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CLINICAL PRACTICE UPDATE

AGA Clinical Practice Update on Management of Short Bowel Syndrome: Expert Review

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Short bowel syndrome (SBS) is a devastating clinicopathological syndrome resulting from the loss of intestinal length due to disease or surgical resection. There is general agreement that a residual small intestinal length of 200 cm or less meets criteria for SBS.[1] although there are reports suggesting that a residual length of 150 cm or less may be more appropriate.[2] The general consequences of SBS including diarrhea, dehydration, electrolyte abnormalities, and weight loss in the longer term are predictable due to loss of digestive and absorptive surface area. Specific consequences are related to the regions of the gastrointestinal tract that are missing, such as vitamin B12 deficiency or bile acid–induced diarrhea from resection of the terminal ileum. Such specific findings provide important clinical clues that can help refine management.

The terms *SBS* and *intestinal failure* (IF) have much in common and are frequently used interchangeably. It is important to emphasize the subtle but important distinction between the 2 terms. IF is defined as the reduction of gut function below the minimum necessary for the absorption of macronutrients or water and electrolytes such that intravenous supplementation, often in the form of parenteral nutrition (PN), is required to maintain health or growth.[3] Most cases of IF are due to SBS, although IF may also be due to other disorders such as chronic intestinal pseudo-obstruction and refractory intestinal malabsorptive syndromes. The focus of this Clinical Practice Update is strictly on SBS, and we will limit further discussion to the clinical characterization and management of SBS only, even if some of the management principles may be applicable to other functional causes of IF.

Methods

This article provides practical advice based on the best available published evidence, taking into account recently published systematic reviews and clinical guidelines. This best practice document is not based on a formal systematic review. The best-practice advice presented in this document is focused on adult patients with SBS and SBS-IF; however, some overlap with the management of pediatric SBS may be present.

This expert review was commissioned and approved by the American Gastroenterological Association (AGA) Institute Clinical Practice Updates Committee and the AGA Governing Board to provide timely guidance on a topic of high clinical importance to the AGA membership, and underwent internal peer review by the Clinical Practice Updates Committee and external peer review through standard procedures of *Clinical Gastroenterology and Hepatology*.

Best Practice Advice 1: Bowel Anatomy

The definition of SBS is underpinned by an accurate estimation and reporting of residual bowel length. Surgeons involved with massive bowel resections should report the residual bowel length, rather than the length of bowel resected. It is only the former that dictates outcome and cannot be reliably estimated if only the length of resected bowel is known. By convention, the length of residual bowel is measured at the time of surgery along the antimesenteric border of unstretched bowel, from the duodenojejunal flexure to the ileocecal junction, the site of any small bowel–colon anastomosis or to the end-ostomy. Based on the presence or absence of residual colon, SBS patients may be classified into 3 groups[4,5]: group 1, end-jejunostomy; group 2, jejunum anastomosed to partial colon (ie, jejunocolonic anastomosis); and group 3, jejuno-ileo-colic anastomosis, retaining entire colon and ileocecal valve (Figure 1). The relationship between residual bowel anatomy and prognosis has been clearly described by Messing et al.[6,7] Group 3 represents the most favorable anatomic phenotype of SBS, while group 1 represents the most severe phenotype, and patients with high-output end-jejunostomies are the most challenging SBS patients to manage. Knowledge of postresection gastrointestinal

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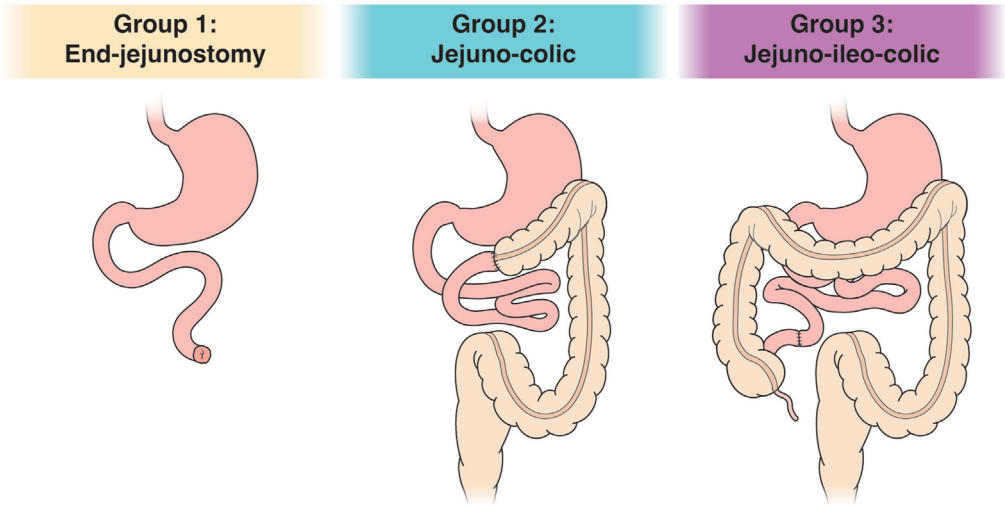

Figure 1. Anatomic classification of SBS.

anatomy also provides an overarching framework for the surgical management of patients with SBS—any surgical opportunity to convert group 1 patients toward group 2 or 3 (ie, by restoring continuity with any remaining small bowel or colon, if present) will generally improve overall prognosis and outcomes subject to presence of any underlying active bowel disease (eg, Crohn's disease or radiation enteritis).[8]

Best Practice Advice 2: Nutrition Assessment

An initial comprehensive nutritional assessment should be performed by a dietitian experienced in SBS on all patients. Supplementary Table 1 lists items that should be assessed.[9] Subsequent long-term monitoring of these patients should include laboratory studies (eg, electrolytes, liver/kidney tests), fluid balance, weight change, serum micronutrient levels, and bone density, as described subsequently.

Fluid and electrolyte problems may affect the course of the patient with SBS, particularly those without a colon who exhibit large enteric losses, and may result in chronic dehydration and kidney disease; nephrolithiasis; sodium deficiency, often without accompanying hyponatremia; and difficult-to-replete hypokalemia, hypomagnesemia, and hypocalcemia.[10] Regular monitoring of renal function and fluid balance should be performed. Patients with SBS should be instructed on the measurement of daily urine output, at a minimum, as periodic assessment can help guide fluid needs; adequate hydration is generally based on a goal urine output of >1 L/d and a urinary sodium concentration >20 mEq/L.

Serial weight measurements are useful to track trends and serve as a warning of compromise in nutrition or hydration status. Because water-soluble vitamins are absorbed in the proximal small bowel, deficiencies in SBS patients are uncommon. In contrast, fat-soluble vitamin and essential fatty acid deficiencies are relatively frequent, and large doses may be required to maintain normal plasma levels. In all patients with SBS on or off PN, serum vitamin and trace element concentrations (Table 1) should be measured at baseline and monitored at least once per year; however, the frequency of monitoring will depend on the presence of existing or prior deficiencies.

A spectrum of metabolic bone disease commonly occurs in patients with SBS because of the effects of the PN, altered bowel anatomy causing malabsorption of macro- and micronutrients, and other underlying patient factors. An assessment of bone density should be undertaken in all SBS patients and repeated periodically every 2–3 years, given the ongoing risk factors present in the patient with SBS, possibly more often in the patient with osteoporosis initiating or changing therapy, at least until stable.[11] Periodic monitoring of calcium, phosphorus, magnesium, vitamin D (25-hydroxyvitamin D), parathyroid hormone status, and presence of metabolic acidosis should also be done.

Best Practice Advice 3: Diet

Dietary therapy in SBS should focus on maintaining compensatory hyperphagia, rather than on excessive dietary restrictions.[12] Because most adult patients with SBS have significant malabsorption, dietary intake must be increased by at least 50% from their estimated needs (ie, hyperphagic diet). The increased quantity of food is best tolerated when consumed throughout the day divided in 5–6 meals. Dietary counseling should be guided by an experienced dietitian and based on the preferences of the patient to ensure high compliance with adjustments based on tolerance as determined by the development of symptoms, stool output, and weight.

Few studies have compared the effects of oral diet composition in patients with SBS and those that have generally involved small, heterogeneous populations of subjects.[13] In those patients with SBS and colon in continuity, a high-carbohydrate (60%), low-fat (20%) diet

Table 1. Vitamin and Mineral Levels to Be Assessed and Typical Supplementation in Short Bowel Syndrome

Micronutrient	Lab Measurement	Typical Supplementation
Vitamin A	Serum retinol	Oral: 5000–50,000 IU daily (sometimes more); IM administration also available
Vitamin B12	Serum vitamin B12, methylmalonic acid	SC/IM: 300–1000 μg monthly; oral and intranasal administration also available
Vitamin C	Serum vitamin C (ascorbic acid)	Oral: 200–500 mg daily; IV administration also available
Vitamin D	Serum 25-hydroxyvitamin D, parathyroid hormone	Oral: 50,000 IU once weekly (or calcitriol 0.25–2 μg daily); IM administration also available
Vitamin E	Serum alpha-tocopherol	Oral: 400 IU up to 3 times daily
Folate	Serum, red blood cell folate	Oral: 1 mg daily
Iron	Serum ferritin, iron, iron-binding capacity	Oral: 100–200 mg once daily or every other day; IV and IM administration also available
Zinc	Serum zinc	Oral: 50 mg elemental zinc (220 mg tablet) once or twice daily
Selenium	Serum selenium	Oral: 100–200 μg daily
Chromium	Serum chromium	Oral: 100–200 μg up to 3 times daily
Multivitamin		Oral: 1–2 capsules daily
Copper	Serum copper	Oral: 2 mg elemental copper daily (higher dose may be needed); IV administration also available
Manganese (in patients on home PN)	Serum manganese	None

IM, intramuscular; IV, intravenous; PN, parenteral nutrition; SC, subcutaneous;

tends to reduce fecal calorie loss, increase overall energy absorption, improve wet weight absorption, and reduce magnesium and calcium loss and oxalate absorption. In contrast, end-jejunostomy patients do not appear to benefit from fat restriction[14,15]; more fat is absorbed when more is consumed. Therefore, while recognizing the limitations in the evidence, SBS patients with a preserved colon should consume a diet high in complex carbohydrates and relatively low in fat. Oxalate restriction (eg, peanuts and baked beans) in those with a colon who are at risk of hyperoxaluria and oxalate stones is essential.

What about enteral (tube) feeding in SBS? Unlike the situation with children with SBS, there are few published reports of the use of enteral nutrition (EN) support in adult patients with SBS, and it appears to be rarely used.[16] Nonetheless, the benefit of tube feeding, albeit over the short term, was demonstrated in a small but elegant randomized crossover study of 15 adult SBS patients that showed that tube feeding, either exclusively or in combination with oral feeding increased net absorption of lipids, protein, and energy in adult SBS patients after the postoperative period.[17] We suggest consideration of EN (ie, tube feeding) in combination with oral feeding in stable patients with SBS-IF and insufficient oral intake despite use of an appetite stimulant (eg, mirtazapine, olanzapine, dronabinol, Megace) when stool output is <2 L/d and the expected gains with tube feeding may allow weaning from PN. For long-term EN, intermittent self-placement of a nasogastric tube (eg, nightly for overnight infusion) by the patient or placement of a percutaneous gastrostomy tube are considerations. Importantly, placement of a percutaneous gastrostomy tube may be technically difficult due to altered anatomy and adhesions often present in SBS. Therefore, a discussion with the patient regarding the risks, benefits, and alternatives of this approach to nutrition support is necessary. Additionally, a temporary trial of nasogastric tube feeding should be considered to ensure tolerance before subjecting the patient to the risk of percutaneous tube placement. Once enteral access has been obtained, slow continuous infusion of the formula into the stomach, rather than bolus administration or infusion directly into the small bowel, is suggested to maximize intestinal transit time and improve nutrient contact time and absorption and reduce diarrhea.[18]

Best Practice Advice 4: PN

Virtually all patients with SBS require PN support in the initial period following resection, and few will be able to discontinue the PN prior to their discharge from the hospital. Although >50% of adults with SBS are able to be weaned completely from PN within 5 years of diagnosis, the probability of eliminating PN use is <6% if not

successfully accomplished in the first 2 years following the individual's last bowel resection.[6,7] Tunneled central venous catheters are preferred for long-term PN as opposed to peripherally inserted central venous catheters because of the higher risk of thrombosis and issues related to self-administration of PN with the latter.[19] Similarly, tunneled catheters are preferred over totally implanted devices (ie, "ports") in the patient on long-term PN patient, as the principal benefit of the port is not realized given that the device needs to continually be accessed and exchanged on a weekly basis. PN should be initiated and adjusted to meet the patient's fluid, electrolyte, energy, protein, and micronutrient needs. Overall energy content and macronutrient composition will depend on the patient's oral intake and the level of repletion required. When ostomy output is high, increased fluid, potassium, magnesium, and zinc losses occur and need to be monitored and replaced appropriately. When calculating PN volume and content, changes in the patient's weight, laboratory results, stool or ostomy output, urine output, and complaints of thirst should be monitored. The amount of PN can be decreased when the patient demonstrates the ability to take oral nutrition without excessive stool or ostomy output and with appropriate weight maintenance or gain. Importantly, the patient with SBS on home PN remains at risk for micronutrient deficiencies and requires periodic monitoring and supplementation alongside PN.[20] In some SBS patients, parenteral fluids without macronutrients may be needed if stool output consistently exceeds fluid intake. During hot summer months, patients receiving PN overnight may occasionally require additional intravenous fluids during the day to prevent dehydration.

Best Practice Advice 5: Oral Rehydration Solution

Because of regional gut differences in water and sodium handling, those patients with SBS without a colon may be "net secretors," in that they lose more water and sodium from their stoma than they take in by mouth.[13] Such patients tend to have $<$100 cm of residual jejunum, and daily jejunostomy output can be $>$4 L. Fluids should be given to compensate for all losses and maintain a urine output of at least 1 L/d.

Because glucose in the gut lumen stimulates sodium absorption across the small intestine, which is followed by anions and water,[21] the sodium and glucose content of oral fluids are important considerations as inappropriate fluids will exacerbate fluid losses in SBS. The oral intake of low sodium, hypotonic (eg, water, tea, coffee, alcohol) and hypertonic (eg, fruit juices and sodas) solutions should be limited to reduce output, particularly in patients with net secretion and a high-output jejunostomy. A major misconception on the part of patients is that they should drink large quantities of water; however, this generally leads to an increase in ostomy output and creates a vicious cycle further exacerbating fluid and electrolyte disturbances. Instead, use of a glucose–electrolyte oral rehydration solution (ORS) to enhance absorption and reduce secretion is preferred.[22] In contrast, most patients with SBS and a colon can usually maintain adequate hydration with hypotonic fluids. Commercially prepared ORS products are widely available, as are recipes for inexpensive, homemade ORS. Importantly, ORSs differ from commercial sports drinks, as the sodium content of ORSs is considerably higher and the sugar content lower.

Best Practice Advice 6: Pharmacologic Therapy

Antimotility and antisecretory agents are frequently necessary to control stool losses in the patient with SBS. Massive enterectomy is associated with gastric hypersecretion and hypergastrinemia, which may last 6–12 months postoperatively. The use of antisecretory medications, including proton pump inhibitors or histamine-2 receptor antagonists, is beneficial in reducing the volume of gastric secretions, the damaging effects of the acid on the upper gut mucosa and the function of pancreatic exocrine enzymes. As gastric acid has a role in suppressing overgrowth of upper gut bacteria, it has been suggested that acid-suppressing agents should be used sparingly beyond 12 months, particularly when there is documented small intestinal bacterial overgrowth, unless there is clear evidence of a persistent beneficial effect on stool volume or dyspeptic symptoms. Some patients may benefit from treatment with the somatostatin analog, octreotide, owing to its effects on reducing the production of a variety of gastrointestinal secretions and slowing jejunal transit,[23,24] although it has not been shown to improve absorption or reduce the need for PN and may in fact inhibit pancreatic enzyme secretion and worsen malabsorption. Octreotide use should generally be reserved for patients with large volume stool losses in whom fluid and electrolyte management is problematic (eg, high-output end-jejunostomy) and should be avoided during the period of intestinal adaptation.

Antidiarrheals work mainly to reduce intestinal motility but also cause a slight reduction in intestinal secretion. Commonly used agents include loperamide, diphenoxylate with atropine, codeine, and tincture of opium. Loperamide should be preferred to opiate drugs because it is not addictive or sedative. Use of antidiarrheals should be guided by objective measurements of effect on stool output. Loperamide and codeine may have a synergistic effect when used together.[25] Because loperamide enters the enterohepatic circulation, which is disrupted in patients with SBS without an ileum, high doses are frequently needed (ie, up to 16 tablets [32 mg]/d). In the setting of SBS, these agents seem to be most effective when administered about 30 minutes

before meals and at bedtime. Clonidine, which can be administered transdermally, has also shown modest benefit in treating high-output stool losses, presumably via its effects on intestinal motility and secretion.[26,27] While antimotility agents may be effective in reducing intestinal transit, in cases in which bowel dilatation has occurred, antimotility agents might worsen diarrhea by encouraging bacterial overgrowth.

In an attempt to improve the depleted bile salt pool resulting from the loss of >100 cm ileum without aggravating stool losses, ox bile supplements have been given and found to improve fat absorption[28]; however, their availability is limited. Given the already diminished bile acid pool in SBS, the use of bile acid sequestrants may worsen steatorrhea, and fat-soluble vitamin losses in SBS and should generally be avoided. Although pancreatic function is reduced in patients receiving only PN and no oral intake and, potentially, during the early hypersecretory period if no antisecretory medications are being administered, at present, we are unaware of any published reports supporting the usefulness of pancreatic enzyme supplementation in SBS.

Best Practice Advice 7: Drug Dosing

Medications in solid dosage forms, such as a tablet, need to undergo disintegration and dissolution, processes occurring in the stomach, duodenum, and proximal jejunum, before being absorbed. As most oral medications are absorbed within the proximal jejunum, they can be used in patients with SBS; however, sustained- and delayed-release medications should be avoided.[29] When applicable, alternative drug delivery methods (eg, liquids, topical) should be considered in SBS, as should the monitoring of medication levels in the blood. The solution in response to a lack of clinical response of a drug will vary and may include escalating the dose, changing to a different dosing schedule or frequency, or changing to a different drug formulation (eg, crushed tablet, capsule, liquid) or route of administration (eg, intravenous, subcutaneous, transdermal).

Best Practice Advice 8: Surgery

The principle of primum non nocere applies even more to patients with SBS, who can ill-afford any inadvertent loss of further bowel length from ill-considered surgery. Surgical intervention in patients with SBS may be of value in 3 different contexts: (1) to recruit unused distal bowel, (2) to augment the function of residual bowel through specific lengthening and tapering operations, or (3) to slow intestinal transit.

An extended and often overlooked role for surgeons involved in managing SBS consists of dealing with complex intra-abdominal problems, whether pre-existent or imminent (eg, massive intra-abdominal desmoid tumors, mesenteric ischemia, complex enterocutaneous fistulae). Such patients are generally better served by a multidisciplinary intestinal rehabilitation team that includes dedicated surgical expertise.[30] It should also be emphasized that the surgical care of patients with SBS often commences at or even before the first operation—by taking all possible surgical measures to prevent the need for massive bowel resection and resulting SBS. Such an approach includes adopting a conservative philosophy in cases of doubtful bowel ischemia, the need for planned second-look operations and avoiding the risk of abdominal compartment syndrome and secondary bowel ischemia through hasty or ill-advised abdominal closure.

In patients with SBS, restoration of intestinal continuity and recruitment of any available distal bowel should be accomplished as soon as safely possible to improve bowel function and reduce the risk of or decrease PN dependency. This philosophic approach is in line with the anatomic guidelines suggested previously, with a shift from group 1 anatomy (jejunostomy or ileostomy) to group 2 or 3 (colon-in-continuity), conferring improved prognosis (Figure 1).[30]

Patients with SBS often have dilated segments of bowel, which occur at sites of suboptimal anastomoses or in watershed areas of blood supply. While bowel dilatation in this scenario can be viewed teleologically as compensatory, providing greater surface area to compensate for loss of length, these dilated segments are also often areas of stasis and set the stage for small intestinal bacterial overgrowth with clinical consequences including worsening of malabsorption and diarrhea. Where bowel length is clearly not a concern (ie, patient unequivocally does not have SBS), a simple tapering enteroplasty to remove a strip of bowel along the antimesenteric border to restore normal caliber is a simple expedient for the problem of segmental bowel dilatation. When dilatation occurs in a patient with SBS receiving PN, the focus is on trying to achieve intestinal tapering without loss of surface area. The choice of operation is mainly between the longitudinal intestinal lengthening and tapering (LILT) operation described by Bianchi[31] and the serial transverse enteroplasty (STEP) operation described by Kim et al (Figures 2 and 3).[32,33] There do not appear to be critical differences in outcomes between the 2 main lengthening operations, and the ultimate choice of operation may come down to surgeon preference. Overall, 50%–60% of patients who undergo such autologous gastrointestinal reconstructive surgery may be able to eventually wean off PN.[34,35] It is important to emphasize that there is no single operation that offers a cure for SBS, but such operations should be employed in the context of a comprehensive multidisciplinary approach to care.

Best Practice Advice 9: Glucagon-like Peptide-2

There is great interest in the use of growth factors in patients with SBS who have been unable to achieve

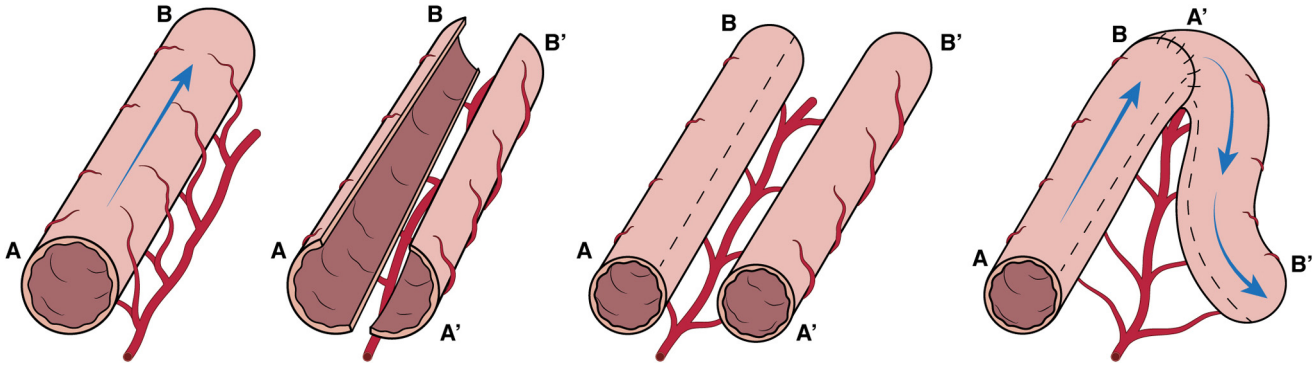

Figure 2. Principle of the Bianchi longitudinal intestinal lengthening and tapering operation: a dilated loop of bowel (AB) is split longitudinally along the antimesenteric border of the bowel, each with essentially half the original blood supply. The 2 hemiloops (AB and A'B') are anastomosed end-end in isoperistaltic fashion, to create a loop of bowel with twice the original length and half the diameter.

enteral independence during the adaptive period despite optimization of diet and medical management. The use of recombinant human growth hormone (Zorbtive; Serono Pharmaceuticals, Rockland, MA) has largely been discontinued due to unacceptable side effects and questionable long-term efficacy.[36] Glucagon-like peptide-2 (GLP-2), secreted in response to postprandial stimulation from the L cells in the distal ileum and right colon, has intestinotrophic effects that aid absorption.[37–39] The very short half-life of native GLP-2 has been extended to allow daily subcutaneous injection in the recombinant molecule, teduglutide (Gattex; Takeda Pharmaceuticals, Tokyo, Japan). Teduglutide can improve intestinal absorptive function and allow PN weaning in patients with SBS-IF, even allowing some patients to achieve enteral autonomy.[40,41] Because teduglutide is a growth factor and has the ability to enhance the growth of colonic and other gastrointestinal polyps as well as accelerate cancer growth, it is contraindicated in patients with active gastrointestinal malignancies, and patients should be screened by colonoscopy before initiating treatment and periodically while on this therapy.[42] As the GLP-2 receptors are primarily expressed in the gastrointestinal tract and brain, the package insert states that the decision to continue teduglutide in patients with nongastrointestinal malignancy should be made based on benefit-risk considerations. We note, however, that the confirmatory Phase 3 STEPS trial (study of teduglutide effectiveness in parenteral nutrition-dependent short bowel syndrome subjects) report to the Food and Drug Administration included the occurrence of 1 liver cancer and 2 lung cancers among trial subjects treated with teduglutide. It is our opinion, therefore, that teduglutide should not be used in patients with active or recent (arbitrarily, 5 years) malignancy, irrespective of location of malignancy. The significant side effects of teduglutide and the cost mandate that teduglutide is employed only after optimizing diet and the more conventional SBS treatments described previously in carefully selected patients with SBS-IF.[43] It is, therefore, important to view and use teduglutide not simply as one more antidiarrheal, even if that is a desired consequence of its pharmacodynamic effects.

Patients with SBS-IF should be carefully informed of the potential benefits and risks associated with growth factor treatments; information should include the probability of weaning from PN, the probability of quality-of-life improvement, the expected duration of treatment, the expected course after cessation of the treatment, the potential adverse effects and risks of the treatment, any

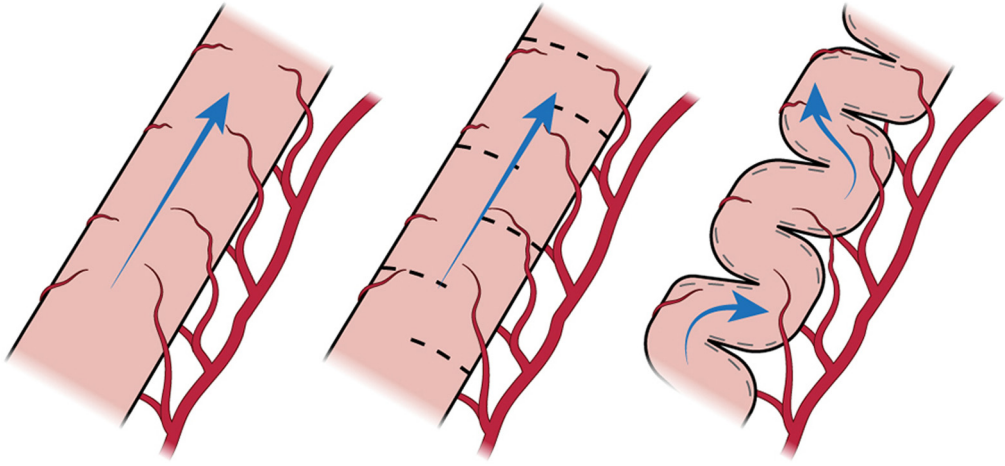

Figure 3. Principle of the serial transverse enteroplasty operation: a dilated loop of bowel is transected partially in a crisscross fashion (usually with a mechanical stapling device), perpendicular to its longitudinal axis, to create a luminal channel of appropriate caliber, without interfering with the blood supply.

cost considerations, and the need to undergo careful and regular monitoring. Importantly, intestinal peptide hormones should only be prescribed by those experienced in the diagnosis and management of patients with SBS and who have the ability and the facilities to objectively evaluate and balance the benefits with the detriments.

Best Practice Advice 10: Prevention of Complications

A variety of complications can affect the patient with SBS. A knowledge of these complications is critical for those caring for these patients to be able to not only identify and treat them when they occur but also to prevent their occurrence whenever possible. These complications may result from the underlying disease, the altered bowel anatomy and physiology, or its treatment, including the need for PN and its associated central venous catheter (Table 2). In some instances, the roles of the altered bowel and PN cannot be clearly separated. It is beyond the scope of this Clinical Practice Update to present each complication but general management approaches and relevant references are noted in Table 2.

Best Practice Advice 11: Referral for Intestinal Transplantation

The Centers for Medicare and Medicaid Services recommends intestinal transplantation (ITX) consideration for patients with IF (ie, refractory PN dependency) and onset of PN failure.[44] PN failure refers to the onset of complications associated with PN, especially the occurrence of progressive intestinal failure–associated liver disease or catheter-related complications such as recurrent catheter-related sepsis or loss of vascular access due

Table 2. Selected Short Bowel Syndrome–Associated Complications

Complication	Management Considerations	References
Central venous catheter Infection Occlusion Thrombosis Breakage	• Aseptic technique during placement and dressing changes • Appropriate catheter and insertion site • Proper catheter care and monitoring for infection • Remove catheter when no longer needed	57-59
Hepatobiliary Steatosis Cholestasis Cirrhosis Cholelithiasis	• Avoid excesses and deficiencies in PN formula • Limit intravenous lipid dose to <1 g/kg/d • Reduce/eliminate soybean-based intravenous lipid emulsion • Use non–soybean-based intravenous lipid emulsion • Cycle PN • Increase oral/enteral intake • Identify/treat sepsis or small intestinal bacterial overgrowth • Prophylactic cholecystectomy when abdominal surgery is being undertaken for other reasons	57,60
Metabolic bone disease Osteoporosis Osteopenia Osteomalacia	• Periodic assessment of bone mineral density • Monitor calcium, magnesium, and vitamin D status and supplement as needed • Correct metabolic acidosis when present • Specific osteoporosis treatments	57,61-63
Kidney injury Nephrolithiasis Oxalosis Acute kidney disease Chronic kidney disease	• Monitor urine output periodically and maintain adequate urine output with increased fluid intake • Low-fat, low-oxalate diet • Potassium citrate supplementation • Calcium carbonate supplementation	57,62
Chronic diarrhea	• Diet/oral fluid modifications • Oral rehydration solution • Antidiarrheal agents • Antisecretory agents (PPI, H2RA) • Somatostatin analogs • Intestinotrophic factor	57,61
Protein energy malnutrition, dehydration, and electrolyte/micronutrient deficiencies	• Monitor vitamin, mineral and electrolyte levels and supplement as needed • Optimize oral diet and fluid intake • Parenteral support (hydration, nutrition) • Intestinotrophic factor	57,61,64

H2RA, histamine-2 receptor antagonist; PN, parenteral nutrition; PPI, proton pump inhibitor.

to thromboses of the central veins. At present, nearly 50% of patients being considered for ITX are also requiring simultaneous liver replacement, indicating late referral for ITX.[44] Because the risk of mortality on the waiting list is much higher for patients requiring simultaneous liver transplantation, it is important that patients with SBS-IF experiencing PN complications are referred early for ITX consideration.[45]

Patients with SBS-IF with high morbidity or low acceptance of PN should also be considered for early listing for intestinal transplantation on a case-by-case basis. Patients with large abdominal desmoid tumors may also be served well by tumor resection with enterectomy if necessary, followed by ITX.[46,47] Such patients and those with severe dysmotility syndromes who have no prospect of PN weaning should be considered early for ITX, even if they do not meet the strict historic criteria of PN failure. In such carefully selected cases, viewing early ITX as standard of care with PN as the safety net in cases of a failed ITX is a paradigm shift in thinking that is gaining some momentum as ITX outcomes improve.[48]

Short- and medium-term outcomes for ITX are steadily improving, with 5-year survival now routinely exceeding 65% at experienced centers.[49] While short- and even medium-term outcomes of ITX have improved steadily over the last decade, the challenge of graft loss in close to half the patients who experience the most severe acute cellular rejection, the high rates of opportunistic and other infections, and the risk of posttransplant lymphoproliferative disease and long-term graft attrition related to poorly understood chronic allograft enteropathy remain impediments to better long term outcomes and, in turn, wider adoption of pre-emptive ITX, before the onset of PN complications.[50]

Best Practice Advice 12: Education and Support for Patients and Caregivers

Patients with SBS and their caregivers should be educated on key issues relevant to SBS. Although long-term PN may result in a restriction of activities and negatively impact daily life, it is also lifesaving and with time and experience, PN patients can modify their lifestyles to minimize the impact of this therapy.[51-53] One example of such a modification is to cycle the PN over 10–14 hours overnight to allow the patient freedom from the infusion pump during the day. Programmable infusion pumps are available that can be carried in a backpack or tote for the individual who needs to infuse PN during the day. Patient support groups such as the Oley Foundation (oley.org) are important sources of information on practical topics (eg, body image and travel), education, and support and may reduce the risk of complications and enhance survival and the quality of life of the patient on either EN or PN support.[54,55]

Because of the relative rarity of SBS, even nonspecialist clinicians care for patients with SBS, far removed from and without a dedicated multidisciplinary team, and may benefit from educational support in the management of these complex patients. One such attempt to provide educational support in the form of a virtual multidisciplinary team of experts is the Learn Intestinal Failure Tele-ECHO (Expanding Community Healthcare Outcomes) (LIFT-ECHO) project.[56] The LIFT-ECHO project leverages the highly successful ECHO project launched by Dr Sanjeev Arora, a hepatologist in New Mexico who first showed the success of such an educational platform in improving outcomes in patients with hepatitis C. The LIFT-ECHO project has evolved into an online educational community providing case-based learning in SBS, IF, and PN, directed at nonspecialist clinicians, with all didactic lectures being archived on the project website (liftecho.org/web).

Conclusion

The care of patients with SBS requires a comprehensive approach and attention to detail. A multidisciplinary approach consisting of dietitians, nurses, surgeons, gastroenterologists or internists, and social workers experienced in the care of patients with IF is essential for the successful management of these patients. Specific dietary intervention combined with medical management and, occasionally, surgical strategies offer the potential of PN reduction, enteral independence, and overall improved quality of life and clinical outcome (Supplementary Figure 1).

Supplementary Material

Note: To access the supplementary material accompanying this article, visit the online version of *Clinical Gastroenterology and Hepatology* at www.cghjournal.org, and at https://doi.org/10.1016/j.cgh.2022.05.032.

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Clinical Practice Update
This expert review was commissioned and approved by the AGA Institute Clinical Practice Updates Committee (CPUC) and the AGA Governing Board to provide timely guidance on a topic of high clinical importance to the AGA membership, and underwent internal peer review by the CPUC and external peer review through standard procedures of Clinical Gastroenterology and Hepatology.

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Kishore Iyer, MBBS: Conception, writing, critical editing/finalizing manuscript
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Conflicts of Interest
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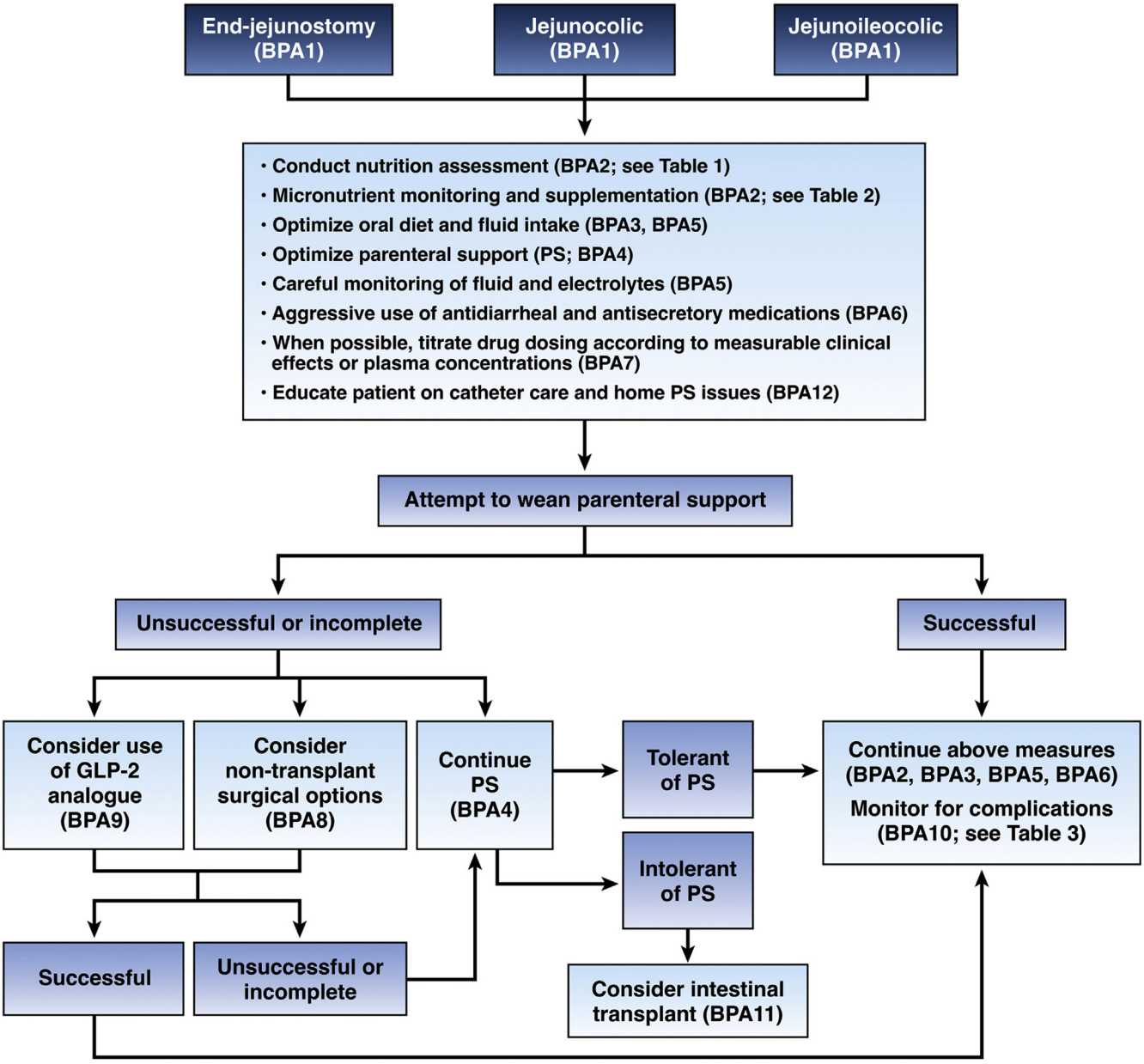

Supplementary Figure 1. Algorithm for the management of the patient with SBS requiring parenteral support (PS). BPA, best practice advice.

Supplementary Table 1. Initial Nutrition Assessment in the Patient With Short Bowel Syndrome

Weight change history
Medication usage including supplements
Presence of gastrointestinal and other symptoms that may affect oral intake or fluid loss
Food diary to determine usual oral diet and daily energy intake
Potential symptoms of micronutrient deficiencies
Pertinent past medical and psychiatric comorbidities
Pertinent surgical history including the presence of bowel complications such as anastomotic strictures, chronic obstruction, enterocutaneous fistulae, and peritoneal drains
Prior/current enteral and/or central venous access device, formula used, route and method of administration, and complications
Physical assessment for signs of dehydration, malnutrition, and micronutrient deficiency
Laboratory studies including complete blood count, chemistry panel, and micronutrient levels
Bone mineral density