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Nutritional management of short-bowel syndrome

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Intestinal failure (IF) is characterized by the reduction of functional gut capacity below the minimum needed for adequate digestion and absorption of nutrients. Short-bowel syndrome (SBS) is the leading cause of IF in children. It is characterized by a compromised bowel absorptive capacity owing to a severely reduced mucosal surface resulting in diarrhoea, fluid and electrolyte imbalance, and malnutrition. SBS usually follows extensive surgical resection leaving the bowel length below a critical value for adequate nutritional supply. In children, the conditions most commonly leading to extensive small-bowel resections are midgut volvulus, necrotizing enterocolitis, intestinal atresia, gastroschisis and extensive aganglionosis in Hirschsprung's disease. Most conditions are often associated with dysfunctional motility in the residual bowel and hence often result in more serious IF.

Important factors (other than the length of bowel remnant) which determine whether or not IF will develop include the underlying diagnosis, the type of segments preserved, the long-term maintenance of a stoma versus a primary anastomosis, the presence of the ileocaecal valve as well as the age of the patient at the time of surgery. Functional gut mass may be assessed by using citrulline plasma levels.

Management of short-bowel syndrome

The management of SBS aims at promoting SB adaptation and villous hyperplasia by enteral (oral or tube) feeding, by providing normal somatic growth with parenteral nutrition (PN), and by optimizing the bowel absorptive surface through non-transplant surgical techniques. PN and home-PN are the mainstay of management based on guidelines from the European Society for Paediatric Gastroenterology Hepatology and Nutrition (ESPGHAN).

Oral feeding or tube feeding

When possible, the gastrointestinal tract should be used for feeding as it is the most physiological and safest way to provide nutrition. PN should ideally not be stopped until adequate intake and growth can be achieved with oral and/or tube feeding alone. The optimal strategies for feeding, including oral versus tube feeding and continuous versus bolus, remain a matter of debate. The advantages of oral feeding, when feasible, include the maintenance of sucking and swallowing functions along with the interest and enjoyment associated with eating, as well as the stimulation of hormones released by the gastrointestinal tract promoting adaptation. For example, oral feeding promotes the release of epidermal growth factor (EGF) from salivary glands, increases gastrointestinal secretion of trophic factors and helps prevent feeding disorders. Sialoadenectomy in animals attenuates ileal villus height, and total protein and DNA content after small-bowel resection.

Type of feeds

Tube or oral feeding should be started as soon as possible after surgery. In an infant, breastfeeding should be encouraged. Human milk may be considered the first choice in this setting, followed by whole-protein feeds, if tolerated. Human milk contains a number of factors believed to support the developing neonate's immune system including nucleotides, immunoglobulin A and leucocytes. Human milk also contains glutamine and growth

factors, such as EGF, which possibly promote bowel adaptation.

Extensively hydrolysed formulas are mostly used when whole-protein feeds are not tolerated, whereas amino acid-based formulas (AABFs) have been used as a last resort. AABF are generally used in the treatment of food allergies or in case of milk protein hydrolysate intolerance. True food allergies have been rarely documented in children with SBS. Two retrospective studies report that the use of an AABF was associated with earlier weaning from PN and also a reduced rate of allergies. However, the very small sample sizes and the lack of a control group in these studies limit the applicability of these findings to all children with SBS and IF.

The gastrointestinal tract, being the most physiological route for feeding, should be used where possible, via the oral and/or (bolus) tube feeding route. This may be beneficial in achieving intestinal adaptation as well as weaning off PN and limiting the rate of complications related to small-intestinal bacterial overgrowth. If breastmilk or polymeric feeds fails, and there is to be one formula for children with intestinal failure, it should contain extensively hydrolysed protein (with a high percentage of short-chain peptides and some amino acids), medium-chain triglycerides (MCTs; 40–60%), long-chain triglycerides (LCTs) [possibly supplemented with *n*-3 polyunsaturated fatty acids (PUFAs)] and maltodextrin. If treatment with such a semi-elemental feed were to fail, an amino acid-based formula could be used.

Tolerance of feeding

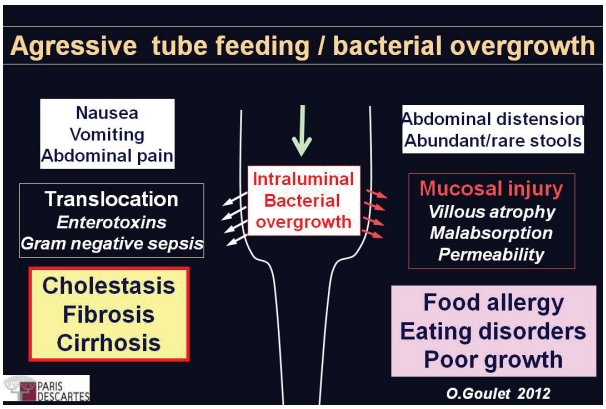
Tolerance of feeding is evaluated by measuring stool output and by the observation of vomiting, irritability and intestinal distension. Stool output should be no more than two-thirds of the feeding volume. Many factors can affect stool volume in SBS, including the length of the residual intestinal segment, the type of segment (the more proximal the resection, and the larger the fluid and sodium losses), the mucosal and endoluminal variables (residual enzymatic activity and absorptive capacity, bacterial overgrowth), the presence of the colon that can absorb large amounts of water, sodium, MCTs and peptides as well as carbohydrates metabolized to short-chain fatty acids. It is important to avoid overfeeding which may worsen fluid, mineral and nutrient malabsorption, and may result in severe perianal skin lesions. Carbohydrate intolerance presents with frequent and liquid stools, the presence of reducing substances and with stool pH < 6. Bile salts malabsorption should be suspected in children with no ileocaecal valve and/or colon, high stool volume, perianal maceration and improvement by using cholestyramine. Fluid losses are often accompanied by sodium loss and depletion; therefore, enteral sodium supplements should be provided with the aim of maintaining urinary sodium > 20 mmol/l, with a sodium : potassium urinary ratio of at least 2 : 1.

Small bowel bacterial overgrowth

Small bowel bacterial overgrowth (SBBO) is common especially in patients without an ileocaecal valve and those having abnormal motility. Common findings in these patients include dilated loops of bowel containing residual non-absorbed nutrients (Figure). SBBO can further cause mucosal inflammation and increased permeability that in turn may lead to sensitization and allergy as well as bacterial translocation, sepsis and cholestasis. Activated Kupffer cells are probably involved in the pathogenesis of intestinal failure-associated liver disease (IFALD). In some patients with dysmotile intestinal loops (intestinal atresia, gastroschisis and necrotizing enterocolitis) and liver disease, aggressive continuous tube feeding is often attempted with the aim of weaning off PN. Over-aggressive feeds may lead to the loss of self-regulation of intake and may also result in abdominal discomfort, intestinal distension and SBBO, with subsequent further mucosal and liver injury. Surgical procedures such as tapering-lengthening or serial-transverse enteroplasty aim not only to enhance the bowel length but also to reduce the diameter of dilated intestinal loops, thus improving its motility and reducing the risk for developing SBBO.

D-Lactic acidosis

D-Lactic acidosis may occur in some children during the process of bowel adaptation and even long after PN weaning. This condition is due to colonic bacterial hypermetabolism as lactobacilli and other bacteria, including *Clostridium perfringens* and *Streptococcus bovis*, when present, may ferment non-absorbed carbohydrate to D-lactic acid which cannot be metabolized by L-lactate dehydrogenase. These microorganisms may proliferate in the acidic environment of the colon that is the result of the metabolism of unabsorbed carbohydrate to short chain fatty acids. D-lactic acidosis presents with encephalopathy (ataxia, blurred speech and



Intestinal microbiota

	Beneficial	Deleterious
Bacterial flora	Microbiota	Bacterial overgrowth
Intestine	Colon	Small bowel
Intestinal barrier	Improved	Increased permeability
Trophic consequences	Hyperplasia Small bowel & colon	Villous atrophy
Mechanisms	SCFA SCFA induced GLP_2	Inflammation Allergy
Outcome	Intestinal autonomy	Liver disease

decreased consciousness) and should be considered when there is a high anion gap metabolic acidosis in the setting of normal serum lactate levels. Preventative measures for D-lactic acidosis includes the reduction of carbohydrate intake, followed by antibiotics (such as metronidazole or cotrimoxazole) when dietary changes fail.

The management of a child with IF-related SBS is complex and should be supervised by a multidisciplinary team, led by experts including gastroenterologists, dietitians, and both transplant and non-transplant surgeons. Patients are at risk of medical, surgical and nutritional complications should be anticipated, so that they can be prevented or managed appropriately. Catheter-associated infections and cholestatic liver disease are important complications that impact on the likelihood of bowel adaptation and long-term survival.

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