4 Assessment and General Management of Intestinal Failure

13 Assessment of Intestinal Failure Patients

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Key points

- Short bowel syndrome is a complex condition, which requires a multidisciplinary approach.
- The intrinsic Characteristics of the remaining intestine after small bowel resection are crucial in determine the functional ability of the bowel.
- To determine the anatomy and the intestinal length of the remaining bowel, review surgical records and obtain radiological studies such as barium enema upper gastrointestinal and small bowel examination (UGI and SBS). These may be followed with esophagogastroduodenoscopy and colonoscopy.
- At the end of the initial assessment, the physician should understand the cause of the intestinal failure, the reasons for feeding intolerance, the likelihood of intestinal anatomy, the patient's nutritional status, and also recognize any acute or chronic complications.

Introduction

Intestinal failure (IF), results from surgical resection, congenital defect, or disease associated loss of absorption and is characterized by the inability to maintain protein-energy, fluid, electrolyte or micronutrient balances when on a normal diet [1]. The causes of IF can be separated by pathogenesis into three different groups: anatomic reduction of the gut length, short bowel syndrome (SBS), the most common cause; neuromuscular diseases involving the GI tract, and, congenital diseases of the intestinal epithelium (Table 13.1). Intestinal adaptation, the progressive recovery from IF that follows a loss of intestinal length, usually allows the restoration of adequate intestinal function within several weeks to months. Only in a small number of children is adaptation inadequate; these continue to be dependent on parenteral nutrition (PN).

The likelihood that a patient with SBS will reestablish enteral autonomy is influenced by the length, location, and function of the remaining bowel. Regardless of multiple attempts to describe this syndrome on the basis of intestinal length alone, it appears that no minimum length of small bowel can be used reproducibly to define the short bowel syndrome. The essential defect is the lack of an adequate mucosal surface to permit enteral nutrition autonomy [2,3]. The purpose of this article is to discuss strategies for the initial assessment of an intestinal failure patient and focuses on the SBS while endeavoring to define the optimal approach for each individual patient.

Assessment

SBS is a complex condition, which requires a multidisciplinary approach including a gastroenterologist, surgeon, nurse practitioner/coordinator, dietitian, as well as a psychological evaluation. To determine the most appropriate therapy, a comprehensive evaluation is necessary. An experienced physician taking care of patients with IF should supervise and guide the

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Table 13.1 Causes of IF.

1. Anatomic reduction of the gut length: SBS the most common cause.
- Normal GI anatomy: Necrotizing enterocolitis, gastroschisis (as a result of resection), multiple resections of Crohn's disease, volvulus, mesenteric arterial embolism, venous thrombosis, volvulus trauma, or tumor resections.
- Congenital anomalies: intestinal atresia, shortened small bowel at birth.

2. Neuromuscular diseases involving the GI tract: Total aganglionosis (long segment Hirschsprung's disease) or chronic intestinal pseudoobstruction.

3. Diseases of the intestinal epithelium: Microvillus atrophy, intestinal epithelial dysplasia, tufting enteropathy, autoimmune enteropathy, radiation enteritis.

management of these complex patients. Treatment options for SBS in children include long-term PN, intestinal rehabilitation, and/or intestinal transplantation. The components of an initial evaluation are identified in Table 13.2.

History and physical evaluation

Evaluating a neonate who has recently suffered from an anatomic reduction of the gut with subsequent SBS is not as difficult as evaluating a patient with a long history of multiple enterotomies and complications.

Table 13.2 Components of an Initial Evaluation.

1. History and physical evaluation
2. Determine the anatomy and bowel length
   - Upper GI and small bowel series.
   - Barium enema.
   - Endoscopic studies: to rule out intestinal stricture/obstruction, to obtain tissue for histology studies, and duodenal fluid for bacterial overgrowth.
3. Ultrasound of the abdomen with Doppler evaluation of hepatic vessels and upper extremities (to determined vascular access)
4. Liver biopsy, if liver disease is associated
5. Chest X-ray.
6. Bone age.
7. Nutritional assessment.
8. Psychological evaluation.
9. Laboratory exams: See table 3.

Important components of the initial history include:

1 A thorough review and summary of past medical record.

2 The cause of SBS, the anatomy and length of the intestine, including a detailed review of prior surgical procedures and any related complications.

3 The number of central lines and the reasons they were changed.

4 The causal microorganism of any prior central line infections (bacteria/fungus) and the clinical consequences associated with each infection (sepsis, thrombosis), including whether intensive care or mechanical ventilation were required.

5 Nutritional assessment.

6 A detailed vaccination status.

7 A complete physical exam (PE): emphasizing hydration and nutritional status (weight, height, basal metabolic index), type of central line, signs of vascular thrombosis, cardiovascular status, dermatologic examination including diaper rash and signs of chronic liver disease.

Bowel anatomy

The intrinsic characteristics of the remaining intestine after small bowel resection are crucial in determining the functional ability of the bowel. Most nutrient absorption occurs within the initial two-thirds of the small bowel. There are three major types of bowel anatomy in SBS: jejunocolic anastomosis, end-jejunostomy, and jejunoileal anastomosis. The third is the least common, but is associated with the best overall prognosis [4]. In general, a jejunal resection is better tolerated, as the ileum can replace the absorptive capacity of the jejunum through adaptation. However, the jejunum cannot adapt the ileal functions. Vitamin B12 and bile acids are exclusively absorbed in the distal ileum [2,4]. In SBS patients, the colon becomes an important digestive organ by absorbing sodium, water, some amino acids, and short chain fatty acids used as energy substrates [2].

The remaining bowel length necessary to prevent PN dependence in adults is approximately 100 cms in the absence of intact and functional colon or 60 cm in the presence of complete functional colon; however,

the degree of adaptation and PN dependence is highly individualized. Adult patients at the greatest nutritional risk are, those with a duodenostomy or with a jejunoileal anastomosis with <35 cm of residual small bowel, a jejunocolic or ileocolic anastomosis with <60 cm of residual small intestine, or an end jejunostomy with <115 cm of residual small bowel [5]. Children with <30 cm of jejunum-ileum, lack of enterocolonic continuity, and lack of feeding tolerance early after birth are associated with failure of weaning from PN [6]. Forty percent of children with <40 cm of residual small bowel and without an ICV remain dependent on parenteral nutrition after 8 years [3].

It is sometimes difficult to determine the anatomy and the intestinal length of the remaining bowel. A good approach is to start by reviewing surgical records and obtaining radiological studies (Table 13.2). If the previous steps are combined with an esophagogastroduodenoscopy (EGD) and a colonoscopy, it is possible to define the anatomy, histology and any anomalies of the remnant intestine. In a recent review of the outcomes of IF, it was observed that 58% of the patients with intestinal stricture/obstruction were misdiagnosed by an UGI and SBS and correctly diagnosed by endoscopic procedures [7].

Nutritional assessment

Nutritional assessment begins with a detailed history of a patient's weight, and parenteral and enteral nutrition intake, either by tube or oral feeding (fluids/kg, calories/kg, macro and micronutrients components). Questions related to bowel functions include diarrhea, nausea, vomiting, bloating, and other factors affecting appetite. Referral to a dietitian for estimation of the type and amount of daily food is appropriated. The physical exam should begin with accurate height and weight measurements, calculating the body mass index. Poor dentition, loss of subcutaneous fat and loss of temporal muscle mass are other signs of weight loss and inadequate nutrition. Measurement of triceps skin fold thickness and midarm circumference may also be followed. Laboratory tests including serum electrolytes, albumin level, prothrombine time, Vitamin B12, and fat soluble vitamins are important.

Evaluation of the small bowel absorptive capacity with functional assays such as D Xylose and 3-O-methylglucose absorption test 6 and 7 and net digestive ratio of nutrients have been developed, but these methods are time consuming and complicated to analyze, more suited for research than for medical screening. Monitoring of weight gain and growth is the most beneficial tool to evaluate nutritional status and nutrition absorption. Efforts to identify malabsorbed nutrients in the stool are not helpful.

Biological assessment of intestinal failure

Serum citrulline, a nonessential amino acid, synthesized exclusively in small intestinal enterocytes, was recently found to be a reliable biochemical marker of small bowel enterocyte mass. Plasma citrulline correlated with remnant small bowel length and net digestive absorption of fat and protein, in patients with SBS, villous-atrophy associated small bowel disease or during intestinal graft rejection with subsequently villous atrophy [8,9]. Citrulline is a marker of small bowel absorptive capacity independent of intestinal inflammation [10]. Whether citrulline levels are predictive of intestinal recovery or not has to be confirmed.

Assessment of complications

It is important to assess the acute and chronic complications to determine prognosis and future management. The most common problems in the acute or chronic stages of IF are diarrhea, fluid and electrolyte abnormalities and nutrient loss. Many of the chronic complications come from the use of PN, including central venous catheter (CVC) infections, central venous thrombosis (CVT), pulmonary embolism (PE), intestinal failure associated-liver disease (IFALD), metabolic complications, anemia, bone demineralization and rickets. Complications not related to PN include bacterial overgrowth, nutritional deficiencies and renal stones [11].

CVC infections and IFALD are a major cause of morbidity and mortality of patients with IF. CVC may be suspected if a patient develops clinical signs of fever, metabolic acidosis, thrombocytopenia, fluctuations in serum bilirubin, glucose instability, or a new onset of recurrent vomiting. Blood cultures taken from both peripheral and central sites are imperative in the presence

Table 13.3 Laboratory Exams.

Day 1
1 CBC with differential and platelet count. Comprehensive metabolic panel, GGT, phosphorous, magnesium, PT/PTT, cholesterol, triglycerides, urinalysis.

Day 2: For patients with concomitant liver disease.
2 HIV1/HIV2 antibody screen
3 Hepatitis Panel: HB/SAG and AB, HCAB, HepA
4 Alpha fetoprotein.
5 Alpha 1 antitrypsin.

Day 3:
Carnitine free and total, zinc, selenium, copper, Vitamin D250H, Vitamin A and E, Vitamin B12.
If liver disease coexists: For children under 1:
1 CMV DNA
2 EBV DNA
3 CMV IgG, IgM
For children over 1:
1 EBV DNA
2 EBV antibodies
3 CMV IgG, IgM

Table 13.4 Indications for Intestinal Transplantation [14].

1. Imminent or evident liver failure as a result of TPN-induced liver injury. A total bilirubin of more than 7 mg/dL, stage 2 fibrosis, or portal hypertension have been recognize as clear indications for liver/intestinal transplantation.
2. Thrombosis of two or more central veins.
3. The development of two or more episodes of systemic sepsis secondary to line infection per year that require hospitalization.
4. A single episode of line-related fungemia, septic shock, or acute respiratory distress.
5. Frequent episodes of severe dehydration despite intravenous fluid supplementation in addition to PN.

of any of these symptoms. Although CVT is a common sequel of central venous catheterization, the majority of patients remain asymptomatic. Clinical symptoms of CVT are swelling in the neck, face, or limbs, prominent superficial veins or pain on starting PN. CVT is confirmed by Echocardiography Doppler Ultrasound, CT scan, and/or venography. Spiral computed tomography is a safe stand-alone test for the diagnosis of PE [12].

Early hepatic dysfunction is asymptomatic, but jaundice is an obvious sign of cholestasis. Clinical signs of advanced IFALD are jaundice, hepatosplenomegaly, collateral vessels, ascites, and signs of coagulopathy. Despite fairly extensive hepatic fibrosis and splenomegaly, esophageal varices are uncommon in SBS patients. The development of hepatic dysfunction should be strongly investigated for other causes of liver disease before assuming the diagnosis of IFALD with specific tests described in Table 13.3, as well as liver ultrasound and liver biopsy [11,13].

At the end of the initial assessment, the physician should understand the cause of IF, the reasons for feeding intolerance, the likelihood of intestinal anatomy, the patient's nutritional status and also recognize any acute or chronic complications. In this way, one can choose the best management approach intestinal rehabilitation or liver small bowel transplantation (Table 13.4).

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