Intestinal failure and short bowel syndrome

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Abstract

Intestinal failure (IF) is a reduction in gut function below the minimum necessary for the absorption of macronutrients and/or water and electrolytes adequate to maintain health. Type I IF is a relatively common acute condition typically occurring in the days after surgery and is usually self-limiting. Type II IF lasts weeks to months, can be reversible but can also progress to type III IF. Chronic, type III, IF is the rarest organ failure. Optimal multidisciplinary management of these complex patients, particularly of those with acute type II IF, is key to their successful physical and psychological long-term survival. Short bowel is one of the main causes of types II and III IF. Depending on the residual small intestine, varying degrees of oral and parenteral support are needed.

Keywords Fistula; high stoma output; intestinal failure; jejunostomy; MRCP; nutrition; short bowel syndrome

Key points

- Intestinal failure (IF) is a rare but important condition with significant morbidity and mortality
- There are three types of IF: type I is acute usually self-limiting IF lasting a few days; type II is acute, lasting weeks to months, is sometimes self-limiting or reversible and occasionally progresses to chronic, type III, IF
- Early identification, optimal fluid and nutrition support by skilled multidisciplinary teams are the key to a good outcome for the patient

Definition

The internationally recognized definition of intestinal failure (IF) is a reduction in gut function below the minimum necessary for the absorption of macronutrients and/or water and electrolytes, such that intravenous supplementation is required to maintain health and or growth.[1] IF is typically divided into types I, II and III (Table 1).

Frequency of intestinal failure

Type I IF is common and thought to occur in up to 15% of patients in the postoperative period. It is usually of short duration and self-limiting.[2] Type II IF is uncommon; in England, around 600–700 individuals have type II IF, with an estimated annual incidence of 9 per million. A proportion of people with type II disease end up with chronic IF and long-term home parenteral nutrition (HPN). The prevalence of patients with HPN in England is approximately 50 per million.

Pathogenesis

Type I IF often occurs in the perioperative period and is in most cases self-limiting.[2] Optimizing fluid management and avoiding sodium and volume overload with early oral nutrition and mobilization are the key to rapid resolution. Type I IF, sometimes termed acute gastrointestinal injury, is also associated with other critical illness, such as head injury, pneumonia or acute pancreatitis, and can occur after cardiac surgery.

Type II IF is uncommon and is accompanied by septic, metabolic and complex nutritional complications (Table 2). It is most often seen as the result of an intra-abdominal catastrophe or radical surgery, such as occurs in mesenteric ischaemia, volvulus and abdominal trauma, or after multiple surgeries for Crohn's disease or complex fistula resection with proximal stoma formation. Type II IF can result from inadvertent intestinal injury in routine surgery with resultant enterocutaneous fistula formation. The management of type II patients requires skilled multidisciplinary teams, including, but not exclusively, critical care, surgery, gastroenterology, radiology and stoma care professionals, dietitians, physiotherapists, microbiologists and pharmacists. Management is aimed at intestinal rehabilitation with a return to exclusive oral or enteral nutrition. This occurs in approximately 40% of patients. Around 50% transition to type III IF requiring prolonged HPN. In-hospital mortality is high in type II IF and has been reported to occur in up to 13% of cases.

Type III IF is chronic IF in a metabolically stable patient;[3] it is the rarest organ failure. It can result from benign or malignant disease (Table 2). It is occasionally reversible, depending on the cause of the IF. Survival in benign disease is good – approximately 80% at 5 years in adults. Two-thirds of adults with IF are working and have a family life. However, type III IF is associated with life-threatening complications such as catheter-related bloodstream infections, venous thrombosis and stenosis, IF-related liver disease and complications related to the underlying disease. In malignant disease, survival depends on the progress of the underlying malignancy.

Approach to the new patient with suspected intestinal failure

History and examination

A careful history should be taken and the medical notes reviewed, with particular reference to current medications, previous radiotherapy, previous operations and operative notes. The operative notes are of particular salience to enable the gastrointestinal anatomy, the residual intestinal length and the proximal length of intestine in continuity with the stoma or fistula to be defined. Establishing premorbid health and nutritional status is

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Summary of type of IF, duration, conditions and initial management

Type	Incidence	Description	Duration	Conditions	Management
I	Common	Acute condition Other organ dysfunction often present Often self-limiting when other organ dysfunction is corrected	Days	Paralytic ileus postoperatively Paralytic ileus as part of multiorgan dysfunction	Support during acute phase Early oral nutrition, early mobilization Good fluid management Stabilization and maintenance of homeostasis Resolution of IF
II	Uncommon	Prolonged acute condition Continuing metabolic instability	Weeks to months	Recurrent abdominal infection with or without fistulization Surgical resection Acute phase of short bowel syndrome	Achievement of steady-state without infection and with no other organ dysfunction Prolonged parenteral nutrition Resolution of IF or progression to chronic IF
III	Rare	Chronic organ failure without concomitant acute organ dysfunction Steady-state condition	Months to years	Short bowel syndrome (multiple aetiologies) Intestinal dysmotility End-stage malignant disease	Maintenance of homeostasis Optimization of nutrition and wound status Restoration of gut integrity where possible

Table 1

helpful. A subjective global assessment is straightforward bedside assessment (Table 3).

An accurate measurement of intake, oral, enteral and parenteral fluids, including drugs, and losses, both gastrointestinal and urinary, is critical for ensuring any deficit is corrected and a steady-state is reached (Table 4).

Investigation and management

Investigation should be directed towards identifying and controlling infection; albumin is a marker of underlying disease status and is a very poor marker of nutritional state. Nutritional status should be optimized. Skin should be cared for to aid wound healing, and imaging should be undertaken to map the anatomy of the residual gastrointestinal tract and/or fistula. Further surgery should be planned carefully with adequate time, usually at least 6 months, given for both physical and psychological healing to occur.

Management of short bowel syndrome

Patients with short bowel syndrome typically have a high-output stoma or high-output diarrhoea if their colon is in continuity with the stoma. In general, if patients have <200 cm of small bowel remaining, additional oral and/or parenteral supplementation can be required.[4] A high output from a stoma or fistula is generally considered to be >2000 ml, or sufficient to cause problems with fluid and electrolyte management or stoma bag care.

Intra-abdominal infection, enteritis (Crohn's disease, radiation), partial or intermittent bowel obstruction, sudden cessation of drugs, opiate withdrawal and prokinetics can contribute to a high output. An Addisonian crisis can also do so, particularly if a patient has been on long-term corticosteroids and these are stopped or not sufficiently increased perioperatively.

There is marked sodium and water depletion and severe thirst; drinking more water can, however, worsen sodium and water loss, further driving the stoma output. Losses of >2 litres/day should be replaced intravenously with 0.9% sodium chloride, to match the sodium and chloride losses (Table 4). Reducing or stopping oral intake will reduce stoma output and demonstrate that the stoma output is driven by oral intake. The aim of treatment is to maintain hydration or body weight and a daily urine volume of at least 800 ml, with a urine sodium concentration of >20 mmol/litre.

Oral hydration solution: with stomal losses of 1200–2000 ml sodium, a balance can be maintained by drinking 1000–1500 ml of glucose–sodium chloride solution with a sodium concentration of at least 90 mmol/litre throughout the day (Table 5). Caution should be exercised with ready-mixed antidiarrhoeal compounds as these are designed for colonic diarrhoea and contain significant amounts of potassium.

Drugs: loperamide and codeine phosphate reduce intestinal motility and decrease water and sodium output by approximately 20–30%. There has been a Medicines and Healthcare products Regulatory Agency alert regarding high doses of loperamide, used almost exclusively for recreational purposes, and cardiac arrhythmias caused by potassium channel inhibition. An electrocardiograph (ECG) should be recorded before starting high-dose loperamide (>4 mg four times per day), and the QT_c interval measured and documented. The ECG should be repeated after starting the high dose, and then every 3 years. The total daily dose should be <80 mg, and it is exceedingly rare for patients to require >32 mg/day.

Proton pump inhibitors and H_2-receptor antagonists can be given to reduce gastric secretion. Omeprazole is absorbed from the duodenum and proximal jejunum. Somatostatin analogues, for example

Pathophysiological classification of IF with the associated main pathophysiological conditions

Condition and causes	Primary mechanism of IF	Additional contributors to IF
Short bowel • Extensive surgical resection for mesenteric infarction (arterial or venous) • Crohn's disease • Radiation enteritis • Surgical complications • Intestinal volvulus • Familial polyposis • Intestinal angiomatosis • Necrotizing enterocolitis • Complicated intussusception • Congenital gastrointestinal malformation	Reduced absorptive mucosal surface	• Increased intestinal losses of fluid and electrolytes • Restricted oral/enteral nutrition (to reduce intestinal losses) • Disease-related hypophagia • Lack of adaptive hyperphagia • Accelerated gastrointestinal transit time • Small bowel bacterial overgrowth
Intestinal fistula • Inflammatory (Crohn's disease, diverticular disease, radiation enteritis, pancreatic disease) • Neoplastic • Iatrogenic • Infection (tuberculosis, actinomycosis) • Trauma • Foreign body	Bypass of large areas of absorptive mucosal surface	• Increased intestinal losses of fluids and electrolytes • Disruption of enterohepatic cycle • Restricted oral/enteral nutrition to reduce fistula output • Impaired intestinal peristalsis • Increased metabolic demand related to concomitant infection and inflammation
Intestinal dysmotility • Chronic intestinal pseudo-obstruction – Primary (neuropathic, myopathic, mesenchymal) – Secondary (collagen vascular diseases, endocrine disorders, neurological disorders, drug-induced, paraneoplastic, radiation, chronic vascular insufficiency)	Restricted oral/enteral nutrition or total fasting from intolerance to feeding-related exacerbations of digestive symptoms or to episodes of non-mechanical intestinal obstruction	• Malabsorption caused by small bowel bacterial overgrowth • Increased intestinal secretion of fluids and electrolytes in the obstructed segments • Increased intestinal losses of fluids and electrolytes from vomiting, gastric drainage and/or diarrhoea
Mechanical obstruction • Obstructing mass (polypoid tumour, intussusception, gallstone, foreign body, bezoar) • Intrinsic bowel lesions (stenosis or strictures, Crohn's disease, drug-induced, anastomotic) • Extrinsic lesions (adhesions, sclerosing peritonitis, hernias, tumour)		• Increased intestinal secretion of fluids and electrolytes in the obstructed segments • Increased intestinal losses of fluids and electrolytes from vomiting or gastric drainage
Extensive small bowel mucosal disease • Microvillous inclusion disease • Autoimmune enteropathy • Intestinal lymphangiectasia • Protein-losing enteropathies • Common variable immune deficiency • Crohn's disease • Coeliac disease • Radiation enteritis • Chemotherapy-induced enteritis • Congenital malabsorptive disorders	Inefficient absorptive and/or nutrient-wasting mucosal surface	• Increased intestinal losses of fluid and electrolytes • Restricted oral/enteral nutrition • Disease-related hypophagia

Table 2

Subjective global assessment

Weight change and height	
Current	Height (cm)
	Weight (kg)
	Overall loss in past 6 months (kg), %
	Change in past 2 weeks (kg), ±%
Dietary intake change	
No change	
Change type	Duration (days)
	Suboptimal solid diet
	Hypocaloric liquids
	Starvation
Supplement	None
	Vitamins
	Minerals
Gastrointestinal symptoms persisting for >2 weeks	
None	
Nausea	
Vomiting	
Diarrhoea	
Pain	At rest
	On eating
Functional capacity	
No dysfunction	
Dysfunction	Duration (days)
	Type
	• Working suboptimally
	• Ambulatory but not working
	• Bed-ridden
Disease and its relation to nutritional requirement	
Primary diagnosis	
Metabolic demand (stress)	No stress
	Moderate stress
	High stress (burns, infection, severe trauma)
Physical status (for each trait specify 0 = normal, 1 = mild deficit, 2 = established deficit)	
	Loss of subcutaneous fat
	Muscle wasting
	Oedema
	Ascites
	Mucosal lesions
	Cutaneous and hair changes
SGA grade	
A, well nourished; B, moderate or suspected protein-energy malnutrition; C, severe protein-energy malnutrition	

Table 3

Oral rehydration solution electrolyte contents

Modified World Health Organization rehydration drink		Alternative oral glucose–sodium chloride drink	
Sodium chloride	60 mmol (3.5 g)	Sodium chloride	120 mmol (7 g)
Sodium bicarbonate	30 mmol (2.5 g)	Glucose	44 mmol (8 g)
Glucose	110 mmol (20 g)	Water	1000 ml
Water	1000 ml		

Table 5

Intestinal length and guide to likely oral or parenteral requirements

Intestinal length (cm)	Intestine to colon	Jejunostomy
0–50	Parenteral nutrition	Parenteral nutrition
51–100	Oral nutrition/ hyperphagia	Parenteral nutrition
100–150	None	Oral nutrition plus oral glucose–sodium chloride ± parenteral sodium chloride with magnesium
150–200	None	Oral nutrition plus oral glucose–sodium chloride

Table 6

octreotide, are occasionally useful; however, they can reduce intestinal adaptation by reducing splanchnic protein synthesis.

Hypomagnesaemia: low magnesium is common and has multiple aetiologies and not just stomal losses. Rehydration to correct secondary hypoaldosteronism is the primary stage. Oral magnesium salts, for example magnesium oxide, magnesium hydroxide and magnesium glycerophosphate, are poorly absorbed and can worsen a high output. Magnaspartate is a recently licensed oral magnesium preparation that seems to be effective and well tolerated.

Nutrition: patients need a high-energy, carbohydrate or lipid, diet. The residual functional length of intestine gives an indication of the likely requirement for parenteral supplementation (Table 6).[4] Early involvement of the multidisciplinary nutrition team, often at a specialist centre, is crucial.

Summary of main gastrointestinal fluid losses and associated electrolyte content

Fluid source	Sodium (mmol/litre)	Potassium (mmol/litre)	Chloride (mmol/litre)	Bicarbonate (mmol/litre)
Gastric (vomiting/nasogastric tube loss)	20–60	14	140	60–80
Biliary (drain/fistula)	145	5	105	40
Pancreatic (drain/fistula)	125–138	8	56	85
Jejunum (fistula/stoma)	140	5	135	8
Ileum (new stoma/high stoma/fistula)	100–140	4–5	75–125	0–30
Ileum (established stoma/low fistula)	50–100	4–5	25–75	0–30
Diarrhoea or colostomy	30–140	30–70		20–80

Table 4

Tertiary interventions: glucagon-like peptide-2 (GLP-2) analogues, for example teduglutide, have been found in trials to reduce the requirements for parenteral nutrition.[5]

Intestinal transplantation and, more rarely, multivisceral transplantation are an option in carefully selected patients. Survival is better for isolated intestinal transplantation, with survival of 80% at 1 year, and reported overall survival of 61% at 15 years. ◆

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TEST YOURSELF

To test your knowledge based on the article you have just read, please complete the questions below. The answers can be found at the end of the issue or online here.

Question 1

A 56-year-old man presented with a stoma output of 1800 ml per 24 hours 10 days after a routine right-hemicolectomy and ileostomy for colorectal cancer. He was eating and drinking.

What is the appropriate first-line management?

A. Octreotide 50 micrograms subcutaneously 8-hourly
B. Make the patient nil-by-mouth to demonstrate the secretory output
C. Discharge the patient home on oral rehydration drinks
D. Loperamide 2 mg 6-hourly titrated to the stoma output
E. Codeine phosphate 30 mg 6-hourly titrated to the stoma output

Question 2

A 30-year-old man presented after extensive small bowel resection, an end-stoma and an ileum–colon mucous fistula for mesenteric venous infarction. He was a body-builder. At surgery, 120 cm of small intestine to the stoma was measured.

Who should he now be referred to?

A. Dietitian
B. Gastroenterologist
C. Pharmacologist
D. Surgeon for immediate stoma reversal
E. Multidisciplinary nutrition team

Question 3

A 74-year-old man presented with abdominal distension, vomiting and no output from his stoma 3 days after emergency left hemicolectomy for perforated diverticular disease. He had no other co-morbidities and his pre-morbid weight was 80 kg.

Investigation
CT scan of the abdomen did not demonstrate mechanical obstruction, collection or leak

What is the most appropriate fluid regimen for him?

A. 3 litres/day 0.9% saline, one bag with 40 mmol potassium chloride 8-hourly bags
B. 3 litres compound sodium lactate given between the hours of 8am and 10pm
C. 3 litres 5% glucose 4 hours per bag, one bag with 40 mmol potassium chloride
D. 1.5 litres 5% glucose, 1 litre 0.18% saline 4% glucose with 40 mmol potassium chloride 6-hourly bags
E. 1 litre 0.18% saline 4% glucose with 20 mmol potassium chloride, 1 litre 0.18% saline 4% glucose with 40 mmol potassium chloride, 500 ml 0.18% saline 4% glucose, given between the hours of 8 am and 10pm