Current Management of Short Bowel Syndrome

Intestinal failure refers to a condition that results from obstruction, dysmotility, surgical resection, congenital defect, or disease-associated loss of absorption and is characterized by the inability to maintain protein-energy, fluid, electrolyte, or micronutrient balance.[1] The short bowel syndrome (SBS) is a type of intestinal failure caused by intestinal resection leading to a shortened intestinal remnant and is characterized by the inability to maintain protein-energy, fluid, electrolyte, or micronutrient balances when on a conventionally accepted, normal diet. SBS accounts for approximately three-fourths of intestinal failure patients in adults and more than one half in children. The pathophysiologic changes that occur in SBS relate primarily to the loss of intestinal absorptive surface and more rapid intestinal transit. The consequences of malabsorption of nutrients include malnutrition, diarrhea, steatorrhea, specific nutrient deficiencies, and fluid and electrolyte imbalance. These patients are at risk for other specific complications, which include an increased incidence of cholelithiasis, gastric hypersecretion, nephrolithiasis, and liver disease.

The history of SBS is one of long-standing interest but more recent advancements. Koeberle[2] reported the first patient surviving massive resection of the small intestine in 1880. The clinical consequences of diarrhea and malabsorption were described by Senn in 1888.[3] Mall[4] reported using reversed intestine segments to improve these symptoms in 1896. Functional adaptation after massive resection was well documented by Flint[5] in 1912. In 1935, Haymond[6] reviewed 257 cases of extensive (>8 feet resected) intestinal resection. He found that only 50 (20%) survived for more than 1 year and further suggested that loss of 50% of the intestine was the upper limit of safety. Simons and Jordan[7] reported 90 SBS patients (<4 feet remaining small intestine) in 1969. Mesenteric vascular disease was the most common diagnosis and mortality remained high. An important milestone was the demonstration by Wilmore and Dudrick in 1968 that parenteral nutrition (PN) would support nutritional

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status in SBS patients.[8] The development of home PN in 1970 revolutionized the long-term care of SBS patients and this modality rapidly became standard therapy.[9,10] More recently, the importance of optimal diet and potential pharmacologic therapy was introduced by Byrne and colleagues,[11] leading to the concept of intestinal rehabilitation. In 1990, Grant and colleagues[12] reported the first long-term survivor of a small bowel transplant who achieved enteral autonomy, ushering in a new avenue of treatment for SBS.

The incidence and prevalence of SBS are estimated to be 3 per million and 4 per million, respectively.[13] Thousands of patients are now surviving with SBS.[14] This condition occurs in approximately 15% of adult patients undergoing intestinal resection, with three fourths of these cases resulting from massive intestinal resection and one fourth from multiple sequential resections.[15] Massive intestinal resection continues to be associated with significant morbidity and mortality rates, which are related primarily to the underlying diseases necessitating resection.[15,16] Approximately 70% of patients who develop SBS are discharged from the hospital.[16] The overall 5-year survival is 75% for those leaving the hospital.[17] This improved survival rate has been achieved primarily by the ability to deliver long-term nutritional support. The overall outcome of these patients is often determined not only by their age and underlying disease but also by complications related to management of SBS.

SBS has been the topic of previous issues of *Current Problems of Surgery*. In 1971, Wright and Tilson highlighted the pathophysiology of SBS as management options were limited at that time.[18] Wilmore and colleagues[19] in 1997 emphasized new therapeutic approaches, particularly promising pharmacologic agents. This led to the concept of intestinal rehabilitation and the development of multidisciplinary teams. The current monograph reviews recent advances in our understanding of pathophysiology, medical management, and surgical therapy, especially intestinal transplantation.

Etiology of the Short Bowel Syndrome

A variety of conditions requiring intestinal resection lead to SBS ( ). The causes of SBS vary by age group. In infants, necrotizing enterocolitis is the most common cause of SBS, followed by intestinal atresia, gastroschisis, and midgut volvulus.[20] In older children, postoperative SBS

TABLE 1. Causes of the short bowel syndrome

Infants	
	Necrotizing enterocolitis
	Intestinal atresia
	Gastroschisis
	Midgut volvulus
Children	
	Cancer
	Postoperative complication
	Trauma
	Motility disorders
Adults	
	Postoperative complications
	Irradiation/cancer
	Mesenteric vascular disease
	Crohn's disease
	Trauma
	Other benign causes

motility disorders requiring resection are also potential mechanisms for SBS in this age group.

The etiology of SBS in adults may be changing. We have found that postoperative SBS, resection performed for complications of previous abdominal operation, has become the most common cause in adults.[21] This occurs after both open and laparoscopic procedures. Although this may be related to infarction secondary to vascular injury, volvulus, or hypotension, it most commonly occurs because of intestinal obstruction. Mesenteric ischemia remains an important predisposing factor. Mesenteric vascular disease is the common mechanism but ischemia secondary to drug abuse and hypercoagulable disorders is increasingly found. Malignancy, with or without radiation treatment, also accounts for a significant number of cases. Crohn's disease remains responsible for a significant number of SBS patients but may be declining with less aggressive resective therapy. Resection for trauma and other benign conditions, such as volvulus and intestinal pseudoobstruction, are other potential causes for SBS.

Prevention of the Short Bowel Syndrome

Prevention of SBS is an important consideration given the morbidity and mortality associated with long-term treatment. Efforts at prevention can be divided into 2 periods: preoperative and intraoperative.

Preoperative strategies to prevent SBS are primarily related to the patient's underlying diagnosis. Postoperative SBS could be minimized by

diagnosing intestinal ischemia in a timely fashion, and approaching the frozen abdomen cautiously.[21] Patients undergoing bariatric procedures should have mesenteric defects closed to prevent internal hernias and this diagnosis should always be entertained when these patients experience abdominal pain.[22] Unsuspected intestinal ischemia is increasingly recognized as a complication of laparoscopic procedures.[23] Intestinal ischemia from mesenteric vascular disease and hypercoagulability must be diagnosed in a timely fashion. This may permit attempts at revascularization. Intestinal viability should be carefully assessed intraoperatively and second-look procedures should be used judiciously.[24] Radiation enteritis can be reduced by minimizing bowel exposed to radiation.[25] Errors in diagnosis, aggressive resectional therapy, and postoperative complications contribute to SBS in patients with Crohn's disease.[26,27] SBS can be minimized in trauma patients by early diagnosis of vascular injuries, use of second-look procedures, and appropriate resuscitation.[28] Bowel-preserving strategies are now being used in infants with necrotizing enterocolitis, which may decrease the incidence of SBS because of this condition.[29]

There are several intraoperative strategies to prevent SBS.[30] The extent of resection should always be minimized where possible. Tapering or lengthening dilated segments and use of stricturoplasty in conditions such as Crohn's disease can obviate the need for resection. Avoiding extensive enterolysis and using cautious resection can prevent SBS in patients with extensive adhesions from conditions, such as radiation enteritis.[25]

Factors Influencing Outcome

The clinical manifestation of SBS varies greatly among patients, depending on intestinal remnant length, location, and function; the status of the remaining digestive organs; and the adaptive capacity of the intestinal remnant (Fig 1). More recently the importance of other patient-related factors has also been recognized, including age, diagnosis, and body mass index.[16,31] Thus, although intestinal length is important, the outcome of SBS is not entirely dependent on a given length of remaining intestine.

Intestinal remnant length is the primary determinant of outcome in SBS. The length of the small intestine reported in adults varies between 12 and 20 feet (360-600 cm), depending on how it is measured and the height and sex of the individual.[32-34] The duodenum measures 10-12 inches (25-30 cm). The length of the small intestine from the ligament of Treitz to the

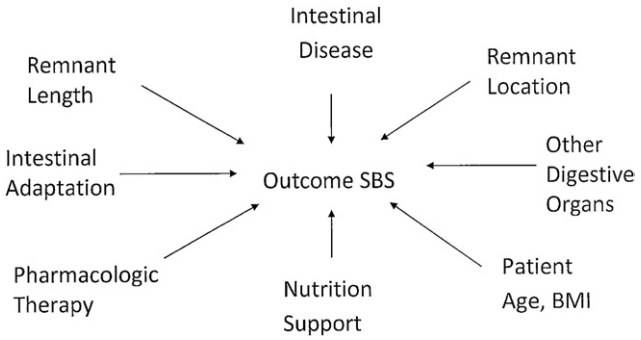

FIG 1. Factors affecting the outcome of short bowel syndrome.

proximal two fifths being jejunum and the distal three fifths being ileum. Resection of up to one half of the small intestine is generally well tolerated. Although adult patients with less than 180 cm of small intestine, approximately one third the normal length, may develop SBS, permanent PN support is likely to be needed in patients with less than 120 cm of intestine remaining without colon in continuity and less than 60 cm remaining with colonic continuity.[17,35]

In infants, the intestinal length is 125 cm at the start of the third trimester and 250 cm at term. The length then doubles again as the child reaches adulthood.[20,36] Children with less than 75 cm small intestine may develop SBS. Children are further classified as short small bowel (>38 cm), very short small bowel (15-38 cm), and ultrashort small bowel (<15 cm).[20] These categories predict those that are most likely to become PN-independent and survival vs need for PN and mortality.

The site of resection is also an important factor. Patients with an ileal remnant generally fare better than those with a jejunal remnant. The ileum has specialized absorptive properties for bile salts and vitamin B_{12}, unique motor properties that prolong transit time, a hormone profile different from the jejunum, and a greater capacity for intestinal adaptation.[37,38] L cells in the terminal ileum secrete several hormones that might influence appetite, gastrointestinal motility, intestinal absorption, and intestinal adaptation, including peptide YY, neurotensin, and glucagon-like peptides 1 and 2 (GLP-1 and GLP-2). The presence of the ileocecal junction improves functional capacity of the intestinal remnant.[38] Although previously this had been largely attributed to a barrier function and transit prolonging property of the

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