Intestinal failure and short bowel syndrome

Victoria Mulcahy

Alastair Forbes

Abstract

Intestinal failure is usefully defined as a reduction of intestinal function such that ordinary fluids and nutrients given by mouth are insufficient, and that at least partial artificial support by the parenteral route is needed to maintain health. Intestinal failure usually follows major resection (e.g. in short bowel syndrome [SBS]), but also occurs when the intact intestine is unable to function owing to inflammation or disorders of motility. In many patients, both causes co-exist. Chronic intestinal failure is relatively rare, with a prevalence of 2–4 per 100,000 and an incidence of 2–4 per 1,000,000. Adaptation occurs in the first months after injury and comprises hyperplasia and hypertrophy; this is potentially responsive to trophic factors. In SBS, intestinal volume losses may exceed 5 L per day. The biggest challenge in SBS management is preventing consumption of spontaneous excess fluid orally, as this is generally sodium-free and drives intestinal secretion. Initial therapeutic steps include intravenous sodium, reduced oral intake of hypotonic fluid, increased sodium intake and high-energy foods. Formula feeds should be polymeric with additional sodium and magnesium. Useful drugs include loperamide, codeine and proton pump inhibitors. Somatostatin agents are not especially helpful, but glucagon-related peptides may become important. Parenteral nutrition should be avoided, if possible, but when essential, follows a routine format. The risk–benefit equation is beginning to favour intestinal transplantation in selected patients, and complete failure of intravenous nutrition should no longer be awaited.

Keywords Crohn's disease; glucagon; intestinal pseudo-obstruction; intestinal transplant; parenteral nutrition; short bowel syndrome

What's new?

- The increasing recognition that nutritional support may be necessary in the absence of an anatomical short bowel is steadily enlarging the number of patients recognized to have intestinal failure
- Patients with advanced malignancy may have a reasonable prognosis with full support, and the presence of active disease is now less likely to be considered a contra-indication to long-term artificial nutrition
- Intestinal transplantation is more widely available, and for some diagnoses the mortality is now no greater than that from long-term home parenteral nutrition

Intestinal failure has been defined as the presence of inadequate functional intestine to maintain health by ordinary food and drink intake (usually extended to include nutrients given enterally). It is similar, but not identical, to short bowel syndrome (SBS), which the definition includes.[1] Intestinal failure usually follows major small bowel resection, but also occurs when intact intestine is unable to function because of severe inflammation or motility disorders. In many patients, both causes are present. The lesser disability of intestinal insufficiency is now proposed to encompass those with milder functional deficits in whom control can be achieved without recourse to parenteral support. The term intestinal failure thus becomes restricted to those who require at least some support via the parenteral route. Established intestinal failure is rare; the prevalence is between 2 and 4 per 100,000 population, and the annual incidence is about one-tenth of this. Crohn's disease used to account for one-third of all long-term cases within the UK, but within recent years this has changed, and now motility problems and ischaemia are the two leading causes of intestinal failure (Figure 1). Intestinal failure is best managed when anticipated (for example, in any patient with an ileostomy and <200 cm of small bowel, with <150 cm of small bowel if it is anastomosed to the colon, or when stoma or fistula output is >1.5 L/day).

Fluid balance and the intestine

Approximate normal intestinal fluid volumes are shown in Table 1. It is clearly an over-simplification to state that all intake and secretion reaches the mid-point of the small bowel and that all fluid absorption is more distal, but net fluid shifts largely follow this paradigm. In most situations, gastrointestinal output is directly proportional to jejunal length; positive fluid balance requires about 100 cm.

The colon is inessential to health and has little direct nutritional impact in normal individuals, but in SBS it becomes crucial for fluid balance, and in this respect it may be equivalent to up to 50 cm of small intestine. The fermentation of non-absorbed carbohydrates by colonic bacteria may also yield additional usable nutrients (as much as 500 kcal/day).

The effluent from a proximal stoma or fistula may exceed 5 L/day. Patients readily develop negative fluid balance, which provokes thirst, and this aggravates the problem because most of these individuals are 'net secretors'. Because of the virtual absence of sodium from ordinary drinks, the inevitable response to thirst and drinking is an increase in output greater than the additional input. This is caused by sodium-linked movement of water across the intestine into the lumen. Recognition of this paradox is vital to the understanding of SBS, and education (of staff and patients) is the most important aspect of management.

Victoria Mulcahy *BSc MBBS MRCP is Specialist Registrar at Norfolk and Norwich University Hospital, UK where she is completing her training in gastroenterology and nutrition. Competing interests: none declared.*

Alastair Forbes *BSc MD FRCP FRCPE FHEA is Professor of Medicine (Gastroenterology and Nutrition) at Norwich Medical School, UK. He also has a background in gastroenterology and nutrition and has specialised in intestinal failure over the past 20 years or more. Competing interests: Professor Forbes has received trial sponsorship from NPS Pharma, and honoraria for speaker-meetings from Baxter, B Braun, Fresenius-Kabi and Nutricia.*

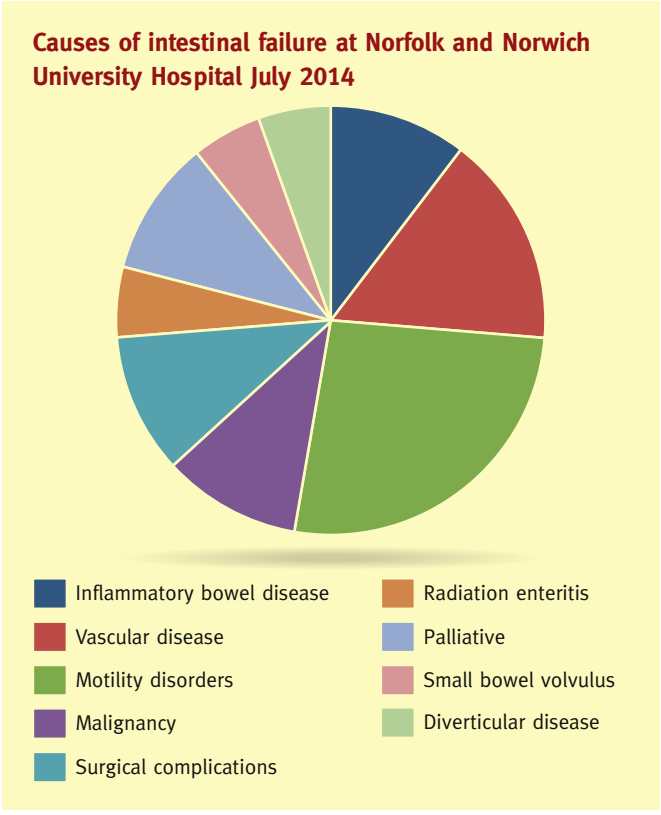

Figure 1

Investigations

In possible intestinal failure, many clinicians rely too heavily on serum electrolytes, plasma osmolality, serum urea or creatinine, and full blood count. These are abnormal in advanced fluid depletion and malnutrition, but are not sensitive. Earlier indications of disturbed fluid balance come from regular estimation of body weight and random urinary sodium. In SBS, urinary sodium is reduced because of the kidney's avidity for sodium so as to maintain an effective circulating volume. Concentrations below 20 mmol/L indicate impending hypovolaemic renal failure and a need for action. Apparently reassuring, but misleading, concentrations over 20 mmol/L occur only in established renal failure and in patients taking diuretics (almost always contraindicated in SBS). Low serum magnesium is common in SBS and is the usual cause of tetany in these patients. A low serum calcium in SBS is generally the result of an uncorrected low magnesium.

Normal gastrointestinal volumes

Food and drink	1500 ml
Saliva	750 ml
Gastric secretions	1250 ml
Biliary secretions	1000 ml
Pancreatic secretions	1000 ml
Jejunal secretions	2500 ml
Total	**8000 ml**
Stool liquid	100–150 ml

Table 1

Management

The principal aims of management are to:

- minimize sodium-poor fluid intake
- reduce gut secretion
- reduce the speed of intestinal transit
- overcome nutritional deficiencies.

It is useful to consider patients with intestinal failure on two parallel algorithms (Figure 2). Most require fluids, electrolytes, and nutrients, but some need daily intravenous fluid, sodium and magnesium with no nutrients, while others (mostly those with motility disorders and without major intestinal losses) may need intravenous nutrients without additional fluids, and can be managed with a parenteral nutrition (PN) infusion three or four times per week.

In the acute situation, it may be necessary to resuscitate the patient with a sodium-based intravenous preparation, but in most cases this is a temporary intervention. Electrolyte solutions such as Hartmann's or Ringer's are preferred to sodium chloride 0.9% to limit the administration of chloride ions, which are less required (other than in uncontrolled vomiting) and difficult for the sick patient to excrete.

The next step is reduction of oral intake of hypotonic fluid, typically to no more than 500 ml of 'free' fluids every 24 hours.

Oral sodium intake should be increased by the addition of extra salt to foods, and/or the use of balanced oral electrolyte solutions; appropriate dietary measures should be instituted. Rigorous attention to this regimen can be expected to reduce gastrointestinal efflux by up to 50%, or by around 2 L. In patients with very high outputs, this is the single most valuable intervention.

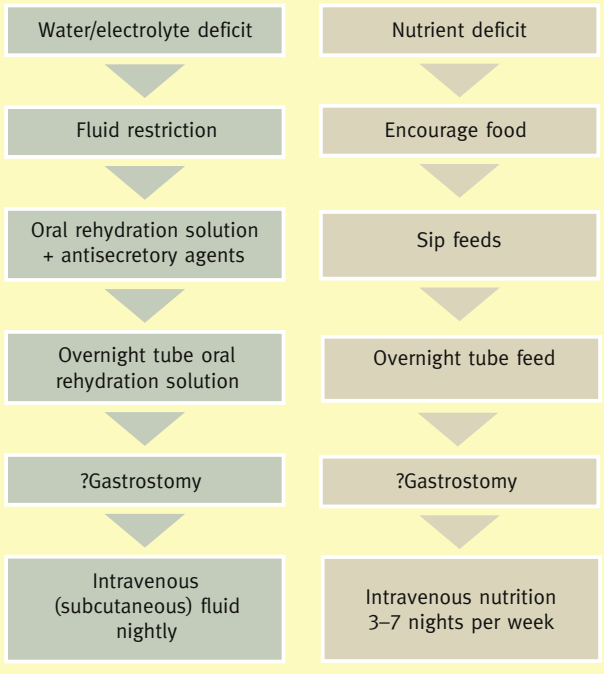

Figure 2