Short Bowel Syndrome in an Infant

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Education Gap

Infants with short bowel syndrome are affected differently depending on individual anatomy. A better understanding of the anatomy and physiology of the intestines will help better predict individual nutritional deficits.

Abstract

Short bowel syndrome (SBS) is a malabsorptive state that may occur either after surgical bowel resection or as the result of congenital bowel anomalies. SBS can incur significant morbidity and mortality including intestinal failure, cholestasis, sepsis, and death. For patients with SBS, management involves a multidisciplinary approach that begins with neonatology, pediatric surgery, nutritionists, pharmacists, and nurses in the NICU and also includes the transition to an intestinal rehabilitation program. The aim of this review is to provide the neonatologist with an overview of the common causes of neonatal SBS, anticipated nutritional deficiencies, complications associated with SBS, and the surgical and medical management of SBS to assist in counseling affected families.

Objectives After completing this article, readers should be able to:

1. Describe the nutritional deficiencies that may occur in infants with small bowel syndrome (SBS) according to the anatomy of their remnant bowel.
2. Explain surgical and medical management for patients with SBS.
3. Recognize the common complications of SBS and their management.
4. Describe differences in intestinal microbiota of patients with SBS compared with healthy infants and how this may play a role in clinical disease.

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ABBREVIATIONS

BSI	bloodstream infection
CVC	central venous catheter
FDA	Food and Drug Administration
GA	gestational age
GI	gastrointestinal
GIR	glucose infusion rates
IF	intestinal failure
IFALD	intestinal failure–associated liver disease
LILT	longitudinal intestinal lengthening and tailoring
NEC	necrotizing enterocolitis
PN	parenteral nutrition
SBBO	small bowel bacterial overgrowth
SBR	small bowel resection
SBS	short bowel syndrome
STEP	serial transverse enteroplasty

INTRODUCTION

Short bowel syndrome (SBS) is a malabsorptive state associated with an insufficient absorptive surface area of the small bowel. SBS most commonly results from a reduction in small bowel length, placing affected infants at risk for intestinal failure (IF). IF is defined as inadequate intestinal absorption of water,

electrolytes, and both macro- and micronutrients, which can lead to dehydration, growth failure, and subsequent dependency on parenteral nutrition (PN). The etiologies of SBS affecting infants include necrotizing enterocolitis (NEC) (35%), complicated meconium ileus (20%), gastroschisis (12.5%), intestinal atresia (10%), and volvulus (10%). (1)

SBS can be divided into 3 anatomic subtypes:

- Type 1: Small bowel resection (SBR) with small bowel anastomosis and intact colon
- Type 2: SBR with partial colonic resection and enterocolonic anastomosis
- Type 3: SBR with a high-output jejunostomy (1)

Patients with type 1 have the best chance of gaining enteral autonomy whereas patients with type 3 present the greatest challenge. It is difficult to predict which patients will develop SBS and/or IF, based on the total length of resected or remaining intestine. The diagnosis of SBS and IF varies among centers, but a commonly used definition is a dependency on PN for a minimum of 90 days after the inciting event. (2)

The most recent population-based estimates of SBS and/or IF incidence rates are from 2004 and 2008, with an estimated 22.1 per 1,000 NICU admissions complicated by SBS and/or IF (3) and 0.7% of infants of less than 1,500 g birthweight who have SBS. (4) The incidence increases with decreasing gestational age (GA) and birthweight, with an estimate of 353.7 per 100,000 live births in infants of less than 37 weeks' gestation compared with 3.5 per 100,000 live births in full-term infants. (3) These estimates may not accurately reflect the current incidence of SBS and/or IF among NICU patients, because trends in resuscitation practices of periviable infants have changed over time (5) and rates of gastroschisis have increased in recent years. (6) Further studies are needed to study the current epidemiology of SBS and IF.

The overall mortality rate of IF is relatively low. A recent meta-analysis estimated an annual mortality rate of 4.5% since 2005, a reduction from 5.9% per year before 2000. (7) Mortality was associated with the development of IF-associated liver disease (IFALD) and sepsis. (7) However, these estimates may underestimate the mortality rate, because the initial insult that renders patients with SBS, such as NEC, has a high mortality rate of up to 16.5% in the perioperative period. (8) This suggests that if patients can survive the initial perioperative period, then they should be expected to survive long term. The ultimate goal for patients with IF is to develop enteral autonomy, but this may take months to years or may eventually require multivisceral organ transplantation. Patients with SBS resulting from NEC achieve enteral autonomy faster than those with SBS from other diagnoses (ie, gastroschisis, intestinal atresia). In one study, 65% of patients with SBS following NEC reached enteral autonomy by 4 years of age compared with 29% of patients with SBS associated with other primary diagnoses. (9) This may be because of patients with NEC having otherwise normally functioning remnant bowel with a greater capacity for adaptation compared with the other congenital bowel anomalies that are often also accompanied by dysmotility. Other prognostic factors include length of remaining small bowel. In a study evaluating time to enteral autonomy, 88% of infants with SBS with more than 50 cm of remnant small bowel achieved autonomy by 1 year, with 96% gaining autonomy by 2 years. (10) For those with less than 50 cm of remnant small bowel, the probability of gaining autonomy was 23% at 1 year, 38% at 2 years, and 71% at 57 months. (10) In addition, patients with primary enterocolonic anastomosis have a greater chance of gaining enteral autonomy. (10)

This review will provide an overview of intestinal anatomy and physiology, which will help clinicians predict specific nutritional deficiencies anticipated in patients with SBS based on their operative reports. In addition, this review will summarize the common complications, surgical and medical management, and long-term outcomes in patients with SBS.

INTESTINAL ANATOMY AND PHYSIOLOGY

The size of a newborn's gastrointestinal (GI) tract varies according to GA and the individual. Studies determining the rate of growth across GAs and time are limited; however, it is estimated that at 24 to 26 weeks' GA, the small bowel and colon measure approximately 70 cm and 23 cm, respectively. (11) By term GA, the small bowel and colon increase to approximately 160 cm and 33 cm, respectively. (11) The intestines continue to grow throughout childhood until reaching adult sizes of about 600 to 700 cm and 150 cm for the small bowel and colon, respectively. (12) The surface area of the small intestines is further increased by the presence of villi and microvilli. Villi are covered by columnar epithelial cells at the tip and are primarily absorptive cells. Crypts are located at the base and are secretory. (13) It is important to understand the different functions and areas of absorption within the intestines to predict which nutritional deficiencies will occur if that portion of the bowel is removed (Fig 1, Table).

The duodenum is the first portion of the small bowel extending from the pylorus to the ligament of Treitz. The duodenum is the primary site for calcium and iron absorption. Up to 80% to 100% of calcium is absorbed through an

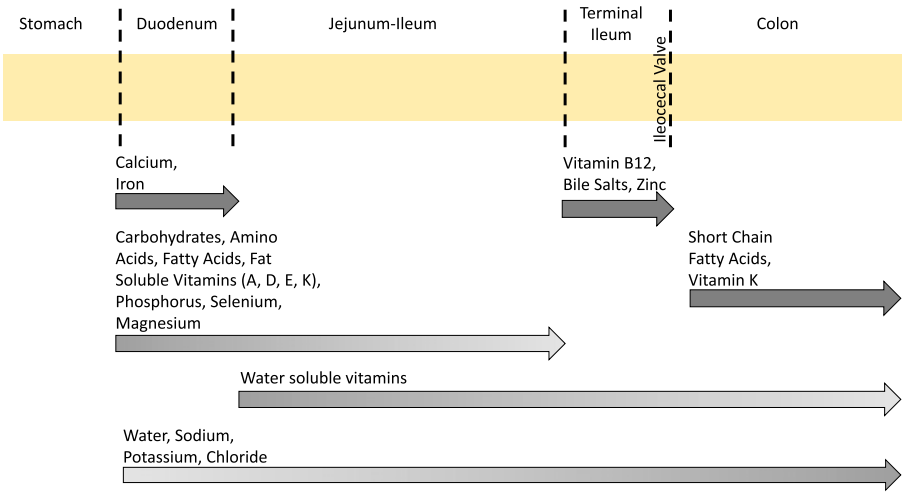

Figure 1. Location of nutrient absorption within the bowel. The color gradient of the arrows indicates the degree of absorption with the greatest level of absorption depicted by the darker color.

active transport system within the duodenum. (14) However, up to 20% to 60% of calcium can be absorbed by the jejunum, but is dependent on a concentration gradient and therefore, absorption at this region is most effective with high levels of oral calcium delivery. Iron is almost exclusively absorbed in the duodenum and can only be excreted through blood loss or intestinal sloughing; therefore, the duodenum is very important for iron homeostasis to meet the needs of erythropoiesis. (14) The duodenum also initiates the digestion of carbohydrates by breaking down starches (amylose and amylopectin) and disaccharides (sucrose and lactose) with further digestion and absorption occurring in the jejunum and ileum. (13) Proteins undergo proteolysis in the proximal small bowel, with nearly 50% of ingested protein being digested and absorbed in the duodenum. In addition, the duodenum is the site of production for a number of GI hormones, including secretin, cholecystokinin, somatostatin, and gastric inhibitory polypeptide, which are important in regulating digestion and absorption throughout the distal small intestine. (14)

The jejunum primarily lies in the central abdomen; although the beginning is designated by the ligament of Treitz, there is no anatomic landmark denoting the end of the jejunum. The transition from jejunum to ileum is marked by a smaller diameter and a thinner mucosal lining and wall. The ileum is primarily located in the hypogastric and pelvic regions and ends at the ileocecal valve. The jejunum and proximal ileum are the primary sites of digestion and absorption of carbohydrates, lipids, and protein as well as fat and water-soluble vitamins. (15) The absorption of nutrients from the proximal to distal small bowel progressively decreases as the luminal diameter narrows and the intercellular junctions go from loose to tight as the GI tract progresses. (16)

The distal ileum contains a number of specialized functions, including the absorption of vitamin B_{12}, zinc, and bile acids. The absorption of B_{12} is dependent on intrinsic factor release from the stomach and binding of B_{12}. The B_{12}-intrinsic factor complex then binds a membrane receptor in the terminal ileum and is absorbed. The complex then dissociates and B_{12} enters the portal circulation. (15) The terminal ileum is also important for the absorption of conjugated bile salts through a sodium-dependent active transport system, which then reenter the portal circulation. (15) Bile salts are important for the digestion and absorption of fat, which occurs primarily in the duodenum and jejunum. Reabsorbed bile salts are stored in the liver and gallbladder until eating stimulates their release. Bile salts that are not resorbed enter the colon and can cause diarrhea by impairing sodium and water absorption. If unabsorbed lipids reach the ileum, they typically stimulate the release of glucagonlike peptide 1 and peptide YY, which cause a delay in gastric emptying that allows for a longer transit time and facilitation of nutrient absorption in the small intestine. (15)

The ileocecal valve regulates the passage of fluids, electrolytes, and nutrients from the small intestine to the colon, and importantly, prevents the reflux of colonic material and bacteria into the small intestine. Although water and electrolytes are absorbed throughout the GI tract, the greatest area of absorption of water and sodium is in the colon. The colon has the tightest intercellular junctions and slowest transit time. Infants who have lost part or all of their colon are at risk for dehydration and inadequate electrolyte absorption.

TABLE. **Potential Nutrient Deficiencies and Suggested Laboratory Monitoring Schedule**

	DEFICIENCY PATHOPHYSIOLOGY	DEFICIENCY SYMPTOMS	LABORATORY MONITORING
Macronutrients			
Carbohydrates	Proximal bowel resection	Growth failure	Blood glucose as indicated
Protein	Proximal bowel resection	Growth failure	Total protein, prealbumin weekly, then every 2–4 weeks when stable
Triglycerides, EFA	Proximal bowel resection, terminal ileal resection, lipid restriction	Growth failure; EFA deficiency: dermatitis, alopecia, thrombocytopenia	Triglycerides weekly, then every 2–4 weeks when stable; triene/tetraene ratio with clinical EFA deficiency or prolonged lipid restriction
Vitamins			
Vitamins A, D, E, K	Fat malabsorption, cholestasis	Vitamin A: Xeropthalmia, blindness Vitamin D: Metabolic bone disease Vitamin E: Hemolytic anemia, myopathy, neuropathy Vitamin K: Hemorrhage	Vitamins A, E, K with evidence of fat malabsorption off PN or clinical signs of deficiency; vitamin 25-hydroxyvitamin D with elevated alkaline phosphatase, hypocalcemia, hypophosphatemia
Vitamin B12	Terminal ileal or gastric resection	Megaloblastic anemia, ataxia	B_{12} level with clinical signs of deficiency
Folate	Duodenal or jejunal resection	Anemia, thrombocytopenia, glossitis stomatitis	Folate level with clinical signs of deficiency
Macrominerals and trace minerals			
Sodium	Small bowel and colonic resection, ostomy fluid losses, diuretics	Growth failure, tissue edema	BMP daily, then 2–3x per week when stable until 2 weeks off PN; urine sodium every 1–2 weeks
Potassium	Small bowel resection, ostomy fluid losses, diuretics	Cardiac dysfunction, muscle cramps	BMP daily, then 2–3x per week when stable until 2 weeks off PN
Chloride	Small bowel resection, terminal ileal or colonic resection, ostomy fluid losses, diuretics	Growth failure, hypokalemic metabolic alkalosis	BMP daily, then 2–3x per week when stable until 2 weeks off PN
Magnesium	Fat malabsorption, presence of high output jejunostomy	Weakness, cardiac dysfunction	Magnesium daily, then 2–3x per week when stable until 2 weeks off PN
Calcium	Fat malabsorption, vitamin D deficiency	Metabolic bone disease, paresthesia, tetany, cardiac dysfunction	BMP and iCa daily, then 2–3x per week when stable until 2 weeks off PN; alkaline phosphatase every 2–4 weeks
Phosphorus	Proximal small bowel resection, vitamin D deficiency	Metabolic bone disease	Phosphorus daily, then 2–3x per week when stable until 2 weeks off PN; alkaline phosphatase every 2–4 weeks

BMP=basic metabolic panel; EFA=essential fatty acids; iCa=ionized calcium; PN=parenteral nutrition.

COMPLICATIONS IN PATIENTS WITH SBS

Patients with SBS require a long-term central venous catheter (CVC) to deliver adequate fluid and calories. Catheter-related complications can be life-threatening and have a major effect on long-term outcomes for these patients. Catheter-related bloodstream infections (BSI) are a significant cause of morbidity and mortality in patients with SBS. In a large retrospective review of 40,723 hospital admissions, 0.2% of the patients had a diagnosis of SBS, but accounted for 20% of all hospital admissions with BSI. (17) Enteric pathogens such as *Escherichia coli* and enterococci accounted for 23% of BSI in children with SBS compared with 12% of BSI in those without SBS. The rate of BSI was also high among SBS patients without a CVC (43%). A leaky gut mucosal barrier contributing to bacterial gut translocation may put these patients at higher risk. In addition to the risk of infection associated with SBS and

need for CVC, patients are at risk for mechanical and thrombotic complications. Ethanol locks for CVC have been introduced as a way to combat these complications because ethanol is both antimicrobial and fibrinolytic. Evidence to support this practice is insufficient, (18) though there is a strong association favoring the use of ethanol locks to prevent catheter-related BSIs and replacement of CVCs. (19)(20) In addition, safety data supporting the routine use of ethanol locks in the neonatal population less than 5 kg are limited, but this may be a potential strategy for older infants and children. Central venous thrombosis can ultimately lead to loss of venous access and venous access sites, preventing the delivery of parental nutrition. This is one of the indications for intestinal transplantation.

Patients with SBS who have IF are at risk for developing cholestatic liver disease also known as IFALD. A biochemical diagnosis of cholestasis is made when a patient's conjugated bilirubin is greater than or equal to 2 mg/dL (34.2 μmol/L), which histologically can range from steatosis to cirrhosis. The incidence of cholestasis is more than 50% in infants receiving PN for longer than 2 months. (21)(22) PN and lipids have been implicated as causative agents in the development of IFALD, but sepsis and lack of enteral feedings also play a role. Patients with SBS and/or IF should be monitored closely for the development of cholestasis, and proactive and reactive approaches to prevent cholestasis should be undertaken. Strategies to reverse cholestasis include lipid reduction (1 mg/kg lipid 1–3 times per week), lipid modification, and advancement of enteral nutrition. Lipid supplementation cannot be stopped completely because of concern for the development of essential fatty acid deficiency.

Until recently, the only commercially available lipid emulsion was derived from soybean oil (Intralipid, Fresenius Kabi, Bad Hamburg, Germany), which contains phytosterols and a greater amount of proinflammatory omega-6 fatty acids, both of which have been implicated in the development of cholestasis. (23)(24) A number of studies have showed the benefits of using alternative lipid emulsions, including fish oil, in the reversal of cholestasis. (25)(26) Fish oil contains a greater amount of omega-3 fatty acids and does not contain phytosterols but does contain α-tocopherol, a potent anti-inflammatory molecule. (27) Previously, lipid emulsions that contained fish oil were only available under a compassionate use protocol through the Food and Drug Administration (FDA) and so were only initiated for patients with severe cholestasis or in cases of impending liver failure. However, in 2016, the FDA approved the use of a combination lipid emulsion that was composed of 30% soy oil, 30% medium-chain triglyceride oil, 25% olive oil, and 15% fish oil (SMOF, Fresenius Kabi) for use in patients with cholestatic liver disease. In 2018, a 100% fish oil–based lipid emulsion (Omegaven, Fresenius Kabi) was approved. Initiation of fish oil–based lipid emulsions could be considered in patients who are predicted to remain dependent on PN for more than 2 weeks, but thus far the evidence for the prevention of cholestasis with these emulsions is insufficient. A caveat is that these alternative lipids are only approved for administration of up to 1 g/kg per day, the same as the lipid reduction strategy with the soy-based lipid emulsion. Although cholestasis in a patient with SBS and IF is most likely related to the disease process and treatment with PN and lipids, it is important to consider other causes of cholestasis as well, in particular biliary atresia, which requires timely surgical intervention.

It can be exceptionally challenging for patients with SBS to grow appropriately, with many experiencing growth failure. (28)(29) Growth is typically monitored as weight gain over time; however, in recent years, there has been a shift to focus more on body composition because linear growth represents lean muscle mass, protein accretion, and organ growth, most importantly, brain growth. Inflammation and prolonged illnesses, both common complications in patients with SBS, have been proposed to disrupt normal linear growth velocity and are associated with stunting, (30) which has been associated with poor neurodevelopmental outcomes and future metabolic syndrome in other patient populations. (31)(32)(33)(34) Pediatric patients with SBS have been found to display evidence of stunting despite supplementation with PN. (29)(35)(36) Poor somatic growth during infancy is of critical importance, because this is normally a period of rapid growth and development; therefore, linear growth in infants with SBS should be monitored closely. The long-term outcomes of stunting during infancy in patients with SBS is unknown, and future studies are needed to determine the effect on neurodevelopmental outcomes and metabolic disorders in patients with SBS in infancy.

Small bowel bacterial overgrowth (SBBO) and intestinal dysbiosis are other significant complications in patients with SBS. SBBO is defined as an increase in the number and species of bacteria within the small intestine, which may cause inflammation, malabsorption, and diarrhea. (37) Factors contributing to the development of SBBO may be loss of the ileocecal valve, causing reflux of colonic bacteria into the small intestine, dysmotility of intestinal contents allowing for bacterial colony multiplication, lack of enteral feedings, and gastric acid suppression with H2 blockers or proton pump inhibitors. (37) Treatment for SBBO includes

antibiotic therapy, most commonly with metronidazole or ciprofloxacin, which may need to be alternated or cycled if symptoms become chronic. SBBO is generally considered an overgrowth of nonpathologic bacteria, but patients with SBS are also at risk of developing intestinal dysbiosis. Patients with intestinal dysbiosis have an overall reduction in bacterial diversity and an increase in the relative abundance of pathogenic bacteria. (38) There has been recent interest in the intestinal microbiota of pediatric patients with SBS, and studies have found that patients with SBS who have IF have an overabundance of pathogenic bacteria, particularly from members of the *Proteobacterium* phylum, with a decrease in the number of beneficial bacteria from the *Firmicute* and *Bacteriodetes* phyla. (35)(36)(39)(40) *Proteobacteria* in the intestinal microbiota has been implicated in a number of disease states, including chronically malnourished children in developing countries, (41)(42) obesity, (43) and type 2 diabetes mellitus. (44) *Proteobacteria* are considered to be a highly inflammatory agent causing a disruption in the intestinal epithelial barrier that allows for bacterial translocation and promoting a chronic host inflammatory state. (45)(46)(47) Further research into how the intestinal microbiome may play a role in many of the complications of SBS, including BSI, cholestasis, intestinal adaptation, malnutrition, and somatic growth failure, are needed as well as investigations of potential therapeutic modifications to the intestinal microbiota in the treatment of SBS (Fig 2).

TREATMENT OF PATIENTS WITH SBS

Surgical Management

The initial approach to surgical management, which requires removal of bowel, involves preservation of as much bowel length as possible. The initial extent of resection is determined by the viability of the bowel, with any area of frankly nonviable bowel removed. The bowel is then either restored to continuity with an end-to-end anastomosis or left in discontinuity. If the bowel is in discontinuity, a proximal stoma can be created with a distal mucous fistula or Hartmann pouch. If bowel viability is a concern, the abdomen can be left open or temporarily closed, with a planned second look operation in 24 to 48 hours. In some patients who are critically unstable, the abdomen may be opened and the entire bowel contents placed in a silo until a more definitive operation can occur when the patient stabilizes. The silo allows decompression of the abdomen, which may improve cardiopulmonary mechanics, thereby improving the patient's clinical condition. Once the bowel is resected, measuring the remaining bowel and noting the presence or absence of the ileocecal valve are useful in prognosticating the severity of SBS.

Subsequent procedures are often necessary in the newborn period, especially if the bowel was placed in discontinuity. The timing of ostomy takedown is variable and depends on patients' clinical condition, their growth and nutrition, and whether they exhibit any signs of IF. Earlier stoma closure has been associated with decreased risk of cholestatic liver disease and helps facilitate intestinal adaptation. However, these patients are at risk for small bowel obstruction secondary to strictures or adhesions. These patients may present with an inability to advance feedings and radiographic evidence of small bowel obstruction, necessitating stricturoplasty, lysis of adhesions, or further resection. Dilated proximal bowel may be dysmotile and tapering of the affected bowel may improve peristalsis and decrease bacterial overgrowth, but also results in the loss of absorptive surface area.

For patients with SBS, long-term management is focused on intestinal rehabilitation and the goal of achieving enteral autonomy. Bowel adaptation is triggered by a substantial loss of enterocyte mass or functional mucosal surface. It begins within 24 hours of resection and continues over the course of the next 2 years. (48) In the first 4 weeks after resection, the remnant intestine experiences increased blood flow, which promotes mucosal hyperplasia, resulting in increased size and number of the crypts and villi. Enteral

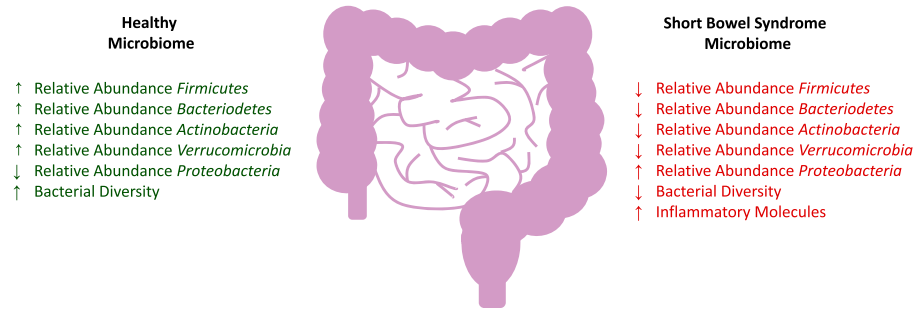

Figure 2. Differences in intestinal microbiome content between the healthy intestine and one with short bowel syndrome.

nutrients help stimulate adaptation by promoting the pancreaticobiliary secretions, stimulating neuroendocrine hormone release, and preventing mucosal atrophy. (49) The ileum has the greatest capacity for adaptation. (48) Intestinal adaptation is important in the ability to gain enteral autonomy. Unfortunately, adaptation can also lead to bowel dilation, dysmotility, and stasis and bacterial overgrowth. Various autologous intestinal reconstructive procedures have been described to increase functional bowel absorptive area and facilitate bowel motility while limiting stasis. The role and timing of these procedures continues to be debated, but they are most often considered after the exhaustion of medical therapies and management. (50)(51)

The earliest described autologous intestinal reconstructive surgery is the longitudinal intestinal lengthening and tailoring procedure (LILT) described by Bianchi. It divides the dilated bowel longitudinally and the 2 "neo" bowel sections are anastomosed isoperistaltically. Criteria for the consideration of Bianchi LILT include dilated bowel with a diameter greater than 3 cm and length greater than 20 cm in patients with SBS who have IF. The mesentery must be dissected into 2 leaves to allow for independent blood supplies for the resulting segments. Using a surgical stapler, the surgeon divides the dilated bowel in half along its longitudinal axis between the mesenteric defect that was created. (16) In theory, the LILT procedure should result in a 50% reduction of intestinal diameter and a doubling of intestinal length. Another procedure known as serial transverse enteroplasty (STEP) was described in 2003. (52) Alternating and opposite transverse staple fires are placed parallel to the mesentery along the length of the dilated bowel, resulting in an elongated zigzag-shaped portion of bowel. The STEP procedure depends on the presence of intestinal dilation. The bowel can be redilated in both the Bianchi LILT and STEP procedures. However, only STEP can be performed as a repeat operation. Multiple long-term studies have compared these 2 procedures. A large single-institution series reported that STEP resulted in decreased need for transplantation, significantly greater increase in bowel length, and a trend toward improved weaning off PN. No survival difference was observed between the 2 procedures. (53) Two other reviews favor STEP as having decreased mortality and decreased rate of intestinal transplantation, whereas LILT was associated with more success in weaning patients off PN. (54)(55)

Patients may be candidates for small bowel or multivisceral organ transplantation if they are unable to attain enteral autonomy, have unsuccessful bowel lengthening procedures, or develop serious complications. Fortunately, the number of organ transplantations in this patient population has decreased over time as a result of advancements in the medical and surgical care of these patients. (56)

Medical Management

In the immediate postoperative period after bowel surgery, significant fluid shifts can occur, resulting in fluid and electrolyte imbalances. Careful laboratory and hemodynamic monitoring is needed to avoid complications such as hypotension and electrolyte derangements. After infants convalesce, they can be started on PN. PN provides the major macronutrients, such as carbohydrates, protein, and lipids, as well as micronutrients such as vitamins and trace elements. Full PN should aim to provide 80 to 100 kcal/kg per day with careful monitoring of growth. As previously described, many patients with SBS experience growth failure. It can be tempting to increase caloric supplementation with carbohydrates in the form of glucose. However, when glucose infusion rates (GIR) are provided in excess of the rate of glucose oxidation (18 g/kg per day in surgical infants, GIR ~12 mg/kg per minute), glucose is converted to fat through the process of lipogenesis, which does not contribute to lean muscle mass and may further increase energy expenditure. (57) This can be especially challenging when lipids are restricted as a result of cholestasis. In this case, it may be preferable to tolerate lower caloric intake while attempting to advance enteral intake and maximize protein delivery as tolerated. Optimal lipid selection has been previously discussed but could include fish oil or combination lipid emulsions.

Enteral nutrition is the preferred route of nutrient provision and should be initiated as soon as clinically indicated. Enteral nutrition stimulates the intestine to release regulatory polypeptides and GI hormones that stimulate gut motility, enzyme release, mucosal growth, and blood flow. (48) The preferred form of enteral nutrition is maternal breast milk, which has been shown to be advantageous over amino acid–based formulas in patients with SBS. (58) Maternal breast milk provides secretory immunoglobulin A, glutamine/glutamate, and growth factors, which may help intestinal adaptation. (59) If maternal breast milk is not available, donor milk is an option, especially for premature low-birthweight infants with a history of NEC.

Infants with SBS may require fortification to achieve desired calories, vitamins, and minerals, especially in premature infants. There is no consensus on what formulas should be used when human milk is unavailable. Limited data are available showing that an elemental amino acid–based formula is better tolerated as the protein is already broken down and more readily available for absorption; (58) however, all commercially available elemental, partial, and

completely hydrolyzed protein formulas contain simple carbohydrates such as corn syrup and lactose as their carbohydrate source, which may contribute to a higher osmotic load and decreased absorption. In patients who do not tolerate advancement of feedings because of the carbohydrate content, feeding advancement should be slowed or a formula with a higher fat content should be used. A promising supplement in patients with SBS is enteral fish oil, which provides a combination of omega-6 and omega-3 fatty acids, and has been shown to improve growth velocity, decrease conjugated bilirubin, and decrease sepsis episodes compared with standard PN. (60) However, further studies evaluating safety and efficacy are needed before this can be recommended in infants with SBS.

Advancement of enteral feedings should be individualized according to the patient's clinical status and dietary needs because there is no consensus on the route or method of delivering enteral nutrition. Some studies advocate for initiating continuous feedings, which has been shown to be tolerated and if delivered slow enough, allows for continuous saturation of carrier proteins. (61)(62) However, bolus feedings are the most physiologic and allow for normal functioning of the extrahepatic biliary tree, (63) but may deliver intermittent high osmotic loads to the intestines, which can worsen diarrhea or ostomy output. Bolus feedings should only be delivered via a gastrostomy tube, whereas continuous feedings can be delivered via either gastrostomy tube or a postpyloric feeding tube. Caution should be taken when considering jejunal feedings because bypassing the duodenum would only further decrease the absorptive area of the remaining intestines and also prevent the release of several GI hormones as previously described. In some cases, a mucous fistula may have been created with the distal portion of the resected bowel. In this case, it would be advisable to refeed the ostomy output to the mucous fistula as clinically tolerated, because this primes the distal GI tract and prevents villus atrophy, may assist in weaning from PN, and helps with reanastomosis in the future. (64)(65)

GI electrolyte losses are common when an ostomy is in place, especially a type 3 high-output jejunostomy. The GI fluid losses can be enhanced by high osmotic loads or fast transit time. In patients with ostomy output approaching 30 to 40 mL/kg per day, feedings should be advanced cautiously, and any further advancement should be held when the output reaches more than 40 mL/kg per day because of the potential for fluid and electrolyte abnormalities. An average of 3.3 mEq/kg per day (3.3 mmol/kg per day) of sodium may be lost through normal ostomy output. Aldosterone is released when sodium losses are increased, which stimulates absorption of sodium in the proximal renal tubules to maintain normal serum sodium levels. (66) Therefore, in the distal renal tubule, sodium is decreased to participate in the sodium-hydrogen ion exchange, which limits the kidney's ability to correct the metabolic acidosis caused by GI bicarbonate loss. (67) A urine sodium less than 10 mEq/L (10 mmol/L) indicates an inadequate amount of sodium reaching the distal renal tubules and requires sodium supplementation. Poor somatic growth has been associated with urine sodium less than 30 mEq/L (30 mmol/L) and therefore additional supplementation should be considered for infants with poor growth and urine sodium less than 30 mEq/L (30 mmol/L). (68)(69)

Patients with SBS may become deficient in a number of nutrients as previously described and may require additional supplementation. A suggested laboratory schedule is provided to monitor for nutritional deficiencies with supplement options (Table). Patients with SBS are also at risk for overaccumulation of copper and manganese in the liver in the presence of cholestasis, leading to toxic levels of copper and manganese. These trace minerals should be removed from the PN prescribed when the conjugated bilirubin is greater than or equal to 2 mg/dL (34.2 μmol/L). The fat-soluble vitamins (A, D, E, and K) can be administered in a water-miscible vehicle for optimal absorption.

There is limited evidence to support the routine use of medications such as pancreatic enzymes, probiotics, bile acids, growth factors, and antimotility and secretory agents in infants with SBS to assist with motility and absorption. (70) In addition, many of these medications would need to be delivered enterally, and their absorption and pharmacokinetics in this population are unknown. However, ursodiol, a bile acid that enhances hepatic bile flow, has been shown to be beneficial in the reversal of cholestasis. (71) This medication must be delivered orally and therefore can only be initiated if the patient is tolerating at least partial enteral nutrition.

A key component of medical management for infants with SBS is the transition from the NICU to a multidisciplinary pediatric intestinal rehabilitation program for long-term care. An intestinal rehabilitation program has been reported to reduce morbidity and mortality. (20)(72) Consultation with the outpatient intestinal rehabilitation team should be initiated before discharge from the NICU to establish care and prepare for a medically complex patient.

CONCLUSIONS

Infants with SBS represent a challenging population that is at risk for a number of complications related to both the

infants' anatomy and the medical therapy used to support them. The risk for specific nutrient deficiencies can be anticipated by knowing the region of resected bowel and understanding the areas of digestion and absorption of macro- and micronutrients, vitamins, and minerals by the small intestine and colon. Despite advances in the care of these patients, SBS is still associated with considerable morbidity, including risk for CVC complications, BSI, growth failure, and liver disease. Further research is needed to continue advancing the medical and surgical care of these patients to optimize outcomes.

American Board of Pediatrics Neonatal-Perinatal Content Specifications

- Recognize the clinical manifestations, diagnosis and management of infants with perforations of the gastrointestinal tract (including gastric and intestinal).
- Know the clinical features, complications and management of short bowel syndrome.

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1. Short bowel syndrome (SBS) is characterized by intestinal malabsorption. In infants, SBS is most commonly secondary to a reduction in small bowel length. Infants with SBS are at risk for intestinal failure, defined as inadequate intestinal absorption of water, electrolytes, and macro- and micronutrients, leading to dehydration, growth failure, and dependency on parenteral nutrition. Which of the following conditions represents the most common cause of SBS in infants?
    A. Necrotizing enterocolitis (NEC).
    B. Complicated meconium ileus.
    C. Gastroschisis.
    D. Intestinal atresia.
    E. Malrotation with volvulus.
2. In infants with SBS, the likelihood of achieving enteral autonomy is dependent on multiple factors, including the etiology of SBS and the length of the remaining small bowel segment. Which of the following statements regarding the outcomes of SBS is correct?
    A. Patients with SBS secondary to intestinal atresia achieve enteral autonomy faster than those with a history of NEC.
    B. Children with greater than 50 cm of small bowel typically achieve enteral autonomy by age 1 year.
    C. Infants with small bowel length less than 50 cm achieve enteral autonomy in only 50% of cases by age 57 months.
    D. Patients with primary enterocolonic anastomosis have a greater chance of achieving enteral autonomy compared with those with a jejunostomy.
    E. The overall annual mortality from intestinal failure is 20%.
3. Nutritional deficiencies can occur in patients with SBS and are dependent on which sections of the bowel are removed. Which of the following statements regarding intestinal physiology is INCORRECT?
    A. The duodenum is the primary site for calcium and iron absorption.
    B. Vitamin B_{12} is absorbed in the proximal ileum after formation of the B_{12}-intrinsic factor complex.
    C. Infants who have lost part or all of their colon are at risk for dehydration and inadequate electrolyte absorption.
    D. Bile salts entering the colon can cause diarrhea by impairing sodium and water absorption.
    E. Unabsorbed lipids in the ileum cause a delay in gastric emptying via release of glucagonlike peptide 1 and peptide YY.
4. Patients with SBS are at risk for multiple complications including catheter-related bloodstream infections (BSIs), intestinal failure–associated liver disease (IFALD), small bowel bacterial overgrowth (SBBO), and growth failure. Which of the following statements regarding the complications of SBS is correct?
    A. *Escherichia coli* and enterococci account for 50% of BSIs in children with SBS.
    B. A conjugated bilirubin level greater than 5 mg/dL (85.5 μmol/L) is used to diagnose IFALD.
    C. The loss of the ileocecal valve is a risk factor for the development of SBBO.
    D. Intestinal dysbiosis is defined as an overgrowth of nonpathologic bacteria.
    E. Children with SBS have poor weight gain but normal linear growth.

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5. For patients with SBS, management is focused on intestinal rehabilitation with the long-term goal of achieving enteral autonomy. Which of the following statements regarding intestinal adaptation is INCORRECT?
    A. Intestinal adaption begins within 24 hours of bowel resection and continues for 2 years.
    B. Increased intestinal blow flow in the month after bowel resection leads to increase in the size and number of crypts and villi.
    C. Enteral nutrients stimulate adaptation by promoting pancreaticobiliary secretions, stimulating neuroendocrine hormone release, and preventing mucosal atrophy.
    D. The jejunum has the greatest capacity for adaptation.
    E. Intestinal adaptation can lead to bowel dilation and dysmotility.

Short Bowel Syndrome in an Infant

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