



Complete Summary

GUIDELINE TITLE

American Gastroenterological Association medical position statement: short bowel syndrome and intestinal transplantation.

BIBLIOGRAPHIC SOURCE(S)

American Gastroenterological Association medical position statement: short bowel syndrome and intestinal transplantation. *Gastroenterology* 2003 Apr;124(4):1105-10. [1 reference] [PubMed](#)

GUIDELINE STATUS

This is the current release of the guideline.

According to the guideline developer, the Clinical Practice Committee meets three times a year to review all American Gastroenterological Association Institute (AGAI) guidelines. This review includes new literature searches of electronic databases followed by expert committee review of new evidence that has emerged since the original publication date.

** REGULATORY ALERT **

FDA WARNING/REGULATORY ALERT

Note from the National Guideline Clearinghouse: This guideline references a drug(s) for which important revised regulatory and/or warning information has been released.

- [June 30, 2008, CellCept \(mycophenolate mofetil\) and Myfortic \(mycophenolate acid\)](#): Novartis and Roche have agreed to include additional labeling revisions to the WARNINGS and ADVERSE REACTIONS sections of the Myfortic and CellCept prescribing information, based on post-marketing data regarding cases of Progressive Multifocal Leukoencephalopathy (PML) in patients treated with these drugs.
- [October 29, 2007, CellCept \(mycophenolate mofetil\)](#): Roche has agreed to include additional labeling revisions to the BOXED WARNING, WARNINGS/Pregnancy and Pregnancy Exposure Prevention, PRECAUTIONS/Information for Patients, and ADVERSE REACTIONS/Postmarketing Experience sections.
- [August 16, 2007, Coumadin \(Warfarin\)](#): Updates to the labeling for Coumadin to include pharmacogenomics information to explain that people's genetic makeup may influence how they respond to the drug.
- [October 6, 2006, Coumadin \(warfarin sodium\)](#): Revisions to the labeling for Coumadin to include a new patient Medication Guide as well as a

reorganization and highlighting of the current safety information to better inform providers and patients.

COMPLETE SUMMARY CONTENT

** REGULATORY ALERT **

SCOPE

METHODOLOGY - including Rating Scheme and Cost Analysis

RECOMMENDATIONS

EVIDENCE SUPPORTING THE RECOMMENDATIONS

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

IMPLEMENTATION OF THE GUIDELINE

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT

CATEGORIES

IDENTIFYING INFORMATION AND AVAILABILITY

DISCLAIMER

SCOPE

DISEASE/CONDITION(S)

- Short bowel syndrome (SBS)
- Complications of total parenteral nutrition (TPN)
- Posttransplant complications of intestinal transplantation

GUIDELINE CATEGORY

Evaluation
Management

CLINICAL SPECIALTY

Gastroenterology
Internal Medicine
Nutrition
Surgery

INTENDED USERS

Advanced Practice Nurses
Nurses
Physicians
Psychologists/Non-physician Behavioral Health Clinicians
Social Workers

GUIDELINE OBJECTIVE(S)

To present recommendations on management of short bowel syndrome (SBS) and intestinal transplantation

TARGET POPULATION

Patients with short bowel syndrome (SBS) including patients undergoing intestinal transplant

INTERVENTIONS AND PRACTICES CONSIDERED

Medical Therapy

1. Total parenteral nutrition (TPN) including dextrose, lipid emulsion, and free amino acid infusions
2. Glucose-polymer-based oral rehydration solutions (ORS)
3. Electrolyte replacement/monitoring
4. Blood glucose and triglycerides monitoring
5. Fluid management
6. H₂ blockers/proton pump inhibitors first 6 months
7. Diarrhea control with (a) anti-motility agents, such as loperamide hydrochloride and diphenoxylate; (b) codeine sulfate, tincture of opium; (c) octreotide

Note: Bile acid supplements are not recommended because they may worsen diarrhea, and cholestyramine is not recommended in patients with >100 cm of ileal resection, because it may worsen steatorrhea

8. Oral calcium supplements
9. Vitamin and other mineral supplements
10. High complex carbohydrate diet
11. Cycle TPN for home
12. Teaching appropriate catheter care
13. TPN weaning
14. Partial TPN support with weaning
15. Low oxalate diet
16. Medium-chain triglycerides (MCT) as part of diet
17. Diagnosis and treatment of TPN complications, including catheter-related complications

Surgical Therapy

1. Nontransplant surgery such as bowel lengthening surgery
2. Intestinal/liver transplantation in patients who have developed complications attributable to intestinal failure and/or long-term TPN therapy
 - *Before transplantation:* complete cardiopulmonary evaluation; determination of associated liver pathology existence including assessment of hepatic aminotransferases, total bilirubin, albumin, international normalized ratio, and liver biopsy; measurement of portal venous pressure; patient education and counseling
 - *After transplantation:* immunosuppressive agents such as tacrolimus, prednisone, mycophenolate mofetil, azathioprine, cyclosporine, Rapamycin, cyclophosphamide, prostaglandin E1, OKT3, antithymocyte globulin (ATG), and daclizumab; gastrointestinal contrast study; initiation of enteric or tube feeding; management of rejection--methylprednisolone, OKT3, thymoglobulin, broad spectrum antibiotics; immunosuppression reassessment; sepsis management;

cytomegalovirus (CMV) prophylaxis and management; prophylaxis of posttransplant lymphoproliferative disorder

MAJOR OUTCOMES CONSIDERED

- Incidence and prevalence of short bowel syndrome (SBS)
- The remaining intestinal adaptation following resection
- Quality of life and survival of short bowel patients
- Complications of long-term total parenteral nutrition (TPN)
- Patient and graft survival after transplantation
- Quality of life in home TPN patients and transplant recipients

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Hand-searches of Published Literature (Secondary Sources)
Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Data and reports were obtained from extensive PubMed and Medline searches using several key words, including short bowel syndrome (SBS), various conditions predisposing to short bowel syndrome, parenteral nutrition, enteral nutrition, relevant specific nutritional deficiencies, intestinal surgery, and intestinal transplantation. In addition, surgical and gastroenterological texts, published national and international scientific meeting abstracts, and the extensive manuscript/abstract files of the authors were reviewed. Expert opinion was sought for the few areas in which no suitable published reports existed (e.g., total parenteral nutrition [TPN] cycling and preparation of the patient for home TPN). Human data and reports were reviewed exclusively.

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Not stated

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Not applicable

METHODS USED TO ANALYZE THE EVIDENCE

Systematic Review with Evidence Tables

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not stated

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Not stated

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Not applicable

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

Internal Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

The American Gastroenterological Association (AGA) Clinical Practice Committee approved the guideline document on August 5, 2002. The AGA Governing Board approved it on November 1, 2002.

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Definition

Short bowel syndrome (SBS) occurs when, after surgery or congenitally, a patient is left with <200 cm of functional small intestine. Absorption is related to the amount of residual intestine; patients at greatest nutritional risk generally have a duodenostomy or a jejunioileal anastomosis with <35 cm of residual small intestine, jejunocolic or ileocolic anastomosis with <60 cm of residual small intestine, or an end jejunostomy with <115 cm of residual small intestine. Patients can be grouped into 2 distinct subgroups: those with colon in continuity and those without colon in continuity. In patients with SBS, the colon becomes an important digestive organ.

Medical Therapy

The most important aspects of medical management of the patient with SBS are provision of adequate macro- and micronutrients and fluid to prevent energy malnutrition, specific nutrient deficiencies and dehydration, and correction and prevention of acid-base disturbances.

Glucose-polymer-based oral rehydration solutions (ORS) with 90-120 mEq/L sodium (Na) should be instituted to decrease dehydration and total parenteral

nutrition (TPN) fluid requirements in patients with residual jejunum ending in a jejunostomy. Several commercial ORS are available, or solutions can be formulated by dissolving sodium chloride (NaCl) (2.5 g), potassium chloride (KCl) (1.5 g), sodium bicarbonate (Na₂CO₂) (2.5 g), and glucose (table sugar, 20 g) in 1 L water. Patients should avoid consumption of plain water and should be encouraged to drink ORS whenever they are thirsty. For patients with residual colon in continuity, ORS may still be of value--provided sufficient Na is present in the diet; the amount of Na in the ORS may not be as critical. For patients with no remaining jejunum, who have residual ileum, the presence of glucose in the ORS is not critical because ileal water absorption is not affected by the presence of glucose.

Magnesium (Mg) deficiency may occur despite a normal serum concentration. It is prudent to measure 24-hour urine Mg. However, Mg replacement is problematic and often requires intravenous infusion. Oral calcium (Ca) supplementation is recommended routinely (800-1200 mg per day). Iron is absorbed in the duodenum and, in the absence of hemorrhage, is not routinely required as a supplement. Phosphorus deficiency is rare; supplementation is rarely required.

Resection of the ileocecal valve may allow colonic bacteria to populate the small intestine, resulting in bacterial overgrowth. This may negatively impact on digestion and nutrient assimilation, because bacteria compete for nutrients with the enterocytes. Diagnosis of bacterial overgrowth may be more difficult using breath tests because of rapid intestinal transit in SBS. Endoscopically obtained small bowel aspirate for culture may be required. Treatment can be undertaken with oral metronidazole, tetracycline, or other antibiotics.

High-dose H₂ antagonists and proton pump inhibitors reduce gastric fluid secretion, and fluid losses during the first 6 months post-enterectomy. Fluid losses usually require long-term control with anti-motility agents, such as loperamide hydrochloride or diphenoxylate (4-16 mg per day). If these are ineffective, especially in patients without colon in continuity or in patients with minimal residual jejunum or duodenum, use of codeine sulfate (15-60 mg two to three times a day) or tincture of opium may be necessary. Rarely, octreotide (100 micrograms subcutaneously [SQ], three times a day, 30 minutes before meals) is required. It should be used only if fluid intravenous requirements are >3 L daily because post-resection intestinal adaptation may be impaired and the risk for cholelithiasis increased. There is insufficient evidence to recommend the use of bile acid supplements to decrease steatorrhea; and they may worsen diarrhea. Cholestyramine is not useful in patients with >100 cm of ileal resection, and it may actually worsen steatorrhea because of the binding of bile salts.

Dietary Management

Typically, patients who have undergone massive enterectomy require TPN, once hemodynamic stability has been achieved, for the first 7-10 days after surgery. Nutritional therapy should be introduced gradually, converting to standard enteral formula as tolerated. The goal is to provide patients with approximately 25-30 kcal/kg per day and 1.0-1.5 g/kg per day of protein. Standard enteral formula is recommended. Nitrogen is the macronutrient least affected by diminished intestinal absorptive surface. Therefore, the utility of peptide-based diets in such patients is generally without merit. Oral intake should be encouraged. There is no

value in separating liquids from solids in the diet or with high fat-low carbohydrate or low fat-high carbohydrate diets except for patients with colon in continuity, in whom soluble fiber intake should be encouraged. Soluble fiber is fermented to short-chain fatty acids by colonic bacteria and serve as an additional energy source. Small amounts of medium-chain triglycerides are absorbed by the colon and may be included in the diet as an additional energy source. Lactose-containing foods should not be restricted in the absence of confirmed lactase deficiency or significant proximal jejunal resection. Dietary oxalate should be restricted in the diet in patients with residual colon in continuity. Oral Ca supplements also may be of value for the prevention of Ca-oxalate nephrolithiasis. Randomized, controlled trials have not shown glutamine and/or growth hormone to improve intestinal absorption. Micronutrients, including water-soluble (B1, B2, B3, B6, B12, biotin, folate, C) and fat-soluble (A, D, E, K) vitamins, and trace elements (zinc [Zn], selenium [Se]) often require supplementation (See Table 1 in the original guideline document for information on vitamin and mineral supplements). Water-soluble vitamin deficiency is rare.

Parenteral Nutrition

Most patients will require TPN, at least initially. For the normally nourished patient, TPN should be supplied at 25-30 kcal/kg per day based on ideal body weight for adults, with greater levels of support for infants and children depending on age. Dextrose is a monohydrate, providing 3.4 kcal/mL. The maximum dextrose infusion rate should be 5-7 mg/kg/min. Blood glucose should be monitored at least daily, optimally four times per day (qid), and should be <180-200 mg/dL; 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 U/g dextrose; subsequent adjustments should be made 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 fluid management issues; 20% lipid emulsion is more calorically dense than dextrose. Generally, the percentage of lipid calories should be increased and the percentage of dextrose calories should be decreased if the amount of supplemental insulin required exceeds 0.2 U/g dextrose, although the serum triglyceride concentration should be kept <700-800 mg/dL, and optimally, <400 mg/dL. Protein is supplied in the form of free amino acids and should be supplied at 1.0-1.5 g/kg per day, based on ideal body weight for adults, with greater levels of support for infants and children depending on age.

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; maximal adaptation may take as long as 1-2 years. For patients who will require TPN at home, the infusion should be compressed to overnight. Typically, this would be during a 10-hour period with an additional 30-60-minute taper period; some patients with fluid management issues will be unable to tolerate this infusion rate. Cycling to overnight infusion should be a gradual process. Once goal infusion volume has been determined (e.g., 1.5, 2.0, 2.5, or 3.0 L for adults; with less volume for infants and children), the total volume should be infused over gradually decreasing time periods (e.g., compress by increments of 2-4 hours). TPN should be infused ideally via a single lumen catheter with its tip positioned in either the superior vena cava (SVC) or inferior vena cava (IVC) to decrease the

risk of infection and thrombosis. Tunneled 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.

The patient's home environment should be evaluated. A room, preferably the bedroom--definitely not a "dirty" room, such as the kitchen or bathroom--should be identified for TPN to be set up prior to use. The patient should be instructed to purchase a small refrigerator to be used solely for TPN storage. A local support group under the umbrella of the Oley Foundation (1-800-776-OLEY) should be contacted. Transition from hospital to home may be smoother if the patient has another patient contact who previously has undergone the same process. The patient should undergo some education about TPN prior to hospital discharge, including the indications for TPN, basic instruction on getting their solutions ready for use (they will need to add their vitamins, insulin, and H₂ blockers if prescribed, and flush catheter), catheter care, dressing changes, and information on their intravenous pump. It is often useful for the patient to meet their home care nurse (who will continue the education process at home until the patient or caregiver is self-sufficient) prior to discharge. The treating physician should have some familiarity with appropriate catheter care and the identification of complications associated with long-term TPN, including catheter-related infections, occlusions, and metabolic complications.

Patients should not be discharged home until their fluid and electrolyte requirements have stabilized. Once home, office visits and laboratory monitoring should initially be more frequent, although the stable patient who has minimal difficulty generally can visit the office and have routine laboratory testing done as infrequently as 3 times a year.

Patients in whom TPN is being weaned, and who acquire <75% of the nutritional needs parenterally, should have vitamin (usually fat-soluble vitamins) and trace metal (zinc [Zn], copper [Cu], selenium [Se]) analyses performed 2-3 times yearly, and whenever possible deficiencies are clinically recognized. Vitamin K is not a constituent of all parenteral multivitamin solutions, although vitamin K is present in the intravenous lipid emulsion. Therefore, the prothrombin time should be regularly monitored, especially in those patients who lack residual colon. During clinic visits, the catheter exit site or skin overlying an implanted infusion port should be examined for warmth, erythema, and tenderness, and the catheter dressing should be examined for purulent exudate, which may signal infection. A properly maintained catheter may remain in place for many years.

Medication Absorption

Oral medication absorption is often impaired and larger doses, intravenous, or sublingual delivery may be required; significant interpatient variability may be observed.

Role of Surgery

Nontransplant Surgery

Restoration of intestinal continuity, such as re-anastomosis of small intestine with colon, should be performed whenever possible, because it can be performed with relatively low morbidity and mortality (often with discontinuation of TPN). Other forms of bowel lengthening surgery have significant associated morbidity and mortality, and therefore should be considered only in select patients.

Intestinal Transplantation

Indications for Transplantation

Thus far, intestinal transplants have been performed only in patients who have developed life-threatening complications attributable to their intestinal failure and/or long-term TPN therapy. Medicare has approved payment for intestinal transplants in patients who fail TPN therapy for one of the following reasons:

1. Impending or overt liver failure (increased serum bilirubin and/or liver enzyme levels, splenomegaly, thrombocytopenia, gastroesophageal varices, coagulopathy, stomal bleeding, hepatic fibrosis, or cirrhosis)
2. Thrombosis of major central venous channels (2 thromboses in subclavian, jugular, or femoral veins). Evidence supporting this indication is weak.
3. Frequent central line-related sepsis (2 episodes of systemic sepsis secondary to line infection per year, 1 episode of line-related fungemia, septic shock, or acute respiratory distress syndrome). Evidence supporting this indication is weak.
4. Frequent severe dehydration

Until better data become available, these parameters are likely to be widely recognized as the indications for intestinal transplantation.

Complications of Long-term TPN That Could Lead to the Need for Intestinal Transplantation

For patients who develop TPN-associated liver disease, investigational studies using metronidazole, oral lecithin, ursodeoxycholic acid, or intravenous choline to treat or prevent the development of TPN-associated liver disease should be considered. Otherwise, no specific therapy is available. Care should be taken to avoid dextrose overfeeding fatty acid deficiency from insufficient intravenous lipid emulsion (minimum of 2%-4% or 4%-8% of nonprotein calories as linoleic acid or lipid emulsion, respectively) and to limit intravenous lipid intake to <2.5 g/kg per day, possibly even to <1 g/kg per day. There is no role for carnitine supplementation.

In patients with end-stage liver disease (ESLD) related to SBS, combined intestine-liver transplant may be the only option; isolated liver transplantation is not recommended. However, carefully selected ESLD patients with significant residual intestine, who are very likely to be weaned from TPN soon after transplant, can achieve successful isolated liver transplantation. Isolated intestine transplantation is not recommended in the setting of ESLD, although outcome data stratifying intestine-only transplant recipients based on their pretransplant liver abnormalities do not exist. It is not yet clear when the TPN-associated hepatic pathological process progresses to the point of irreversibility. Patients with SBS who progress to ESLD are placed on the waiting list for a combined intestine-

liver transplant, but have an extremely high mortality rate exceeding all other solid organ transplant waiting lists, including isolated liver transplants.

With proper catheter care techniques, the rate of catheter-related infections and thrombosis can be minimized. Calcium phosphate compatibility should be monitored in the TPN solution to prevent non-thrombotic catheter occlusion. Prior catheter thrombosis is a risk factor for development of SVC/IVC syndrome in the future; therefore, warfarin anticoagulation should be undertaken in patients with prior catheter thrombosis in the absence of catheter malposition as the cause. Typically, TPN catheters are first placed in the SVC by accessing either the internal jugular, brachial, or subclavian veins. If these veins are no longer accessible, the catheters are usually placed in the IVC via the femoral or saphenous veins. True loss of catheter insertion sites is extremely rare; clinicians often prematurely determine that a patient has no suitable venous access. When all the usual central veins have been exhausted, alternatives include translumbar or transhepatic access to the IVC, and thoracotomy with direct placement of an intra-atrial catheter, among others.

Current Management of the Intestinal Transplant Patient

Standards of care for intestinal transplantation are still evolving and will continue to evolve until outcomes are comparable to those seen with other solid organ transplants. All patients should have a complete cardiopulmonary evaluation. If intestinal failure resulted from mesenteric thrombosis, the etiology should be sought. Metastatic malignancies and active or uncontrolled systemic infections, including human immunodeficiency virus, exclude transplantation.

It is essential to determine if associated liver pathology exists in patients being evaluated for potential intestinal transplantation; etiologies other than TPN or malabsorption should be considered. Hepatic aminotransferases, total bilirubin, albumin, international normalized ratio, and platelet count should be determined, and liver biopsy should be performed. Portal venous pressure should be measured to exclude portal hypertension, although normal results may be deceiving in patients who have had major intestinal resections, since most portal inflow will be missing. Patients and their families should meet with a social worker, psychiatrist, and a financial/insurance counselor who understand the complex medical, psychologic, and social issues involved with organ transplantation. Living donation should be considered to eliminate waiting time, optimize human lymphocyte antigen (HLA) matching, and simplify coordination of donor-recipient procedures.

After the transplant, intestinal recipients require life-long immunosuppression. While immunosuppressive regimens vary between individual centers, tacrolimus and prednisone are generally included. Other agents may include mycophenolate mofetil, azathioprine, cyclosporine, Rapamycin, cyclophosphamide, prostaglandin E1, OKT3, antithymocyte globulin (ATG), and daclizumab. In the early post-transplant period, blood tacrolimus concentrations should be 20-30 ng/mL. Simultaneous transplantation of donor bone marrow has been performed in an attempt to induce recipient hyporesponsiveness to donor tissues. This approach has not clearly altered outcomes in intestinal transplant patients, although long-term effects remain to be seen.

Following transplantation, multi-visceral recipients require monitoring in an intensive care unit, while isolated intestine recipients can usually go directly to a non-intensive care unit bed. Within the first week post transplant, if a gastrointestinal contrast study has excluded an anastomotic leak, enteric feeding should be initiated. If the patient is not ready or willing to eat, tube feedings should be started. Once initiated, enteral nutrition is gradually increased until nutritional goals are met, at which time TPN can be discontinued. Some patients may require only fluid supplements. Many infants with congenital SBS may require tube feeding because of reluctance to eat. Thought to be due to these infants never having learned to eat, this phenomenon requires long-term re-education and psychologic counseling.

Rejection is the most common cause of graft loss in intestinal transplant recipients. While clinical signs are frequently unreliable, fever and gastrointestinal symptoms (bloating, cramping, diarrhea, increased stomal output) are often present. There are no reliable biochemical markers of rejection. Small intestinal endoscopy with multiple biopsies (6 total biopsies) initially should be performed biweekly; because early rejection may not be endoscopically apparent, biopsy specimens should be obtained even from normal appearing mucosa. Increased intestinal permeability (diethylenetriaminepentaacetic acid nuclear medicine scan) also has a high correlation with acute rejection. Cytomegalovirus (CMV) enteritis can resemble rejection clinically and histologically; therefore, intestinal allograft biopsy specimens should be reviewed by a pathologist experienced with intestinal transplantation. When rejection is diagnosed, high-dose intravenous corticosteroids should be administered for 3 days (methylprednisolone 500 mg daily or equivalent). In severe or corticosteroid-resistant rejection, antibody therapy (OKT3, thymoglobulin) for this agent may be used for up to 14 days. Because of the theoretical potential for bacterial translocation from injured bowel, broad-spectrum antibiotics should be administered during rejection. After rejection is reversed, the adequacy of maintenance immunosuppression should be reassessed.

The high level of immunosuppression necessary to prevent intestinal rejection contributes to many post-transplant complications, including sepsis. Unless intestinal rejection underlies sepsis, immunosuppression should be reduced. In progressive or persistent sepsis, immunosuppression withdrawal and removal of the intestinal graft should be considered before sepsis is irreversible. CMV enteritis can also lead to graft loss; CMV-positive donors are avoided in CMV-negative recipients, and CMV prophylaxis with ganciclovir should be continued for a prolonged period after transplant. If CMV infection is suspected (fever, flu-like symptoms, diarrhea, headache, leucopenia), confirmation should be obtained by an antigenemia assay, and ganciclovir and/or CMV-specific hyperimmune globulin should be initiated. Post-transplant lymphoproliferative disorders are responsible for 14% of post-transplant deaths. Because the Epstein-Barr virus (EBV), the causative agent in post-transplant lymphoproliferative disorders, is ubiquitous in the adult population, use of EBV negative donors is impractical. However, ongoing surveillance of peripheral blood EBV load can provide guidance for preemptive therapy with acyclovir and/or hyperimmune globulin.

CLINICAL ALGORITHM(S)

The original guideline document contains the following clinical algorithms:

- Diagnosis and treatment of catheter occlusion
- Catheter-related infection algorithm for diagnosis and treatment
- Management of short bowel syndrome

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of supporting evidence is not specifically stated for each recommendation. The guideline is accompanied by a comprehensive literature review. The available data used for that review were based on retrospective analysis of case series (type II-3 or type III data) and were often few in number, because of the rareness of the covered diseases, where randomized controlled trials had been undertaken (type 1 and type IIb data), and the studies were described in detail.

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

The appropriate management of patients with short bowel syndrome

POTENTIAL HARMS

- Complications of total parenteral nutrition (TPN) such as catheter occlusion, catheter-related infection, liver complications, biliary complications, D-lactic acidosis, renal dysfunction, metabolic bone disease, memory deficits, and neurological problems
- Posttransplant complications such as acute rejection, chronic rejection, and posttransplant lymphoproliferative disease (PLPD)

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

IMPLEMENTATION TOOLS

Clinical Algorithm

For information about [availability](#), see the "Availability of Companion Documents" and "Patient Resources" fields below.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Getting Better
Living with Illness

IOM DOMAIN

Effectiveness
Patient-centeredness
Safety

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

American Gastroenterological Association medical position statement: short bowel syndrome and intestinal transplantation. Gastroenterology 2003 Apr;124(4):1105-10. [1 reference] [PubMed](#)

ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

2003 Apr

GUIDELINE DEVELOPER(S)

American Gastroenterological Association Institute - Medical Specialty Society

SOURCE(S) OF FUNDING

American Gastroenterological Association Institute

GUIDELINE COMMITTEE

American Gastroenterological Association Patient Care Committee

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

Not stated

FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

Not stated

GUIDELINE STATUS

This is the current release of the guideline.

According to the guideline developer, the Clinical Practice Committee meets three times a year to review all American Gastroenterological Association Institute (AGAI) guidelines. This review includes new literature searches of electronic databases followed by expert committee review of new evidence that has emerged since the original publication date.

GUIDELINE AVAILABILITY

Electronic copies: Available from the [American Gastroenterological Association Institute \(AGAI\) Gastroenterology journal Web site](#).

Print copies: Available from American Gastroenterological Association Institute, 4930 Del Ray Avenue, Bethesda, MD 20814.

AVAILABILITY OF COMPANION DOCUMENTS

The following is available:

- AGA technical review on short bowel syndrome and intestinal transplantation. Gastroenterology 2003 Apr;124(4):1111-34.

Electronic copies: Available from the [American Gastroenterological Association Institute \(AGAI\) Gastroenterology journal Web site](#).

Print copies: Available from American Gastroenterological Association Institute, 4930 Del Ray Avenue, Bethesda, MD 20814.

PATIENT RESOURCES

None available

NGC STATUS

This summary was completed by ECRI on August 20, 2003. This summary was updated by ECRI on March 15, 2005 following release of a public health advisory from the U.S. Food and Drug Administration regarding the use of Elidel. This summary was updated by ECRI on March 6, 2007 following the FDA advisory on Coumadin (warfarin sodium). This summary was updated by ECRI Institute on September 7, 2007 following the revised U.S. Food and Drug Administration (FDA) advisory on Coumadin (warfarin). This summary was updated by ECRI Institute on November 6, 2007, following the U.S. Food and Drug Administration advisory on CellCept (mycophenolate mofetil). This summary was updated by ECRI Institute on July 8, 2008, following the updated U.S. Food and Drug Administration (FDA) advisory on CellCept (mycophenolate mofetil) and Myfortic (mycophenolate acid).

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