# Short-Bowel Syndrome: A Clinical Update

22

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## Introduction

During the second half of the twentieth century, the basic laboratory development and subsequent successful clinical application of the techniques of total parenteral nutrition (TPN) had a transformative effect on the modern practice of medicine, surgery, pediatrics, and many of their subspecialties. Arguably, none of the benefits of this technique has been more fundamental and lifesaving than the resultant developments and advances in the metabolic management, nutritional support, innovative operative procedures, and pathophysiologic understanding of patients with the short-bowel syndrome (SBS) following massive intestinal resection. Furthermore, primarily because of the remarkable salvage of most of these patients with this critically severe life-threatening situation, it has eventually been recognized that an even broader spectrum of disorders of alimentary tract functions could be identified in addition to the dramatic endgame of SBS; it has been recognized that the patients with these various intestinal dysfunctions deserve our special basic and clinical attention, investigations, and attempts to prevent, ameliorate, or cure them. As a result, the concept of intestinal failure inevitably and logically arose and continues to evolve as knowledge and experience regarding these often-complex alimentary tract problems and their management are generated or acquired [1].

Intestinal failure has had a multitude of definitions, which will likely undergo additional revisions as knowledge of this deceptively simple but tremendously complex and adaptable organ system and the variations in the types, extents, and degrees of failures of its multiple components accumulates with further study. Simply stated, intestinal failure is a condition characterized by deficient, inadequate, ineffective, or absent performance of the appropriate and expected intestinal functions essential for the efficacious and optimal absorption of the fluids and nutrients required to maintain the normal physiologic activities of the body cell mass. However, intestinal failure encompasses a broad spectrum of variety and severity of signs, symptoms, presentations, and responses to therapeutic interventions; its precise definition is difficult and virtually impossible to standardize to "one-size-fits-all" situations. Moreover, its clinical description usually has more practical relevance and usefulness for specific optimal management than does its broad definition. In this regard, intestinal failure is analogous, for example, to cardiac failure, pulmonary failure, renal failure, circulatory failure (shock), and other organ/system failures in that it can present, advance, respond, and adapt in myriad ways to challenge both the patient and the caregivers attempting to ameliorate, manage, and support the patient throughout the various stages of intestinal failure. Attempts to define intestinal failure more precisely by a single, comprehensive, and uniformly accurate statement is, in reality, a futile academic endeavor of limited utility to the practitioner. A summative description of the clinical picture and the relevant laboratory data in each individual patient will ordinarily be of the most value in formulating a management plan specifically best suited for each case. These complex problems are not routine or common, and their management and resolution require persistent, conscientious, dedicated, intensive attention to detail, together with skill, knowledge, experience, judgment, wisdom, and resilience if optimal outcomes are to be achieved [1].

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Short-bowel syndrome is a form of intestinal failure usually consisting of an inadequate length of intestine that results following massive bowel resection. SBS is a clinical entity characterized primarily by intractable diarrhea, steatorrhea, dehydration, malnutrition, weight loss, and malabsorption of fats, minerals, and other macronutrients and micronutrients and not a situation merely defined anatomically by a specific length of remaining functioning small intestine. Subsequent adverse consequences of SBS include hypovolemia, hypoalbuminemia, hypokalemia, hypocalcemia, hypomagnesemia, hypozincemia, hypocupricemia, essential fatty acid and vitamin deficiencies, anemias, hyperoxaluria, and metabolic acidosis. The formation of kidney stones or gallstones can also often accompany SBS. The actual clinical presentation and progression of the patient with SBS depends on several factors, including the following:

1. The extent of the bowel resection;
2. The site(s) of the resection;
3. The presence or absence of the ileocecal valve;
4. The residual function of the remaining small bowel, stomach, pancreas, biliary tree, and colon;
5. The capacity or potential of the intestinal remnant for adaptation;
6. The primary nature and status of the disease, disorder, or trauma that precipitated the loss of the small bowel;
7. The type, extent, location, and activity of any residual disease in the intestinal remnant; and
8. The general condition of the organ systems and body cell mass of the patient [2–8].

The minimum length of small bowel sufficient for adequate digestion and absorption is controversial. Standardization of the adaptive potential of the residual bowel is difficult because of the variable absorptive capacity of the remaining remnants, the wide variation in the length of the normal small intestine, and the difficulty in obtaining reproducible measurements of the length of the remaining bowel following massive resection. The nutritional and metabolic status, overall general health and function, and age of the patient are important collateral factors. Depending on the state of contraction or relaxation of the intestinal musculature, intraoperative estimates of the length of the normal, intact, small intestine in the adult vary from 260 to 800 cm (approximately 8–26 ft). On the other hand, the mean length of normal small intestine measured during life is 350 cm (11–12 ft), and postmortem it is 600 cm (20 ft) [6]. Because of this large variability, it is virtually impossible to determine the exact initial length of the remaining small bowel, and it is difficult to estimate the percentage of the total length of small bowel represented by the segment remaining following massive intestinal resection. Moreover, many surgeons often only measure the length of the resected small bowel, rather than also measuring the length of the remaining intestinal segment, which is the critically important functional and prognostic measurement. In addition, they then often fail to describe accurately the nature, condition, and extent of the remaining small bowel in the patient's medical record for future reference. Furthermore, because inflamed intestine generally shortens after operation, the absorptive functions following massive small bowel resection often do not correlate well with the original intraoperative estimated or measured length of the remaining intestine [6–8].

Because of the rather ample functional reserve capacity of the small bowel, short segmental resections of the small intestine usually do not result in significant problems with digestion and absorption [8–10]. Indeed, resection of as much as 40 % of the small intestine is usually well tolerated, provided that the duodenum, the distal half of the ileum, and the ileocecal valve are spared [11]. On the other hand, resection of 50 % or more of the small intestine usually results in significant malabsorption initially but can be tolerated eventually without extraordinary pharmacological or parenteral or enteral nutritional support. However, resection of 75 % or more of the small intestine usually leaves the patient with 70–100 cm (2–3 ft) of remaining intestine, resulting in a degree of SBS that can significantly impair the ability of the patient to maintain normal nutrition and metabolism. Such patients will likely require special nutritional management on a long-term or permanent basis, especially with the loss of the terminal ileum and the ileocecal valve, if normal body cell mass and function are to be preserved or restored [7].

The severity of symptoms and signs following massive small bowel resection is related both to the extent of the resection and the specific anatomic sites of the resected small bowel [12]. However, the minimal residual small intestinal absorptive surface required to sustain life without permanent parenteral nutritional support appears to vary somewhat with each patient [13, 14]. Development of effective TPN has revolutionized the treatment of SBS by allowing maintenance of adequate nutrition indefinitely or until the remaining bowel can adapt maximally to oral or enteral feeding, thus reducing the morbidity and mortality significantly [15–20]. Prolonged survival has now been achieved in a number of patients having only an intact duodenum and 15 cm (6 in.) of residual jejunum, with or without all or part of the colon [4, 10, 21]. If approximately 60 cm (2 ft) of jejunum or ileum remain functional in addition to the entire duodenum, survival has been the rule rather than the exception [21].

Preservation of the ileocecal valve is of paramount importance during massive small bowel resection and, by significantly increasing the duration of the intestinal transit time, allows a longer exposure time of the intestinal chyme to the residual absorptive surface of the mucosa. Salvage of the ileocecal valve, whenever possible, has the clearly beneficial effect of increasing the absorptive capacity of the remaining small bowel to approximately twice that anticipated for the same length of comparable small bowel

without an intact ileocecal valve. Primarily as a result of mucosal hyperplasia and villous hypertrophy, absorption by the residual intestinal segments of patients with SBS can increase as much as fourfold. Therefore, in a patient with an intact ileocecal valve, the total cumulative absorptive capability of the remaining bowel potentially can be increased maximally about eightfold. This amount of adaptive absorptive recovery function often approaches normal intestinal capacity [7, 21].

The most common clinical conditions that precipitate massive small bowel resections are those that compromise the vascular supply of the small intestine [22–24]. These include venous thrombosis and arterial occlusion as a consequence of primary vascular disease, heart failure with attendant mesenteric low-flow state, various coagulopathies, volvulus, malrotation of the gut, and internal or external herniation of the bowel with strangulation. SBS can also occur as a result of necrotizing enterocolitis or massive atresia of the small intestine in newborn infants, at times associated with gastroschisis or omphalocele. Inflammatory bowel disease involving large segments of the small bowel, or recurrent exacerbations of inflammatory bowel disease over a long period of time, can eventually result in SBS secondary to massive or multiple intestinal resections. Excision of retroperitoneal malignancies that involve the celiac or superior mesenteric vessels can mandate secondary resection of most or the entire small bowel to accomplish palliation or cure. Major abdominal blunt or sharp trauma involving transection, disruption, or avulsion of the mesenteric vasculature can also result in ischemic necrosis of large segments of the small bowel, resulting in SBS. Postirradiation or postoperative complications such as extensive severe radiation enteritis, multiple small bowel fistulas, multiple bowel obstruction procedures, and intestinal gangrene can also result in irreversible SBS.

Some of these conditions or situations are accompanied by, result in, or result from complex abdominal wall defects. For example, in neonates, gastroschisis is a congenital anomaly that not only is comprised of a defect in the closure of the abdominal wall but also is frequently associated with other developmental intestinal deformities, such as extensive or multiple small bowel atresias or mesenteric vascular abnormalities that result in the "apple peel" or "Christmas tree" mesentery anomalies. Omphaloceles, sometimes ruptured during the birthing process, are accompanied not only by an underdeveloped and contracted peritoneal cavity causing a "loss of domain" of the extra-abdominal small intestine, but also by atretic segments of bowel and an abdominal wall defect in the region of the umbilical cord. Surgical correction of these problems is obviously required, and the extent and nature of the procedure or procedures vary with the magnitude and complexity of each individual situation, ranging from simple closure of the abdominal wall defect, with or without resection of an accompanying atretic segment of bowel, to a compound or composite operative and nonoperative management plan of a multifaceted or variegated nature to restore both the integrity of the abdominal wall and the anatomical and functional continuity of the intestinal tract. The most difficult or complex of these conglomerate situations not only can pose formidable challenges to the neonatology and pediatric surgery teams but also can represent the highest level of personal and professional accomplishment when optimal outcomes result from their combined skills, efforts, and acumen. Obviously, nutritional and metabolic management and support must be intricately and masterfully interwoven judiciously with surgical operative talent, ingenuity, and timing; it must be continued, persistently and conscientiously, throughout the recovery and rehabilitative periods until optimal organ, system, and body cell mass functions are achieved or restored for the patient.

In adults, the recent era of abdominal compartment syndrome and the treatment or decompression of intraperitoneal hypertension by "open abdomen" measures or temporary intestinal coverage by various reconstructive operative techniques, using native tissues or various artificial or despeciated substitute products for abdominal closure, has been accompanied by a significant incidence of fistula formation, bowel obstructions, herniations, recurrent operations, and so on. At times, the prolonged treatment periods necessary to salvage and rehabilitate these patients, together with the multiple associated complications, not only have challenged surgeons technically to restore abdominal wall integrity but also have required their understanding of the physiologic and metabolic states of the patients that will enable them to restore and maintain intestinal continuity and function. This occurs while dealing with multiple enteroatmospheric fistulas ("the surgeon's nightmare"), multiple intestinal resections, functional or anatomical intestinal failure or SBS, combined with the ever-present need to maintain optimal nutritional status to promote immunocompetence; combat infection; heal anastomoses and wounds; support normal organ, system, and body cell mass functions; and preserve life itself [25, 26]. The problems for most such patients result from acute major traumatic injuries, in which portions of the abdominal wall might be lost, destroyed, or devitalized, in addition to injuries to other organ systems. However, these complex abdominal wall/SBS catastrophic situations can also arise following nontraumatic gastrointestinal (GI) tract perforations secondary to a variety of inflammatory or neoplastic disorders, mesenteric infarctions of the intestine, anastomotic leaks, various intraperitoneal abscesses, abdominal wound disruptions, and so on, often coupled with or compounded by hypoproteinemic malnutrition as a contributing, precipitating factor, as a comorbidity, or as a secondary complication of SBS or other intestinal failure [26].

## Pathophysiology of Short-Bowel Syndrome

The intestinal absorption of water, electrolytes and other specific nutrients is dependent primarily on the extent and site of the small bowel resection. The intestinal phase of digestion occurs initially in the duodenum, where pancreatic enzymes and bile acids promote digestion of all nutrients and enhance fat absorption. It is highly uncommon for the duodenum to be resected together with extensive segments of the small bowel, primarily because of the differences in blood supply; however, total duodenectomy, when it occurs, leads to malabsorption of calcium, folic acid, and iron [2]. Proteins, carbohydrates, and fats are absorbed virtually completely in the 150 cm of the jejunum; therefore, only small quantities of these nutrients or their derivatives ordinarily reach the ileum [27].

The small intestine acquires and handles a total of about 8 L of fluid daily, including dietary ingestion and endogenous secretions. Normally, approximately 80 % of the intraluminal water transported is absorbed in the small bowel, leaving approximately 1.5 L of fluid to traverse the colon. The colon usually absorbs about 1–2 L of fluid, having maximal absorptive capacity of approximately 6 L of fluid per day [28]. Because the ileum and colon have a large capacity for absorbing excess fluid and electrolytes, proximal small bowel (jejunal) resections only rarely result in diarrhea. On the other hand, extensive or total resection of the ileum results in a greater potential for malabsorption and resultant diarrhea. Not only will such resections increase the volume of fluid reaching the colon, but also, depending on the length of ileum resected, bile salt diarrhea (cholorrhea) or steatorrhea may ensue, with subsequent losses of essential fatty acids and fat-soluble vitamins. If the ileocecal valve has been resected, transit time is likely to decrease, and bacterial colonization of the small bowel will eventually be more likely to occur, further aggravating cholorrhea and steatorrhea.

As the length of ileal or colonic resections increases, essential absorptive surface area is lost, resulting in proportionally increased dehydration, hypovolemia, and electrolyte derangements. If the colon remains in continuity with the remaining small bowel following massive intestinal resection, malabsorbed bile salts can be deconjugated by colonic bacteria, stimulating increased colonic fluid secretion and further compounding existing diarrhea. Following extensive ileal resection, the enterohepatic circulation is interrupted, and irreversible loss of bile salts results, with or without the colon in continuity. Although the excess fecal losses stimulate hepatic synthesis of bile salts, a higher incidence of cholelithiasis occurs in these patients. Because the transit time in the ileum is usually slower than in the jejunum, residual intestinal transit is slowed, and fecal output is diminished as the length of remaining ileum increases.

Following extensive small bowel resections, intestinal lactase activity might decrease, resulting in lactose intolerance [29]. The presence of unhydrolyzed lactose causes increased hyperosmolality in the intestinal lumen. Moreover, fermentation of lactose by colonic bacteria produces a large amount of lactic acid, which can further aggravate osmotic diarrhea [2]. The water-soluble vitamins (vitamin B complex and C) and minerals ($Ca^{2+}$, $Fe^{3+}$, $Cu^{2+}$) are absorbed in the proximal small intestine, whereas magnesium diffuses passively throughout the entire small bowel [2]. On the other hand, the ileum is the only absorption site for vitamin $B_{12}$ and bile salts. Resection of the jejunum with preservation of the ileum produces no permanent impairments of protein, carbohydrate, and electrolyte absorption [30]. The ileum can compensate for most absorptive functions, but not for the secretion of jejunal enterohormones. Following jejunal resections, diminished secretions of cholecystokinin and secretin decrease gallbladder contraction and emptying and pancreatic exocrine secretions. In addition, after jejunal resection, gastric hypersecretion is greater than after ileal resection. This results from the loss of inhibitory hormones such as gastric inhibitory polypeptide (GIP) and vasoactive intestinal polypeptide (VIP), which are secreted in the jejunum, thus causing gastrin levels to rise, stimulating gastric hypersectretion [31]. Significant gastric hypersecretion can be documented within 24 h postoperatively, and the gastric and small bowel mucosa can be injured by the accentuated high gastric acid output, causing gastritis, ulceration, and bleeding. Subsequently, the high salt and acid load secreted by the stomach, together with the inactivation of digestive enzymes by the inordinately low intraluminal intestinal pH, serves to compound the other causes of diarrhea associated with SBS.

Ordinarily, the colon is a major site of water and electrolyte absorption, and as the ileal effluent increases, the colon may increase its absorptive capacity to three to five times normal [32]. Moreover, the colon has a moderate capacity to absorb other nutrients, and concomitant colon resections can adversely affect the symptomatic and nutritional courses of patients with massive small bowel resections. Malabsorbed carbohydrates that reach the colon are fermented there by indigenous bacteria to yield short-chain fatty acids, principally acetate, butyrate, and propionate [33, 34]. These short-chain fatty acids can be absorbed by the colon in quantities representing up to 500 cal per day and can enter the portal circulation to serve as a fuel source [35, 36]. Although retention of the colon is highly desirable during massive bowel resections, its presence can be associated with potential complications. In addition to cholorrheic diarrhea, a patient with a massive small bowel resection and an intact colon often develops hyperoxaluria and a tendency to form calcium oxalate renal stones. These

result from the increased absorption of dietary oxalate, which is normally rendered insoluble by binding with calcium in the intestinal lumen and therefore is ordinarily unabsorbable. However, in patients with SBS and steatorrhea, intestinal calcium ion is bound preferentially to the increased quantities of unabsorbed fatty acids, leading to decreased binding, and thus an increased colonic absorption of unbound oxalate [12].

Finally, preservation of the ileocecal valve is important in preventing abnormal metabolic sequelae because the ileocecal valve not only slows intestinal transit and passage of chyme into the colon but also to a large extent prevents bacterial reflux and passage from the colon into the small bowel. Nutrients that reach the colonic lumen, especially vitamin $B_{12}$, become substrates for bacterial metabolism rather than being absorbed into the circulation by the mucosa [2]. Furthermore, bacterial overgrowth in the small bowel in patients with SBS appears to increase the incidence of liver dysfunction [37].

## Nutritional and Metabolic Management of Short-Bowel Syndrome

In the metabolic and nutritional management of patients with SBS, three different but overlapping therapeutic periods having rather distinctive characteristics can be designated arbitrarily (Table 22.1) [38]. During the first 2 months (immediate and early postoperative period), the clinical picture and course are dominated by problems related to fluid and electrolyte balance; adjustments of organ blood flow patterns, especially the portal venous flow; and other effects of the major operative insult and its accompanying specific and general complications. During the second period, from about 2 months up to 2 years postoperatively (bowel adaptation period), efforts are directed toward defining maximum oral feeding tolerances for various nutrient substrates, encouraging and maximizing intestinal and bowel adaptation, and determining and formulating the most effective patient-specific feeding regimens. Usually within 2 years, 90–95 % of the bowel adaptation potential has been accomplished, and only 5–10 % further improvement in absorption and bowel adaptation can be anticipated. The third period (long-term management period) constitutes the period after 2 years, when nutritional and metabolic stability have ordinarily occurred. By this time, the patient has either adapted maximally so that nutrition and metabolic homeostasis can be achieved entirely with oral feeding, or the patient is committed to receiving specialized supplemental or complete nutritional support for the remaining life span, either by ambulatory home TPN or specially prepared enteral or oral feedings [7].

**Table 22.1** Synopsis of short-bowel syndrome management [7, 21, 38]

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| **Immediate postoperative period (First 2 months)** |
| *Fluid and electrolyte replacement* |
| Lactated Ringer's solution |
| Dextrose 5 % in water |
| Human serum albumin (low salt) |
| $K^+$, $Ca^{++}$, $Mg^{++}$ supplementation |
| Strict intake and output |
| Daily body weight |
| Graduated metabolic monitoring |
| *Antacid therapy (optional prn)* |
| (30–60 mL via nasogastric [N-G] tube q 2 h; |
| clamp N-G tube 20 min) |
| Mylanta liquid |
| Camalox suspension |
| Amphogel suspension |
| Gelusil liquid |
| *Antisecretory/antimotility therapy* |
| Cimetidine, 300 mg IV q 6 h |
| Ranitidine, 150 mg IV q 12 h |
| Famotidine, 20 mg IV q 12 h |
| Pantoprazole, 40 mg IV daily |
| Codeine, 60 mg IM q 4 h |
| Loperamide, 4–16 mg po daily |
| Lomotil, 20 mg po q 6 h |
| Hyoscyamine sulfate, 0.125 mg sc q 4 h |
| Cholestyramine 4 g po q 8 h |
| *Total parenteral nutrition* |
| 1 L on second postoperative day |
| Gradually increase dosage as tolerated |
| Supplemental fluids, electrolytes, and colloids as needed |
| **Bowel adaptation period (First 2 years)** |
| *Progression of oral diet* |
| Water, tea, broth |
| Simple salt solutions |
| Simple sugar solutions |
| Combined salt/sugar solutions |
| Dilute chemically defined diets |
| High carbohydrate, high protein |
| Modified fiber, low-fat diet |
| Near-normal, normal diet |
| *Enteral supplementation* |
| Coconut oil, 30 mL po tid |
| Safflower oil, 30 mL po tid |
| Multiple vitamins, 1 mL bid |
| Ferrous sulfate, 1 mL tid |
| Ca gluconate, 6–8 g/day |
| Na bicarbonate, 8–12 g/day |
| *Parenteral supplementation* |
| Electrolytes, trace elements |
| Divalent cations (Mg, Zn, Cu, Se) |
| Vitamin $B_{12}$, Vitamin K, folic acid |
| Albumin, packed red cells |
| Fat emulsion |

(continued)

**Table 22.1** (continued)

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|---|
| *Antisecretory/antimotility* (Refer to items for the first 2 months for additional agents) |
| Famotidine, 20 mg po q 12 h |
| Pro-Banthine, 15 mg po q 4–6 h |
| Dicyclomine, 20 mg po q 6 h |
| Omeprazole, 20 mg po q day |
| Deodorized tincture of opium, 10–30 gtts q 4 h |
| Codeine, 30–60 mg po q 4 h |
| Paregoric, 5–10 mL po q 4 h |
| *Growth hormone/glutamine* [54, 55] |
| **Long-term management (After 2 years)** |
| *Apply previous principles* |
| As indicated individually |
| *Ambulatory home TPN (total parenteral nutrition)* |
| Supplemental or total continuous, cyclic or intermittent |
| *Surgical management* |
| Treat operative complications |
| Drain abscesses |
| Resect fistulas |
| Lyse adhesions |
| Reduce obstructions |
| Restore bowel continuity |
| Probable cholecystectomy |
| *Intestinal lengthening* [70–75] |
| *Intestinal transplantation* [77–81] |

## Immediate Postoperative Period

During the immediate postoperative period, for up to 2 months, virtually all nutrients, including water, electrolytes, fats, proteins, carbohydrates, and all vitamins and trace elements, are absorbed from the GI tract poorly, unpredictably, or not at all [38]. Fluid losses via the GI tract are greatest during the first few days following massive small intestinal resection, and anal or stomal effluent frequently reaches volumes in excess of 5 L per 24 h. To minimize life-threatening dehydration, hypovolemia, hypotension, electrolyte imbalances, and other related potential problems, vigorous fluid and electrolyte replacement therapy must be instituted promptly and judiciously. Frequent measurements of vital signs, fluid intake and output, and central venous pressure, together with regular determinations of hematologic and biochemical indices, are mandatory in monitoring the patient during this period of rapid metabolic change and instability. All patients with SBS exhibit some abnormalities in their liver profiles, and the vast majority of them experience at least transient hyperbilirubinemia [38]. This has been advocated by some to be secondary to the translocation of microorganisms or their toxins through the ischemic or gangrenous intestinal mucosa into the portal vein and thence to the liver [39, 40]. Others attributed the hyperbilirubinemia to impaired blood flow to the liver through the portal vein by as much as 50 % as a result of greatly diminished mesenteric venous return secondary to the massive small bowel resection [41]. Still others attributed this phenomenon to a combination of both factors or other etiologies [42]. Broad-spectrum anaerobic and aerobic antibiotic therapy should be instituted empirically and maintained for several days to 1 week following massive intestinal resection.

Typical patient management efforts during this period are directed toward achievement of four primary goals: fluid and electrolyte replacement, antisecretory/antimotility therapy, antacid therapy, and TPN. During the first 24–48 h, replacement therapy usually consists of 5 % dextrose in lactated Ringer's solution administered intravenously concomitantly with appropriate amounts of potassium chloride or acetate, calcium chloride or gluconate, magnesium sulfate, and fat- and water-soluble vitamins. If there is no evidence of sepsis, low-salt human albumin (12.5–25 g) usually is added exogenously to the intravenous regimen every 8 h for the first 24–48 h postoperatively to maintain normal plasma albumin concentrations and normal plasma colloid oncotic pressure. It is our opinion and experience that maintenance of optimal intravascular colloid osmotic pressure with normal albumin and erythrocyte concentrations reduces intestinal mucosal edema and enhances fluid and nutrient absorption while reducing losses as diarrhea. In patients with severe diarrhea, zinc losses can increase to as much as 15 mg/day, and appropriate aggressive parenteral replacement is required [43].

Antiacid therapy can reduce the increased tendency for peptic ulceration, which commonly occurs following massive small bowel resection. Antacids are given through a nasogastric tube, if one is in place, every 2 h in doses of 30–60 mL, and the tube is then clamped for 20 min before reapplying suction. Alternatively, or concomitantly, liquid sucralfate can be given by mouth or via the nasogastric tube in a dose of 1 g every 6 h, clamping the tube for 20 min after each dose. To counteract the hypergastrinemia and associated gastric hypersecretion that follows massive small bowel resection in the majority of patients, an $H_2$ receptor blocker is infused intravenously [44]. The intravenous administration of 300–600 mg of cimetidine every 6 h can have a profound effect on reducing gastric acid and intestinal fluid production. Alternatively, 150 mg ranitidine can be given intravenously every 12 h, 20 mg famotidine can be given intravenously every 12 h, or an intravenous form of a proton pump inhibitor, pantoprazole, can be given daily in 40-mg doses. In selected patients with short bowel, somatostatin analog (octreotide) has reduced fecal losses when administered in a dosage of 50–150 μg IV or subcutaneously every 6 h [45, 46]. If diarrhea persists despite these measures, an opiate can be prescribed. Preferably, codeine is given intramuscularly in doses of 60 mg every 4 h. Improvement in fluid and electrolyte management can also be achieved in selected patients with stomal access to a distal

defunctionalized bowel loop by reinfusing the chyme from the proximal stoma into the distal bowel segment [47]. Later in the course of the postoperative period, when the patient is tolerating liquids by mouth, antimotility therapy can be achieved by giving 4–16 mg loperamide orally in divided doses daily, 4 g cholestyramine every 4–8 h, or 20 mg diphenoxylate every 6 h. Codeine (30–60 mg), 5–10 mL paregoric, or 10–30 drops deodorized tincture of opium (DTO) every 4 h orally can be used to impede bowel motility. The major advantages of DTO are that it is readily absorbed by the upper alimentary tract, and the patient's bowel hypermotility and diarrhea can be titrated to tolerable therapeutic levels by adjusting the dosage up or down a few drops at a time to optimize dose effectiveness and to minimize undesirable side effects [7, 21].

By the second or third postoperative day, the patient's cardiovascular and pulmonary status have usually stabilized sufficiently to allow TPN to be initiated [7, 21]. The average adult patient can usually tolerate 2 L of TPN solution daily administered by central vein. By titrating levels of plasma glucose and glycosuria, the daily nutrient intake can be increased gradually to desired levels or to patient tolerance. In a patient with diabetes mellitus or who is glucose intolerant, crystalline regular human insulin is added to the TPN solution in dosages up to 60 units per 1,000 cal as needed. Following an operation of the magnitude of massive small bowel resection, patients may require up to 3,000 mL of TPN solution (about 3,000 cal) per day initially for a few days to maintain nutritional and metabolic homeostasis. Supplemental fluid and electrolyte infusions might be necessary for several days or weeks to replace excessive losses as diarrhea. The patient is offered a clear liquid diet as soon as the postoperative condition is stabilized, and fecal output is controlled with antidiarrheal medications. It may take several days to several weeks before the patient is able to discontinue TPN support in favor of oral or enteral feedings. It is essential to provide adequate nutritional supplementation with TPN for as long as the patient requires such support to maintain optimal nutritional status. The TPN ration is reduced gradually in an equivalent reciprocal manner as oral intakes and intestinal absorption of required nutrients are increased. The patient's diet is advanced slowly and gradually to a low-lactose, low-fat, high-protein, high-carbohydrate composition according to individual tolerances to the nutrient substrates and to the water volume and osmolality of the dietary regimen [7, 21, 48].

## Bowel Adaptation Period

During the period of bowel adaptation from 2 months to 2 years postoperatively, the patient is allowed to consume increasing amounts of water, simple salt solutions, and simple carbohydrates [7, 21]. Various fruit and other flavorings can be added to 5 % dextrose in lactated Ringer's solution as a relatively inexpensive and practical oral nutrient and fluid replacement solution. Gradually, dilute solutions of chemically defined diets containing simple amino acids and short-chain peptides are given as tolerated in increasing volumes and concentrations as bowel adaptation progresses toward a normal or near-normal diet consisting of high carbohydrate, high protein, and low fat and comprised of food most preferred by the patient as the next stage of nutritional rehabilitation. Alternatively, the major nutrients can be provided as required in commercially prepared modular feedings tailored to the needs of individual patients until ordinary food is well tolerated. All essential vitamins, trace elements, essential fatty acids, and minerals are initially supplied in the patient's balanced intravenous nutrient ration. Subsequently, the oral diet may be supplemented most economically by short- and medium-chain triglycerides in the form of coconut oil, 30 mL two or three times daily; essential fatty acids as safflower oil, 30 mL two or three times daily; multiple fat- and water-soluble vitamins in pediatric liquid form, 1 mL twice daily; vitamin $B_{12}$, 1 mg intramuscularly every 4 weeks; folic acid, 15 mg intramuscularly weekly; and vitamin K, 10 mg intramuscularly weekly. Some patients may require supplemental iron, which can be administered initially by deep intramuscular injection as iron dextran according to the recommended patient-specific dosage schedule or as an intravenous infusion after testing the patient for sensitivity [7, 21]. Alternatively, an oral liquid iron preparation can be given one to three times daily, while closely monitoring iron indices and liver function tests.

A strong tendency for patients with SBS to develop metabolic acidosis usually requires the use of sodium bicarbonate tablets, powder, wafers, or liquid in doses of 8–12 g/day for as long as 18–24 months, but usually not for fewer than 6 months [7, 21]. It is often helpful to alternate the form of sodium bicarbonate prescribed to encourage maximal patient compliance. Because of the difficulty in absorbing adequate dietary calcium, supplemental calcium gluconate should also be prescribed as tablets, wafers, powder, or liquid in doses of 6–8 g/day. As bowel adaptation progresses, the doses of sodium bicarbonate and calcium gluconate can be decreased concomitantly or discontinued as restorative goals are attained. However, such oral supplements might be necessary for as long as 2 years or more in some patients to maintain homeostasis. Occasionally, on the other hand, a patient may become severely acidotic (pH 7.0–7.2) as a result of obviously copious diarrhea, but sometimes more subtly, and may require urgent or emergency intravenous infusion of sodium bicarbonate. Usually, the patient responds promptly to the therapy within a few hours and without untoward sequelae. Rarely, calcium gluconate must be given intravenously as a supplement to correct recalcitrant

hypocalcemia (<8.0 mg/dL). It is important to maintain normal serum albumin levels in patients with hypocalcemia. Dietary advancement and nutrient supplementation must obviously be individualized for each patient, and an effective nutrition support team can be helpful in maintaining and monitoring these complex patients. When solid foods are given, they should be dry and followed 1 h later with isotonic fluids, rather than giving solids and liquids together at the same time. This practice is followed to minimize diarrhea and to improve nutrient absorption. Lactose intolerance should be anticipated and treated as required with a low-lactose diet or 125–250 mg lactase by mouth. Clearly, milk products should be avoided as much as possible if intolerance persists [7, 21].

As progress occurs during the bowel adaptation period of management of the SBS, fat can be increased in the diet as tolerated, and supplementation with short- and medium-chain triglycerides and essential fatty acids may no longer be necessary [7, 21]. Serum-free fatty acid levels and triene-to-tetraene ratios are monitored periodically to determine the efficacy of treatment and the need for supplementation. Contrary to early reports, high-fat diets apparently are comparable to high-carbohydrate diets when evaluated in reference to calories absorbed, blood chemistries, stool or stomal output, urine output, and electrolyte excretions [47]. However, it has been suggested that enteral intake of fat should approach 50–100 % greater than expected goals to compensate for malabsorbed nutrients [43]. Patients who cannot tolerate or utilize a normal oral diet should be given a trial of continuous administration of enteral formula. Low-residue, polymeric, chemically defined, or elemental diets offer the putative advantage of high absorbability in the patient with a short bowel. However, some investigators have recently shown no differences in caloric absorption, stomal output, or electrolyte loss among elemental, polymeric, and normal diets in patients with SBS [7, 21, 49–51].

Depending on the results of periodic hematologic and biochemical studies, adjustments are made in the patient's intake of sodium, potassium, chloride, and calcium [52]. In addition, intermittent supplemental infusions of solutions containing magnesium, zinc, copper, and selenium might be required. As malabsorption and diarrhea become less troublesome, the vitamin and trace element requirements may be satisfied by multivitamin capsules, tablets, or chewable tablets containing therapeutic doses of vitamins or minerals, one dose twice daily. Relatively large amounts of magnesium, zinc, vitamin C, and vitamin B complex can be administered in the form of several commercially available therapeutic vitamin and mineral preparations [7, 21, 38]. It is especially important to avoid thiamine deficiency (Wernicke's syndrome).

In some patients, it might be necessary periodically to correct individual nutrient substrate deficiencies intramuscularly or intravenously for prolonged periods of time. Intermittent infusions of human serum albumin and packed erythrocytes might be required to treat recalcitrant hypoalbuminemia and anemia and to restore the plasma albumin level and the hematocrit to normal. Cholestyramine can be administered to counteract bile salt diarrhea if indicated, but intraluminal cholestyramine itself can cause or aggravate diarrhea. Fatty acid, electrolyte, trace element, vitamin, and acid–base imbalances must be promptly corrected enterally or parenterally as required when manifested clinically or by laboratory assessment. Serum vitamin $B_{12}$ levels must be monitored and vitamin $B_{12}$ deficiency corrected immediately. Hyperoxaluria should be assessed regularly, and if documented, foods containing high levels of oxalate, such as chocolate, spinach, celery, carrots, tea, and colas, should be restricted [7, 21].

In patients with severe forms of SBS, in whom little or no small intestine is present distal to the duodenum or in whom the remaining small intestine has residual disease, hypermotility and recalcitrant or intractable diarrhea may require continuous long-term antimotility/antisecretory treatment with oral or parenteral forms and dosages of the previously described pharmaceutical agents. Additional oral medications that have been helpful in selected patients include omeprazole, 20 mg daily; propantheline bromide, 15 mg every 4–6 h; dicyclomine hydrochloride, 20–40 mg every 6 h; and hyoscyamine sulfate, 0.125–0.250 mg every 4–6 h as needed [7, 21].

## Long-Term Management Period

Long-term management of SBS can be accomplished successfully in most patients by conscientious attention to the principles and practices outlined previously. However, in a few patients who have undergone massive small bowel resection, TPN or supplemental parenteral nutrition must be provided in a continuous or cyclic manner for extended periods of time and sometimes for life. The metabolic management and nutritional therapy of patients with SBS must be tailored specifically to each patient, and the clinical responses following massive intestinal resections depend on many and varied factors. Patients with SBS pass through several stages of nutritional and metabolic support during their recovery, convalescence, and rehabilitation. Most of them can ultimately be maintained on a normal or near-normal diet. However, depending on the adaptability of their remaining bowel, they may have to settle for receiving their nutritional requirements by one or more of the following options:

1. A modified oral diet;
2. An oral diet supplemented with intravenous fluid or electrolytes;

3. An oral diet supplemented with enteral feedings;
4. An enteral diet entirely;
5. An oral diet supplemented with enteral feedings and parenteral nutrition;
6. An enteral diet supplemented with oral feedings;
7. An oral diet supplemented with parenteral nutrition;
8. An enteral diet supplemented with parenteral nutrition;
9. An enteral diet supplemented with parenteral nutrition and oral feedings;
10. A primarily parenteral nutrition regimen supplemented with variable oral or enteral diets; and
11. Total parenteral nutrition virtually entirely, but with trophic oral feedings as tolerated to stimulate intestinal adaptation and immunocompetence.

Almost every patient with SBS eventually develops gallstones, most usually requiring cholecystectomy within 2 years following massive intestinal resection if the gallbladder had not been previously removed. Indeed, the high propensity of patients who have undergone massive intestinal resection to develop stones in the gallbladder has stimulated some physicians to advocate cholecystectomy prophylactically at the time of bowel resection [53]. However, gallstone formation in the common bile duct and elsewhere in the biliary tree is also increased in these patients even after cholecystectomy. Therefore, long-term surveillance with periodic abdominal ultrasonography might be useful in identifying and monitoring echogenic changes in the gallbladder and biliary tree in patients with a short bowel [7, 21].

Finally, some otherwise-stable patients occasionally develop recalcitrant diarrhea secondary to colonization or bacterial overgrowth of the residual small bowel segment, requiring periodic stool culture and bacterial antigen studies followed by parenteral treatment with appropriate antibiotics [7, 21].

## Growth Hormone, Glutamine, and Modified Diet

An extensive study has been completed to determine if growth hormone or nutrients, given alone or together, could enhance absorption from the small bowel after massive intestinal resection, especially in patients who continue to experience malabsorption and require long-term parenteral nutrition [54]. The effects of a high-carbohydrate, low-fat diet, the amino acid glutamine, and growth hormone, administered alone or in combination, were studied in 47 adult patients with SBS who were dependent on TPN to some extent for an average of 6 years. The average age of the patient was 46 years; and the average residual small bowel length was 50 cm in those with all or a portion of the colon remaining, and it was 102 cm in those with no colon remaining. During the 28 days of therapy, recombinant growth hormone was given by subcutaneous injection at a dose ranging from 0.03 to 0.14 mg/kg/day (average dose 0.11 mg/kg/day). Supplemental glutamine was provided by both the parenteral and enteral routes. The parenteral glutamine dosage averaged 0.6 g/kg/day, whereas a standard daily dose of 30 g glutamine was administered orally in six equal portions of 5 g mixed with a hypotonic cold beverage. In addition to the growth hormone and glutamine, all patients underwent extensive diet modification and nutritional education, the details of which have been reported extensively elsewhere [55]. On completion of the 4-week protocol, growth hormone was discontinued, and the patients were discharged home on 30 g/day oral glutamine and the modified oral diet [7, 21].

The initial balance studies indicated improvement in absorption of protein by 39 %, accompanied by a 33 % decrease in stool output with the regimen. In evaluation of the long-term results, averaging 1 year and extending as long as 5 years, 40 % of those studied remained off TPN, and an additional 40 % reduced their TPN requirements, with no change in TPN requirements in the remaining 20 %. These changes had occurred in a subset of patients that had previously failed to adapt to the provision of enteral nutrients, and this therapy may offer an alternative to long-term dependence on TPN for some patients with severe SBS. Subsequently, a more comprehensive clinical study of greater than 300 patients has been reported by the same group of investigators [56, 57]. However, growth hormone alone has not been shown to be beneficial consistently in other randomized, blind, placebo-controlled, crossover studies, and the Bryne et al. study results have not been reproduced by other investigators [58–60]. These conflicting data emphasize the need for further clinical studies to evaluate the effects of trophic agents on intestinal adaptation [61]. Both growth hormone and glutamine are available for clinical use, but growth hormone generally is not used routinely or often because of its high cost, side effects, and questionable efficacy [58, 62].

A recent review article on the management options in SBS reported that administration of glucagon-like peptide-2 (GLP-2) to patients following major small bowel resection improved intestinal adaptation and nutrient absorption [63]. Teduglutide, an enzyme-resistant GLP-2 analog, has shown promise in preventing intestinal injury, restoring mucosal integrity, increasing villous height, enhancing intestinal absorptive function, and increasing lean body mass, based on data from ongoing clinical trials in patients with SBS [64–69]. However, further studies and the completion of current phase III trials are necessary to determine the appropriate dosage (high vs. low) and length of treatment required for these patients to gain optimal benefits from the administration of this agent [63, 64].

## Surgical Considerations

Total parenteral nutrition is the mainstay of early and sometimes late management of SBS [56]. Prior to the widespread use of TPN, patients often survived the initial surgical insult of massive small bowel resection and its early complications only to die ultimately of fluid, electrolyte, and nutritional imbalances. Today, however, patients can usually be managed successfully and often rehabilitated with the judicious use of TPN. In this regard, the surgeon is required to insert, maintain, and supervise a temporary and subsequently a permanent indwelling central venous catheter or catheter port for administration of TPN solutions [7, 21].

Massive small bowel resection is associated with a prompt and inordinate increase in the secretion of gastrin and gastric acid. The resulting hypersecretion can readily cause or aggravate existing gastritis, ulceration, bleeding, diarrhea, and fluid and electrolyte depletion. Because the hypersecretion is thought to be mediated hormonally, truncal vagotomy and pyloroplasty have been performed in human beings with good results [2]. Now that effective $H_2$ receptor blockers have been developed for clinical use, the surgical treatment of hypersecretion is seldom indicated or required. Currently, vagotomy or other acid-reducing operations should be reserved only for those SBS patients who develop complicated peptic ulceration problems resistant to conservative medical therapy. Partial or total gastric resections in patients with SBS should be avoided assiduously.

In patients with SBS following massive intestinal resection, parenteral nutrition should be given for at least 6–12 months to ensure that optimal bowel adaptation has occurred before contemplating the use of any surgical procedures to increase absorption of nutrients [39]. In most SBS patients, sufficient bowel adaptation occurs during the first year following massive intestinal resection so that parenteral nutrition can be discontinued, and contemplated surgical interventions can be avoided [7, 21].

Thompson has recently reviewed his extensive operative experience with adjunctive management of SBS patients [58]. He posited that if an adult with SBS develops intestinal dilation, it usually is secondary to obstruction, either secondary to recurrent intra-abdominal adhesions or at the site of a previous anastomosis. Bacterial overgrowth often develops in dilated, relatively hypotonic bowel and compounds the malabsorption secondary to SBS. Although conservative management is preferable initially, surgery is usually required to relieve intestinal obstruction, which may include lysis of adhesions, stricturoplasty, or minimal segmental resections only as absolutely necessary [58, 70]. Dilation of the intestinal remnant occurs more frequently in children than in adults and appears to have a basis that is more adaptive in nature than pathologic [58, 71]. In patients with adequate bowel length, longitudinal taper enteroplasties have been used to restore the dilated lumen diameter toward normal. Tapering enteroplasties may be either resective or imbricating, with no significant differences between the approaches [58, 71]. Lengthening procedures are not performed on obstructed bowel in an effort to "create length," but rather to relieve the functional obstruction and to allow the bowel transit to return toward normal. To restore luminal diameter, Thompson and others have found the so-called intestinal-lengthening procedures to be the optimal treatment [58, 71]. Although easiest to describe as lengthening, Thompson stated that these procedures actually more truly represent an attempt to optimize the ratio of volume to surface area of the intestine to improve contact time between luminal contents and absorption surface [58]. The initial operative approach was longitudinal lengthening via the Bianchi procedure, which involves meticulous dissection of the mesentery of the bowel segment to allocate terminal blood vessels to either side of the bowel wall [58, 71–73]. Longitudinal transection of the bowel is then performed, usually with a stapling device, which creates two parallel vascularized limbs of a smaller caliber, which can then be anastomosed effectively to lengthen the intestinal remnant through which the chyme must flow [58, 71, 72]. More than 100 cases have been reported, mostly in children, with overall improved nutrition in approximately 80 % of patients [58, 71]. Complications have been reported after 20 % of procedures, with the complications not surprisingly including ischemia, anastomotic leaks, and recurrent dilation [58, 71]. However, follow-up for up to 10 years suggests that long-term benefits occurred in 50 % of patients, while 10 % ultimately underwent intestinal transplantation [58, 71].

An alternative method of lengthening, serial transverse enteroplasty (STEP), has been introduced, consisting of repeated applications of a linear stapling device from opposite directions in a zigzag fashion, which divides the bowel about 50 % of its diameter from either the mesenteric and antimesenteric sides or transversely [58, 73, 74]. Thompson indicated that this procedure ideally involves complete release of adhesions from the duodenum to the colon, and then a combination of tapering enteroplasties or STEP enteroplasties restore a uniform bowel lumen appropriate for the size of the patient. He typically required a bowel diameter of at least 4 cm before performing a STEP enteroplasty to maintain a subsequent lumen diameter of about 2 cm [58]. Motility can be somewhat slow to return, and in general, the full benefit of a STEP taper procedure is not often realized until 8–12 weeks after surgery [58]. More than 70 cases of STEP have been reported in the literature, with clinical improvement in 80 % of patients; 5 % underwent subsequent intestinal transplantation [71].

Thompson summarized his experience with these procedures as follows:

> Our experience with the STEP technique has been quite favorable, and it has now become our procedure of choice [58, 73, 75]. We found that 58 % of 64 patients undergoing either the Bianchi procedure or STEP were able to discontinue Parenteral Nutrition (PN). This correlated with the length gained and total length after the procedure. Overall clinical outcome is similar with STEP and Bianchi procedures. STEP avoids the difficult dissection along the mesenteric border required of the Bianchi procedure and the end to end anastomosis. While bowel may have to be more dilated to use this technique, it is more feasible in challenging areas such as near the ligament of Treitz. There are no prohibitions to performing either repeat STEP procedures or tapering enteroplasties at later operations [58].

Attempts to ameliorate the untoward effects of SBS surgically by interposing isoperistaltic or antiperistaltic bowel segments, intestinal valves, or recirculating loops; by pacing the intestine electrically; by growing new intestinal mucosa; and by transplanting small intestine have been of limited additional value to date [76]. Therefore, no operative procedure for adjunctive management of SBS currently is sufficiently safe and effective to recommend its routine use [58, 73]. Long-term parenteral nutrition remains the cornerstone of successful management of SBS, and its judicious use is recommended in appropriate amounts and formulations for as long as needed not only to ensure maximal GI adaptation and nutritional rehabilitation of the patient but also to support the optimal size and function of the body cell mass [7].

## Intestinal Transplantation in Patients with Short-Bowel Syndrome

Since the early 2000s, intestinal transplantation has been increasingly applied as a rescue therapy for patients with life-threatening complications of SBS and other forms of intestinal failure [63]. When the complications include portal hypertension or progressive liver failure, patients with SBS become candidates for combined liver/small intestine transplantation [58, 77]. The generally accepted indications for intestinal transplantation include recurrent sepsis, loss of central venous access, and development of progressive liver disease. Intestinal transplants have also been used following extensive resection of retroperitoneal neoplasms such as desmoids, fibromas, and neuroendocrine tumors, during which the superior mesenteric artery and its dependent bowel are sacrificed in deference to potential cure [58].

To date, almost 2,000 intestinal transplants have been performed in the United States, approximately 75 % of which have been in recipients under 18 years of age [58, 77]. One-year graft survival rates are currently as high as 89 % in adults aged 18–34 and as low as 64 % in children under 1 year of age [58, 77]. Graft survival drops at 5 years, with published rates ranging as low as 31 % in children under 1 year of age to as high as 69 % in children aged 6–10 years of age [58, 77]. Patient survival rates are similar at 1 and 5 years after transplant [58, 77–79]. Chronic rejection and infectious complications remain important determinants of survival, and improvements in outcomes since the mid-1990s have in large part been related to improved pediatric critical care and to judicious management of immunosuppression to reduce the incidence of opportunistic infections and post-transplant lymphoproliferative disorder [58, 79]. Overall, it is increasingly being recognized that the treatment of intestinal failure involves both nutritional and metabolic rehabilitation and transplantation, and that these approaches are complementary rather than competitive or contradictory [58].

Information regarding long-term nutritional outcome and quality of life (QOL) is continually emerging [58, 77, 80, 81]. Approximately one-third of patients undergoing intestinal transplantation require parenteral nutrition at discharge; however, at 1 year, 90 % are independent of it [58, 77]. QOL has been improved in almost all areas, but particularly related to digestive function, vocational abilities, medical compliance, optimism, and energy [58, 80]. On the other hand, this should be interpreted cautiously in view of more recent studies suggesting that QOL in SBS transplant patients remains lower than in nontransplant controls [58, 81].

Of all of the surgical approaches to SBS, intestinal transplantation has the greatest potential for treating selected patients with SBS, in terms of both the number of patients who might benefit and the functional improvement achieved [58]. With greater experience and improved results, it is hoped that this therapy can be extended to a larger number of patients with SBS [58, 63]. Thompson recommended that patients with high-risk complications of intestinal failure be referred early to a center specializing in intestinal transplantation so that patients might be carefully managed and monitored by an experienced team and, if needed, listed for transplant prior to development of complications that preclude the operation [58].

## Summary and Conclusions

Short-bowel syndrome is a form of intestinal failure following massive intestinal resection for a variety of conditions in which the remaining length of small bowel has inadequate capabilities for the absorption of the required water,

macronutrients, and micronutrients to support optimal health, functions, and performance of the body cell mass. Some of these conditions or situations are accompanied by, result in, or result from complex abdominal wall defects. Notable are the clinical scenarios that often accompany the treatment of abdominal compartment syndrome by the various "open abdomen" techniques. The complex pathophysiology of SBS was summarized together with its clinical consequences. Nutritional and metabolic management of SBS can be characterized arbitrarily by three overlapping periods of therapy, which were discussed in some detail and have withstood the tests of time for a few decades. This was followed by a summation of the more recent efforts to enhance intestinal absorption by incorporating the use of growth hormone, teduglutide, glutamine, and other nutraceuticals, in combination with dietary modifications, in attempts to reduce or obviate the use of long-term parenteral nutrition in selected patients while promoting maximal adaptation of the intestine. Surgical considerations in the adjunctive management of SBS were discussed as potential means of enhancing intestinal absorption. Increasing the exposure of the intestinal chyme to the mucosal enterocytes by decreasing intestinal transit and overcoming functional bowel obstructions with a variety of specialized surgical procedures has been helpful in appropriate patients. Of all of the surgical approaches to SBS management, intestinal transplantation may well have the greatest promise in terms of restoring GI tract function to normal as this field of endeavor continues to advance and improve its long-term outcomes. Finally, parenteral nutrition remains the cornerstone of optimally successful management of SBS, and its judicious use and monitoring by expert, experienced, dedicated nutrition support teams can ensure safe, effective, and maximal GI adaptation and nutritional rehabilitation of the patient while maintaining the optimal size and function of the body cell mass.

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