enteral autonomy on their own bowel, and with a full good quality of life. Autologous bowel reconstruction (including expansion, lengthening and reversed segments) enhances the natural adaptive process towards increased absorption.**

The short bowel state may be loosely defined as a residual length of autologous bowel whose absorptive capability is insufficient to sustain survival and growth on enteral nutrition. Long-term survival for patients with established short bowel will depend on the type, physical condition and natural adaptive response of the residual autologous bowel. The ileum has a greater adaptability than the jejunum, and the ileo-caecal valve and a longer colonic length are invaluable assets.

## The problem

Most short bowel patients will have lost the midgut supplied by the superior mesenteric vascular pedicle. Following antenatal volvulus complicating malrotation or gastroschisis, the child is born with a residual 10–20% of the small bowel presenting as a high jejunal atresia with an obstructed, markedly dilated, peristaltic jejunum of short length, and an unused left colon. Despite active peristalsis in the dilated proximal jejunum, a jejuno-colic anastomosis is associated with a failure of propulsion, with intraluminal stasis leading to sepsis and to portal and systemic bacterial translocation. Following postnatal small bowel loss the residual jejunum is initially of normal diameter and demonstrates a rapid transit with insufficient absorption and major loss of fluid, electrolytes and nutrients. It is relevant to note that the gut-associated lymphoid tissue, specifically the intraepithelial lymphocytes, the ileal Peyer's patches and the mesenteric lymph nodes, plays a major immunological role and that its absence is also of consequence.

Before starting treatment the child's parents need to appreciate that without prolonged parenteral nutrition and effective medical and surgical management their child is effectively non-viable and that patience, persistence and multiple surgery will be required.

## Considerations in management

The natural adaptation response is a slow and steady process that is said to be maximal within 1–2 years but

that frequently continues for prolonged intervals, e.g. 4–7 years, before enteral autonomy is established (Williamson, 1978a,b). The adaptive process also affects the immune component within the bowel, and as villous hypertrophy and cellular hyperplasia occur so also is there an improvement in gut-related immunity. During this lengthy adaptive period survival depends on a combination of parenteral and enteral nutrition.

Preservation of venous access for placement of feeding catheters, avoidance of central line sepsis and intraluminal bowel sepsis, and prevention of cholestasis and progressive toxic liver injury (Iyer et al, 1998) are critical aspects to the child's survival and care. Concomitantly a well-structured programme of autologous gastrointestinal reconstruction attempts to develop and restructure the residual bowel, optimizing its adaptive potential towards enteral autonomy sufficient to support survival and growth (Bianchi, 2006). Aggressive pharmacological manipulation of bowel activity to slow transit (loperamide, codeine) and to increase mucosal absorption (clonidine) is also relevant.

The alternative to autologous reconstruction is bowel transplantation, which instantly provides sufficient bowel for adequate nutrient absorption, but carries significant and potentially serious concerns from acute rejection, post-transplant lymphoproliferative disease and chronic rejection. Better immunotherapy has improved organ and patient survival; however, post-transplant life requires lifelong intensive monitoring to avoid serious life-threatening complications or loss of the transplanted bowel. Until tolerance between the donor bowel and the recipient can be reliably induced, the option of bowel transplantation should only arise once all other treatment modalities have failed and/or when the patient's quality of life is unacceptable.

## Bowel reconstructive surgery

Several procedures (*Table 1*) have been proposed for improving absorption from the remaining bowel.

## Vagotomy with pyloroplasty

Vagotomy with pyloroplasty was designed to reduce acid output thus favourably altering the intraluminal pH. This procedure does not contribute greatly to increasing absorption but has the advantage of countering the ten-

Mr A Bianchi is Consultant Specialist Paediatric and Neonatal Surgeon, Royal Manchester Children's Hospital, Manchester

Correspondence to: Mr A Bianchi, 1 Elleray Road, Middleton, Manchester M24 1NY

dency to duodenal ulceration driven by the hypergastrinaemia and hyperacidity that accompanies the short bowel state.

**Colon interposition**

Colon interposition, proposed by Hatcher et al (1973), interposes a length of isoperistaltic colon prejejunally between the duodenum and the jejunum, taking advantage of the colon’s fluid and electrolyte absorptive ability and its lesser motility, for slower delivery of a more concentrated nutrient to the small bowel absorptive surface. The relative safety of the procedure with no loss of bowel and its ease of reversibility, recommend its consideration when sufficient colonic length is available. Despite reports of successful outcomes (Glick et al, 1984), results have been inconsistent with little certainty as to the effective length for interposition.

**Antiperistaltic (reversed) segments**

Antiperistaltic (reversed) segments attempt to delay nutrient transit thereby increasing mucosal contact time and aiding absorption (Thompson and Rikkers, 1987). Surgery involves an in-situ 180° rotation of a length of small bowel on its axis without impairing its blood supply. Single segments are commonly located at the distal end of the jejunum just proximal to the anastomosis to the colon. Multiple reversed segments are placed at different levels along the small bowel. Although the literature suggests single 3 cm lengths for children and 10–15 cm for adults (Pigot et al, 1990), the critical length, position and number for an effective reverse are likely dependant on the unique individual circumstances. Experience with antiperistaltic (reversed) segments has been limited (Panis et al, 1997), and results have been variable with an inconsistency in outcome that has not encouraged their use.

**Semi-obstructive valves**

Semi-obstructive valves are commonly placed between the jejunum and the colon to slow transit and increase nutrient contact with the absorptive small bowel. Several designs have been proposed including intussusception and reimplantation models. The valve’s effectiveness depends on the degree of subacute obstruction that it induces. Georgeson proposed a nipple valve, wrapping a layer of foreign material around the bowel sufficiently tightly to delay passage and induce proximal bowel dilatation (Collins et al, 1995; Vernon and Georgeson, 2001).

Valve construction, with or without foreign materials, carries an inherent risk of further reducing valuable absorptive bowel length at the time of valve resection. Significantly the degree of obstruction is fixed and may be insufficient or may eventually lead to stasis and sepsis within the proximal dilating bowel. Reported results have been variable, but inconsistency in outcome and inability to vary the degree of obstruction have been detracting factors.

**Table 1. Autologous gastrointestinal reconstructive techniques**

Transit delay	Colon interposition
	Semiobstructive intestinal valves
	Reversed (antiperistaltic) segments
Improved propulsion	Tailoring
	Plication (de Lorimier)
Bowel expansion	Nipple valve (Georgeson)
	Controlled tube occlusion (Bianchi)
Bowel lengthening	Longitudinal – LILT (Bianchi)
	Serial transverse enteroplasty (Kim)
	Horizontal-isolated bowel segment (Kimura)
	Composite bowel loop (Bianchi)
Combined techniques	Sequential lengthening (Georgeson)
	LILT and reversed segment(s)
	Expansion and LILT and reversed segments
	Other combinations

LILT = longitudinal intestinal lengthening and tailoring

**Tailoring and plication**

Tailoring and plication by reduction towards normal diameter are designed to counteract the ineffective propulsion and consequent stasis with sepsis that develop within grossly dilated small bowel loops. Both techniques involve the antimesenteric border of the dilated loop, which is excised (tailoring) or imbricated (plication) to reduce bowel diameter. The downside to tailoring is the loss of a large amount of potentially valuable absorptive mucosa, reducing or eliminating the child’s ability to achieve enteral autonomy. Plication attempts to preserve the mucosa by turning it inwards into the bowel lumen (de Lorimier and Harrison, 1983). To avoid recurrence of the dilatation it is necessary to excise the antimesenteric seromuscular layer, which may alter the mucosal absorptive characteristics. Both tailoring and plication fail to increase bowel length or to delay transit. When sufficient bowel length is available, tailoring of the dilated bowel is preferable to plication; however, on its own neither technique is of major value to the management of the short bowel state.

**Bowel tailoring with lengthening**

Bowel tailoring with lengthening tailors the dilated small bowel to aid propulsion, preserves all absorptive mucosa and uses the tailored segment to increase small bowel length. Nutrient mucosal contact time is increased and bowel adaptation is enhanced. Various techniques are now available:

**Longitudinal intestinal lengthening and tailoring**

Longitudinal intestinal lengthening and tailoring (LILT) divides the dilated small bowel loop longitudinally along

its mesenteric and antimesenteric borders with each hemisegment remaining fully vascularized by its ipsilateral mesenteric vessels (Bianchi, 1980). The hemisegments are rolled into hemiloops and anastomosed isoperistaltically to halve the original diameter and double the length. LILT, first applied successfully clinically by Boeckman and Traylor (1981), has now been used extensively with minimal morbidity and promising outcome (Bianchi, 1995; Waag et al, 1999). Neonates with bowel lengths of >30 cm and particularly with an ileocaecal valve and longer lengths of colon are likely to benefit most. For others, a lesser dependence on parenteral nutrition represents a welcome reduction in morbidity and a better quality of life.

### **Isolated bowel segment (Iowa models)**

Proposed by Kimura and Soper (1990, 1993), isolated bowel segment depends on the development of a new blood supply to the antimesenteric border of the dilated jejunum through adhesions to the liver and the abdominal wall. The seromuscular layer of the dilated jejunum is peeled back to expose the submucosa that is then applied to the scarified surface of the liver and abdominal wall. At a subsequent procedure the dilated jejunum is divided horizontally and the bowel segments are tubularized and anastomosed isoperistaltically to increase small bowel length and reduce diameter without loss of absorptive mucosa.

### **Composite bowel loops**

These are constructed by grafting small bowel mucosa to a mucosally denuded seromuscular flap that is raised from the greater curvature of the stomach on the right gastroepiploic vessels, or to a segment of mucosally denuded colon supplied by the colonic mesentery (Bianchi et al, 1996). These vascularized seromuscular surfaces are applied to the exposed submucosa at the antimesenteric border of the dilated jejunum. Some 6 weeks later the dilated jejunum is divided horizontally and the hemisegments are tubularized and anastomosed isoperistaltically to increase total bowel length. Success depends on sufficient development of a new blood supply across the graft interface.

### **Serial transverse enteroplasty**

In 2003 Kim et al proposed serial transverse enteroplasty (STEP) to reduce dilated small bowel diameter and to increase length without the mesenteric dissection inherent to LILT (Kim et al, 2003). Using a longitudinal stapler and working serially alternately from each side, the dilated bowel is partially divided transversely to create a concertina effect. Early results suggest that this technique is a welcome addition to the management of children with short bowel. When bowel length is adequate but dilatation is a problem, STEP can be used primarily as the sole tailoring and lengthening procedure. If residual bowel length is more critical STEP is better considered after LILT for adding further to bowel length.

### **Bowel expansion**

If only short lengths of small bowel are available or, as in postnatal midgut volvulus, when the residual jejunum is of normal diameter, it is necessary to develop more bowel to increase the absorptive surface area and to facilitate reconstructive procedures. Noting that bowel dilatation occurred proximal to a chronic obstruction Georgeson et al (1994) placed a semiobstructive nipple valve at the jejunocolic anastomosis, and followed this with LILT once dilatation had occurred.

Georgeson et al's contribution was seminal in introducing the concept of bowel expansion. To overcome the fixed obstruction inherent to the nipple valve and the loss of valuable absorptive bowel at eventual valve resection, Bianchi introduced controlled bowel expansion (Bianchi, 2006) by intermittent occlusion of a large tube (Malecot 16F) placed in the jejunum and brought out onto the abdominal wall. This technique has the added advantages of minimal early surgery for the vulnerable short gut child with an immature liver, and of providing access and controlled drainage of the small bowel lumen to avoid stasis and sepsis. Collected jejunostomy drainage (nutrients, bile and pancreatic enzymes) is recycled down a second tube placed in the distal bowel to develop its absorptive potential.

### **Combined procedures**

In sequential lengthening Georgeson et al (1994) followed bowel expansion with their nipple valve with simultaneous Kimura isolated bowel segment and Bianchi LILT to maximize bowel length. Similarly in one patient Bianchi (1995) combined the composite gastrojejunal loop with LILT to triple the length of the small bowel. Unfortunately this child died from end-stage liver failure before establishing enteral autonomy. Two subsequent occasions of controlled bowel expansion by jejunostomy tube occlusion were followed by simultaneous LILT and a 5 cm reversed (antiperistaltic) segment at the jejunocolic anastomosis. Both children achieved enteral autonomy, but the simultaneous use of two techniques made it difficult to determine whether this was a direct result of this combination.

On a further occasion, over a 3-year period, reconstructive procedures were undertaken separately and at intervals with careful assessment of enteral absorption and parenteral nutrition dependence after each event. A 6-month period of controlled bowel expansion by jejunostomy tube occlusion was followed by LILT. After 1.5 years, when there was no further improvement in enteral absorption and parenteral nutrition was necessary on three nights each week, two 12 cm jejunal segments were reversed at separate points along the lengthened small bowel. Parenteral requirements again decreased progressively until full enteral autonomy sufficient to sustain survival and growth was established 1 year later. These cases are the subject of a future detailed publication. Other combinations of

bowel reconstructive techniques are possible and could usefully include the STEP procedure for additional length after LILT.

## Discussion

The management of the child with short bowel should be dictated by the long-term goal of survival and growth with good quality of life and minimal morbidity or mortality. This is only achievable with the patient's own bowel. The path is long and arduous requiring careful attention to preservation of venous access for central feeding catheters for life-supporting parenteral nutrition for several months or years. Protection of the liver from sepsis and from parenteral nutrition related toxicity (Iyer et al, 1998), specifically cholestasis-inducing plant phyosterols in fat solutions (intralipid), is essential to survival and to bowel adaptation (Iyer et al, 2004).

Autologous bowel reconstructive techniques (*Table 1*) are safe, and when applied in a structured manner as part of a patient-orientated individualized plan, have been shown to enhance the natural adaptive response and to improve absorption over increased lengths of propulsive small bowel. Techniques that may be relatively ineffective when used alone can prove significant when applied in combination within a structured management plan that also includes pharmacological manipulation of bowel activity and absorption. Thus, in the words of Bernard Messing (personal communication, 2004) 'even short lengths of bowel have great value'. When autologous bowel is not available or is insufficient in spite of all attempts at bowel reconstruction, it is appropriate to offer bowel transplantation with its attendant ever-present risks from aggressive immunosuppression (sepsis, lymphoproliferative disease) and from acute and chronic rejection. Until such time bowel transplantation is premature.

The child or adult with a short residual small bowel can now realistically hope for a return to enteral autonomy with good quality of life. He or she requires high-level multidisciplinary specialist services over a prolonged period determined by the natural adaptive response of the bowel. These services are best coordinated by a dedicated team leader in an intestinal failure centre (Bianchi, 2006), who is responsible for implementing an agreed individualized management plan to be delivered largely by local carers, thereby enabling the patient to retain a family, social and working life. Experience gained from a larger number of patients would allow intestinal failure centres to become the focus for advice, to direct expensive resources, and to stimulate research towards a better future for the unfortunate patient with short bowel.

*Conflict of interest: none.*

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## KEY POINTS

- Autologous gastrointestinal reconstruction of the residual short bowel offers a realistic hope for a return to enteral autonomy and good quality of life.
- Short bowel patients need careful attention to central venous access and protection from sepsis and liver toxicity during long-term parenteral nutrition.
- Multidisciplinary specialist management should be guided by a dedicated intestinal failure centre, but delivered by the local care team keeping the patient within his/her family and work environment.
- Autologous gastrointestinal reconstruction combines multiple safe surgical techniques (expansion, lengthening, and antiperistaltic segments) with medical therapy within a structured individualized plan over a long time frame.
- Bowel transplantation still carries major life-threatening risks and should be offered only when all other options have failed.