Guidelines for management of patients with a short bowel

J Nightingale, J M Woodward on behalf of the Small Bowel and Nutrition Committee of the British Society of Gastroenterology

1.0 FORMULATION OF GUIDELINES

1.1 Aim
These guidelines aim to help clinicians manage patients who have had an intestinal resection that leaves a short length (about 2 m or less) of small bowel remaining.

1.2 Development
The preliminary guidelines were compiled from the literature and a first document was drafted by Dr J Nightingale and modified by members of the Small Bowel and Nutrition Committee under the chairmanship of Dr B Jones. A section on “intestinal transplantation” was written by Dr Woodward and added with the approval of the Small Bowel and Nutrition Committee. The resulting document was shown to clinicians at the intestinal units of Hope and St Mark’s Hospitals. Professor A Forbes made recommendations, which have been incorporated. The article was reviewed by the patient organisation PINNT (patients on intravenous or nasogastric nutritional therapy) and modifications made to result in the current document.

The guidelines conform to the North of England evidence based guidelines development project. The grading of each recommendation is dependant on the category of evidence supporting it. Recommendations based on the level of evidence are presented and graded as:

- A: requires at least one randomised controlled trial of good quality addressing the topic of recommendation (evidence categories Ia and Ib);
- B: requires the availability of clinical studies without randomisation on the topic of recommendation (evidence categories IIa, IIb and III); and
- C: requires evidence from expert committee reports or opinions or clinical experience of respected authorities in the absence of directly applicable clinical studies of good quality (evidence category IV).

1.3 Scheduled review
The content and evidence base for these guidelines should be reviewed within five years of publication. We recommend that these guidelines are audited and request feedback from all users.

1.4 Service delivery
Patients with a short bowel are not common but should be managed by a multidisciplinary team headed by a clinician with expertise in managing these patients. If managed appropriately, there may be an improved quality of safe care and also considerable cost savings. On occasions, patients thought to need long term parenteral nutrition may be weaned from it with appropriate advice.

As patients (particularly with a short bowel and jejunostomy) may rapidly become dehydrated or septic (if having parenteral nutrition), they and indeed any patient needing artificial nutritional support should have rapid access to medical expertise (advice, clinics, or inpatient treatment).

1.5 Patients’ experience
- Patients with a short bowel should each be managed as an individual; they are all different in diagnosis, remaining bowel length/function, and psychosocial characteristics.
- Patients will become experts in coping with their condition and management. All decisions should all be made in conjunction with them. They are often more knowledgeable about their condition than the clinicians, nurses, and dieticians, and this should be respected.
- Facilities for looking after these patients should be able to deal with the physical, emotional, psychological, social, and quality of life issues.
- Techniques needed for home parenteral nutrition should be taught by competent, patient, and keen staff who can convey the confidence required to undertake the therapy successfully and safely.
- Patients need to know that the aseptic technique for parenteral nutrition will be used whenever their feeding line is accessed, which is vital for safety and peace of mind.
- Patients should be referred rapidly to places of expertise if management is difficult or unsuccessful. There should be clinicians, specialist nurses, and dieticians available to discuss or see the patients at all times. Healthcare professionals should be familiar to the patient and know their history, thus eliminating the need for time consuming explanations.
- There should ideally be dedicated beds for nutrition patients to ensure they are not cared for by healthcare professionals unfamiliar with their specialist needs.
- A 24 hour helpline should be in place so that emergencies are dealt with immediately and appropriately.

Abbreviations: PINNT, patients on intravenous or nasogastric nutritional therapy; IF, intestinal failure; GLP-2, glucagon-like peptide 2
Bowel length measurements are from the duodenojejunal flexure and can be made at surgery or with the use of an opsiometer\(^2\) tracing out the long axis of the bowel on a contrast study that shows all of the remaining small bowel.

2.5 Patients with irreversible intestinal failure expected to die prematurely on parenteral nutrition should be referred for consideration of intestinal transplantation where appropriate

3.0 INTRODUCTION TO INTESTINAL FAILURE AND SHORT BOWEL

Intestinal failure (IF) occurs when there is reduced intestinal absorption so that macronutrient and/or water and electrolyte supplements are needed to maintain health and/or growth.\(^7\) If untreated, undernutrition and/or dehydration result. IF may be defined and quantified by balance study techniques; however, only few centres have the facilities for these difficult metabolic studies, and therefore nutrient/fluid requirements determine whether IF is termed severe, moderate, or mild (fig 1). Severe is when parenteral, moderate when enteral, and mild when oral nutritional fluid supplements are needed.\(^*\) A patient may change, due to compensatory mechanisms (for example, functional adaptation), from having severe to mild IF with time. IF can be acute or chronic (fig 2) and encompasses a wide variety of underlying medical and surgical conditions. Chronic IF most commonly results from a bowel resection that leaves a short residual length of small bowel.

Severe IF is a complex clinical and physiological situation that requires an experienced multidisciplinary input. These patients may be most appropriately managed in a centre with special interest and expertise in IF.

These guidelines aim to help practicing clinicians manage patients with IF due to a short bowel. Complications of the short bowel are covered but specific complications relating to therapy (for example, catheter related sepsis, hepatic or bone complications of parenteral nutrition) are not included. While bowel length is often unknown, patients who have had several ileal resections and have diarrhoea and weight loss may be treated as jejunum-colon patients, and patients with ileostomy diarrhoea, dehydration, hypomagnesaemia, and undernutrition may be treated as jejunostomy patients.

4.0 SHORT BOWEL

Normal human small intestinal length, measured from the duodenojejunal flexure at autopsy or surgery, varies from about 275 cm to 850 cm, and tends to be shorter in women. After intestinal resection it is important to refer to the remaining length of small intestine measured at surgery or with an opsiometer\(^2\) rather than to the amount removed. In general, nutritional/fluid supplements are likely to be needed if less than 200 cm of small bowel remains.

2.1 Aims of treatment in patients with a short bowel

- To provide the nutrition, water, and electrolytes necessary to maintain health, with normal body weight or growth.
- To use oral/enteral nutrition in preference to parenteral nutrition.
- To reduce the complications resulting from the underlying disease, intestinal failure, and/or nutritional/fluid support.
- To achieve a good quality of life.

2.2 Patients with a short bowel and an intact ileum and colon rarely need long term enteral or parenteral nutrition.

2.3 Patients with a short bowel (due to loss of ileum) and a retained functional colon

Gradual undernutrition dominates the clinical picture. Due to adaptation, nutritional requirements may reduce with time.

- May need parenteral nutrition if less than 50 cm small intestine remains (grade B).
- Need a high carbohydrate low oxalate diet. The volume of food may increase diarrhoea (grade A).

2.4 Patients with a jejunostomy

Fluid and electrolyte losses dominate the clinical picture. Adaptation does not occur so nutritional and fluid requirements do not reduce with time.

- If less than 100 cm of jejunum remains, parenteral saline, and if less than 75 cm, parenteral nutrition and saline are likely to be needed in the long term (grade B).
- If less than 200 cm of jejunum remains, oral hypotonic fluids may need to be restricted and a glucose-saline supplement (sodium concentration of about 100 mmol/l approximating to the concentration in jejunostomy fluid) is sipped to reduce stomal losses of sodium (grade B).
- Hypomagnesaemia is common and is treated by correcting sodium depletion, oral or intravenous magnesium supplements, and occasionally with oral 1 alpha hydroxycholecalciferol (grade C).
- Jejunal output may be further reduced by drugs that reduce motility (loperamide) or, if the bowel is very short (less than 100 cm), drugs that reduce gastric acid secretion (H\(_2\) antagonists, proton pump inhibitors, or somatostatin analogues) (grade B).

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Severe IF is a complex clinical and physiological situation that requires an experienced multidisciplinary input. These patients may be most appropriately managed in a centre with special interest and expertise in IF.

These guidelines aim to help practicing clinicians manage patients with IF due to a short bowel. Complications of the short bowel are covered but specific complications relating to therapy (for example, catheter related sepsis, hepatic or bone complications of parenteral nutrition) are not included. While bowel length is often unknown, patients who have had several ileal resections and have diarrhoea and weight loss may be treated as jejunum-colon patients, and patients with ileostomy diarrhoea, dehydration, hypomagnesaemia, and undernutrition may be treated as jejunostomy patients.
There are three main types of patient with a short bowel: those who have had a jejunoileal resection and a jejunocolic anastomosis (jejunum-colon); those who have had a predominantly jejunal resection, and have more than 10 cm of terminal ileum and the colon remaining (jejunum-ileum); and those who have had a jejunoileal resection, colectomy, and formation of a stoma (jejunostomy). Jejunum-ileum patients are uncommon and rarely have problems of undernutrition and therefore do not often need nutritional support. When jejunoileum patients are seen, with undernutrition or severe diarrhoea after eating/drinking, they are managed in the same way as jejunum-colon patients. Jejunum-colon and jejunostomy patients are most commonly encountered. 7–9 The most common reasons for a short bowel in adults are Crohn’s disease, superior mesenteric artery thrombosis, and irradiation damage (table 1). 7–9 A short bowel more commonly arises in women (67%) than men, 7 possibly because women start with a shorter length of small intestine than men.

Jejunum-colon patients often appear well after their resection except for diarrhoea/steatorrhoea, but in the following months may lose weight and become severely undernourished. Patients with a jejunostomy have problems of dehydration immediately after surgery due to large stomal water and sodium losses. This jejunal output is greatest after infection, 100 mmol/l is reached. 14 Jejunal mucosa is ‘leaky’ and rapid sodium efflux of sodium from the plasma into the bowel lumen, 15 concentration of less than 90 mmol/l is drunk there is a net efflux of sodium from the plasma into the bowel lumen, 15 until a luminal sodium concentration of approximately 100 mmol/l is reached.

Gastric acid hypersecretion may occur in the first two weeks after a small bowel resection 26 but there is no evidence that it occurs in the long term in humans.

4.1 Physiological consequences
The problems after major intestinal resection are due to both normal and altered physiology.

4.1.1 Gastrointestinal motility
Gastric emptying and small bowel transit for liquid is normal in jejunum-colon patients but fast in patients with a jejunostomy as the ileal and colonic braking mechanisms in jejunum-colon patients but fast in patients with a jejunostomy the patient may lose more fluid than is taken by mouth. 14 Jejunal mucosa is ‘leaky’ and rapid sodium fluxes occur across it. If water or any solution with a sodium concentration of less than 90 mmol/l is drunk there is a net efflux of sodium from the plasma into the bowel lumen, 15 until a luminal sodium concentration of approximately 100 mmol/l is reached.

4.1.2 Gastrointestinal secretions
Daily gastrointestinal secretions are made up of 0.5 litre of saliva, 2.0 litres of gastric juice, and 1.5 litres of pancreaticobiliary secretions plus passive jejunal secretions to render the lumen isotonic during passage and digestion of ingested nutrients. The majority of the fluid is reabsorbed in the upper jejunum. Jejunum-colon patients can reabsorb unabsorbed fluid in their colon but this is not the case for jejunostomy patients who lose much salt and water from their stoma. If less than 100 cm of jejunum remains proximal to a jejunostomy the patient may lose more fluid than is taken by mouth. 14 Jejunal mucosa is ‘leaky’ and rapid sodium fluxes occur across it. If water or any solution with a sodium concentration of less than 90 mmol/l is drunk there is a net efflux of sodium from the plasma into the bowel lumen, 15 until a luminal sodium concentration of approximately 100 mmol/l is reached.

4.1.3 Absorptive functions
Vitamin B12 17, 18 and fat malabsorption 19 occurs when more than 60–100 cm of terminal ileum have been resected. Increased hepatic synthesis of bile salts cannot compensate for the loss of ileal surface area. Unabsorbed bile salts may contribute to colonic secretion in patients with a remaining colon.

Magnesium deficiency occurs due to reduced absorption because of chelation with unabsorbed fatty acids in the bowel lumen 20 and to increased renal excretion (consequent on secondary hyperaldosteronism). 21, 22 Hypomagnesaemia reduces the secretion and function of parathormone, 23 so directly increasing renal magnesium loss and indirectly by reducing the manufacture of 1,25 hydroxy-vitamin D which normally increases jejunal magnesium absorption. 24

4.1.4 Adapative processes
Patients with a short bowel do eat more food than normal (hyperphagia). 25 Intestinal adaptation is the process that attempts to restore the total gut absorption of macronutrients, macrominerals, and water to that prior to intestinal resection. 26 This occurs partly by increasing the absorptive area of the remaining bowel (structural adaptation) and/or by slowing gastrointestinal transit (functional adaptation).

**Table 1** Common causes of a short bowel

<table>
<thead>
<tr>
<th>Jejunum-colon</th>
<th>Jejunostomy</th>
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<tbody>
<tr>
<td>Crohn’s disease</td>
<td>Crohn’s disease</td>
</tr>
<tr>
<td>Mesenteric ischaemia</td>
<td>Ulcerative colitis</td>
</tr>
<tr>
<td>Irradiation</td>
<td>Irradiation</td>
</tr>
<tr>
<td>Small bowel volvulus</td>
<td>Mesenteric ischaemia</td>
</tr>
<tr>
<td>Adhesions</td>
<td>Desmoid</td>
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</table>

Figure 2 Reasons for intestinal failure.

www.gutjnl.com
probably due to high circulating peptide YY and GLP-2 levels. 11 12 There is increased jejunal absorption of macronutrients (for example, glucose), water, sodium, and calcium with time, 29–32 and an increased chance of the patient being able to stop parenteral nutrition. 7 8 31 Thus these patients may show a gradual reduction in nutritional requirements with time.

Jejunostomy

Although intestinal adaptation occurs in the months after creation of an ileostomy, there is no evidence for any structural 32 or functional 33 adaptation at any time in patients with a jejunostomy. Thus the nutritional and fluid needs of a patient with a jejunostomy are unlikely to change with time.

4.2 Clinical assessment

Clinical assessment of a patient with a short bowel includes assessment of water, sodium, magnesium, and nutritional status. Knowledge of residual small bowel length allows predictions to be made about the long term fluid/nutritional support that may be needed (table 2). The state of hydration and magnesium balance is of immediate importance to those with a jejunostomy.

Water and sodium deficiency (most common in jejunostomy patients) may result in thirst, hypotension, and pre-renal failure. Daily body weight and accurate fluid balance (including stomal output) are the most important measurements. Serum creatinine, potassium, and magnesium, and urinary sodium may be measured every 1–2 days initially, then once or twice a week and, if long term, at home every 2–3 months. Water and sodium deficiencies are detected by an abrupt fall in body weight, postural systolic hypotension, low urine volume and, if severe, by a rising serum creatinine and urea. The most helpful early measure of sodium depletion is the sodium concentration in a random urine sample. A concentration of less than 10 mmol/l suggests sodium depletion.

Magnesium depletion is common, especially in patients with a high stomal output. A serum value of less than 0.6 mmol/l may give rise to symptoms.

Current nutritional status may be assessed by recording the following (significantly low): body mass index (<18.5 kg/m²), percentage weight loss (<10%), or mid-arm muscle circumference (<19 cm in a woman, <22 cm in a man). 36 37

5.0 PROBLEMS AND TREATMENT OF JEJUNUM-COLON PATIENTS

Immediately following surgical resection that results in a short bowel, nutritional and fluid support are given, with parenteral nutrition being used early, even when enough bowel is thought to remain. This helps surgical repair, ileus recovery, and prevents vitamin/mineral deficiencies. Initially, a proton pump inhibitor is started and continued for six months as gastric acid hypersecretion may occur. 34

In jejunum-colon patients the problems of undernutrition, diarrhoea due to malabsorption, and vitamin/mineral deficiencies dominate the clinical picture. These are the classical features initially described as constituting the “short bowel syndrome”.

5.1 Undernutrition: protein-energy malnutrition

Undernutrition only becomes apparent slowly and should be prevented from occurring by predicting its likelihood and from knowledge of the residual length of bowel remaining (table 2). All patients who can be maintained on an oral diet need to consume more energy than normal subjects because 50% or more of the energy from the diet may be malabsorbed. Patients can achieve this by eating more high energy food, having oral sip-feeds, or high energy enteral feeds given at night through a nasogastric or gastrostomy tube. There are rarely any problems inserting a percutaneous endoscopic gastrostomy (PEG) into patients with Crohn’s disease providing that there is no distal obstruction. 35 Once weight is regained, a nocturnal feed can be reduced or stopped and sip-feeds during the day may be adequate. Only if these measures fail and the patient continues to lose weight, or fails to regain lost weight, is parenteral nutrition given. Parenteral nutrition may only be needed for a few weeks or months before oral supplements are adequate.

In the long term, parenteral nutrition is needed if a patient absorbs less than one third of the oral energy intake, 36 37 if there are high energy requirements and absorption is 30–60%, or if increasing oral/enteral nutrient intake causes a socially unacceptably amount of diarrhoea or a large volume of stomal output.

In jejunum-colon patients, unabsorbed long chain fatty acids in the colon are likely to reduce transit time 44 and reduce water and sodium absorption 45 so making diarrhoea worse. In addition, they are toxic to bacteria and so reduce carbohydrate fermentation. 46 They bind to calcium and magnesium, increasing stool losses, and they increase oxalate absorption so predisposing to the formation of renal stones (see below).

Theoretically, a low fat diet is ideal for patients after a small bowel resection 46 but in practice it is hard to implement. Fat yields twice as much energy as comparable weights of carbohydrate and makes food more palatable. A high carbohydrate/low fat diet involves eating a large volume of food. A low fat diet may increase calcium, magnesium, and zinc absorption 47 48 but makes essential fatty acid deficiency more likely. 49 Sunflower oil may be rubbed into the skin to ensure adequate amounts of essential fatty acids enter the body. 50 If a diet is high in monosaccharides D (−) lactic acidosis may occur (see 5.5). Medium chain triglycerides are an alternative source of energy and are absorbed from the small and large bowel. 50 51

In order to increase energy absorption and reduce the risk of renal stones and D-lactic acidosis, patients with a retained colon need a large total energy intake with a diet high in carbohydrates (polysaccharides), normal (not restricted) in fat (long chain triglycerides), and low in oxalate.

5.1.1 Conjugated bile acid treatment

Cholylarsarcosine has been used as bile acid replacement therapy but with variable improvement in fat and calcium absorption. 52 53 There is little evidence for its use.

5.2 Salt, water, and magnesium depletion

The colon has a large capacity to absorb sodium and water; thus patients with a short bowel and a preserved colon are rarely in negative water and sodium balance and rarely need water or sodium supplements. 7 54 55 Although the colon
secretes potassium, a low serum potassium level rarely occurs.7 If sodium depleted, a glucose-saline drink can be sipped during the day, as for patients with a jejunostomy. A low serum magnesium is less common than in patients with a jejunostomy but is treated in the same way.

5.3 Vitamin and mineral deficiencies
Most patients require long term B12 treatment.17 18 Selenium deficiency is common and patients may need larger amounts than normal subjects.19–22 Zinc deficiency is uncommon unless stool volumes are large.23 Vitamins A, D, E, and K, and essential fatty acids may need to be replaced.

5.4 Diarrhoea
Oral intake determines the volume of stool passed, so limiting food intake will reduce diarrhoea but will exacerbate the problems of undernutrition. A patient may require parenteral nutrition to allow them to eat less and so reduce their diarrhoea.

Diarrhoea is treated in the same way as for patients with a jejunostomy, with loperamide 2–8 mg given half an hour before food and occasionally codeine phosphate is also added (30–60 mg half an hour before food).

If 100 cm or more of terminal ileum have been resected, bile salt malabsorption may contribute to diarrhoea and occasionally this is helped by cholestyramine. Cholestyramine has the additional advantage of reducing oxalate absorption, but by further reducing the bile salt pool will increase fat malabsorption.24 Although gastric antisecretory drugs may reduce diarrhoea shortly after surgery, they may not be effective in the long term.

5.5 Confusion
In addition to the many common general medical causes of confusion (for example, hypoxia, hepatic, renal or cardiac failure, sepsis, hypoglycaemia, alcohol, or other drugs), other specific causes should be sought in a patient with a short bowel. Hypomagnesaemia may cause mild confusion when the serum magnesium level is very low (less than 0.2 mmol/l). Thiamine deficiency can cause a Wernicke/Korsakoff psychosis which responds rapidly to large regular doses of thiamine. Two other specific causes of confusion in patients with a short bowel are D(−) lactic acidosis and hyperammonaemia.

D(−) lactic acidosis only occurs in patients with a short bowel and a preserved colon. Colonic bacteria may degrade a surplus of fermentable carbohydrate to form D(−) lactic acid which is absorbed but not easily metabolised.25 In addition to a metabolic acidosis with a large anion gap, increased concentrations of D(−) lactate are found in blood and urine. Treatment involves restricting mono and oligosaccharides and encouraging the more slowly digestible polysaccharides (starch), thiamine supplements, and broad spectrum antibiotics. In rare cases the patient may need to fast while receiving parenteral nutrition.

Confusion may be due to hyperammonaemia (also occurs in jejunostomy patients); this results because ammonia cannot be detoxified. The small amount of intestine remaining cannot manufacture adequate citrulline to detoxify ammonia via the urea cycle. The increase in blood ammonia is a problem if there is concomitant renal impairment, as the excess ammonia cannot be excreted. By giving arginine (an intermediary in the urea cycle), hyperammonaemia can be corrected.26–28

5.6 Drug absorption
Omeprazole can be absorbed in the duodenum/upper jejunum and only if less than 50 cm of jejunum remains are problems likely to occur. Many drugs will be incompletely absorbed by patients with a short bowel and may be needed in much higher amounts than usual (for example, thyroxine, warfarin,29 and digoxin but may need to be given intravenously. If a patient is taking warfarin, blood for the international normalised ratio should be taken at the same time of day relative to a lipid infusion. As the enterohepatic circulation, around which loperamide circulates, is disrupted, higher doses than usual may need to be given.

5.7 Gall stones
Gall stones are common (45%) (same prevalence in jejunostomy patients) and are more common in men.7 Such stones probably result from gall bladder stasis in which biliary sludge develops and subsequently forms calcium bilirubinate stones.

Therapies to prevent the formation of biliary sludge and hence gall stones include periodic intravenous infusions of amino acids or enteral feed, cholecystokinin injections, non-steroidal anti-inflammatory drugs, ursodeoxycholic acid, and reducing the formation of the more lithogenic secondary bile acids by either increasing bowel transit or by inhibiting bowel bacteria (for example, metronidazole). Some units advocate prophylactic cholecystectomy whenever a large resection of the small intestine is performed.

5.8 Renal stones
Jejunum-colon patients have a 25% chance of developing symptomatic calcium oxalate renal stones. Jejunum-colon patients may have diarrhoea, which is malodorous and bulky due to steatorrhoea.

6.0 PROBLEMS AND TREATMENT OF JEJUNOSTOMY PATIENTS
The major differences between jejunum-colon and jejunostomy patients are that jejunostomy patients lose much salt and water from their stoma, commonly have hypomagnesaemia but they do not have significant bacterial fermentation occurring in the bowel lumen. Many other problems are similar (e.g. Bl deficiency, confusion, drug absorption and gall stones).

6.1 Salt and water depletion
Patients with a jejunostomy have a large volume of stoma output, which is greater after eating or drinking. Each litre of jejunostomy fluid contains about 100 mmol/l of sodium.30 The effluent from a jejunostomy or ileostomy contains relatively little potassium (approximately 15 mmol/l). Potassium balance is not often a problem and net loss
through the stoma occurs only when less than 50 cm of jejunum remains. A low serum potassium level is most commonly due to sodium depletion with secondary hyperaldosteronism and thus greater than normal urinary losses of potassium. Hypokalaemia can also be due to hypomagnesaemia, which causes dysfunction of many of the potassium transport systems and increases renal potassium excretion; this hypokalaemia is resistant to potassium treatment but responds to magnesium replacement.55

6.1.1 Management of a high output jejunostomy/ileostomy (table 3)

It is helpful both in predicting outcome and in choosing the type of nutritional support if the remaining length of bowel is known. Contrast studies (small bowel meal or enema) may help. Examination of the output (colour/consistency/24 h volume) may also give an indication of the type of stoma. The principles described here are the same for a high output ileostomy.

Exclude other causes of a high output

A stoma may produce a high output if there is intra-abdominal sepsis, partial/intermittent bowel obstruction, enteritis (for example, clostridium or salmonella), recurrent disease in the remaining bowel (for example, Crohn’s disease or irradiation), suddenly stopping drugs (for example, steroids or opiates), or giving prokinetics (for example, metoclopramide).

Treatment of a high output jejunostomy

If there is marked sodium and water depletion and severe thirst, it is often necessary to establish equilibrium by giving intravenous normal saline (2–4 l/day), keeping the patient “nil by mouth”, which will also demonstrate that output is driven by their oral intake. Over 2–3 days intravenous saline is gradually withdrawn while food and restricted oral fluids are reintroduced. Great care must be taken not to administer too much fluid, which will readily cause oedema (partly due to high circulating aldosterone levels). The aims of treatment are to maintain hydration/body weight and a daily urine volume of at least 800 ml with a sodium concentration greater than 20 mmol/l.

To correct hypokalaemia in patients with a high output stoma, sodium/water depletion must be corrected, and serum magnesium brought into the normal range. It is uncommon for potassium supplements to be needed.

Restrict oral fluids. It is a common mistake for patients to be encouraged to drink oral hypotonic solutions to quench their thirst but this causes large stomal sodium losses. Hypertonic fluids, which may contain sorbitol or glucose, can also cause stomal losses of water and sodium. Treatment for high output from a jejunostomy (ileostomy or high fistula) begins with the patient restricting the total amount of oral hypertonic fluid (water, tea, coffee, fruit juices, alcohol, or dilute salt solutions) and also of hypertonic fluids (fruit juices, Coca cola, and most commercial sip feeds) to less than 500 ml daily. To make up the rest of the fluid requirement, the patient is encouraged to drink a glucose-saline replacement solution (90 mmol/l sodium or more) (table 4). Many patients at home with marginally high stoma outputs (1–1.5 litres) will be helped by a combination of oral fluid restriction (less than 1 litre per day) and addition of salt to their diet.

Often patients are advised to take liquids and solids at different times (no liquid for half an hour before and after food) however there is no published evidence that this reduces stomal output or increases macro or micronutrient absorption.55

Drink oral glucose-saline solution. Patients with stomal losses of less than 1200 ml daily can usually maintain sodium balance by adding extra salt to the limit of palatability at the table and when cooking. When stoma losses are in the range 1200–2000 ml, or sometimes more, it is possible for a patient to maintain sodium balance by taking a glucose-saline solution or salt capsules.54 56 In hot weather, due to water and sodium loss in sweat, patients with a stoma are more likely to have problems of dehydration.

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**Table 3** Summary of management of a patient with a jejunostomy

- Exclude/treat causes other than a short bowel (for example, infection or extraluminal), partial obstruction, abrupt stopping of drugs) (grade C)
- Correct dehydration with intravenous saline while the patient takes nothing by mouth for 24–48 hours. This stops thirst and thus the desire to drink (grade C)
- Reduce oral hypotonic fluids to 500 ml/day. This is the most important measure (grade B)
- Give glucose/saline solution to sip (sodium concentration at least 90 mmol/l). Most stomal/fistula losses (except from the colon) have a sodium concentration of about 100 mmol/l (grade B)
- Add sodium chloride to any liquid feeds to make the sodium concentration near to 100 mmol/l while keeping osmolality near 300 mosmol/kg (grade B)
- Give drugs to reduce motility; loperamide 2–8 mg (non-sedative and non-addictive) before food. Occasionally, addition of codeine phosphate further reduces stomal output (grade B)
- If there is not “secretory” output (generally more than 3 l/24 hours), drugs that reduce gastric acid secretion (H2 antagonists or proton pump inhibitors) or if unable to absorb oral drugs, octreotide can reduce stomal output by 1–2 l/24 hours (grade A)
- Other measures include:
  - Separating solids and liquids (that is, having no drink for half an hour before or after food) (grade C)
  - Using salt capsules instead of glucose/saline solution (grade B)
  - A trial of fludrocortisone if the ileum remains (grade B)
- Correct hypomagnesaemia (see table 6)
  - Intravenous magnesium sulphate initially then oral magnesium oxide and/or 1-alpha cholecalciferol (grade B)

*The same management applies for management of high output enterocutaneous fistula (providing there is no ongoing intra-abdominal sepsis) and for “ileostomy diarrhoea”.

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**Table 4** Replacement solutions for treatment in high output jejunostomy

<table>
<thead>
<tr>
<th>Modified WHO choleria rehydration solution</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sodium chloride</td>
</tr>
<tr>
<td>Sodium bicarbonate (or citrate)</td>
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<tr>
<td>Glucose</td>
</tr>
<tr>
<td>Tap water</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>Alternative rehydration solution</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sodium chloride</td>
</tr>
<tr>
<td>Glucose</td>
</tr>
<tr>
<td>Tap water</td>
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</tbody>
</table>
Drugs that reduce gastric acid secretion (for example, the H₂ antagonist or proton pump inhibitors or the somatostatin antagonists or oral loperamide and codeine phosphate reduce intestinal motility or secretions. They may be needed. As the intestinal output, especially in net “secretors” of intestinal fluid is relatively constant at approximately 90 mmol/l throughout the day. The World Health Organization (WHO) cholera solution has a sodium concentration of 90 mmol/l, and is commonly used (without the potassium chloride) if the sodium content of fluid is maintained in the long term.

The patient is encouraged to sip a litre or more of one of these solutions in small quantities throughout the day. To improve palatability the solution may be chilled or flavoured with fruit juice.

Sodium chloride capsules (500 mg each) are effective when taken in large amounts (14/24 h) but can cause some patients to feel sick and even vomit. If an enteral feed is given, sodium chloride needs to be added to make the total sodium concentration of the feed 100 mmol/l while keeping osmolality near to 300 mosmol/kg.

Drug therapy. If restricting oral fluids and giving a glucose-saline solution to drink are not adequate drugs may be needed. As the intestinal output, especially in net “secretors” rises after meals, it is important to give the drugs before food. Drugs used to reduce jejuno-stomy output act to reduce either intestinal motility or secretions.

- Antimotility (anti-diarrhoeal) drugs

Loperamide and codeine phosphate reduce intestinal motility and thus decrease water and sodium output from an ileostomy by approximately 20–30%. Loperamide is preferred to opiates (for example, codeine phosphate) as it is not sedative, addictive, and does not cause fat malabsorption.

Oral loperamide 4 mg taken four times a day was more effective in reducing the weight and sodium content of ileostomy fluid than codeine phosphate 60 mg taken four times a day but the effect of both together may be greater.

Loperamide circulates through the enterohepatic circulation, which is severely disrupted in patients with a short bowel, and small bowel transit may be very rapid. Thus high doses of loperamide (for example, 12–24 mg at a time) may be needed. Lomotil is occasionally used but anti-cholinergic effects (for example, dry mouth) may be a problem.

If these or any tablets/capsules emerge unchanged in stool/stomal output, they can be crushed, opened, mixed with water, or put on food.

- Antisecretory drugs

Drugs that reduce gastric acid secretion (for example, the H₂ antagonists or proton pump inhibitors or the somatostatin analogue octreotide) are most commonly used.

Cimetidine (400 mg orally or intravenously four times a day), ranitidine (300 mg orally twice daily), and omeprazole (40 mg orally once a day or intravenously twice a day) reduce jejuno-stomy output, particularly in those with a net secretory output and generally in those with an output exceeding 2 litres daily. Omeprazole is readily absorbed in the duodenum and upper small bowel, but if less than 50 cm of jejunum remains, it may need to be given intravenously (some units administer it orally with sodium bicarbonate). These drugs, which inhibit gastric acid secretion, are as effective as octreotide (50 μg subcutaneously twice daily) in reducing the volume of stomal output. They do not change the absorption of energy, carbohydrate, lipid, nitrogen, or divalent cations, and do not reduce jejuno-stomy output enough to reduce the severity of IF (that is, prevent the need for parenteral fluid and electrolyte replacement).

In adults, octreotide reduces ileostomy diarrhoea and large volume jejuno-stomy output. The greatest reductions in intestinal output are in those with a net secretory output, and the volume of parenteral supplements needed may be reduced. The reduction in sodium output parallels that of intestinal output while magnesium balance is unchanged.

Total energy and nitrogen absorption are not significantly changed while fat absorption may be unchanged or reduced. The effect of octreotide is maintained in the long term.

Mineralocorticoids (for example, 2 mg oral fludrocortisone or 2 mg intravenous d-aldoysterone or high dose hydrocortisone) may reduce stomal output in patients with a retained ileum.

Intravenous therapy. Some patients will not maintain hydration with the above measures and intravenous or subcutaneous saline may be needed. Half to one litre of saline may be given subcutaneously (with 4 mmol magnesium sulphate) if only needed 1–3 times a week and intravenously if more frequently. An intravenous line is likely to be a tunnelled cuffed (long term) central line through which a parenteral feed can also be given if undernutrition is or is likely to be a problem. The saline bag may have 4–12 mmol magnesium sulphate added.

6.2 Hypomagnesaemia (table 5)

Rehydration to correct secondary hyperaldosteronism is the most important first step. Most magnesium salts are poorly absorbed and may worsen diarrhoea/stomal output. Magnesium oxide is commonly given and contains more elemental magnesium than other salts, is insoluble in water and alcohol, but soluble in dilute acid. In the stomach it is converted to magnesium chloride. It is given as gelatine capsules of 4 mmol magnesium oxide (160 mg of MgO) to a total of 12–24 mmol daily. Magnesium oxide is usually given at night when intestinal transit is assumed to be slowest and hence there is more time for absorption.

If oral magnesium supplements do not normalise magnesium levels, oral 1-alpha hydroxy-cholecalciferol in a gradually increasing dose (every 2–4 weeks) of 0.25–9.00 μg daily may improve magnesium balance bit regular monitoring of serum calcium is necessary to avoid hypercalcaemia.

Occasionally magnesium is given as an intravenous or subcutaneous infusion, usually with saline.

6.3 Undernutrition: protein-energy malnutrition

Patients with a jejunostomy absorb a constant proportion of nitrogen, energy, and fat from their diet.

As the sodium content of jejuno-stomy (or ileostomy) effluent is relatively constant at approximately 90 mmol/l and as there is coupled absorption of sodium and glucose in the jejunum, patients are advised to sip a glucose-saline solution with a sodium concentration of at least 90 mmol/l throughout the day. The World Health Organization (WHO) chlorella solution has a sodium concentration of 90 mmol/l and is commonly used (without the potassium chloride) if the sodium content of fluid is maintained in the long term.

The patient is encouraged to sip a litre or more of one of these solutions in small quantities throughout the day. To improve palatability the solution may be chilled or flavoured with fruit juice.

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Patients with a jejunostomy absorb a constant proportion of nitrogen, energy, and fat from their diet.
fat in the diet increases energy density, keeps the diet osmolality low, increases palatability, and provides essential fatty acids. It does raise fat excretion but does not usually increase stomal output, nor make output offensive. There is no advantage in giving a diet of small molecules (for example, an elemental diet), which causes a feed to be hyperosmolar and usually contains little sodium, so stomal losses of water and sodium increase. As stomal sodium losses are approximately 100 mmol/l, any diet will need added sodium chloride.

These patients need a diet of high energy (carbohydrate or lipid) in which osmolality is kept low using large molecules (polysaccharides, protein, and triglycerides) and to which sodium chloride is added to give the meal/liquid feed a total sodium concentration of 90–120 mmol/l and an osmolality of approximately 300 mosmol/kg.

6.4 Social problems

Effluent from a small bowel stoma, unlike from a colostomy, is not offensive but large volumes of fluid may cause practical difficulties with emptying the bag and embarrassment with occasional leakage.

A summary of the problems of a short bowel are given in table 6.

### 7.0 ALTERNATIVE TREATMENTS

#### 7.1 Growth factors

Four randomised placebo controlled studies have been performed using growth hormone to stimulate mucosal growth. In three studies there was no significant increase in absorption but one showed a small improvement in nutrient absorption. As plasma levels of GLP-2, which causes villus growth, are low in patients with a jejunostomy, GLP-2 has been given as a subcutaneous injection and a small increase in nutrient absorption occurred. An analogue of GLP-2 that is resistant to degradation (teduglutide) has shown a more pronounced effect in increasing intestinal fluid absorption, and drug induced mucosal growth was demonstrated for the first time in humans.

#### 7.2 Surgical treatments

Surgical treatments other than transplantation have tried to slow intestinal transit or increase the surface area for absorption. Favourable results have been reported from reversal of a 10 cm segment of small intestine.

#### 7.3 Intestinal transplantation

Transplantation has now become a standard operation for end stage liver, kidney, heart, or lung failure. Intestinal transplantation is possible for patients with IF and over 1200 such operations have now been performed worldwide. In many cases the patient may require a liver-small bowel graft or a “multivisceral” graft, including other organs such as the stomach and pancreas. Unlike renal failure, where transplantation is preferable to long term extracorporeal renal support, intestinal transplantation cannot yet be recommended as an alternative therapy for patients stably maintained on intravenous nutrition. This is due both to the excellent outcomes reported overall for long term parenteral nutrition and the challenges posed by intestinal transplantation. The intestine is a difficult organ to transplant due to its immunogenicity, large population of donor immune cells present within the graft, and its non-sterile contents. Rejection causes barrier failure and bacterial translocation so that sepsis may occur at a time when increased immunosuppression is required and the patient may rapidly become too unwell to consider graft removal. Significant recent advances have made intestinal transplantation a more acceptable procedure. The development of tacrolimus based immunosuppressive regimens in the 1990s more than doubled the number of survivors following intestinal transplantation compared with the use of cyclosporin in the previous decade. The resulting increase in lymphoproliferative disease resulting from heavy non-specific immunosuppression has led to revision of immunosuppressive regimens over the last few years, with further benefits in patient outcome due to the use of antilymphocyte induction therapy. Furthermore, while it was considered an option of last resort, initially only the sickest patients were transplanted with predictably poor results—identifying patients likely to benefit from transplantation and offering the operation at an earlier stage in the disease results in improved survival.

Intestinal transplantation is currently associated with approximately 80% one year survival and approaching 50% five year survival, with the majority of survivors being free of parenteral nutrition. Studies indicate a considerable increase in the quality of life following transplantation, which has been found to be better than for patients on long term parenteral nutrition with complications of therapy, and equivalent to that of patients with uncomplicated IF. Direct comparison of the results of intestinal transplantation with long term survival rates on home parenteral nutrition are invalid, as transplantation is only currently considered for selected patients likely to experience a poor outcome on intravenous feeding. Clearly, patients with IF comprise a heterogeneous group and only recently have attempts been made to identify those patients that have impaired survival while receiving long term parenteral nutrition. Those most at risk of poor outcomes on parenteral nutrition include patients with very short residual small intestinal length (<50 cm), those with an end jejunostomy, and patients with motility disorders.

<table>
<thead>
<tr>
<th>Table 6</th>
<th>Summary of the problems of a short bowel</th>
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</thead>
<tbody>
<tr>
<td><strong>Presentation</strong></td>
<td>Gradual, diarrhoea and undernutrition</td>
</tr>
<tr>
<td><strong>Water, sodium, and magnesium depletion</strong></td>
<td>Uncommon in the long-term</td>
</tr>
<tr>
<td><strong>Nutrient malabsorption</strong></td>
<td>Common*</td>
</tr>
<tr>
<td><strong>D (–) lactic acidosis</strong></td>
<td>Occasionally</td>
</tr>
<tr>
<td><strong>Renal stones (calcium oxalate)</strong></td>
<td>25%</td>
</tr>
<tr>
<td><strong>Gall stones (pigment)</strong></td>
<td>45%</td>
</tr>
<tr>
<td><strong>Adaptation</strong></td>
<td>Functional adaptation</td>
</tr>
<tr>
<td><strong>Social problems</strong></td>
<td>Diarrhoea</td>
</tr>
</tbody>
</table>

*Bacterial fermentation of carbohydrate salvages some energy, but D (–) lactic acidosis can occur if the diet is high in mono and oligosaccharides.*
Most deaths of parenterally fed patients are attributable to the underlying disease and with some exceptions (for instance, liver and small bowel transplantation for mesenteric infarction due to an inherited thrombophilic disorder), the deaths preventable by transplantation are those caused by complications of long term parenteral nutrition. These include infection related to the indwelling venous feeding catheter accounting for up to 70% of parenteral nutrition related deaths, 124–137 and liver complications. 156–159 Certain types of patients appear to be at increased risk of line related sepsis, including those requiring high doses of opiates on a regular basis and those with a stoma. 150 160 161 Unfortunately, sepsis, including those requiring high doses of opiates on a regular basis and those with a stoma. 150 160 161 Similarly, venous thromboses and occlusions preventing adequate access occur infrequently 154 and it is impossible to predict the rate at which loss of vascular access may occur. The extent to which serious liver complications occur as a result of parenteral nutrition is controversial. Alterations in biochemical liver function are common, 145 but the proportion of parenteral nutrition related deaths attributable to liver disease varies in adults from 0% to 22%. 146 Identification of those groups of patients at most risk of major complications on parenteral nutrition and likely to benefit from intestinal transplantation therefore remains a high research priority.

Adult intestinal transplantation in the UK is carried out in two national centres—at Addenbrooke’s Hospital in Cambridge and in St James’ Hospital in Leeds, linked respectively to the Intestinal Failure Units at St Mark’s and the Hope Hospital for joint assessment of candidates. Intestinal transplants in children are performed at the Birmingham Children’s Hospital. Survival values are comparable with those reported in international series 125 but to date only 14 adult patients have received intestinal grafts in this country. Compared with other European and North American transplant centres, fewer patients are referred for intestinal transplantation in the UK and often too late to consider the operation. 147 As for all organ transplantation programmes, early discussion with a view to referral for assessment is essential. For instance, while lack of vascular access for intravenous nutrition is an indication of intestinal transplantation, it must be remembered that adequate central venous access is still required for a successful operative outcome. Furthermore, patients may have to wait a considerable length of time for donor organs to become available. As outcomes of intestinal transplantation continue to improve, its indications will evolve, but the current major criteria for referral for consideration of intestinal transplantation are listed in table 7.

Table 7  Current indications for referral of adults to an intestinal transplantation centre in the UK

<table>
<thead>
<tr>
<th>Complications of parenteral nutrition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Liver disease (portal hypertension, bridging hepatic fibrosis, or cirrhosis) due to parenteral nutrition— irreversible despite referral and management by an established parenteral nutrition centre.</td>
</tr>
<tr>
<td>Progressive or compromised vascular access for parenteral feeding—loss of all but two major venous access points (one of which should be above the diaphragm).</td>
</tr>
<tr>
<td>Recurrent or life-threatening central line sepsis (including fungal sepsis).</td>
</tr>
<tr>
<td>Inadequate maintenance on parenteral nutrition for any other reason—for instance, inability to manage hydration/nutrition status despite parenteral nutrition.</td>
</tr>
<tr>
<td>High risk conditions</td>
</tr>
<tr>
<td>Requirement for extensive evisceration (that is, desmoid tumours, trauma, rare selected malignancies including neuroendocrine tumours)</td>
</tr>
</tbody>
</table>

8.0 REFERENCES


Intestinal transplant registry: http://www.intestinaltransplantregistry@uhn.on.ca.


