The Surgical Management of Short Bowel Syndrome

Edward M. Barksdale, Jr, MD and Ala Stanford, MD

Address
Intestinal Care Center, Department of Pediatric Surgery, Children's Hospital of Pittsburgh, University of Pittsburgh School of Medicine, 3705 Fifth Avenue, 4A-485, Pittsburgh, PA 15213, USA.
E-mail: Barkem@chp.edu

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Short bowel syndrome is a clinical entity that results from a diverse group of congenital and acquired conditions in the pediatric population. The pathophysiology of this syndrome is characterized by malabsorption, malnutrition, and metabolic disturbances. The vast majority of children with this condition undergo spontaneous adaptation of the intestinal remnant and achieve enteral nutritional autonomy. However, a small portion of pediatric patients develop intestinal failure and require long-term or permanent dependence on total parenteral nutrition. These children may benefit from surgical interventions that facilitate intestinal adaptation. Such adjunctive procedures facilitate nutrient absorption by improving motility, prolonging intestinal transit, and/or increasing mucosal contact time. In selected patients, this may allow them to be weaned from parenteral nutritional support or to have it discontinued. The purpose of this review is to discuss the various surgical techniques for the management of short bowel syndrome in children, along with their indications, complications, and outcomes.

Introduction

Short bowel syndrome (SBS) is an imprecisely defined clinical condition characterized by the malabsorption and malnutrition associated with extensive intestinal loss or dysfunction. Despite attempts to characterize this syndrome on the basis of intestinal length, there appears to be no minimum small bowel length that defines SBS. The fundamental defect is the lack of adequate mucosal surface area to achieve enteral nutritional autonomy. The resulting malabsorption of protein, fats, carbohydrates, micronutrients, fluids, and electrolytes is referred to as SBS. Although patients with SBS undergo a physiologic compensatory response following resection, leading to improved nutrient absorption, they also develop pathophysiologic consequences to intestinal loss that include gastric hypersecretion, intestinal dysmotility, biliary lithiasis, liver disease, osteomalacia, nephrolithiasis, and metabolic disturbance [1••,2••,3••,4••]. In children, growth failure and cognitive impairment may be additional long-term sequelae [5,6]. These disorders may have a significant negative impact on the morbidity and mortality of patients with SBS. Physicians who manage the care of patients with SBS are therefore in a race to promote adaptation while limiting side effects of this multifaceted disease. Surgery may be an important adjunctive strategy in the coordinated care of these complex patients.

In the pediatric population, the causes of SBS are primarily related to neonatal conditions such as necrotizing enterocolitis, intestinal atresia, midgut volvulus, and aganglionosis. Crohn's disease and trauma are more common causes in older children and teenagers [1••,3••]. Intestinal pseudoobstruction, microvillus inclusion disease, and other mucosal abnormalities may produce SBS pathophysiology in the setting of normal intestinal length [7]. The adult small bowel length ranges from 350 to 600 cm, whereas the normal small bowel length in term infants ranges from 200 to 250 cm. The last trimester of pregnancy is associated with a doubling of intestinal length, which may be a significant factor in the prognosis of premature infants with SBS [8,9]. Although many studies suggest that term infants with greater than 20 cm of small bowel and an ileocecal valve (ICV) or 40 cm of bowel without the ICV can be successfully weaned from parenteral support [2••,10••,11]. Our recent long-term review of over 250 patients within the Intestinal Care Center at Children's Hospital of Pittsburgh suggests that a length of 40 to 60 cm of normally functioning small bowel is necessary to achieve long-term nutritional autonomy. Anecdotal reports chronicle the survival of pediatric patients with ultrashort (<20 cm) segments of small bowel; however, longitudinal follow-up on these patients beyond 3 to 5 years of life has not been reported [12–14]. Although length of the bowel is an important criterion for prolonged survival, the outcome of patients with SBS is not solely determined by length. In fact, the prognosis in this syndrome is primarily determined by the interplay of six major factors: the extent of intestinal resection; the anatomic site of resected intestine; the presence or absence of an ICV; the functional capacities of remaining small and large bowel; the functional adaptations occurring in the remaining small and large bowel; and the functional capacities of the liver, gallbladder, and pancreas [1••,2••,15].

Major advances in the medical management of SBS during the last three decades have led to prolonged survival and diminished complications. More than 75% of children undergo spontaneous adaptation following massive resection with medical management alone [2••,4••]. This management involves meticulous attention to the details of fluid, electrolyte, and nutritional care. Currently, surgery is primarily reserved for those patients who fail to achieve adequate spontaneous adaptation, develop life-threatening complications, or have ultrashort (inadequate) initial bowel lengths and are unable to be weaned from total parenteral nutrition (TPN). The purpose of this report is to review the process of intestinal adaptation; to summarize the various operative strategies available to treat pediatric patients with refractory SBS; and to present an algorithm for the prospective management of children with this syndrome.

Intestinal Adaptation

Following extensive small bowel resection, the intestinal remnant undergoes an adaptive response that results in morphologic and physiologic changes that lead to improved nutrient absorption. At the ultrastructural level, the small bowel mucosal changes are characterized by increases in villus height, crypt depth, enterocyte number, and enterocyte proliferation. Coincident with these ultrastructural alterations is the development of luminal dilation, intestinal lengthening, and altered motility. Consequently, the absorption of nutrients per unit area of mucosa increases dramatically over baseline levels. This intricately coordinated process, known as intestinal adaptation, begins within 48 hours of surgery, but it may take from 1 to 2 years to complete [16–19].

Intestinal adaptation is under the complex regulation of pleiotropic humoral factors (*eg*, glucagon, insulin-like growth factor-1, neurotensin, and peptide-YY) and intraluminal factors (*eg*, gastrin, cholecystokinin, neurotensin, polyamines, epidermal growth factor, and ornithine ketoglutarate). However, the most important stimulant to intestinal adaptation is exposure of the intestinal lumen to enteral nutrients [16,17,19]. This fact increases the significance of timely surgical intervention as a component of the proadaptive therapeutic management plan. The goal of therapy during this period is to maximize enteral absorption of nutrients by decreasing gastrointestinal secretions and controlling diarrhea. Combined enteral and parenteral nutritional support and the judicious use of pharmacologic agents to delay intestinal transit have become the mainstay of treatment during this phase. The role of surgery in patients undergoing adaptation is primarily adjunctive. Major interventions should be reserved for the treatment of significant surgical complications, or the provision of intravenous or enteral access that may promote adaptation. In fact, surgery may be unnecessary and could delay the adaptation process [3••,19,20]. However, certain patients who do not achieve adequate adaptation to meet their metabolic needs require long-term parenteral nutritional support. These patients have intestinal failure and are candidates for early surgical intervention. The consensus of most authors is that surgery in SBS should not occur for at least 1 year or until intestinal adaptation is complete. The primary strategic goals of surgical intervention in SBS are to optimize intestinal function, slow intestinal motility, and increase mucosal surface area.

Strategies to Optimize Intestinal Function

Restoration of intestinal continuity

Many patients who undergo surgery for acute, life-threatening conditions are left with proximally diverting stomas [3••]. At the initial intervention, the presence of severe associated conditions, hemodynamic instability, or significant peritoneal contamination may mandate stoma placement as the most expeditious and prudent strategy. The restoration of gastrointestinal continuity is a critical factor in promoting the adaptive response after resection [2••,3••,4••,18]. The principal advantage of stoma closure is that it makes the entire mucosal surface area available for nutrient absorption. The distal defunctionalized segments of small bowel undergo mucosal atrophy. Reestablishing the flow of intraluminal nutrients and trophic factors facilitates absorption and adaptation in this segment of bowel [3••,16]. Moreover, the colon, if present, may contribute directly to the patient's nutritional status by its capacity to absorb short-chain fatty acids (SCFA). Carbohydrates that reach the colonic mucosa undergo bacterial fermentation to the SCFA, acetate, butyrate, and propionate. These molecules can be directly absorbed by the colonic mucosa and enter the portal circulation, where they are used as a source of caloric substrate. SCFA also stimulate water and sodium absorption from the colon, which may ameliorate the fluid and electrolyte losses associated with SBS. Further data also suggest that SCFA may be trophic to the colonic mucosa. The proadaptive and nutritional roles of SCFA in SBS were underappreciated until recently. In addition, the increased surface area for intestinal absorption will slow intestinal transit by improving absorption [4••,21].

There are, however, some disadvantages to early stoma closure. Significant fluid shifts, secretory diarrhea, perineal complications, and dietary restrictions are common. The increased delivery of bile salts and acids to the colon may stimulate steatorrhea and excess fluid losses. The irritant nature of this diarrhea may result in significant cutaneous perineal irritation [21]. Nguyen *et al*. [23] reported that approximately 60% of their patients without a stoma had major dietary restrictions, compared with 33% of patients with a stoma. Also, there is a significant risk of calcium oxalate renal stones in those patients with colon in continuity. Reliable preoperative evaluation of which patients should undergo stoma closure is not currently available. Often, however, there is no way of gauging the benefits of early stoma closure without performing the procedure in

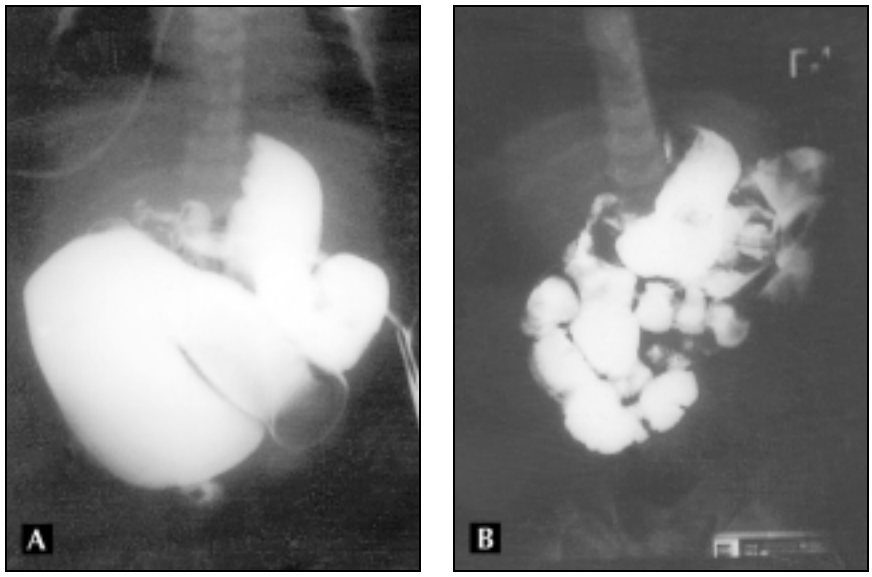

Figure 1. Upper gastrointestinal contrast studies demonstrate compensatory small bowel dilation in a child with SBS (**A**), and appearance following Bianchi bowel lengthening in the same patient (**B**). Note decreased caliber and increased length of the small bowel.

these patients. Attempts at distal reinfusion of stomal effluent into a mucous fistula, when present, or measurement of the polyamine content of the enterostomy effluent have been used to evaluate outcome but with poor reliability [21]. At our institution, we have found that patients with greater than 40 cc/kg/d of stomal effluent and more than 30% colon resections often fare poorly due to significant perineal complications and fluid and electrolyte disturbances. Our practice is to close stomas promptly once the patient's metabolic, physiologic, and anatomic state will tolerate it. Patients with 1° motility disorders (intestinal pseudoobstruction) often achieve no benefit from stoma closure.

Stricturoplasty or segmental bowel resection

Occasionally patients develop areas of intestinal narrowing or stenosis secondary to late ischemic effects, as seen in necrotizing enterocolitis, midgut volvulus, or Crohn's disease. These stenotic regions lead to bowel dilation and associated intestinal dysmotility. Bacterial overgrowth may predominate proximally, and mucosal atrophy may be characteristic distally. These stenotic areas of bowel should be revised either by stricturoplasty, in patients with limited length, or by limited resection in patients with adequate bowel length [3••].

Strategies to Improve Intestinal Motility

Intestinal plication and tapering

The natural proadaptive response of the small bowel following massive resection is to undergo segmental dilation (Fig. 1). This dilation is most pronounced in the ileum [18]. Occasionally these dilated segments have disordered peristalsis leading to stasis, bacterial overgrowth, and malabsorptive diarrhea. DeLorimier *et al.* [24] first characterized this observation fluoroscopically. They demonstrated that dilated bowel had diminished peristaltic pressures that resulted in poor prograde flow of luminal contents and stagnation. Antibiotic therapy using metronidazole alternating with colistin is often useful in these children. Patients with massive dilation, however, may develop refractory bacterial overgrowth despite antibiotic therapy and require surgical intervention. Surgical procedures designed to taper or resect segments improve functional absorption of nutrients, although some mucosal surface area may be lost. This fact, however, limits the utility of segmental resection in those SBS patients with very short segments of bowel.

DeLorimier and Harrison [25] proposed an intestinal plication technique to reduce the diameter of the bowel wall without compromising mucosal surface area. In this technique, the bowel is folded into the lumen, and the antimesenteric serosal surfaces are imbricated. This procedure results in mucosal preservation and improved absorption, while simultaneously reducing the diameter of the bowel. Unfortunately, these plicated segments are prone to breakdown. Subsequently, bowel dilation and obstruction may recur. This technique has been modified (by Kimura, refer to section titled "Methods to Increase Mucosal Surface Area") through performance of antimesenteric serosal stripping at the site of bowel wall imbrication to prevent such breakdown [1••,2••,18].

An alternative to intestinal plication is tapering enteroplasty. This procedure involves partial resection of the antimesenteric border of the dilated bowel, preserving the mesenteric portion as a tubularized segment of smaller diameter. This may be accomplished in either a hand-sewn manner or by a gastrointestinal anastomosis (GIA) stapling device. Blood supply to the remaining tubularized antimesenteric border is preserved (Fig. 2). This approach is a viable surgical alternative for treating segmental bowel dilation as seen in jejunoileal atresias [18,21,25].

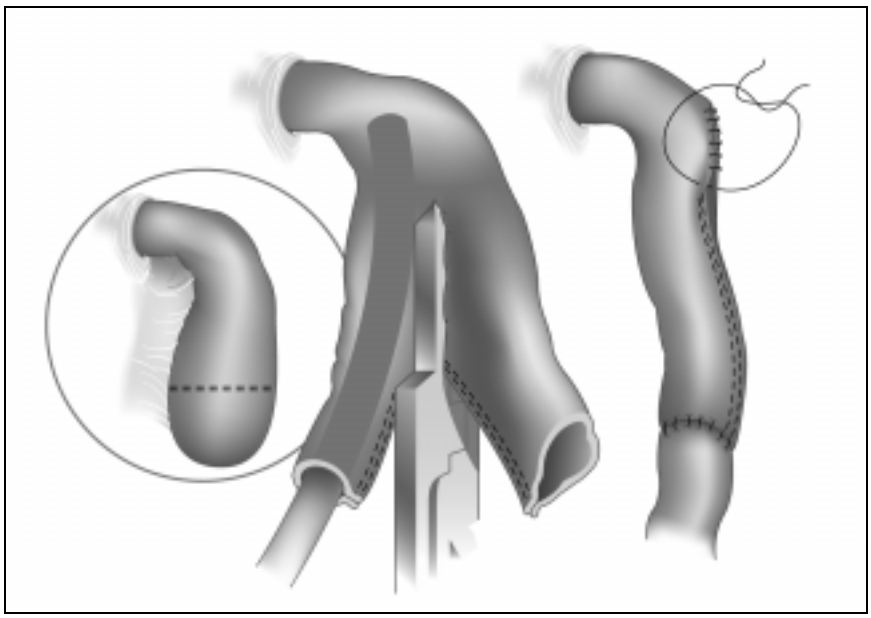

Figure 2. Tapering enteroplasty is shown.

Strategies to Slow Intestinal Motility

Artificial intestinal valves and sphincters

Recognition of the prognostic significance of the ICV by Wilmore and others in the late 1960s and early 1970s led to the proliferation of interest in the construction of a neovalve for treatment of patients with refractory SBS or intestinal failure [10••,15,18]. The ICV serves both as a one-way barrier to the retrograde reflux of colonic contents and bacteria into the ileum and a "brake" to delay distal intestinal motility. Malfunction of this valve leads to both intestinal dysmotility and bacterial backwash. The transient reflux of bacteria into the small bowel may further potentiate the malabsorption associated with SBS [18]. Animal studies demonstrate delayed transit, improved absorption, and augmented growth following nipple valve construction. In addition to these improvements, such valves have also been demonstrated to increase intestinal crypt cell proliferation and mucosal thickness.

Although numerous operations have been devised to create a neo-ICV that will slow intestinal motility and eliminate colonic small bowel reflux, none appears to have outstanding effectiveness. Some of these procedures include submucosal tunneling of the proximal bowel into the distal bowel lumen; serosal muscular stripping of a circumferential proximal bowel segment followed by suturing of the muscular layers; and placement of a prosthetic cuff around the circumference of the bowel followed by intussusception of the proximal segment of bowel into the distal segment [2••,3••,15].

Numerous technical challenges may arise in the construction of a neovalve. Many of these procedures lead to either functional bowel obstructions or significant alteration in motility. Considerable patient-to-patient variability has been reported with regard to the optimal valve length necessary to retard transit without inducing a functional bowel obstruction. It appears that the maximum valve length in children should be less than 3 cm. Most recently, Georgeson *et al*. [26] have used valve construction in conjunction with bowel lengthening procedures to perform sequential bowel elongation.

Reversed (antiperistaltic) small intestinal segments

During the late 1960s and early 1970s, extensive experience was gained with the use of reversed segments of small bowel to impede intestinal peristalsis in patients with diarrhea following vagotomy [3••]. Numerous anecdotal reports suggested a potential role for this procedure in patients with SBS. In principle, the antiperistaltic segment serves as a physiologic brake that delays the increased intestinal transit associated with SBS by competing with or altering the proximal-to-distal conduction of the normal myoelectric pattern of peristalsis [3••,15]. Priebe [27] reported the potential utility of reversed segments in beagle puppies that had undergone extensive bowel resection. Although these studies demonstrated short-term increases in water, nitrogen, and fat absorption, sustained responses beyond 7 months were not observed. Antiperistaltic segments have been clinically applied with considerable variability of response. Favorable responses have been reported in more than 70% of adult patients treated with this approach; however, there has been a predominance of failure in the pediatric patients. In a similar manner to the animal studies, initial improvement is followed by the lack of a sustained effect. Although numerous reports suggest a benefit in human studies, no long-term follow-up data are available at the present time [1••,2••,3••,4••,18].

The variability of response to this procedure in part may reflect the lack of uniformity in length of reversed segments in the patients. Recent developments reveal that the ideal length of reversed segment of small bowel appears to be

approximately 10 to 15 cm of distal bowel segment in the adult or 3 cm in the infant. The goal of this procedure is to provide short-term improvement in intestinal nutrient absorption while adaptation is occurring [18]. This technique is primarily an adjunctive strategy used to accelerate adaptation. It has limited utility for the long-term management of patients with SBS.

Colonic Interposition

Recognition of the segmental irregular peristaltic activity of the colon has led to the proposition that a colonic segment interposed between two lengths of small bowel might lead to improved nutrient and electrolyte absorption. Hutcher *et al*. [28] first studied this possibility in a canine model of SBS with a 90% small bowel resection. The experimental animals that underwent interposition alone developed delayed peristalsis, improved weight gain, less stool output, and decreased stool fat excretion, compared with the control group. Lloyd [29] demonstrated that isoperistaltic colonic interposition segments prolonged intestinal transit without creating obstruction. Clinical application of this procedure in children has been favorable but highly variable. Several groups have reported improved nutrient absorption and ability to wean children with SBS from parenteral support [30,31]. The principal complications of this procedure appear to be the development of bowel obstruction and eosinophilic colitis in the colonic segment [18]. The overall procedure-related morbidity and mortality appear to be low in these patients. The use of antiperistaltic segments has been reported, but with less consistency in results [3••].

Reversed Electrical Intestinal Pacing

Reversed electrical intestinal pacing may be a plausible treatment alternative for managing the hyperperistalsis seen in SBS. This concept for treatment emerged as an adaptation of the principle that the heart could be paced in patients with irregular rhythms [32]. Control of the bowel pacemaker in SBS patients with hyperperistalsis would function to slow transit time. The motor or pacemaker mechanism in the bowel is thought to originate in the duodenum. The impulse migrates distally to the ileum, inducing a coordinated propulsive contraction. Through manipulation of the intestinal pacemaker, intestinal transit may be slowed, subsequently increasing nutrient and water absorption by prolonging the mucosal exposure to luminal contents. This manipulation of the proximal pacemaker appears to require duodenal transsection.

British investigators developed a complex canine model to study the role of intestinal pacing in the 1970s. Following extensive bowel resection, duodenal division, translocation, and retrograde electrical pacing, they found that retrograde pacing increased absorption of water, glucose, and sodium while decreasing potassium excretion in dogs with SBS [33,34]. Similar procedures were performed in other animal models with analogous results [35,36]. However, this approach has not been employed in humans because of its anticipated high morbidity and mortality rates. Alternative techniques using distally placed electrodes without bowel transsection provide a more reasonable strategy for pacing. Evidence also suggests that pacing in humans is temporary and does not persist long term [32].

More recently, investigators at the Lynn Institute for Health Care Research in Oklahoma have shown entrainment of the intestinal slow waves in dogs [37]. Four pairs of intraluminal intestinal electrodes were placed in the jejunum of the animals, and intestinal slow waves were recorded. Intestinal pacing was achieved in seconds in this model. These authors conclude that intestinal pacing is an effective and efficient method to entrain slow waves and a potential approach for treatment of intestinal motor disorders associated with myoelectrical abnormalities [37]. These methods await clinical trial in humans.

Methods to Increase Mucosal Surface Area

Neomucosa

Neomucosa was first described as distinct histologic changes arising from intestinal defects repaired with serosa from normal bowel. The mucosa adjacent to an intestinal defect proliferated and lined the serosa, which had formed the new bowel wall. Lillemoe *et al*. [38] first studied this phenomenon in the 1980s. They found that mucosa would develop on distal ileal defects that were patched with a skeletal muscle flap. Furthermore, this neomucosa adherent to the muscle was similar to normal ileal mucosa by microscopic and electrophysiologic criteria. Further experimental work by Thompson [39] extended these observations in a canine model. Intestinal patching resulted in the ingrowth of neomucosa and slowed transit time but had deleterious effects on absorption and nutritional status. Thompson [39] suggested that this effect might be secondary to inhibition of intestinal adaptation and gastric hypersecretion in patched animals. Further studies in rabbits combined lengthening procedures with patching to slow transit time and generate neomucosa, with quite favorable outcome [40].

Choi *et al*. [41] and Kaihara *et al*. [42] have developed tissue-engineered intestine. This tubular structure, composed of a microporous biodegradable polymer, was "seeded" or inserted with rat intestinal epithelial units for several weeks. After seeding, the polymer was implanted into the abdominal cavity of adult male rats following 75% small bowel resection. Three weeks after implantation, the unit polymer constructs were anastomosed to jejunum in a side-to-side fashion. These anastomosed segments achieved a high patency rate for up to 36 weeks. The tissue-engineered intestine increased in size and was lined with well-developed neomucosa for the duration of the study. Although histologic analysis showed formation of neomucosa, characterized by columnar epithelium with goblet and Paneth cells, and the

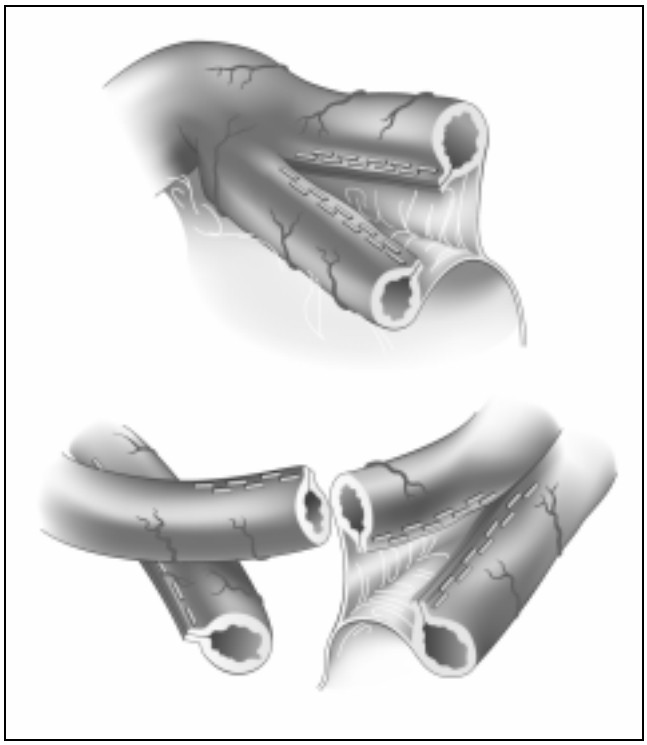

Figure 3. The Bianchi bowel lengthening procedure (isoperistaltic) is depicted.

electrophysiology of neomucosa measured by Ussing-chamber exhibited similar transepithelial resistance as adult ileal mucosa, the absorptive capacity of nutrients and water has not yet been demonstrated. This model and technique show great promise for the future treatment of SBS but currently remain on the horizon of care.

Autologous bowel lengthening procedure (Bianchi)

The fundamental problem in SBS is the lack of mucosal surface area to provide sufficient intestinal nutrient absorption. The Bianchi autologous intestinal lengthening technique exploits the hemicircumferential segmental blood supply of the intestine to allow a segment of dilated small bowel to be doubled in length [43]. The mesenteric blood vessels bifurcate immediately prior to their junction with the bowel wall, allowing for the longitudinal division of the bowel into two lumens of half the circumference based on the bifurcated blood supply. These two new lengths of intestine can be sewn together in an end-to-end fashion to create two isoperistaltic segments (Fig. 3). Consequently, the bowel is both lengthened and, probably more importantly, tapered [2••,16,19,44]. Following several months of readjustment, delayed peristalsis results in improved nutrient absorption and decreased parenteral nutrient requirements. Numerous authors have reported short-term studies demonstrating significant improvement in intestinal function following the Bianchi procedure, but these reports have not been standardized for length of residual bowel or underlying cause of the SBS. Thompson *et al*. [44] reported on 16 children ranging in age from 2 months to 19 years with intestinal remnant length of 23 to 120 cm. They found that, at 1 and 5 years, more than 40% of their patients were on enteral nutrition alone. This procedure was also associated with decreased episodes of bacterial overgrowth. Other authors have demonstrated that, following an initial decline in function with the Bianchi procedure, the stool frequency decreases dramatically [16,29]. Complications following this procedure are not uncommon, with adhesive small bowel obstruction and anastomotic strictures occurring more frequently. Long-term recurrent dilation of the lengthened segments has been noted by others. This problem may be treated with tapering enteroplasty or with repeated lengthening procedures [16].

Kimura procedure (isolated bowel segments)

An alternative technique of intestinal lengthening is the Kimura procedure, which utilizes the parasitization of blood supply from the abdominal wall or liver by the antimesenteric surface of the bowel, followed by transverse division of the bowel into two lumen [45,46]. One of these segments of bowel has a blood supply that is isolated from the mesentery. This staged procedure initially involves an antimesenteric serosal stripping of the dilated intestinal segment, followed by its suture to abdominal wall or liver that has been deperitonealized. Over the ensuing few months, collateral blood supply develops, allowing horizontal division of the bowel into two parallel structures that can be anastomosed. This procedure is particularly useful in situations in which the mesentery is very short or tethered, limiting the ability to perform a Bianchi division [44,45]. Many critics of this procedure emphasize the potential lack of portal nutrient flow; however, this does not appear to have been problematic in patients who underwent this procedure. Other critics of this procedure note that the extensive dissection and ectopic placement of the bowel makes subsequent operations more difficult. No large series has been reported with the Kimura technique as the primary approach to care.

Sequential autologous lengthening procedures (Georgeson)

Georgeson *et al*. [26] have developed a technique of sequential bowel lengthening that employs aspects of several remedial operations for SBS to be used in patients with intestinal failure. In those patients who have an intestinal remnant of normal diameter, they induce intestinal dilation by constructing a valve. This promotes bowel dilation and increases in mucosal mass, which, in a few patients, may facilitate spontaneous adaptation. Following 3 to 9 months of progressive intestinal dilation, a Bianchi procedure is performed. This operation is associated with a significant increase in motility and absorption. In a few patients with very short lengths of bowel, a subsequent Kimura procedure is performed to lengthen the bowel further [46]. Bianchi [47] has reported similar modifications of this approach using composite gastrointestinal serosal

(mucosectomized) patch grafts in conjunction with autologous bowel lengthening to further increase the functional absorptive area of the bowel in patients with ultrashort segments of bowel. Long-term follow-up on these patients has not yet been reported, but preliminary data are encouraging.

Surgical Management

Although general consensus suggests that surgery should not be performed in the first year after resection, precise timing and indications for surgery after this time remain elusive. The principal indications for surgical intervention in children with SBS are the failure to progress in efforts to wean the patient from TPN, and the presence of life-threatening complications such as TPN-related liver disease, recurrent central line sepsis, and limited venous access. At our institution, we have evaluated and followed more than 250 pediatric patients with SBS since 1997, many of whom have intestinal failure. This experience has led us to develop an early aggressive posture not only for patients with medically refractory SBS or evolving complications, but more importantly during the first year of life in those patients with ultrashort segment disease. We define ultrashort bowel syndrome as an intestinal remnant of less than 10% of the expected bowel length with an ICV, or less than 20% without an ICV. These patients, in our view, may often benefit from adjunctive proadaptive surgery such as stoma closure, bowel resection, tapering, or lengthening procedures in the first year, especially if there is significant intestinal dilation and dysmotility. The choice of operation in SBS is influenced by three principal factors: intestinal remnant length, intestinal function, and caliber of the intestinal remnant.

Prior to surgery, a thorough assessment of the patient's underlying medical status, intestinal anatomy, and associated medical conditions should be made. End-stage liver disease and coincident coagulopathy are major considerations that would preclude palliative surgical procedures. Strong consideration should be given to referral for intestinal transplantation in these patients. Review of previous operative notes and recent bowel radiographs is critical in the strategic planning for rehabilitative bowel surgery. This is particularly important if the surgeon for this procedure did not perform or participate in the previous procedures. Immediately prior to surgery, we obtain a full gastrointestinal series with small bowel follow-through to identify any areas of stenosis or dilation and overall transit time. This study also allows assessment of bowel length and caliber. Barium is favored because of its density and ability to assess subtleties that may be missed with water-soluble contrast. This contrast media may become too dilute within the dilated bowel segments. In patients with SBS and suspected motility disorders of the colon, we also favor motility studies to assess for areas of segmental or global dysmotility that would be amenable to tapering or resection. The goals of the preoperative workup are to determine the suitability of a candidate for surgery and to assess risk factors that would complicate or contraindicate surgery, including bowel length, dilation, colon length and motility, and hepatobiliary disease.

A preoperative plan can then be formulated based on the algorithm presented in Figure 4. At exploration, full mobilization of the small bowel with lysis of adhesions is necessary. The bowel should be measured for length along its antimesenteric border from a standard reference point such as the ligament of Treitz when present or the pylorus in cases of malrotation, nonrotation, or other conditions in which there is no ligament of Treitz. Areas of dilation or changes in bowel caliber should be noted. Inspection of the gallbladder for stones and of the liver for significant cholestasis should be made. Liver biopsy should be considered almost a routine component of surgery in these patients to asses the degree of cholestasis. Strong consideration should be given to performing cholecystectomy in these patients even if they are asymptomatic because biliary sludge or small stones may induce subclinical choledochal or pancreatic obstruction.

In general terms, if the bowel is very short and segmentally dilated and the mesentery is not thickened, we would perform a Bianchi procedure. However, if there is no dilation in this setting, we would consider a colonic interposition as our next option. We have limited experience with valve techniques but might consider placement of a prosthetic cuff temporarily in such patients to achieve dilation prior to a future Bianchi procedure. If the bowel length is adequate, we would perform tapering enteroplasty.

Conclusions

SBS is a complex and multifaceted condition that can have significant morbidity and mortality within the pediatric population. The extensive functional reserve of the gut in children, and their improved ability to achieve spontaneous adaptation, favor less aggressive initial approaches to SBS care. Medical management, namely TPN, may have deleterious, life-threatening effects. Although the role of surgery is primarily adjunctive, it may offer great therapeutic benefit in selected patients. This benefit may be to promote complete adaptation or to decrease parenteral nutrition requirements. Numerous procedures are available in the armamentarium of the modern surgeon to combat this deadly syndrome; however, none of these operations can be applied universally (Fig. 4). Current assessments of efficacy and outcome of various surgical procedures are limited by the small number of these patients in any one center, the diversity of the diseases leading to SBS, the lack of uniform definition of the syndrome, and the lack of adequate risk stratification for intestinal failure. The care of these patients is sufficiently complex to require the coordinated interdisciplinary efforts of the pediatric gastroenterologist, pediatric surgeon, transplant surgeon, nutrition support nurse, and dietician. Nationally, there is a great need to

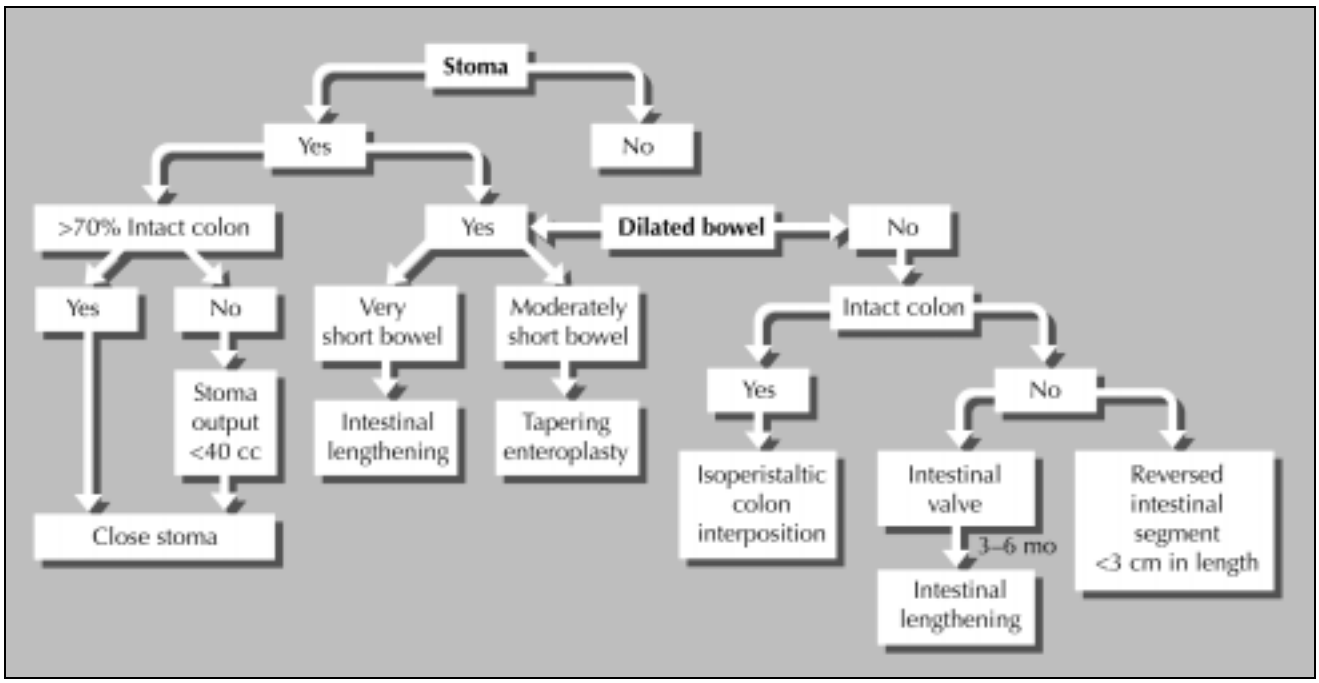

Figure 4. Algorithm for management of short bowel syndrome.

develop a registry and consortium of centers of excellence to focus on the care of these highly complex patients through stratification by disease severity and standardization of treatment protocols.

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