The normal human small intestine ranges between 3 and 8 m in length, depending on whether measurement is made by radiologic or surgical techniques or at autopsy when the intestine may be desicated.1–5 Short bowel syndrome is defined in adults as <200 cm of small intestine. Although usually acquired because of one or more enterectomies, short bowel syndrome may also be congenitally acquired. Nutrient, electrolyte, and fluid absorption is proportional to the amount of residual intestine. The degree of intestinal function is better described in terms of energy absorption/loss rather than length of residual intestine.6 Intestinal failure may be defined as the inability to sustain adequate nutritional, electrolyte, or hydration status in the absence of specialized nutritional support. This requires an increase in oral intake because absorption is compromised. Clinically, nutrient assimilation is a function of intake and absorption. As such, some patients with short bowel syndrome will not have sufficient loss of functional capacity as to develop intestinal failure. Significant individual variability in jejunal absorption efficiency may be encountered.7

The patients with the greatest risk for development of dehydration, generalized protein-calorie malnutrition, and multiple nutrient deficiencies are those with a duodenostomy or jejunoileal anastomosis and <35 cm of residual small intestine; jejunocolic or ileocolic anastomosis, and <60 cm of residual small intestine; or end jejunostomy with <115 cm of residual small intestine.6,8–11 Those patients with residual colon in continuity will have enhanced energy and fluid absorption. Hence, such patients can tolerate greater loss of small intestine and retain their nutritional autonomy.

**Incidence and Prevalence of Short Bowel Syndrome**

It is unclear how many individuals in the United States suffer from short bowel syndrome, but, based on the numbers in Europe, the incidence is probably approximately 2 individuals per million.12 More recent data from 1993 indicated that the incidence and prevalence of home parenteral nutrition, for which short bowel syndrome was the most prevalent indication, had increased slightly to 2 or 3 per year per million inhabitants and 4 per year per million, respectively.13,14 The most recent European survey, in 1997, indicated that the incidence of home total parenteral nutrition (TPN) had remained approximately 3 per million and that the prevalence had increased to 4 per million.15 The largest single group of patients who received home TPN were those with short bowel syndrome (35%). In comparison, the most recent data for incidence and prevalence in the United States is from 1992. At that time, it was estimated based on extrapolated data from the Oley Foundation Home TPN Registry that approximately 40,000 patients required TPN each year.16 Approximately 26% of the patients in the Oley Registry had short bowel syndrome, although some patients with a primary TPN indication of malignancy or radiation enteritis may have had short bowel syndrome as well. Patients with short bowel syndrome who did not require home TPN or were successfully weaned from home TPN are not reflected in these statistics. Approximately half of the short bowel patients who initially require TPN can be weaned off of TPN successfully in optimal settings.8 Therefore, the number of patients with short bowel syndrome may be substantially greater than previously estimated. A registry of short bowel patients, including those who require TPN permanently, transiently, and not at all, should be implemented.

**Etiology of Short Bowel Syndrome**

Short bowel syndrome may be a congenital or acquired condition. Infants may be born with congenital jejunal or ileal atresia. Otherwise, short bowel syndrome results from surgical resection of bowel. This is usually related to multiple resections for recurrent Crohn’s disease, massive enterectomy made necessary because of a catastrophic vascular event such as mesenteric arterial embolism, venous thrombosis, volvulus, trauma, or tumor resection in adults, and, in children, gastrochisis,
necrotizing enterocolitis (NEC), volvulus, and extensive aganglionosis. Functional short bowel syndrome may also occur in cases of severe malabsorption in which the bowel length is often intact. Such conditions may include chronic intestinal pseudo-obstruction syndrome, refractory sprue, radiation enteritis, and congenital villus atrophy. Severe nutrient and fluid malabsorption occurs following extensive small intestinal resection. Patients with less than 100 cm of jejunum remaining generally have a net secretory response to food.17

Patients can be grouped into 2 distinct subgroups: those with intact colon in continuity and those without colon in continuity. The colon becomes an important digestive organ in patients with short bowel syndrome. Sodium, water, and some amino acids are absorbed in the colon9,18–20 as well as energy from absorbed short-chain fatty acids.

**Postresectional Adaptation**

The intestine adapts as well to ensure more efficient absorption per unit length. Following massive enterectomy, the intestine hypertrophies, and nutrient absorption becomes more efficient. Although there are limited human data, observations in patients with massive enterectomy as well as in the functional short bowel patient (JI-bypass) indicate that the intestine lengthens some, but more importantly, diameter and villus height increase with a resultant increase in absorptive surface.21–24 This process may evolve over 1 or 2 years.6,9,25 Patients may be taught to adapt to their decreased absorption as well by dramatically increasing their food intake (hyperphagia).

The presence or absence of the colon and ileocecal valve; location (jejunum versus ileum), health, and length of residual bowel; mucosal blood flow; patient age; and comorbid conditions such as Crohn’s disease, radiation enteritis, carcinoma, or pseudo-obstruction are important determinants in the functional adaptation process and clinical outcome.6,26,27 Although the length of remaining bowel necessary to prevent dependence on TPN is approximately 100 cm in the absence of an intact and functional colon, or 60 cm in the presence of a completely functional colon,6,9,28 the degree of adaptation and TPN dependence may be highly individualized. Adaptation to full enteral nutrition has been reported with as little as 10 cm of residual intestine in infants.25 Patients with a jejunostomy are at increased risk for TPN dependence, and those with a jejunal-ileal anastomosis are less likely to be TPN dependent.

Animal models have suggested that enteroglucagon, glucagon peptide II, epidermal growth factor, growth hormone, cholecystokinin, gastrin, insulin, and neurotensin are involved in postresection intestinal adaptation.29 There are little data on the role of either increased endogenous hormonal release or exogenous hormone supplementation during the intestinal adaptation phase in humans.

Residual ileum is able to adapt and to assume the role of macronutrient absorption when jejunum has been resected. However, the specialized cells of the terminal ileum in which vitamin B-12/intrinsic factor receptors are located and in which bile salts are reabsorbed cannot be replaced by jejunal hypertrophy.

**Medical Therapy of Short Bowel Syndrome**

The goal of medical therapy is for the patient to resume work and a normal lifestyle, or as normal of one as possible. This is undertaken via the use of specific measures to decrease gradually the requirement for TPN and, at best, to eliminate its need. The most important aspects of the medical management of the patient with short bowel syndrome are to provide adequate nutrition, including both macro- and micronutrients, to prevent energy malnutrition and specific nutrient deficiencies, to provide sufficient fluid to prevent dehydration, and to correct and prevent acid-base disturbances. Most macro-nutrients, including carbohydrate, nitrogen, and fat, are absorbed within the first 100 cm and up to 150 cm of jejunum.30

**Macronutrient Assimilation and Dietary Therapy**

Typically, patients who have undergone massive enterectomy require TPN for the first 7–10 days. The immediate goals during this period are survival and hemodynamic stability. Nutritional therapy should not be introduced until the patient is hemodynamically stable and fluid management issues are relatively stable. Patients should be provided approximately 25–35 kcal/kg/day and 1.0–1.5 kg/day of protein. It remains controversial whether intake should be based on actual body weight or ideal body weight.31 Enteral nutrition should be started as soon as possible once hemodynamic stability has been achieved; standard enteral formula is recommended. Enteral nutrition should be instituted gradually as tolerated. Once patients are able to eat, they should be encouraged to eat a regular diet but modified as described below. Patients should also be encouraged to adapt their diet to eat substantially more than what was usual prior to their catastrophic event (hyperphagia).32
nutrient, electrolyte, or mineral absorption or fecal volume or fecal weight.19

Proteins/Amino Acids

Dietary protein is first digested by pancreatic, gastric, and intestinal proteases and then absorbed as di- and tripeptides. Therefore, it was reasoned that if dietary protein were provided in a predigested form, it would be more readily absorbed. However, absorption of nitrogenous macronutrients (or proteins) is least affected by the decreased intestinal absorptive surface in patients with short bowel syndrome. Therefore, the utility of peptide-based diets in such patients is theoretically without merit.

McIntyre et al fed 7 patients with an end-jejunostomy (60–150 cm residual small bowel) a peptide-based or an essentially isocaloric and isonitrogenous polymeric formula. Energy, carbohydrate, nitrogen, fat, electrolyte, fluid, and mineral absorption and stool weight were similar regardless of the enteral formula provided.39 Uncontrolled data from Levy et al support these findings.34 A small study of 6 patients (90–150 cm of residual jejunum and end-jejunostomy) reported by Cosnes et al, however, suggested that nitrogen absorption may be modestly improved with the use of a peptide-based diet, although similar to previous studies, energy, other macronutrient, electrolyte, mineral, and fluid absorption were unaffected.35 These studies were all very small, the study populations somewhat heterogeneous, and the various peptides used in the formulas differed significantly, as did the type and amount of fat (long-chain triglycerides vs medium-chain triglycerides). It is therefore difficult to make definitive comparisons among studies.

The amino acid glutamine, together with glucose, is the preferred fuel for the small intestinal enterocyte.36 Rodent TPN models suggested that either parenteral or enteral glutamine supplements could effect more rapid and more significant bowel adaptation following massive enterectomy.37,38 Therefore, it was thought that glutamine supplementation in humans would have a similar effect. Although an early case series of 10 patients who were beyond the usual 1–2 year adaptation period suggested that glutamine, combined with growth hormone supplementation, and a high complex carbohydrate diet could result in decreased stool output and increased absorption of energy, protein, carbohydrate, sodium, and water,39 2 subsequent, double-blinded, randomized placebo-controlled trials failed to confirm any of these effects.40,41 In addition, Scaplando et al showed that glutamine and growth hormone supplementation did not lead to morphologic changes in the intestine.40 Glutamine-supplemented oral rehydration solution (see fluid and electrolyte management) was associated with decreased Na absorption and a trend toward decreased fluid absorption in a small controlled trial in 6 patients.41 All of the patients studied by Byrne et al39 had colon in continuity; it is likely the treatment-associated increase in energy absorption was related solely to the increased complex carbohydrate diet as discussed above. Treatment with growth hormone and glutamine in the setting of short bowel syndrome has been associated with significantly increased extracellular fluid and peripheral edema.40,42,43 A recent study reported by Seguy et al showed that low-dose growth hormone (0.05 mg/kg/day) was associated with a modest increase in carbohydrate and nitrogen absorption.44 A recent randomized, placebo (glutamine)-controlled study indicated growth hormone administration, when used outside the natural adaptation period, led to the ability to reduce parenteral fluid and nutrition requirements, although absorptive studies were not undertaken.45 Neither growth hormone nor glutamine have been studied in humans when used in the initial 1–2-year periadaptive period. Therefore, treatment with glutamine and/or growth hormone cannot be recommended at the present time. However, it may be that use of such exogenous growth factors may have their greatest utility if used during the normal adaptive period. There are no human studies to test this hypothesis.

Lipid

Lipid digestion may be impaired because of impaired solubility because micelle formation will be impaired when ileal bile salt malabsorption occurs in the setting of ileal resection (>100 cm).46 Treatment with ox bile supplements has been attempted in 3 patients in an attempt to increase duodenal bile salt concentration to stimulate micellar lipid solubilization.47–49 This therapy has been associated with significantly increased fecal volume, at least in those patients with intact colon. A preliminary, open-labeled study of 4 patients (2 with colon in continuity) indicated that treatment with the conjugated bile acid cholyxarscine (6 g/day) was associated with an increase in fat absorption of 17 ± 3 g/day without any effect on stool wet weight.50 Cholyxarscine is a conjugated bile acid and is resistant to colonic bacterial deconjugation. Another case series of 4 subjects had similar findings.51 However, 1 of the 4 patients experienced a significant increase in wet stool output, and nausea developed in another patient. Cholestyramine should not be used in patients with >100 cm of ileal resection because it may actually worsen steatorrhea because of the binding of dietary lipid.52 Regardless of whether a high- or low-fat diet is used, the percentage of fat absorption remains stable, and stool
weight is unaffected by the fat content of meals. However, because of the energy density of fat (9.0 kcal/g) when compared with carbohydrate (4.0 kcal/g), it is an important dietary energy source. In addition, in short bowel syndrome patients, up to 65% of dietary carbohydrate may be malabsorbed and lost in the feces without degradation by colonic bacteria.

A mixed long-chain triglyceride:medium-chain triglyceride (LCT:MCT) diet was studied in 19 patients. Those patients with intact colon absorbed 96% ± 3% of C8 fatty acids and 87% ± 6% of C10 fatty acids, and those patients without colon absorbed 63% ± 25% of C8 and 57% ± 28% of C10, (P = .007 for C8 and P = .004 for C10). MCT contains 8.3 kcal/g. Energy absorption (approximately 2.1 MJ/day; 500 kcal/day) was significantly increased in patients with colon, although the LCT:MCT diet did not result in increased energy absorption when compared with the LCT-based diet in patients with an end-jejunostomy or ileostomy. These patients also had increased fecal output with a mixed LCT:MCT diet. Some, but not all LCT, can be replaced by MCT in the diet because the essential fatty acid linoleic acid is a constituent of LCT and is not found in MCT. Excessive MCT intake may result in nausea, vomiting, and ketosis. In a short bowel patient eating 10.5 MJ/day (2500 kcal/day), approximately 1.5–3 MJ/day (360–720 kcal/day; 40–80 g) of LCT can be replaced with MCT.

Carbohydrates

The proximal jejunum rarely requires resection in patients that undergo massive enterectomy. Most intestinal dissacharidases are present in highest concentration in the very proximal small intestine, and, hence, patients with more distal resection are not likely to benefit from a lactose-free diet. In a study of 14 short bowel patients, a lactose-free diet was compared with a diet containing 20 g/day of lactose (still with <4 g milk). Lactose absorption, breath hydrogen, subjective symptoms of flatulence, and diarrhea were similar regardless of the diet. These data confirmed the findings of an earlier controlled study in 17 short bowel patients, which found that lactose absorption was enhanced when provided in yogurt rather than via milk. Regardless, in the absence of significant jejunal resection, patients with short bowel syndrome should not be provided lactose-restricted diets. Such diets are also generally low in calcium. Even patients with an end-jejunostomy will generally tolerate a glass of milk (20–25 g lactose).

The Role of the Complex Carbohydrate Diet

Soluble nonstarch polysaccharides and some starches are not generally absorbed by the small intestine. Soluble fiber dissolves in water and is found primarily in oatmeal, oat bran, psyllium (Metamucil, Proctor & Gamble, Cincinnati, OH; Konsyl, Konsyl Pharmaceuticals, Easton, MD), barley, artichokes, strawberries, legumes, prunes, grapefruit, and squash in descending order of concentration. Soluble fiber and starches pass undigested into the colon in which they are fermented by enteric bacteria into hydrogen and methane (hence patient “gas” complaints) but also into short-chain fatty acids (SCFAs), including butyrate, propionate, and acetate. SCFAs are the preferred fuel for colonic enterocytes. Therefore, the colon becomes a digestive organ in patients with short bowel syndrome. Approximately 75 mmol SCFA are produced from 10 g unabsorbed carbohydrate. Patients with short bowel syndrome but intact colon in continuity were able to decrease fecal energy loss by 1.3–3.1 MJ/day (310–740 kcal) when they were fed a diet consisting of 60% carbohydrates. Colonic metabolism of unabsorbed carbohydrate was indicated by decreased fecal carbohydrate losses in the patients with colon in continuity. The intact colon may absorb up to 2.2–4.9 MJ (525–1170 kcal) of energy daily from dietary fiber. Colonic energy absorption may also increase some during the postresection adaptation phase, related to increased colonic bacterial carbohydrate fermentation. The increased fermentation may result from increased colonic bacteria and/or an increase in the concentration or activity of various enzymes including β-galactosidase during the adaptation period. Because SCFAs stimulate sodium and water absorption, patients might be expected to have decreased fecal fluid and sodium loss as adaptation progresses, although this has not been observed clinically.

Vitamins

Micronutrients often require supplementation. It is unusual for water-soluble vitamin deficiencies to develop in short bowel patients in the absence of TPN (except in those who have proximal jejunosomies or duodenostomies) because these are absorbed in the proximal jejunum. Thiamine deficiency has been described, especially during a recent parenteral vitamin shortage. Patients may present with Wernicke’s encephalopathy, beriberi, and severe metabolic alkalosis. Whole blood thiamine concentration may not be indicative of deficiency as it reflects recent nutritional intake. Erythrocyte transketolase activity should be measured and empiric therapy begun with 100 mg parenteral thiamine daily. Biotin deficiency has rarely been reported in patients with short bowel syndrome. Scaly dermatitis, alopecia, lethargy, hypotonia, and lactic acidosis have been described. Therapy consists of parenteral biotin supplemen-
Fat-soluble vitamin deficiency (A, D, and E) is more common in patients with short bowel syndrome because of fat maldigestion related to decreased bile salt reabsorption and decreased micelle formation. Cholestyramine may cause fat-soluble vitamin deficiency because of its binding of bile salts. Night blindness and xerophthalmia may be described in short bowel syndrome. If not recognized, vitamin A deficiency may progress to corneal ulceration and later permanent visual loss may develop. The serum vitamin A concentration should be monitored in patients who do not require parenteral vitamin supplementation. If a low serum vitamin A concentration is detected, therapy should be started with 10,000–50,000 units daily, administered either orally or parenterally.

Vitamin D deficiency manifests in osteomalacia. Usually, dietary intake is a relatively unimportant source of vitamin D because the majority is endogenously synthesized from 7-dehydrocholesterol via ultraviolet light. However, because enterohepatic circulation is disrupted in patients who have undergone significant ileal resections, deficiency may result.

Vitamin E deficiency may manifest in hemolysis and various neurologic deficits. Because serum vitamin E concentration reflects serum total lipid concentration, which may be low in short bowel patients who have steatorrhea, low serum vitamin E concentration alone may not be indicative of a deficient state; the serum vitamin E:total lipid ratio should be calculated. Most vitamin K is synthesized by colonic bacteria (60%), although dietary intake accounts for approximately 40% of requirements; deficiency is therefore uncommon in patients with intact colon. However, vitamin K deficiency is frequent in patients who have no residual colon or who have taken broad-spectrum antibiotics recently. The requirement is approximately 1 mg daily. Vitamin K was not a constituent in adult multivitamins for TPN until recently, although it has in children.

**Trace Metals**

Significant amounts of zinc and selenium are lost with diarrhea. A significant amount of zinc is also lost in small bowel effluent (12 mg/L small intestinal fluid and 16 mg/L stool). Zinc deficiency has been associated with growth abnormalities, delayed wound healing, and cellular immunity dysfunction. Patients in whom zinc deficiency is suspected should be treated empirically with oral zinc sulfate (220–440 mg daily) or parenteral zinc supplements if they require TPN. Serum and leukocyte measurements of zinc concentration, although helpful, may be unreliable. Selenium deficiency has been associated with cardiomyopathy; peripheral neuropathy, proximal muscle weakness, and pain; whitening of the hair; and macrocytosis. Serum selenium is a reliable indicator of selenium status, and, if low, oral or parenteral supplementation should be provided. Although there are 3 reported possible cases of chromium deficiency in patients requiring long-term TPN, deficiency has not been reported in short bowel patients who do not require TPN, and, therefore, routine supplementation is not recommended. Chromium is a necessary cofactor for insulin’s effects in peripheral tissue. However, available evidence suggests that there is sufficient chromium present in the TPN solutions as a contaminant, and supplemental chromium may result in the possibility of nephrotoxicity. Copper deficiency is very rare in the patient with short bowel syndrome. Deficiency may result in microcytic anemia, neuropathy, and decreased fertility.

**Medication Absorption**

Medication absorption is often impaired, just as is nutrient absorption in patients with short bowel syndrome, although significant interpatient variability may be observed. The oral or enteral route for medication delivery should be used whenever possible to avoid additional manipulations of the TPN catheter, which are associated with increased infection risk. The degree to which a medication is malabsorbed is dependent on the residual small bowel surface area and its health (for example, whether or not active Crohn’s disease is present) and morphologic and physiologic factors, including the presence or absence of the terminal ileum (especially important for vitamin B-12 and bile salt absorption, which are necessary for the absorption of lipid-soluble medications such as cyclosporin) or the presence of an acidic or alkaline environment (which may be caused by the use of H2 blockers in TPN or proton pump inhibitors). Many, but not all, medications are absorbed in the jejunum. Therefore, absorption will be minimally impacted for most medications in the absence of decreased intestinal transit time, which will decrease mucosal contact time. Most of the available data on oral medication absorption in patients with short bowel syndrome are in the form of isolated case reports. The sublingual route
for medication delivery may be preferable for some medications.

**Fluid and Electrolyte Management**

Massive enterectomy is associated with transient gastric hypersecretion and hypergastrinemia, generally lasting for up to 6 months following resection. H₂ antagonists and proton pump inhibitors are useful to reduce gastric fluid secretion and, therefore, also reduce fluid losses during this period. However, because absorption of orally dosed medications may be impaired, either large doses or intravenous delivery may be required. Although fluid losses are decreased, macronutrient and electrolyte absorption are not affected by the increased gastric secretion. Reduction of gastric acid output is also important to help prevent the deconjugation of bile acids in the duodenum and decreased pancreatic lipase excretion to enhance fat digestion.

Fluid losses usually require chronic control with antimitoty agents such as loperamide hydrochloride or diphenoxylate. Typical doses are 4–16 mg/day. If these are ineffective, especially in patients with no colon in continuity or those who have a minimum of residual jejunum or duodenum, codeine sulfate or tincture of opium may be necessary. The usual dose for codeine is 15–60 mg, 2 or 3 times daily. Rarely, patients will require treatment with octreotide. Octreotide’s mechanism of action is unclear, but it may be useful to slow intestinal transit time, which will increase water and sodium absorption. Daily jejunostomy volume was reduced from 8.1 ± 1.8 to 4.8 ± 0.7 L/day using a dose of 100 μg, subcutaneous, 3 times daily, of octreotide 30 minutes before meals in one open-labeled study of 9 patients with end-jejuno stomies. However, octreotide use does not result in TPN discontinuation, and it reduces splanchnic protein synthesis, thereby reducing use does not result in TPN discontinuation, and it reduces splanchnic protein synthesis, thereby reducing

**Glucose-polymer-based oral rehydration solutions** should be used to improve hydration and to decrease TPN fluid requirements in patients with residual jejunum ending in a jejunostomy. Patients with <100 cm of residual jejunum have significant risk for dehydration because they secrete more sodium (Na) and fluid than they orally consume. Because the jejunum is permeable to Na and chloride (Cl), passively absorbed solutions with high NaCl concentration are readily absorbed. Glucose promotes salt and water absorption by solvent drag and via stimulation of sodium-linked transport. However, although Na and water are absorbed from hypotonic isotonic solutions delivered into the jejunum, absorption is superior with more hypertonic fluids. Therefore, hypotonic oral rehydration solutions may be of benefit in patients with an intact gastrointestinal tract and acute infectious diarrhea but may not be of use in the short bowel patients. Ingestion of hypotonic fluids may be associated with increased Na loss. Several commercially available oral rehydration solution (ORS) formulas are available, although probably the best, and certainly the least expensive, is that recommended by the World Health Organization (WHO). This can be formulated by dissolving the following in 1 liter of tap water: NaCl (2.5 g), KCl (1.5 g), Na₂CO₃ (2.5 g), and glucose (table sugar, 20 g). Only the KCl requires a physician prescription. Most, if not all, of the commercially available ORS have substantially less sodium (in the range of 45–50 mmol/l). Less NaCl may be added, but the optimal Na concentration should be at least 90–100 mmol/L, which is the usual concentration of small bowel effluent. ORS solutions with lower sodium concentrations result in increased sodium losses. Therefore, patients with short bowel syndrome should be cautioned against consumption of plain water and should be encouraged to drink ORS whenever they are thirsty. More recent evidence has shown that hypotonic (160 mosm/kg) ORS leads to decreased intraluminal duodenjal fluid flow rate in normal volunteers, although the effect on gastrointestinal fluid losses or fluid status in patients with short bowel has not been evaluated.

ORS may still be useful even in patients with residual colon in continuity, but, provided sufficient sodium is present in the diet, the amount of Na in the ORS may not be as critical because the colon readily absorbs Na and water against a steep electrochemical gradient. The presence of glucose in the ORS is not critical in patients with no remaining jejunum, but who have residual ileum, because ileal water absorption is not affected by the presence of glucose.

In addition to Na losses, significant quantities of magnesium (Mg) are lost in jejunal or ileal effluent. Given that magnesium deficiency may develop despite a normal serum magnesium concentration, it is prudent to measure 24-hour urine Mg loss. The median 24-hour urine Mg in normal volunteers in one study was 127 mg (vs 19 mg for magnesium deficient short bowel patients).
Mg deficiency may cause calcium (Ca) deficiency because hypomagnesemia impairs parathyroid hormone release. In addition, the majority of patients with short bowel syndrome who do not require TPN are in negative Ca balance. Therefore, in the absence of TPN, oral Ca supplementation is recommended routinely (800–1200 mg/day). Mg replacement is problematic. Attempts with oral MgO or oral consumption of injectable Mg are generally not successful and have been associated with increased fecal loss because of their cathartic effect. Although Mg gluconate is water soluble, Mg has not generally been a constituent of ORS. Therefore, some patients may require periodic parenteral Mg infusion despite the absence of a TPN or intravenous fluid requirement otherwise. Iron is absorbed in the duodenum and, therefore, in the absence of hemorrhage, is not routinely required as a supplement. Phosphorous deficiency is not well described in short bowel syndrome, and, therefore, supplementation is rarely, if ever, required.

Pharmacologic Enhancement of Intestinal Adaptation

Glucagon-like peptide 2 (GLP-2) is another hormone secreted from L cells (as well as from pancreatic A cells). Postprandial serum GLP-2 concentration is not unexpectedly depressed in patients who have had extensive small bowel resection, especially including ileum. It has been postulated that the relative lack of jejunal hypertrophy following ileal resection may be at least in part related to the resection of GLP-2-producing L cells, although these investigations were undertaken in patients some 2 years following their resection without a baseline comparison. Studies have not been undertaken in patients immediately following resection. In a similar patient population, Jeppesen et al suggested that subcutaneous injection of GLP-2 resulted in modestly increased nutrient and fluid absorption. More recently, a longer-acting analogue of GLP-2 has shown similar effects.

Dietary Restriction

Normally, oxalate in the diet binds to dietary calcium and is excreted in stool. However, in the presence of significant fat malabsorption, dietary calcium preferentially binds to free fatty acids, rendering the oxalate free to pass into the colon in which it is readily absorbed; dietary oxalate is absorbed only to a minimal extent in the small intestine. Oxalate absorption may also be enhanced in the colon because of increased permeability because of injury caused by malabsorbed bile salts, which flow into the colon. Oxalate is renally filtered and then bound to calcium once absorbed in the colon. This results in hyperoxaluria with subsequent calcium oxalate nephrocalcinosis and nephrolithiasis. Therefore, patients with short bowel syndrome with colon in continuity should be prescribed an oxalate-restricted diet. Because calcium does bind to oxalate, oral Ca supplements may be used to prevent Ca-oxalate nephrolithiasis. Hyperoxaluria may also develop in any patient receiving TPN because the vitamin C contained in the TPN solution is metabolized to oxalate when exposed to light. TPN should therefore be shielded from light, but it remains unclear whether dietary oxalate should be restricted in patients without colon in continuity who require TPN.

Providing Parenteral Nutrition/Macronutrients

Most patients with short bowel syndrome will require TPN, at least initially. For the normally nourished patient, TPN should be supplied at 25–30 kcal/kg/day based on ideal body weight for adults, with greater levels of support for infants and children depending on age. Dextrose is a monohydrate and, as such, provides 3.4 kcal/mL (vs 4.0 kcal/mL for most carbohydrates). The maximum dextrose infusion rate should be 5–7 mg/min. Blood glucose concentration should be monitored at least daily (optimally 4 times a day) and should be below 160 mg/dL. If blood glucose exceeds this level, glucose spills in the urine, energy is lost, dehydration may ensue, and infection risk is increased. The addition of regular insulin to the TPN solution may be required. If insulin is required, it should be added to the TPN bag at an initial dose of 0.1 units per gram of dextrose, with subsequent adjustments as necessary. Intravenous lipids are generally used to provide 20–30% of infused calories, although a greater percentage of lipid may be used in the patient with significant glucose intolerance or difficult fluid management; 20% lipid emulsion is more calorically dense than dextrose. Generally, the percentage of lipid calories should be increased and the percentage of dextrose calories decreased if the amount of supplemental insulin required exceeds 0.2 units per gram of dextrose. The serum triglyceride concentration should be kept under 700–800 mg/dL and, optimally, <400 mg/dL. Protein is supplied in the form of amino acids and should be supplied at 1.0–1.5 g/kg/day, based on ideal body weight for adults; greater levels of support are required for infants and children depending on their age.
Getting the Patient Ready for Home TPN

Initially, TPN is infused continuously while postoperative complications are addressed and metabolic issues stabilized. Attempts should be made, when appropriate, to wean patients who have sufficient absorptive capacity as discussed above, being mindful that maximal adaptation requires up to 1–2 years. For patients who will require TPN at home, the TPN infusion should be compressed to nighttime infusion. Typically, the infusion would run over a 10–12-hour period with an additional 30–60-minute taper period; some patients with fluid management issues from renal failure or congestive heart failure will be unable to tolerate this infusion rate and will require either a longer infusion time or less volume. Because rapid carbohydrate and fluid infusion may be associated with intra- and extracellular fluid shifts, some patients may develop extremity cramping from extracellular potassium deficiency, which may be corrected by slowing the infusion rate. Pancreatic insulin secretion requires some time to adapt to the significant dextrose infusion; therefore, the cycling of TPN to nighttime use should be a gradual process. The infusion time for TPN is generally compressed by 2–4 hr/day, depending on the total infused volume and electrolyte and glucose measurements. TPN should be infused ideally via a single lumen catheter, with its tip positioned in either the superior or inferior vena cava to decrease the risk of infection and thrombosis.128,129 Tunneled catheters, including Hickman, Broviac, or Groshong catheters; implantable ports; or percutaneously inserted central catheters (PICCs) should be used at home, although the experience with PICCs for >1 year at home is minimal. To qualify for Medicare reimbursement, home TPN must be required for at least 3 months, fat malabsorption must be documented, and enteral feeding must have failed.

Discussion

The initial management of the patient with newly developed short bowel syndrome is complex. The goals are to correct and prevent nutritional deficiencies; to maintain normal nutritional status and growth, especially in children; to prevent complications associated with short bowel syndrome; to decrease diarrhea; and to improve quality of life. To do this effectively, one should assess the patient’s absorptive status and nutrient losses, assess length of residual intestine, reanastomose residual colon so that it remains in continuity with the residual small intestine, evaluate specific or at-risk nutrient deficiencies, identify risk factors for medical complications, and evaluate the home and work environments to determine quality of life.

Highly selected patients with dilated segments of small intestine may benefit from intestinal lengthening procedures; however, these surgeries remain controversial, often with poor outcomes, and therefore should not be considered part of the initial management of patients with short bowel syndrome. Similarly, because many patients may be successfully weaned from home TPN using the conservative medical measures discussed herein, small intestinal transplantation is inappropriate in the initial management of the patient with short bowel syndrome. It should be reserved for the patient who develops irreversible malabsorption-associated liver disease; other indications remain controversial and are not supported by the available data.130

With appropriate medical and dietary therapy, many patients with short bowel syndrome can be weaned from parenteral nutrition. For those who cannot, the administration of intestinal mucosal growth factors, currently investigational, may prove useful.

References


Received July 29, 2004. Accepted July 14, 2005.
Address requests for reprints to: Alan L. Buchman, MD, MSPH, Northwestern University Medical School, Division of Gastroenterology and Hepatology, Feinberg School of Medicine, 676 N. St. Clair Street, Suite 1400, Chicago, Illinois 60611. e-mail: a-buchman@northwestern.edu; fax: (312) 695-4514.