Short Bowel Syndrome: A Review of Management Options

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Abstract

Extensive resection of the intestinal tract frequently results in inadequate digestion and/or absorption of nutrients, a condition known as short bowel syndrome (SBS). This challenging condition demands a dedicated multidisciplinary team effort to overcome the morbidity and mortality in these patients. With advances in critical care management, more and more patients survive the immediate morbidity of massive intestinal resection to present with SBS. Several therapies, including parenteral nutrition (PN), bowel rehabilitation and surgical procedures to reconstruct bowel have been used in these patients. Novel dietary approaches, pharmacotherapy and timely surgical interventions have all added to the improved outcome in these patients. However, these treatments only partially correct the underlying problem of reduced bowel function and have limited success resulting in 30% to 50% mortality rates. However, increasing experience and encouraging results of intestinal transplantation has added a new dimension to the management of SBS. Literature available on SBS is exhaustive but inconclusive. We conducted a review of scientific literature and electronic media with search terms 'short bowel syndrome, advances in SBS and SBS' and attempted to give a comprehensive account on this topic with emphasis on the recent advances in its management.

Keywords: Intestinal adaptation, intestinal failure, malabsorption, short bowel syndrome, total parenteral nutrition

Short bowel syndrome (SBS) is an intestinal failure resulting from an inadequate length of intestine following intestinal resection. Intestinal failure refers to a condition that results in inadequate digestion or absorption of nutrients or both, so that an individual becomes malnourished and requires specialized medical and nutritional support.[1]

The prevalence of SBS is 3-4 per million.[1] It occurs in about 15% of adult patients who undergo intestinal resection, with 3/4th of these cases resulting from massive intestinal resection and 1/4th from multiple sequential resections.[2] About 70% of patients in whom SBS develops are discharged from the hospital and a similar percentage remain alive a year later.[3] This improved survival rate has been achieved primarily by the ability to deliver long-term nutritional support.

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ETIOLOGY AND PATHOPHYSIOLOGY

Several conditions requiring intestinal resection lead to SBS. In a reported series of 210 cases, these conditions included postoperative 52 (25%), irradiation/cancer 51 (24%), mesenteric vascular disease 46 (22%), Crohn's disease 34 (16%) and other benign causes 27 (13%).[4] The manifestations of SBS are due to:

2. Loss of site-specific transport processes
3. Loss of site-specific endocrine cells and gastrointestinal (GI) hormones
4. Loss of ileocecal valve

The major consequence of extensive intestinal resection is loss of absorptive surface area, which results in malabsorption of macro and micronutrients, electrolytes and water.[5] Most macronutrients are absorbed in the proximal 100–150 cm of intestine.[6] Specific micronutrients are absorbed from specific areas of small intestine. Intestinal remnant length is the primary determinant of outcome in patients with SBS. Resection of up to half of small intestine is generally well tolerated. SBS is likely to develop in patients with loss of two-thirds length of small intestine. Permanent total PN (TPN) support is likely to be needed in patients with less than 120 cm of intestine without colon in continuity and less than 60 cm with colonic continuity.[7] Besides, malabsorption of macro and micronutrients with a loss of intestinal absorptive surface area results in water and electrolyte malabsorption, which manifests as voluminous diarrhea, hypovolemia, hyponatremia and hypokalemia.

The absorption of some compounds is restricted to certain areas of small intestine. Iron, phosphorus and water soluble vitamins are predominantly absorbed in proximal small intestine. As most patients with SBS have intact duodenum and proximal jejunum, deficiencies of these entities are rare but tend to develop calcium and magnesium deficiency.[8] Having lost part or whole of the ileum, vitamin B12 and bile salt malabsorption also develops. Even hormones in the GI mucosa are distributed in a site specific manner. Gastrin, cholecystokinin, secretin, gastric inhibitory polypeptide and motilin are produced by endocrine cells in proximal gastrointestinal tract (GIT). In SBS, the status of these hormones remains intact. Glucagon-like peptide (GLP) 1 and 2, neurotensin, and peptide YY are produced in ileum and proximal colon. In SBS, deficiency of these hormones is common and this results in rapid gastric emptying, shortened intestinal transit and hypergastrinemia.[9,10] The presence of ileocecal junction improves the functional capacity of intestinal remnant.[11] Although previously this had been attributed to a barrier function and transit prolonging property of ileocecal valve, this advantage may actually be related to the specialized property of the terminal ileum itself.

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INTESTINAL ADAPTATION

The small intestine is able to adapt to compensate for the reduction in absorptive surface area caused by intestinal resection. This process occurs in the first couple of years following resection.[12] This adaptive response results from changes in the intestinal structure, motility and function. Structural adaptation following intestinal resection involves all the layers of the intestine.[13] The process is characterized by crypt cell proliferation, lengthening of the villi, increase in the ratio of the crypts to villi, increase in microvilli along the epithelial surface and an overall increase in the mucosal weight. The thickness and length of muscle layers increase as a result of hyperplasia.

Intestinal motor activity is also altered by intestinal resection.[14] Motor adaptation seems to be more prominent in the jejunum than in the ileum. There is disrupted motor activity in the first few months after resection followed by adaptation. Studies demonstrate a shorter duration of migrating motor complex cycle and fed pattern after resection. [15] Functional adaptation results in improved absorption by individual enterocytes.[1] This process is facilitated by structural and motor adaptation which results in prolonged intestinal transit time.

The mechanism of intestinal adaptation is not entirely understood. The degree of intestinal adaptation is related to the extent and site of intestinal resection.[13] Adaptation is greater with extensive intestinal resection and ileum has a greater adaptive capacity than jejunum. Factors which influence intestinal adaptation include GI regulatory peptides, growth factors, hormones, cytokines, and tissue factors which include immunity, blood flow and neural influences.

MEDICAL MANAGEMENT

The early management of a patient with SBS is that of a critically ill surgical patient who has recently undergone intestinal resection and other concomitant procedures. Thus, control of sepsis, maintenance of fluid and electrolyte balance and initiation of nutritional support are important in the early management of these patients. For patients who have survived this early phase, the primary goals of management are to maintain adequate nutritional status and prevent development of complications related to both underlying pathophysiology and nutritional therapy.

Maintenance of nutritional status

This is the primary objective in the management of SBS. Fluid and electrolyte losses from the GIT may be great in the early postoperative period and must be monitored and replaced. TPN will be required in the early postoperative period and enteral nutrition should be initiated as soon as possible.

Patients with limited ileal resection (less than 100 cm) with or without right hemicolectomy can resume intake of solid food in late postoperative phase. These patients may develop diarrhea or steatorrhea with consumption of a regular diet due to fat malabsorption, which in turn can lead to deficiencies of fat soluble vitamins, vitamin B12, calcium and magnesium. The deficiencies of these nutrients should be looked for and these nutrients should be supplemented if needed. Maintenance of nutritional status becomes all the more important in the setting of diarrhea, which is quite common in SBS and may be due to gastric acid hypersecretion, rapid intestinal transit time and fat malabsorption. H2 blockers, proton pump inhibitors (PPI), antidiarrheals, cholestyramine and octreotide have all been used to control diarrhea. Octreotide acts by slowing intestinal transit and increasing sodium and water reabsorption,[16] but carries potential risk of decreasing splanchnic protein synthesis, thereby inhibiting intestinal adaptation and also a risk of cholelithiasis. These medications should be taken one hour before meals and their effect on diarrheal volume should be evaluated before they are recommended for long-term treatment.

Glucose polymer-based oral rehydration salts (ORS) are recommended for patients to improve hydration and thereby reduce TPN requirements. Glucose and sodium are absorbed through the same active transport mechanisms and stimulate the absorption of each other. In addition, glucose promotes sodium and water absorption by means of solvent drag.

Dietary management and special diets

Patients with SBS should be encouraged to eat substantially more than usual (a hyperphagic diet) to compensate malabsorption. Patients should be encouraged to eat small portions throughout the day rather than at defined meal times. Those with colonic continuity should be provided with a high complex carbohydrate diet containing starch, non-starch polysaccharides and soluble fibers. These food stuffs which are typically not absorbed by human small intestine get fermented by colonic bacteria into butyrate, acetate and propionate. Butyrate is the preferred fuel for colocyte.[17] Studies have indicated that up to 525 to 1170 kcals per day can be absorbed from an intact colon from fermentation of unabsorbed carbohydrates and soluble fibers.[18] The amount of energy absorbed is proportional to the length of residual colon and may increase as a part of adaptive response to enterectomy.

Treatment of steatorrhea associated with ileal resection

Fat maldigestion due to bile salt malabsorption occurs when more than 100 cm of terminal ileum has been resected. Various therapeutic options have been suggested for the treatment of the resulting steatorrhea. Use of bile salt replacement therapy with ox bile or a synthetic conjugated bile acid (cholesarcosine) has been reported.[18] The bile acid sequestering agent cholestyramine may be useful in decreasing bile salt related diarrhea in patients with less than 100 cm of terminal ileum loss, but may worsen steatorrhea in those patients who have undergone a more significant resection, because of its binding with dietary lipid.[19] Also cholestyramine interferes with absorption of

high fat intake is associated with malabsorption of divalent cations, delayed gastric emptying, early satiety and increased water loss from colon. Since medium chain triglycerides (MCT) are absorbed in colon, dietary supplementation with MCT may lead to increased energy consumption.[20,21] Limitations of MCT include the fact that they do not provide essential fatty acids (FAs) and can cause nausea, vomiting and ketosis.

Another important aspect of dietary management is to provide a diet that will maximize intestinal adaptive response.[22] Provision of fat and dietary fibers may be particularly important in this regard. Long and short chain FA appear to have a greater trophic effect on the intestine than medium chain FA do. Although these nutrients directly stimulate intestinal adaptation, they also bring about intestinal adaptation through endocrine and paracrine effects.

Pharmacologic therapy for SBS is a rapidly expanding area of investigation. Recent evidence suggests that provision of appropriate diet, nutritional supplements such as glutamine and growth factors such as growth hormone improves intestinal absorption and perhaps modifies the adaptive response in patients with established SBS.[23] Currently GLP-2 appear to have the most promising results.[24]

Home parenteral nutrition

Home parenteral nutrition is an option for patients who require long-term TPN. To prepare the patient for home TPN, the regime should be compressed gradually in 2 to 4 h daily increments so that the total volume can be infused over a 10–12-h period, typically over night. The TPN infusion is generally tapered off over a 30–60-min period to avoid hypoglycemia. Additional fluid allowances may be needed for patients with a permanent jejunostomy. The TPN solutions should be infused into a central vein such as superior or inferior venacava through a tunneled catheter to decrease the risks of infection and thrombosis.[25]

Prevention of complications

Complications in SBS could be related to either the underlying pathology or the nutritional therapy. Among patients who require long-term TPN for survival, sepsis and liver disease related to TPN are important factors governing morbidity and mortality. The incidence of sepsis varies from 0.1 to 0.3 episodes per patient per year of TPN. Sepsis may be associated with catheter thrombosis. In cases with catheter-related sepsis an attempt at line sterilization before removal is appropriate when infections are caused by coagulase-negative staphylococci and gram-negative bacteria.

End-stage liver disease develops in about 15% of patients on long-term TPN and is associated with a survival time of about 1 year without liver transplantation.[26] The etiology of TPN-associated liver disease is not completely understood and seems to be multifactorial. This is reversible in initial stages, but ultimately leads to severe steatosis, cholestasis and cirrhosis. The liver function tests (LFTs) of patients on long-term TPN should be monitored regularly and patients with abnormal LFT should undergo ultrasound evaluation of gall bladder and bile ducts and should have a liver biopsy performed, as appropriate. TPN-induced liver disease can be minimized by providing high calories enterally, avoiding over feeding, using mixed ‘fuels’ (less than 30% fat), preventing specific nutrient deficiencies, treating bacterial growth and preventing recurrent sepsis. Ursodeoxycholic acid administration may be beneficial.

Metabolic complications in SBS include hypocalcemia, hypomagnesemia and fat soluble vitamin deficiencies. A specific problem is D-lactic acidosis, which results from bacterial fermentation of unabsorbed nutrients particularly simple sugars. The diagnosis is suggested by unexplained metabolic acidosis and associated neurological symptoms. Treatment includes minimizing overall caloric intake or instituting a low carbohydrate diet. Administration of intestinal antibiotics may be appropriate.

Cholelithiasis occurs in 30–40% of patients with intestinal insufficiency.[27] Factors that predispose to gall stone formation include altered hepatic bile metabolism and secretion, gall bladder stasis and malabsorption of bile acids. Long-term TPN is an important contributing factor. The risk for cholelithiasis increases significantly if less than 120 cm

of cholelithiasis can be minimized by providing enteric nutrition whenever feasible. Cholelithiasis among patients on TPN can be prevented by intermittent cholecystokinin injections and administration of intravenous lipids both of which prevents gall bladder stasis. Several authors recommend prophylactic cholecystectomy in these patients when laparotomy is undertaken for other reasons.[28]

Calcium oxalate stones are formed as a result of increased oxalate absorption from the colon.[28] Nephrolithiasis is more common among patients with an intact colon and can be prevented by maintaining the patient on a diet low in oxalate, minimizing intra luminal fat, supplementing diet with calcium orally and maintaining a high urinary volume. Cholestyramine which binds to oxalic acid in colon is another potential agent, which can be used for treatment.

Gastric hypersecretion can be a serious problem in SBS and is due to parietal cell hyperplasia and hypergastrinemia. In addition to malabsorption and diarrhea, gastric hypersecretion can cause or flare up peptic ulcer disease. H2 receptor antagonists or PPI can be tried with good results. Few intractable cases may need surgical intervention. A highly selective vagotomy may be the most desirable procedure if feasible.[29]

Bacterial overgrowth can occur among patients with SBS. Causes include impaired intestinal motility, stasis and achlorohydria. Bacterial overgrowth results in impaired bile absorption, vitamin B12 deficiency and diarrhea and may require long-term administration of intestinal antibiotics. Various drugs are used to treat or control the complications of SBS and are outlined in .

Commonly used drugs in short bowel syndrome

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SURGICAL MANAGEMENT

The primary goal of surgical therapy for SBS is to increase the intestinal absorptive capacity and can be achieved by:

Preserving the existing intestine

An abdominal reoperation is required in about half of the patients with SBS. Intestinal problems are the most common indications.[30] The strategy in such a reoperation should be to avoid resection and preserve the existing length of intestinal remnant. The procedures that can be employed as alternatives to resection in such instances include (1) stricturoplasty for benign strictures and (2) serosal patching for certain strictures and chronic perforations. When resection becomes unavoidable, an end to end anastomosis is preferred to prevent blind loops and maximize functional length of intestine.

Improving the intestinal function

The functioning of existing intestine can be enhanced by improving the motility and slowing the intestinal transit.

Improving the intestinal motility

The motility of intestinal remnant in SBS deteriorates over a period of time due to dilatation of intestine. This dilatation could be due to chronic unresolved obstruction or intestinal adaptation. All attempts should be made to relieve any obstruction. As the dilated segment cannot generate sufficient intraluminal pressures during peristalsis, it should be narrowed. This procedure is called “tapering enteroplasty”. The preferred methods of tapering enteroplasty are (1) simple imbrication of redundant bowel and (2) longitudinal transection and removal of part of the circumference of intestine along the antimesenteric border. Tapering enteroplasty does improve intestinal function in patients with SBS.[31]

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