CHAPTER 71
SHORT BOWEL SYNDROME

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Introduction

Short bowel syndrome (SBS) is defined as intestinal failure due to a loss of intestine resulting in inadequate length of bowel for maintaining the nutrition and hydration of the individual without either intravenous or oral supplementation.

With some reported exceptions, the minimum length of small bowel required for infant survival on enteral feeds is generally 25 cm in the presence of an intact ileocaecal valve (ICV) and colon, and 40 cm without an ICV and large bowel. Note that norms of intestinal length vary considerably, with a range of 250 cm to 300 cm of small bowel at term. The estimated length in a preterm infant of 26 and 32 weeks gestation is 70 cm and 120 cm, respectively. Thus, gestational age is an important factor. However, an infant is considered to have SBS when he or she behaves as if SBS is present, and the infant should be treated as such.

The intestine has the ability to adapt over time such that up to half of patients with SBS who initially require total parenteral nutrition (TPN) may be weaned off and gradually gain independence from TPN. However, in the absence of the availability of TPN, SBS carries a dismal prognosis. The management of SBS is resource-intense, requiring the availability of intensive care, TPN, and expert medical and surgical intervention. Even in optimal settings, infants and children with SBS suffer extensive morbidity and mortality. In most countries in Africa where TPN is not available, the outcomes are poor.

Demographics

Short bowel syndrome is mercifully rare. In Europe, the incidence is estimated to be approximately 2 per million. According to Gupta et al.,[1] the incidence of SBS in neonates is around 3 per 100,000 births per year. In Africa, the incidence is unknown because survival is close to zero.

Aetiology

Some of the common causes of short bowel syndrome are shown in Table 71.1. SBS can be congenital, but is more generally acquired from surgical resection of bowel. Of the congenital bowel atresias type 3b ("apple peel" type) and type 4 (multiple atresias, "string of sausages" type) are most likely to result in SBS. Functional SBS can also occur where there is severe malabsorption despite adequate bowel length or intact bowel.

Table 71.1: Common causes of short bowel syndrome.

Congenital	Acquired
Atresia Gastroschisis Hirschsprungs disease (long segment)	Midgut volvulus Mesenteric infarction (e.g., sickle cell crisis) Necrotising enterocolitis; adhesive band obstruction/strangulation Trauma

Pathophysiology

The effects of loss of bowel length depend on the type and length of bowel remaining.[1]

Small bowel motility is three times slower in the ileum than in the jejunum. The ileo-caecal valve also slows transit. The colon has the slowest transit time, between 24 and 150 hours. The efficiency of salt and water absorption also varies in the different parts of the intestine. The jejunum is very inefficient, with an efficiency of water absorption of 44% compared to 70% in the ileum and greater than 90% in the colon. The corresponding estimates for efficiency of salt absorption are 13% in the jejunum, 72% in the ileum, and greater than 90% in the colon.

Jejunum

If the jejunum alone is lost, there is no permanent defect in absorption—the ileum will take over. However, the jejunum normally releases the hormones cholecystokinin, serotonin, gastric inhibitory peptide, and secretin. Lower secretion of these hormones due to absence of the jejunum will result in decreased pancreatic secretion, gallbladder contraction, and gastric hypersecretion.

Ileum

The ileum is unique in absorbing vitamin B_{12} and bile salts. Absorption of nutrients takes place throughout the small bowel. Ileal resection results in decreased transit times and salt and water absorption. A larger than normal volume of fluid and electrolytes enters the colon. The reabsorption of bile salts is decreased and so their synthesis is increased. The flow of unabsorbed bile salts into the colon irritates the colon and impairs its ability to absorb salt and water, leading to choleretic diarrhoea. The depletion of the bile salt pool leads to cholelithiasis and fat malabsorption, resulting in steatorrhoea.

The degree of fat and carbohydrate malabsorption is related to the length of ileum resected. There is also reduced absorption of essential minerals, including calcium, magnesium, zinc, and phosphorus. Furthermore, there is an excessive loss of zinc in diarrhoea, leading to immune deficiency.

Ileocaecal Valve

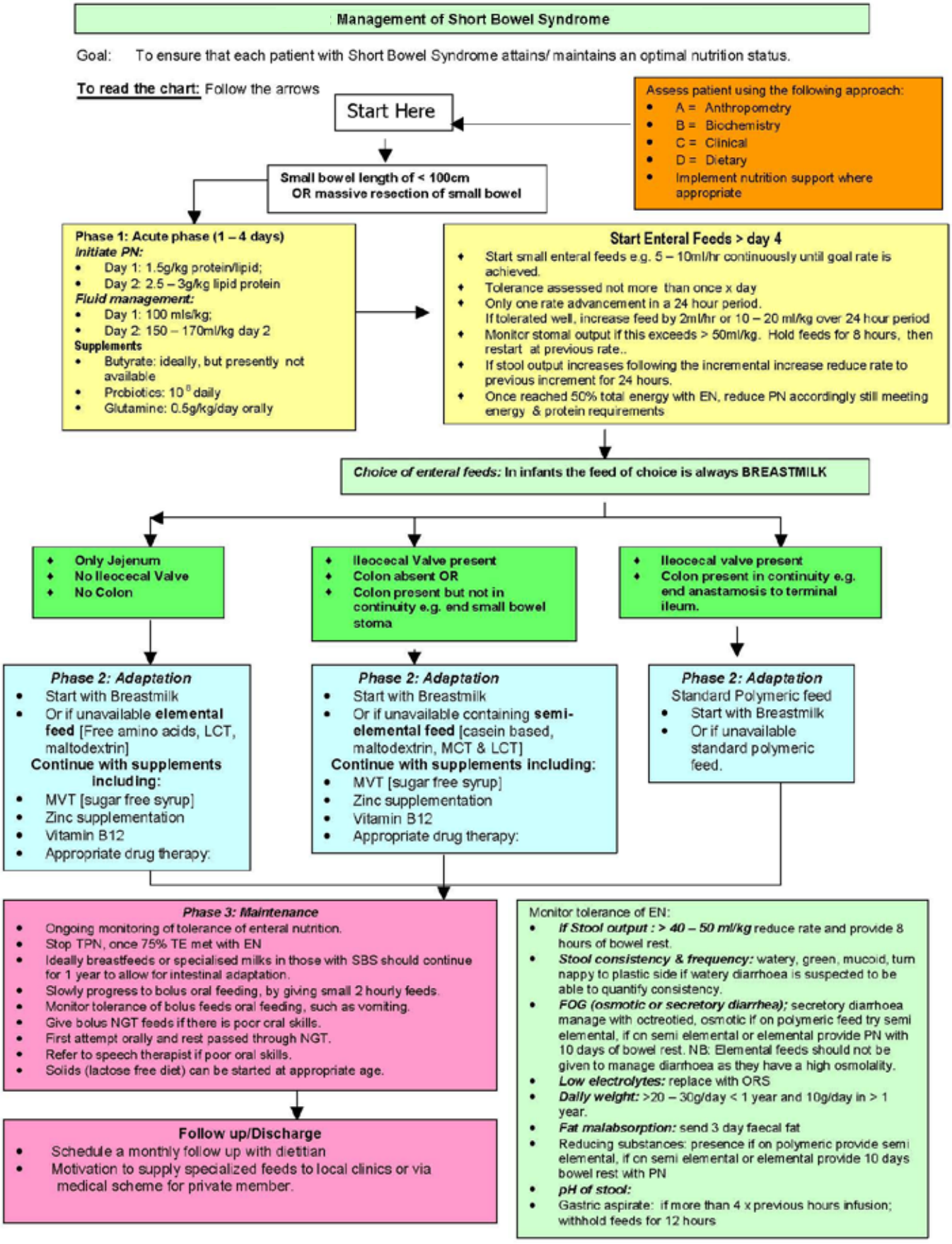
Loss of the ICV reduces gut transit time. The loss of this barrier between the small and large bowel also leads to bacterial overgrowth of the remaining gut.

Colon

It is thought that the colon in SBS is important for driving adaptation of the gut as well as assuming an increased importance in absorption of water, potassium, and sodium as well as carbohydrates. Without the colon and ICV a longer length of ileum is required for survival.

Intestinal Adaptation

Intestinal adaptation[2] is the process whereby the intestine adjusts to its loss of length through hyperplasia of the mucosal surface in an effort to increase its absorptive capacity. The bowel dilates, lengthens, and thickens to increase the efficiency of absorption per unit length. There is an increase in the number of cells in the proliferating zones of the crypts, and villus height increases, resulting in an increased surface area for absorption. Histological evidence of adaptation can be seen from 48 hours after bowel resection, and diarrhoea is seen to decrease over the first 3 months. Early adaptation may take years and is greater with proximal than distal bowel resection.



Source: Adapted from Marino, L. Western Cape Guidelines for Management of short bowel syndrome.

Figure 71.1: Algorithm for the management of short bowel syndrome to ensure that each patient attains and maintains optimal nutrition.

Note: EN = enteral nutrition; FOG = faecal osmolar gap; LCT = long-chain triglycerides; MCT = medium chain triglycerides; MVT = multivitamins; NB = nota bene (note well); NGT = nasogastric tube; ORS =oral rehydration solution; PN = parenteral nutrition; TE = total energy (requirements); TPN = total parenteral nutrition.

Adaptation is driven by the increased load of fatty acids, carbohydrates, and proteins on the enteroglucagon-producing cells found in the ileum. Enteroglucagon stimulates ornithine decarboxylase, which in turn stimulates crypt cell proliferation. In animal models, other factors (e.g., glutamine, epidermal growth factor, cholecystokinin, and somatostatin) have also been shown to be involved in intestinal adaptation, although there is little evidence so far that their clinical use increases adaptation in humans.

The colon can become a digestive organ in patients with SBS. Bacteria in the colon can ferment undigested starch and fibres into short chain fatty acids, which are the preferred fuel for colonocytes. An intact colon will increase its energy absorption during the adaptive phase postoperatively by increasing the fermentation of carbohydrates.

The caloric requirement per kilogram also decreases with age, particularly after 1 year of age, which also contributes towards adapting to a shorter bowel length.

Management

The initial and primary consideration in the immediate period following extensive bowel resection[3] concerns fluid and electrolyte balance, even before calories. Gastric hypersecretion in the early period requires control with an H_2 receptor antagonist or proton pump inhibitor. Patients should be initially kept NPO (nothing by mouth) and have a nasogastric tube placed on free drainage, as well as a urinary catheter placed for monitoring fluid balance. All patients will require intravenous fluids to replace fluid losses.

Sodium and potassium chloride are the most important ions to closely monitor and replace. An infusion of normal saline (0.9%) with potassium chloride should be used to replace millilitre for millilitre measured enteral (stoma and nasogastric) fluid losses. Additional amounts of sodium and potassium may need to be given separately to avoid deficiency. Urine output should be monitored, and an adequate urine output maintained. Urinary sodium levels, where available can also be used to monitor sodium loss. A urine sodium level >30 mmol/l should be maintained.

Nutritional therapy should not be introduced until the patient is haemodynamically stable and fluid management is relatively stable, which is likely to be a few days after bowel resection.

Nearly all patients with SBS will require parenteral nutrition to survive the period while the bowel adapts. Avoidance of TPN depends on the anatomy of the remaining bowel; patients with an intact colon are the most likely to be able to survive without TPN.

Oral feeds can be started at the same time as parenteral feed and gradually increased as tolerated by the patient. Parenteral nutrition is then decreased as enteral feeding is increased.

In infants, breast milk with oral sodium and vitamin supplements can be used.

The initial oral treatment in older children should be with oral rehydration solution and a gradual introduction of carbohydrates. Children with SBS will require a diet high in calories from both fat and carbohydrates to provide sufficient calories despite malabsorption. They will also need supplements of potassium, sodium, magnesium, calcium, fat-soluble vitamins (large doses of vitamins A, D, and E), and zinc. Sodium is vital because it stimulates the bowel to absorb, promoting adaptation. Vitamin B_{12} injections are specifically required with the loss of the distal ileum.

Loperamide can be used to slow intestinal transit and decrease diarrhoea. In those patients with a stoma, effluent from the proximal bowel stoma can be introduced down the mucous fistula to promote bowel growth and adaptation of the distal bowel before stoma closure.

As feed is introduced, those patients who will tolerate enteral feeding and those who will be dependent on parenteral nutrition will become apparent. Bowel adaptation can take months or years, so survival of these patients will depend on funding for, and availability of, home parenteral nutrition.

Other medications used are cholestyramine to reduce the irritant effect of bile salts on the colon, ursodeoxycholic acid to reduce cholestasis, and intermittent use of oral antibiotics to reduce bacterial overgrowth (see algorithm in Figure 71.1).

Surgical Options

The main aims of surgery for SBS are to correct mechanical obstruction in order to decrease bacterial overgrowth, and to maximise bowel length. More recently, intestinal transplant has become a reality in selected centres worldwide. Stomas should be closed as early as possible so that all potentially functional bowel is used.

Tapering

Isolated dilated stagnant sections of bowel are a site for bacterial overgrowth. If symptoms of bacterial overgrowth are present, then dilated segments should be treated with tapering, especially in the duodenum and jejunum. This procedure involves excision of the antimesenteric border of the dilated portion of bowel. This enables more effective peristalsis, thus reducing stasis and bacterial overgrowth. Inversion placation has been used in an attempt to preserve mucosa but tends to unravel despite technical modifications such as seromuscular stripping of the inverted segment.

Bowel Lengthening

Bowel lengthening relies on the presence of dilated bowel resulting from intestinal adaptation and should therefore be reserved until 6 months to 1 year following initial bowel resection.

The two main bowel-lengthening procedures are Bianchi's longitudinal intestinal lengthening and tailoring (LILT)[4] and serial transverse entroplasty (STEP)[5].

Bianchi's LILT procedure (Figure 71.2) makes use of the bifurcation of the mesenteric vessels at the mesenteric border of the small bowel. The bowel is divided longitudinally between the mesenteric and antimesenteric borders along its dual blood supply, dividing the bowel into two limbs, each with a blood supply. These two limbs are then closed and anastomosed end to end, thus doubling that length of bowel.

In one series, 9 of 20 patients survived with this procedure, 7 of whom were able to wean off TPN. Factors associated with success were lack of liver failure and presence of at least 40 cm of intestine before the doubling procedure.

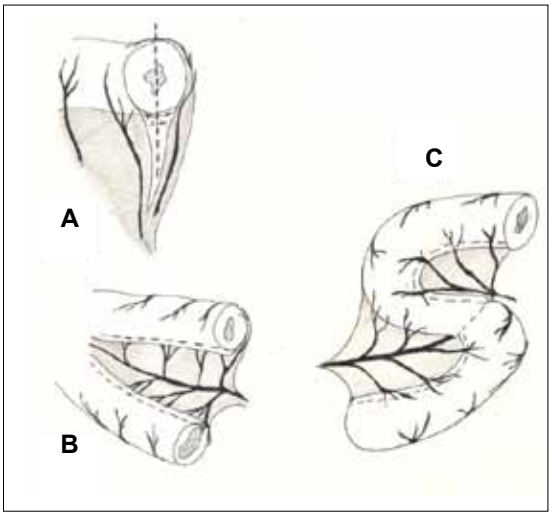


Figure 70.2: Bianchi's LILT procedure: (A) A natural plane between the leaves of the mesentery is found by dividing the bowel as shown (dotted line) and using upwards and outwards traction on the divided bowel. The bowel and mesentery are divided into two along the length so each hemisegment of bowel has a leaf of mesentery with blood supply. (B) The two hemisegments are then tubularised by using a continuous horizontal mattress 5/0 absorbable suture. (C) The opposite ends of the two new bowel segments are apposed and anastomosed in an S-shape with the bowel overlying the mesentery.

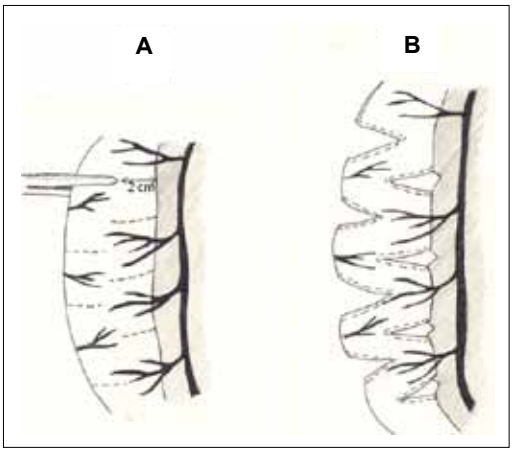


Figure 70.3: The STEP procedure: (A) A GIA stapler is used transversely across the dilated bowel from the antimesenteric border, leaving 2 cm of the bowel diameter uncut. The next cut is taken too distally this time from the antimesenteric border via a small gap created in the mesentery. (B) The GIA stapler is used down the bowel, alternating between the mesenteric and antimesenteric border as shown (dotted lines). Care is taken to keep the GIA stapler perpendicular to the mesentery to preserve the blood supply.

The STEP procedure is shown in Figure 71.3. STEP registry figures show a mean pre-STEP bowel length of 68 cm achieving a mean post-STEP bowel length of 115 cm. The percentage of enteral feeding increased from a mean of 33% preoperatively to a mean of 63% postoperatively.

Complications of bowel-lengthening procedures are high, including anastomotic and staple line leaks, bowel obstruction from adhesions or ischaemic strictures, bleeding, abscess formation, and death.

The limitations of bowel-lengthening procedures have led some authors to advocate that they should be reserved for those patients who, after 6 months of bowel adaptation, are tolerating more than half of their feeds enterally and would therefore have a greater chance of successfully becoming fully enterally fed following a lengthening procedure.

Intestinal Transplant

Intestinal transplant is offered in only a few centres worldwide. It is not an alternative to long-term TPN. It is reserved only for patients who are unable to have TPN, usually due to TPN-related liver disease or difficulty with venous access for TPN administration.

Intestinal transplant may involve (1) isolated bowel, for those with good liver function and normal motility; (2) bowel plus liver, for those with liver disease; or (3) multivisceral, which includes liver, bowel, stomach, and pancreas, for those with multiple abdominal organ failure and dysmotile bowel. The most frequent transplant performed for children with SBS is a liver plus bowel transplant. This procedure is currently limited to children weighing more than 5 kg due to the lack of size-matched donors.

Isolated liver transplants can be offered to some infants with early-onset liver failure but with sufficient bowel length such that adaptation could be expected.

The 5-year survival is approximately 70%; however, around 25% of patients die while on the waiting list for a transplant.

Prognosis and Outcomes of SBS

Long-term survival without TPN depends on the remaining bowel length. With TPN availability, survival is related to complications of TPN rather than to bowel length. The overall mortality of infants with SBS on TPN is 10–25%.

The two main causes of death and long-term morbidity in patients with short gut on TPN are liver failure and sepsis. In children, liver failure is secondary to intrahepatic cholestasis. This is most common in those who are entirely TPN dependent with no enteral feeding. It is also seen more frequently in neonates who are premature, have low birth weight, and have recurrent sepsis. The other main cause of death—septicaemia—arises because a complete lack of enteral nutrition results in bacterial overgrowth of the bowel and mucosal atrophy with impaired mucosal immunity, leading to an increased incidence of sepsis. Recurrent septicaemia is also related to central venous catheters. Early enteral feeding is therefore vital if these complications are to be reduced.

Even after discharge from hospital on full enteral feeds, infants are at risk during episodes of enteric infection, when rapid fluid and electrolyte loss may require emergency admission for intravenous rehydration. The management of a patient with SBS requires a multidisciplinary team, including paediatrician, surgeon, community nurse, dietitian, and pharmacist.

An audit of 63 patients with SBS seen at the Red Cross Children's Hospital between 1998 and 2006 revealed the following: The mean gestational age was 32 weeks (range 25–40 weeks). The most frequent causes were necrotising enterocolitis (NEC) (40%), along with intestinal atresia, midgut volvulus, intestinal aganglionosis, and gastroschisis. Overall, mortality was 36.5% (23/63). The mean number of days on parenteral nutrition was 95 (range 30–420 days).

Ethical Issues

The annual cost of care of a patient with SBS on parenteral nutrition has been estimated at between $100,000 and $150,000, making such care beyond the reach of all but a few. Treatment of patients with significant bowel loss in resource-poor settings is likely to be limited to those who attain enteral feeding quickly and have sufficient bowel function to require only increased oral calories and vitamin and mineral supplementation. There is therefore a need to counsel parents before surgery for bowel conditions that can potentially lead to short gut syndrome.

Evidence-Based Research

Table 71.2 presents a comparison of intestinal-lengthening procedures for patients with SBS.

Table 71.2: Evidence-based research.

Title	Comparison of intestinal lengthening procedures for patients with short bowel syndrome
Authors	Sudan D, Thompson J, Botha J, Grant W, Antonson D, Raynor S, Langnas A
Institution	Department of Surgery, Nebraska Medical Center, Omaha, Nebraska, USA
Reference	Ann Surg 2007; 246(4):593–601; discussion 601–604
Problem	Outcome of bowel-lengthening procedures.
Intervention	A review of the clinical results of 24 years of intestinal lengthening procedures at one institution.
Comparison/ control (quality of evidence)	A retrospective review of a single centre experience comparing the outcome of two intestinal-lengthening procedures (Bianchi and STEP) in terms of survival, total parenteral nutrition (TPN) weaning, and complications.
Outcome/ effect	This review involved 64 patients, including 14 adults, who underwent 43 Bianchi and 34 STEP procedures between 1982 and 2007. Three patients had prior isolated liver transplants. The median (range) remnant bowel length before first lengthening was 45 (11–150) cm overall (Bianchi = 44 cm; STEP = 45 cm); and 68 (20–250) cm after lengthening (Bianchi = 68 cm; STEP = 65 cm). Actual survival is 91% overall (Bianchi, 88%; STEP, 95%), with a median follow-up of 3.8 years (Bianchi = 5.9 years; STEP = 1.7 years). Average enteral caloric intake in paediatric patients was 15 kcal/kg before lengthening and 85 kcal/kg at 1 year after lengthening. Sixty-nine percent of patients were off TPN at the most recent follow-up, including 8 who were weaned from TPN after intestinal transplantation. Liver disease (when present) was reversed in 80%. Surgical complications occurred in 10%, more commonly requiring reoperation after Bianchi than STEP. Intestinal transplantation salvage was required in 14% at a median of 2.9 years (range = 8 months to 20.7 years) after lengthening.
Historical significance/ comments	Surgical lengthening with both Bianchi and STEP procedures results in improvement in enteral nutrition, reverses complications of TPN, and avoids intestinal transplantation in the majority, with few surgical complications. Intestinal transplantation can salvage most patients who later develop life-threatening complications or fail to wean TPN.

Key Summary Points

1. Short bowel syndrome may be congenital or acquired.
2. The effect of bowel resection depends on the site and length of bowel resected and the bowel remaining.
3. Intestinal adaptation takes place by means of bowel dilatation, lengthening, and thickening to increase the efficiency of absorption per unit of length.
4. Medical management is mainly supportive along with fluid and electrolyte balance, nutritional support with or without TPN, and infection control.
5. Surgical options include tapering, bowel lengthening using the Bianchi approach, the STEP procedure, or bowel transplant.
6. Long-term survival without TPN depends on the remaining bowel length. With TPN availability, survival is related to complications of TPN rather than to bowel length.

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