

# Short Bowel Syndrome

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Short bowel syndrome (SBS) may be defined as any disease, traumatic injury, or vascular accident which leaves less than 200 cm of viable small bowel or results in a loss of 50% or more of the small intestine.<sup>[1]</sup>

## Pathophysiology<sup>[1]</sup>

Loss of large amounts of small intestinal absorptive mucosa results in malabsorption of water, electrolytes and nutrients. It also results in failure of resorption of the liquid produced by the gastrointestinal tract itself. This scenario is being increasingly referred to as 'intestinal failure'.<sup>[2]</sup>

Some 8,000-9,000 ml of fluid per day are produced, of which the normally functioning bowel retains all but 100-200 ml. One feature of small bowel syndrome is thus the production of large amounts of liquid stool. The situation may be compounded by colonisation with bacteria. One study of the bowel flora of patients who developed SBS after bowel resection showed a predominance of *Lactobacillus mucosae*.<sup>[3]</sup>

Loss of significant lengths of ileum results in reduction of bile salt reabsorption, resulting in steatorrhea.

SBS is not an inevitable consequence of loss of a significant amount of bowel. Other factors which come into play include:

- The length of premorbid bowel.
- Which segment of bowel is lost.
- The age of the patient at the time the bowel was lost.
- The remaining length of small and large intestine.
- Whether the ileocaecal valve is present or absent.

Three physiological changes may be identified.

- Acute phase - immediately after bowel resection. It is associated with malnutrition and fluid and electrolyte loss.
- Adaptation phase - 2-4 days after bowel resection, histological changes result in improved absorption and may influence the amount of parenteral nutrition required.
- Maintenance phase - the absorptive capacity of the bowel recovers to a greater or lesser extent. The degree of recovery will dictate the management of the patient in terms of the need for parenteral nutrition and nutritional supplements.

## Epidemiology<sup>[1]</sup>

The patient profile has changed over the years. Bowel strangulation and midgut volvulus were common aetiologies in the first decades of the twentieth century. By the 1950s and 1960s, the common causes had become mesenteric vascular accidents, including thrombosis and embolism of the superior mesenteric artery. The most common aetiology in adults is currently *Crohn's disease*.

Extrapolated figures of home parenteral nutrition centres and single-centre studies suggest an incidence of 2-5 per million.<sup>[4]</sup>

## Presentation<sup>[4]</sup>

### History

The presenting history is usually diarrhoea and this may be coupled with weight loss, fatigue, malaise and lethargy.

The past history may be highly significant and may include Crohn's disease, or a history of several intestinal resections, a major abdominal catastrophe or vascular accident (eg midgut volvulus or embolus of the superior mesenteric vessels).

Vitamin and mineral deficiency can cause:

- **Night blindness** and xerophthalmia (vitamin A).
- Paraesthesias (vitamins D and E, calcium, magnesium).
- Tetany (vitamins D and E, calcium, magnesium).
- Ataxic gait and visual disturbance due to retinopathy (vitamin E).
- Spontaneous bruising or prolonged bleeding (vitamin K).
- Dyspnoea, exertion and lethargy (**vitamin B12**, folic acid, iron).
- Anorexia.

## Examination

Several features giving a clue to the diagnosis may be revealed by examination. These relate to the following nutritional abnormalities:

- Protein loss and malnutrition - there may be temporal wasting, loss of digital muscle mass, peripheral oedema, dry flaky skin, blunt lingual papillae and prominent ridges in the nails. Poor **growth** performance in children is characteristic.
- Deficiency of essential fatty acids (linoleic and linolenic) - growth restriction, dermatitis, alopecia.
- **Vitamin A deficiency** - corneal ulcerations, growth delays.
- B complex vitamins in general - stomatitis, cheilosis and glossitis.
- Vitamin B1 deficiency - oedema, tachycardia, ophthalmoplegia, depressed deep tendon reflexes.
- Vitamin B6 deficiency - peripheral neuropathies (also a feature of B12 deficiency) and seizures.
- Vitamin D depletion - poor growth, bowed extremities.
- Vitamin E deficiency - if severe, this can result in ataxia, oedema and depressed deep tendon reflexes.
- Vitamin K deficiency - petechiae, ecchymoses, purpura, or outright bleeding diatheses.
- Iron deficiency - pallor, spooned nails, glossitis.
- **Zinc deficiency** - angular stomatitis, poor wound healing, alopecia, scaly erythematous rash around the mouth, eye, nose and perineum.

## Differential diagnosis

This may be wide-ranging and depending on presentation may include:

- Causes of chronic diarrhoea
- Causes of malabsorption
- Causes of growth failure

## Investigations<sup>[4]</sup>

### Laboratory tests

- FBC - to check for anaemia; the type of anaemia may give a clue as to the nature of any deficiency (ie macrocytic or microcytic).
- Albumin - low albumin may indicate poor nutritional status whilst a high level may be seen in dehydration.
- LFTs - persistently elevated liver enzymes may indicate parenchymal damage. This should be differentiated from the transient elevation seen in patients on parenteral nutrition.
- Electrolytes - these may indicate dehydration and also need to be monitored in patients on parenteral nutrition in order for any abnormal levels to be corrected.
- Creatinine - may signal deteriorating renal function and suggest a change in nutritional support.
- Serum calcium, magnesium and phosphorus - to detect deficiencies which can affect many metabolic processes.
- Serum vitamin, mineral and trace element levels - to detect any deficiency.

- Coagulation profile - INR, prothrombin time (PT) and activated partial thromboplastin time (aPTT) should be assessed to exclude coagulopathy associated with hepatic dysfunction, especially in patients being considered for surgery.

## Imaging

- Plain abdominal Xray - to exclude signs of ileus or obstruction and provide information on bowel status.
- Upper gastrointestinal series with small bowel follow-through - to identify areas of stricture and assess the appearance of small bowel mucosa. In the recovery stage, dilation of the bowel indicates that the adaptation phase has been reached.
- Abdominal CT scan with contrast - useful for identifying bowel obstruction, imaging the liver (and any consequent **cirrhosis** or early signs of liver dysfunction).
- Abdominal ultrasound - to exclude biliary sludge or gallstones, which may be associated with short bowel syndrome (SBS).

## Other tests

- Bone densitometry - to exclude metabolic bone disease which may develop as a consequence of calcium and **vitamin D deficiency**.
- Liver biopsy - in patients with abnormal LFTs. A variety of associated conditions may be found, including **cholestasis**, fatty change, non-alcoholic steatohepatitis and cirrhosis.

## Associated diseases<sup>[4]</sup>

### Adults

- Crohn's disease
- **Radiation enteritis**
- Mesenteric vascular accidents
- Trauma
- Recurrent **intestinal obstruction**

### Children

- **Necrotising enterocolitis** (the most common cause).
- **Ischaemic bowel** infarction.
- Intestinal atresias.
- **Intestinal volvulus**.
- Congenital short small bowel.<sup>[5]</sup>
- **Gastroschisis**.
- Meconium peritonitis.

## Management<sup>[1]</sup> <sup>[2]</sup>

### Medical

- Most patients are treated with **total parenteral nutrition** (TPN) in the early stages.
- A decision is then taken as to whether the patient needs limited specialised enteral therapy or prolonged TPN. This will depend on the length and function of the remaining intestine and the mechanisms influencing transit time, including the functioning of the ileocaecal valve and colon. A combination of TPN and enteral feeding is often tried as this facilitates the discontinuation of TPN at the earliest possible stage.
- Haemodynamic considerations are important and the volume of fluid replaced should be calculated according to the amount obtained from nasogastric suctioning and stool plus 300-500 ml for insensible losses. This volume should be added to the patient's usual daily maintenance volume. Urine output should be at least 1 litre daily.
- Parenteral nutrition should provide adequate protein, calories, macronutrients and micronutrients. The time for adaptation, when improved absorption occurs, is controversial. Some authorities hold that maximal adaptation can take up to a year, others that little improvement is to be obtained after three months.

- Most patients will require 7-10 days of parenteral feeding but some patients may require much longer. Nutrient absorption is not complete in patients who have lost half or more of their small bowel, Patients with less than 60 cm of small bowel remaining, loss of the ileum and ileocaecal valve and loss of the colon are likely to require lifelong parenteral therapy.
- Excessive fluid and electrolyte loss (usually occurring in the first week after extensive intestinal resection) requires resuscitation with fluids, parenteral nutrition or both. Subsequent fluid replacement should be dictated by the amount of stool or ostomy output.
- Levels of vitamins A, D, E and K and trace metals should be measured at regular intervals and replaced as appropriate.
- TPN at home is usually cycled over a 10-12-hour overnight period with 30-60 minutes for tapering off. Patients can thus move around and continue to work and go on holiday. Patients need to be extensively educated about how to recognise infection and minimise the risk.
- Hypersecretion - usually noted within the first twelve months of resection - should be treated with an H<sub>2</sub>-antagonist or proton pump inhibitor.
- The introduction of enteral feeding as soon as possible encourages bowel adaptation and may reduce the amount of time that parenteral feeding is required.<sup>[6]</sup> Small amounts of oral food are introduced gradually.
- Patients who have lost large amounts of ileum or colon may have significant fluid loss and may require prolonged intravenous fluid therapy.
- Pharmacological bowel compensation has been explored as a way of increasing bowel adaptation and improving the chances of patients being released from dependency on parenteral nutrition. Such approaches include the use of **somatropin** (recombinant human **growth hormone**) and a high-carbohydrate diet. In recalcitrant cases, the somatostatin analogue **octreotide** in doses of 100 micrograms subcutaneously three times a day can significantly reduce diarrhoea.

## Surgical

The treatment of short bowel syndrome (SBS) has to be individualised according to the clinical status of individual patients and the decision on whether to resort to surgery and what operation to perform, depends upon many factors. In general, surgery is indicated as a last resort when all other therapeutic options, including parenteral and enteral feeding and pharmacological bowel compensation, have been tried. In some patients, operation may be required because of complications of prolonged parenteral nutrition or stasis of enteric content and bacterial overgrowth.

Surgery can be divided broadly into non-transplant surgery, transplant surgery without organ transplantation and combined intestinal and organ transplantation.

- **Non-transplant surgery:** a variety of procedures have been tried to a greater or lesser degree of success. These include connecting residual small bowel to the colon to maintain intestinal integrity, intestinal lengthening and tailoring (the Bianchi procedure), tapering for dilated segments, strictureplasty, creation of intestinal valves or reversed bowel segments to reduce rapid transit time. It is likely that as these procedures improve, this type of surgery will become less of a 'last resort' and more of an option for those patients faced with long-term parenteral feeding.<sup>[7]</sup><sup>[8]</sup> One study of 53 patients undergoing the Bianchi procedure reported a high survival rate, weight gain and a high quality of life.<sup>[9]</sup> Bowel lengthening is technically feasible in over half of adult patients with SBS.

- **Transplant surgery:**

### Indications for transplant surgery

As considered by the two UK specialist centres (Addenbrooke's Hospital in Cambridge and St James' Hospital in Leeds).<sup>[2]</sup>

- Complications of parenteral nutrition.
  - Liver disease (portal hypertension, bridging hepatic fibrosis, or cirrhosis) due to parenteral nutrition.
  - irreversible, despite referral and management by an established parenteral nutrition centre.
  - Progressively compromised vascular access for parenteral feeding loss of all but two major venous access points (one of which should be above the diaphragm).
  - Recurrent or life-threatening central line sepsis (including fungal sepsis).
  - Inadequate maintenance on parenteral nutrition for any other reason - for instance, inability to manage hydration/nutrition status despite parenteral nutrition.
  - High-risk conditions.
  - Requirement for extensive evisceration (that is, desmoid tumours, trauma, rare selected malignancies, including neuro-endocrine tumours).
- **Intestinal transplantation:** this is currently considered inferior to TPN in most cases but long-term TPN is not without its problems. Advancement of surgical techniques, new immunosuppressive techniques, improvement of postsurgical management, adequate timing of transplantation and refined selection of candidates may all help to make intestinal transplantation a viable management rather than a last resort.<sup>[10]</sup> Such considerations are particularly relevant in children, in whom freedom from a life of TPN is particularly important.<sup>[11]</sup>
  - **Isolated intestinal transplantation:** this is mainly indicated for patients who have reasonable liver function or significant liver disease that has not progressed to cirrhosis. It may also be appropriate for patients who have significant fluid losses and who have episodes of frequent, severe dehydration despite appropriate medical management.
  - **Combined intestinal and organ transplantation:** this is indicated for patients with SBS and end-stage liver failure. Renal transplantation may also be combined in appropriate patients.

## Complications<sup>[1]</sup> <sup>[2]</sup>

- Infection related to the indwelling venous feeding catheter.
- Thrombosis precluding adequate access for feeding.
- Complications of any underlying condition - eg liver disease.

General complications of surgery include haemorrhage, wound sepsis, postoperative pulmonary dysfunction, renal failure and pulmonary embolism.

Complications of non-transplant surgery include:

- Bowel obstruction.
- Bowel necrosis.
- Bowel dysmotility and dysfunction.
- Anastomotic disruption.
- Stasis of intestinal contents with or without bacterial overgrowth.

Patients who undergo transplant surgery are at risk of all of the above but, in addition, may also develop:

- Acute rejection.
- Chronic rejection.
- Hepatic, portal, or mesenteric vein thrombosis.
- Systemic sepsis with ordinary pathogens or opportunistic organisms (eg, cytomegalovirus).
- Lymphoproliferative disorders or malignancies.

## Prognosis<sup>[1]</sup>

- The most common cause of death of patients on TPN is liver failure, consequent upon chronic hepatic parenchymal damage.
- The 4-year survival rate of patients dependent on parenteral nutrition is about 70%. Many patients die from severe septic complications or run out of venous access. Pharmacological bowel compensation is allowing increasing numbers of patients to avoid long-term parenteral nutrition.
- Studies of non-transplant surgical procedures suggest that clinical improvement occurs in 80% of patients.
- Early postoperative complications can result in a high mortality rate (30% in some studies). However, of patients who survive this period, 80-90% are still alive after one year and 60% after four years.
- One study of 74 patients found that, at the end of one year, 74.3% were still on TPN. 11.8% were switched to enteral nutritional support.

## Further reading & references

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