Complications of short bowel syndrome

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Abstract

Short bowel syndrome is the commonest reversible cause of intestinal failure. Most of the children are started on parenteral nutrition (PN) after surgery to enable growth and allow time for intestinal adaptation i.e. a process whereby the shorter length of bowel is able to achieve complete function as if the entire length of bowel is present. With advances in management, the majority of children with short bowel syndrome are able to discontinue PN and establish on full enteral feeds. This article mainly focuses on the complications of short bowel syndrome that need to be avoided in order for intestinal adaptation to progress and the child to be established on enteral feeds/oral diet.

Keywords central venous catheter; gastroschisis; intestinal failure; intestinal failure associated liver disease; intestinal transplantation; necrotising enterocolitis; parenteral nutrition; short bowel syndrome

Introduction

Short bowel syndrome (SBS) and intestinal failure are mostly used interchangeably but strictly, SBS should only be used to refer to patients who suffer from malabsorption from a shortened bowel length due to congenital or due to surgical resection. Intestinal failure is a more encompassing term which includes SBS as well as enteropathies and dysmotility malabsorptive syndromes.

The hallmark of short bowel syndrome or intestinal failure is its malabsorptive state leading to fluid, electrolytes and nutrient derangements. Most of the children are established on parenteral nutrition (PN) until they can be established on enteral feeds by a process called intestinal adaptation. PN is discussed in detail in the article *Long term parenteral nutrition* in this issue.

Children are usually discharged on home PN and strategies to achieve intestinal adaptation can be done on an outpatient basis with intermittent inpatient admissions. The process of intestinal adaptation may take months or years to establish. Resultant complications may develop during the process and can be grouped into three general categories: anatomical, nutrient deficiencies and therapy related (Table 1).

Nutritional management

Fluid requirements and total body sodium deficit

Fluid and electrolyte imbalance is common in SBS, with GI water and salt losses highly individual, being dependent on both anatomical and functional factors. In a child with a high jejunostomy, for example, there may be massive fluid loss, with high sodium, potassium and bicarbonate content. This needs to be carefully considered in children and appropriate adjustments need to be made to PN. Total body sodium deficits is seen especially in infants below 32 weeks gestation due to renal salt wasting from impaired proximal and distal tubule reabsorption. Moreover, sodium deficit inhibits DNA synthesis so a positive sodium balance is required for growth. Patients with a stoma are also prone to developing a sodium deficiency due to electrolyte losses through the stoma. As filtered sodium excretion fraction may be difficult to obtain, a low spot urinary sodium (less than 10 or urinary sodium is less than twice that of the urinary potassium value) is an alternative to indicate sodium retention and hence total body sodium deficit. Enteral or parenteral supplementation of 1–2 mmol/kg of sodium chloride is recommended initially.

Carbohydrate malabsorption

Even though glucose absorption and sucrose hydrolysis increases significantly after bowel resection, carbohydrate malabsorption is one of the most common problems after small bowel resection. This may be due to decreased intestinal transit time from short length of bowel and dysmotility causing less contact time for enzymatic breakdown as well as transporter facilitated uptake of monosaccharides. This same mechanism might account for decrease formation of short chain fatty acids (acetate, propionate ad butyrate) by colonic bacteria, the lack of which has been shown to be associated with diversion colitis.

Carbohydrate malabsorption usually manifests as watery stools and/or an increase in the frequency of stools. Stool pH and reducing substances are useful investigations. A faecal pH less than 5.5 is indicative of carbohydrate malabsorption and a positive reducing substance suggests the presence of malabsorption of reducing sugars (glucose, galactose, fructose, maltose and sucrose). Excoriation in the perianal region might be a sign of acidic stools. Depending on the clinical state of the patient, a reduction in the carbohydrate component or a complete cessation of feeds is required to reduce symptoms. If, after commencing feeds the child develops diarrhoea, modular feeds (individualised milk feeds) and dietetic review can be helpful. A low osmolar feed may be required particularly if the ileum is completely resected or a high jejunostomy in children with short bowel syndrome.

Fat and fat soluble vitamins malabsorption

As the ileum is the site for enterohepatic recycling of bile acids, its resection will affect its main function of fat absorption and emulsification of fat. Bile acids which are not recycled are loss distally and can cause bile acid diarrhoea in the colon. Under normal physiological situations undigested fats reach the ileum, and induce activation of the so-called 'ileal brake'. This mechanism serves to decrease appetite and increase small bowel transit time but will be lost following ileal resection thereby potentiating intestinal hurry and fat malabsorption. Dietetic manipulation of feeds to reduce the amount of long chain triglyceride (LCT) and increase the amount of medium chain triglyceride (MCT) may help in symptom control whilst still maintaining caloric provision. Care needs to be taken as excessive MCT can cause diarrhoea and ketosis.

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Complications associated with short bowel syndrome

Anatomical	Nutrient deficiencies	Therapy related
Stoma prolapse		Catheter related sepsis
Anastomotic stenosis	Fluid and electrolyte	Catheter related thrombosis
Anastomotic ulceration	Carbohydrate malabsorption	Breach in catheter integrity
Dysmotility – altered transit time	Fat malabsorption	Intestinal failure associated liver disease
altered enterohepatic reabsorption	Total body sodium deficit	
of bile acid small bowel bacterial	Iron deficiency	
overgrowth hyperoxaluria metabolic	Fat soluble vitamins deficiency	
bone disease	Vitamin B12 deficiency	
	Zinc deficiency	

Table 1

Calcium, phosphate and vitamin D

Bone demineralisation is often a slow evolving problem. Therefore vitamin D, calcium and phosphate deficiencies are often not detected until late in the piece. Decreased bowel length causes decreased vitamin D absorption and thereby increases calcium and vitamin D losses in steatorrhoea. This is especially the case in ileal resection where fat soluble vitamins are absorbed. In addition, most intestinal failure patients are hospital bound with little sunlight to help with vitamin D synthesis.

These deficiencies is a particular problem in the preterm neonates as their in utero calcium retention rate is about 100–120 mg/kg/day and due to the solubility of calcium and phosphate (PO_4) in PN, this cannot be achieved. Diuretics are commonly used in these infants and will exacerbate the problem further by promotingrenal calcium excretion.

It is important to note that unfortified breast milk will not supply the necessary calcium and phosphate requirement. With absorption of calcium enterally being variable (between 30 and 60% for infant formulas and 60% for human milk), this will also need to be taken into consideration when dealing with patients with short bowel syndrome.

A combination of alkaline phosphatase (ALP), PTH and bone radiographs might help in elucidating the extent of bone demineralisation with an ALP more than 800 IU/Litre triggering a more extensive investigation. Care needs to be taken as vitamin C and K deficiencies can also lead to bone demineralisation.

Iron deficiency

Iron is important in both the formation of haemoglobin and in ensuring optimal neurodevelopment. Premature infants with SBS are particularly at risk of iron deficiency as they have not accumulated the iron stores. Term infants have 3–6 months iron stores but this will also be depleted in infants solely dependent on parenteral nutrition for greater than the 3 month period. Frequent blood sampling for full blood count, electrolyte and micronutrient estimation also contributes to iron deficiency as this removes available iron for the recycling pool.

Supplementation of iron via PN poses difficulties as ferrous (Fe2+) iron is oxidised to ferric (Fe3+) iron, forming insoluble complexes and free radicals in the presence of light and oxygen. In patients fully dependent on parenteral nutrition, parenteral iron supplementation might be necessary to provide adequate iron pool.

Although iron is absorbed in duodenum and proximal jejunum, patients on partial enteral nutrition might have iron absorption disturbed due to phytates and/or fibre in formulas as well as the lack of enhancing factors such as vitamin C.

Micronutrient deficiencies/excess

Zinc: Zinc is mostly absorbed in the duodenum and jejunum. Patients with a stoma, fistula and persistent diarrhoea are at risk of deficiency and should be supplemented in addition to normal metabolic requirements as zinc is important in growth and cell turnover.

Copper: controversies surrounds the optimum provision of copper in patients with SBS. As copper is excreted in the biliary tract and patients with SBS frequently have liver dysfunction there may be impairment of biliary flow and therefore a decrease in copper requirements. However, clinical studies provide conflicting evidence.

Non – transplant surgery for anatomical complications

When the large intestine is in situ, children with SBS and stomas need to be reconnected and established in continuity to make progress. Children with SBS can have dilated loops of bowel secondary to intestinal adaptation or due to other anatomical reasons like strictures and adhesions. If there is failure to advance feeds despite dietary manipulation of feeds, a contrast study may be needed to evaluate the cause and degree of dilatation of the bowel.

Strictuoplasty ± lengthening procedures (STEP- serial transverse enteroplasty procedure or LILT procedure-longitudinal intestinal lengthening procedure) may be necessary depending on the clinical condition of the child. These decisions are best made by multidisciplinary Intestinal Failure team and the operating paediatric surgeon.

Small bowel bacterial overgrowth

The concept of small bowel bacterial overgrowth (SBBO) stems from the view that with the absence of the ileocaecal valve, colonic bacteria are able to translocate to distal or even proximal small bowel thereby affecting nutrient absorption as well as development of symptoms depending on the metabolic process. SBBO can present rather non-specifically with vomiting, diarrhoea, abdominal bloating and abdominal pain with variable degrees of encephalopathy.

D-lactate acidosis (DLA) occurs due to the production of D-isomer of lactate by anaerobic bacteria from enteral carbohydrate. Strictly