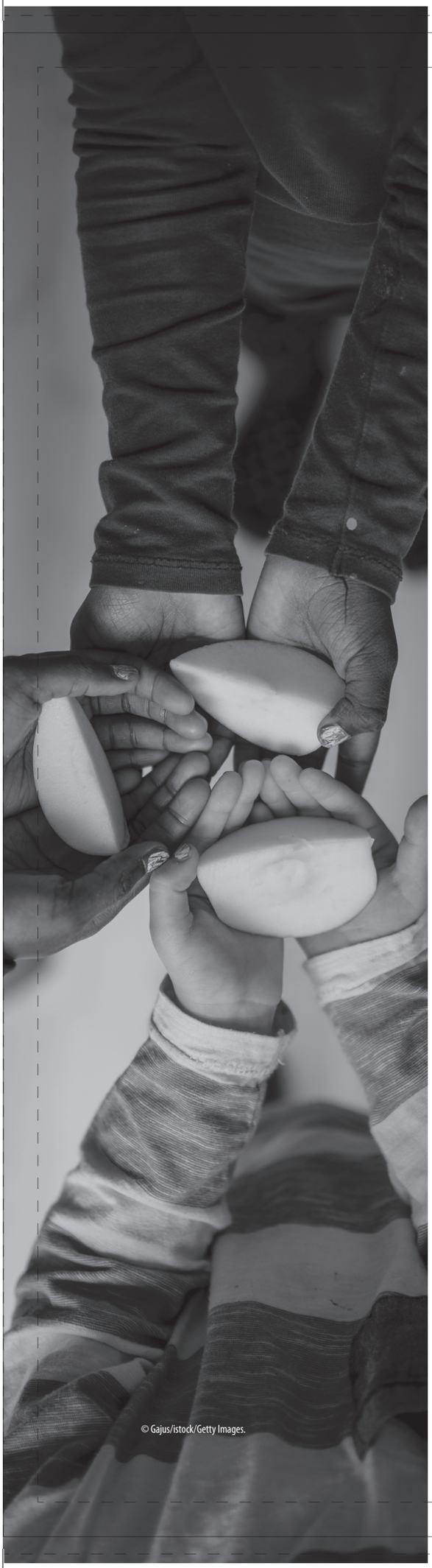

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SECTION V

Pediatric Medical Nutrition Therapy

CHAPTER 12

Gastrointestinal Disorders

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LEARNING OBJECTIVES

- Explain how to assess, diagnose, and manage common pediatric gastrointestinal nutrition related topics such as celiac disease, lactose intolerance, inflammatory bowel disease, and pancreatitis
- Discuss normal digestion and absorption in the gastrointestinal tract
- Identify diagnosis related at-risk macro and micronutrients
- Interpret common diagnostic tests for pediatric gastrointestinal disorders

Introduction

The gastrointestinal tract is an organ system that stretches from the mouth to the anus. It is a large muscular tube designed to ingest, digest, and absorb nutrients before expelling waste as feces. Functions of the gastrointestinal tract may be disrupted by disease, injury, chemotherapeutic agents, antibiotics, parasites, environmental toxins, and/or bacterial overgrowth, resulting in alterations in nutritional requirements. Many nutrients are absorbed throughout the intestinal tract, whereas others are absorbed only at specific sites. Absorption of the latter class of nutrients is particularly vulnerable to disease or surgical resection. **FIGURE 12.1** graphically portrays the principal sites of absorption of macro- and micronutrients, vitamins, and minerals.

Symptoms of gastrointestinal disease can arise from disorders located in a specific region of the bowel, the entire bowel, or distant sites (for example, vomiting can occur due to pyloric stenosis, gastroenteritis, or a brain tumor). Common pediatric disorders are listed in **TABLE 12.1** and common diagnostic tests are presented in **TABLE 12.2**. Many tests to evaluate gastrointestinal function as well as the presence or absence of disease are available.

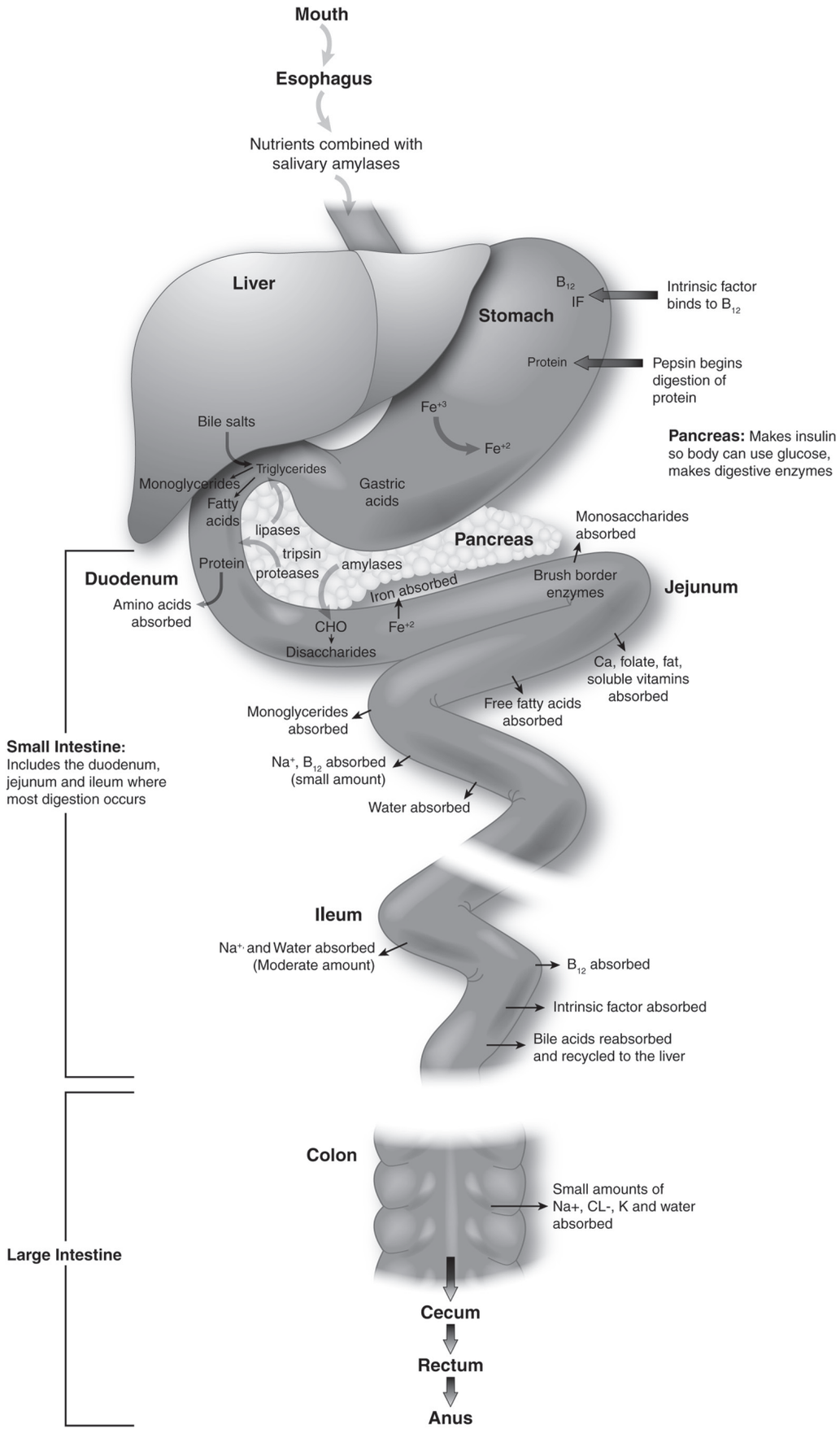

FIGURE 12.1 Normal Digestion and Absorption in the GI Tract.

TABLE 12.1 Common Pediatric Gastrointestinal Disorders

Presenting Symptom	Differential Diagnosis	Treatment
Stomach and Esophagus		
Vomiting/regurgitation	Congenital anomaly of the gastrointestinal tract	Surgery
	Gastroesophageal reflux	Infants: positioning, medications such as antacids, H2 blockers, and proton pump inhibitors (PPI). If preceding fails, consider surgical treatment.
		All ages: medications, antacids, H2 blockers, PPI, avoid caffeine-containing foods and other personal triggers
	Eosinophilic esophagitis	Elimination diet, swallowed steroids
	Eosinophilic gastritis	Steroids, immunosuppressive medication
	Peptic disease	Medications such as antacids, H2 blockers, and PPI; avoid caffeine-containing foods and other personal triggers
	H. pylori	Antibiotics, PPI
	Gastroparesis	Prokinetics, diet changes such as multiple small low-fat meals per day, or postpyloric feeds
Dysphagia (choking after eating), odynophagia (pain with swallowing)	Congenital anomalies, strictures, webs	Surgery
	Eosinophilic esophagitis	Elimination diet, swallowed steroids
	Esophageal spasms/dysmotility	Calcium channel blockers and nitrates; avoid extreme temperatures in foods
	Peptic strictures	Medications such as antacids, H2 blockers, and PPI; dilation
Liver and Pancreas		
Jaundice	Extrahepatic biliary tract obstruction, such as biliary atresia	Surgical correction; diet/formula with medium chain triglycerides (MCT), fat-soluble vitamin supplementation, choleretic agents such as ursodeoxycholate
	Autoimmune hepatitis	Steroids, evaluation for fat malabsorption, fat-soluble vitamin supplementation, protein restriction only if encephalopathic
Jaundice with recurrent abdominal pain	Gallstones	Surgery
	Choledochal cyst	Surgery
Nausea, vomiting, abdominal pain	Pancreatitis	NPO; if severe or prolonged course expected then postpyloric tube feeds or parenteral nutrition; pain control, H2 blockers; when clinically able, resume low-fat oral diet
	Pancreatic pseudocyst	Monitor cyst size; if cyst increases with enteral nutrition, may require parenteral nutrition

(*continues*)

TABLE 12.1 Common Pediatric Gastrointestinal Disorders (*continued*)

Presenting Symptom	Differential Diagnosis	Treatment
Chronic diarrhea, failure to thrive	Pancreatic insufficiency, such as cystic fibrosis	Enzyme replacement therapy, fat-soluble vitamin supplementation, high calorie balanced diet
	Cholestatic disease	Diet/formula with MCT, fat-soluble vitamin supplementation
Small Bowel and Colon		
Anemia, gastrointestinal bleeding	Congenital malformations, such as Meckel's diverticulum, duplication cysts	Surgery
Vomiting	Food allergies	Hydrolysate formula, elimination diet
	Infectious enteropathies	Oral rehydration solutions, followed by lactose and/or sucrose restrictions
Diarrhea in neonatal period	Congenital disorders of carbohydrate absorption and transport	Restriction of the problematic carbohydrate, balanced nutrition, vitamin/mineral supplementation, enzyme replacement
Diarrhea, perioral and perianal rash	Zinc deficiency	Zinc supplementation
Diarrhea	Food allergies	Elemental formula and/or elimination diet
	Infectious enteropathies	Intravenous fluids, oral rehydration solutions, followed by lactose and/or sucrose restrictions if clinically indicated
	Crohn's disease	Enteral feeds for therapy and/or malnutrition, replete iron, fat-soluble vitamins, and zinc as necessary; monitor vitamin B_{12} if severe ileal disease or resection
	Ulcerative colitis	Enteral feeds for weight gain, replete iron as necessary, low-residue diet if strictures
	Celiac disease	Gluten-free diet
	Short bowel syndrome	Parenteral nutrition progressing to enteral nutrition to oral feeds; vitamin and mineral supplements specific to patient's condition
	Fructose intolerance	Dietary restrictions of fructose-containing foods
	Lactose intolerance	Dietary restrictions of lactose-containing foods
Diarrhea, normal growth pattern	Irritable bowel syndrome, chronic nonspecific diarrhea, toddler's diarrhea	Normal diet for age, increased soluble fiber intake, decreased intake of sorbitol-containing beverages (apple and pear juice) and other personal triggers

Presenting Symptom	Differential Diagnosis	Treatment
Abdominal distention/pain	Celiac disease	Gluten-free diet
	Short bowel syndrome	Total parenteral nutrition progressing to MCT-predominate hydrolysate formula; vitamin and mineral supplements
	Functional constipation	Complete bowel clean-out using saline enemas, mineral oil, Miralax; high-fiber diet and adequate fluids; bowel habit training
	Congenital disorders of carbohydrate absorption and transport	Restriction of the problematic carbohydrate, balanced nutrition, vitamin/mineral supplementation, enzyme replacement
	Fructose intolerance	Dietary restrictions of fructose-containing foods
	Lactose intolerance	Dietary restrictions of lactose-containing foods
Constipation	Hirschsprung's disease; post-NEC strictures	Surgery
	Functional constipation	Complete bowel clean-out using saline enemas, mineral oil, Miralax; high-fiber diet and adequate fluids; bowel habit training

TABLE 12.2 Common Diagnostic Tests for Pediatric Gastrointestinal Disorders

Test	Description	Useful to Help Diagnose
Barium enema	Barium sulfate administered by enema; colonic lumen and mucosa visualized by fluoroscopy.	■ Colonic strictures and obstructions ■ Hirschsprung's disease ■ Polyps
Barium swallow	Barium sulfate administered orally; upper gastrointestinal tract is visualized by fluoroscopy.	■ Aspiration ■ Dysmotility disorders ■ Hiatal hernias ■ Strictures ■ Varices
Breath hydrogen test	Oral administration of sugar and expiratory collection of hydrogen as an indirect measure of bacterial fermentation of unabsorbed carbohydrate.	■ Fructose malabsorption ■ Lactose malabsorption ■ Bacterial overgrowth
DXA (dual energy X-ray absorptiometry)	Measures bone density in the spine, hip, or forearm.	■ Osteomalacia ■ Osteopenia ■ Osteoporosis
Colonoscopy	Insertion of flexible fiber optic tube via anus into large bowel; visual examination of colonic lining, biopsies obtained.	■ Colitis ■ Polyps

(*continues*)

TABLE 12.2 Common Diagnostic Tests for Pediatric Gastrointestinal Disorders (*continued*)

Test	Description	Useful to Help Diagnose
CT (computed tomography scan) of abdomen	Multiple radiographs of abdomen with or without intraluminal and/or intravenous contrast; computer reconstructs multiple images to generate "slices" through the abdomen.	■ Areas of inflammation (e.g., abscess) ■ Blood vessel anatomy and obstructions ■ Organ size and consistency ■ Tumors
EGD (esophagogastroduodenoscopy)	Fiber optic tube is inserted into upper gastrointestinal tract allowing mucosal lining of upper GI tract to be visualized and biopsies to be taken.	■ Celiac disease ■ Duodenitis ■ Esophagitis, including eosinophilic esophagitis (EE) ■ Gastritis ■ Peptic ulcer disease
Fecal fat test	Concurrent 3-day diet record of fat intake and stool collection; comparison as percentage of total fat in 24 hours excreted in stool. Malabsorption indicated in children if greater than 7% of fat is excreted; for infants less than 6 months of age if greater than 15% of fat is excreted.	■ Fat malabsorption ■ Pancreatic insufficiency
pH probe	Tube with pH sensor is inserted into esophagus for 24 hours with feeding at regulated intervals.	■ Gold standard for gastroesophageal reflux
Scintigraphy ("milk scan")	Barium ingested with X-rays capturing movement through upper GI tract	■ Delayed gastric emptying ■ Pulmonary aspiration
Ultrasound	Can be of all abdominal organs or individual organs such as the stomach, intestines, gallbladder, liver, spleen, pancreas, kidney, and bladder	■ Anatomical abnormalities ■ Cysts ■ Obstructions ■ Stones ■ Tumors
Upper GI/upper GI with small bowel follow-through:	Barium ingested with X-ray monitoring of path in GI tract to duodenum or to ileum if small bowel follow through.	■ Anatomical abnormalities ■ Inflammation ■ Tumors
X-ray of abdomen		■ Bowel dilatation or obstruction ■ Calcified gall bladder stones ■ Gas patterns and free air ■ Presence of stool in GI tract ■ Pnuematosis ■ Toxic megacolon

Data from Corkins MR, Scolapino J. Diarrhea. In: Merritt R, ed. *The ASPEN Nutrition Support Practice Manual,* 2nd ed. Silver Spring, MD: ASPEN; 2005:207–210.[1]
Graham-Maar RC, French HM, Piccoli DA. Gastroenterology. In: Frank G, Shah SS, Catallozzi M, Zaoutis LB, eds. *The Philadelphia Guide: Inpatient Pediatrics.* Philadelphia, PA: Lippincott, Williams, and Wilkins; 2003:100–115.[2]
Leonberg BL. *ADA Pocket Guide to Pediatric Nutrition Assessment.* Chicago, Il: American Dietetic Association; 2008:106.[3]

Common gastrointestinal problems will be discussed in the first section of this chapter. These include acute diarrhea, chronic diarrhea, constipation, gastroesophageal reflux, and lactose intolerance. Discussions of celiac disease, inflammatory bowel disease, pancreatitis, cholestatic liver disease, liver transplant, short bowel syndrome, and intestinal transplant will be presented later.

Diarrhea

Diarrheal illnesses in children follow a continuum from acute to chronic or persistent diarrhea. Acute diarrhea can be a common side effect of gastroenteritis, infectious diseases, foodborne illness, and chronic gastrointestinal diseases like Crohn's Disease and irritable bowel syndrome. Diarrhea has been defined by the World Health Organization as passage of three or more loose, watery stools per day or as 10 mL/kg liquid stool per day.[1,4] A child having diarrhea for 3–7 days is among the most common reasons for seeking the assistance of a pediatrician and is estimated to cost at least $1.5 billion yearly for evaluating and treating the population cared for.[5] Incidence of infectious diarrhea is estimated at approximately 99 million new cases each year in the United States.[6] Foodborne illness is a major cause of these cases.

Chronic or persistent diarrhea has been defined as "the passage of ≥3 watery stools per day for >2 weeks in a child who either fails to gain or loses weight." [7] Persistent diarrhea has many triggers, including acute diarrhea caused by an enteric infection, and more recently HIV infection and AIDS.[7] Chronic or persistent diarrhea (also called intractable diarrhea of infancy) has been thought of as a nutritional disorder, and certainly requires nutritional treatment for recovery. As with all diarrheal illnesses in infants, chronic diarrhea is dangerous if not treated promptly and appropriately, because it can result in dehydration and severe malnutrition.[8] Recent reports indicate that the incidence of chronic diarrhea has declined in the United States over the past two decades due to better treatment of acute diarrheal episodes.[9]

Anatomy, Physiology, Pathology & Diagnosis

Diarrhea is classified into five categories including: osmotic, secretory, dysmotility, malabsorption, and inflammatory. These classifications can be acute or chronic based on origin of causality.

Osmotic diarrhea occurs when active particles in the intestine pull water into the lumen in an attempt to normalize osmolality. As water passes through the lumen the bowel is unable to reabsorb fluid due to the presence of the nonabsorbable solutes. Stool volume is typically less than 1 liter per day.[10] Diarrhea stops when the dietary cause or solute particle is removed. Osmotic diarrhea can be caused by both dietary and pharmacological agents outlined below:

- Maldigestion of nutrients such as fat and carbohydrates
- Excessive sorbitol of >10 g/d (found in many liquid medications)[10]
- Medications including antihypertensives, cholinergics, glucose-lowering agents, anti-inflammatory drugs, laxatives, chemotherapy agents[10]
- Prokinetics (most common when receiving combination therapy of both erythromycin & metoclopramide)[10]
- Excessive fructose, lactose, or simple sugar
- Enteral formula feeding
- Overfeeding in infants and young children
- Food allergies
- "Starvation stools," defined as loose stool caused by a prolonged period of consuming only clear liquids[11]
- Laxatives

Secretory diarrhea results from excessive fluid and electrolyte secretions into the intestine. Increased fluid drawn into the intestinal lumen exceeds the absorption capacity of the bowel. Stool volume is typically greater than 1 liter per day.[10] The difference in the underlying disease is the factor causing the excessive secretions. Secretory diarrhea does not resolve when the patient is indicated as having nil per os, or nothing by mouth, status. Causes of secretory diarrhea are outlined below:

- Foodborne illnesses including *Escherichia coli*, clostridium difficile, and salmonella
- Medications, including antibiotic-associated diarrhea[10]
- Hormone-producing tumors
- Excessive prostaglandin production
- Excessive amounts of bile acids or unabsorbed fatty acids in the colon
- Rotavirus or *Clostridium difficile* ova and parasites
- Crohn's or celiac disease[12]
- Acquired immune deficiency syndrome[12]
- Pancreatic insufficiency or Cystic Fibrosis
- Zinc Deficiency

Diarrhea from dysmotility occurs when gastric, small bowel, or colonic motility alterations cause rapid transit time throughout the bowel. Diarrhea alternates with constipation and intermittent abdominal pain is present. Common causes include irritable bowel syndrome or gastrectomy. *Diarrhea from malabsorption* is caused by damage to the bowel or loss of bowel absorption causing maldigestion of nutrients. Symptoms include bloating, excessive gas, steatorrhea, and weight loss. Malabsorptive diarrhea can be caused from short bowel syndrome, bacterial overgrowth, pancreatic insufficiency, and **congenital-sucrose iso-maltase deficiency** (CSID).

Inflammatory diarrhea is due to intestinal inflammation that impairs the absorption of water and electrolytes. Characteristics include the presence of mucus, blood, and/or proteins in stool as well as an elevated white count. Intestinal lumen damage and small bowel disease are causes of inflammatory diarrhea.

An initial diagnosis of diarrhea is based on the degree of stool output and examination of stool cultures evaluating presence of blood, microorganisms, ova and parasites, leukocytes, and lactoferrin. Additional procedures to evaluate intestinal structure and function including upper endoscopy, flexible sigmoidoscopy or colonoscopy and bowel biopsy may be used to assist in diagnoses.

Nutrition Screening

Nutrition screening in pediatric patients with acute diarrhea should be consistent with clinical malnutrition (undernutrition) risk screening as defined by the **Academy of Nutrition and Dietetics** (AND) and the **American Society for Parenteral and Enteral Nutrition** (ASPEN).[13]

Nutrition Assessment

Patients with acute and chronic diarrhea presenting with signs of severe dehydration, poor oral intake, or malnutrition should be referred to a pediatric **registered dietitian/nutritionist** (RDN) with expertise in gastrointestinal disorders in order to receive individualized nutrition assessment. Nutrition assessment for patients with diarrhea consists of obtaining a thorough diet history with emphasis placed on fluid, fiber, and food intake or breastmilk/formula intake. If infection as a cause is excluded, other etiologies must be considered. Additional dietary history including food allergy, dietary protein intolerances, wheat intolerance, and lactose or other disaccharide intolerances should be gathered. In addition to nutritional intake, one should evaluate changes in appetite, as well as nausea and vomiting since these may coincide with underlying causes of diarrhea. Close attention should also be paid to anthropometric assessment, growth trends, electrolyte levels, and zinc deficiency with need for supplementation.

Nutrition Diagnostic Statements (PES)

- Altered GI function (NC-1.4) related to excessive intake of poorly absorbed carbohydrates as evidenced by cramping and loose stools.
- Food- and nutrition-related knowledge deficit (NB-1.1) related to frequent intake of apple juice and products containing sorbitol as evidenced by cramping and loose stools.
- Inadequate mineral intake: zinc (NI-5.10.1.8) related to environmental causes as evidence by altered GI function: diarrhea.

Management

The **American Academy of Pediatrics** (AAP) and the **Centers for Disease Control and Prevention** (CDC) have issued practice parameters and guidelines delineating treatment depending on the presence of dehydration.[5,14,15] "Gut rest," in which food is restricted, is an outdated concept and may result in malnutrition. General principles of diarrhea management are replacing fluid and electrolyte losses and nutritional therapy with early age-appropriate feeding.[4]

Replacing fluid and electrolyte losses is the primary medical management for acute diarrhea. Standard **oral rehydration solutions** (ORS) have been shown to replete electrolytes and fluid yet have not been shown to decrease fecal volume or duration of diarrhea. The investigative use of lower osmolality ORS have found some success in decreasing fecal volume and diarrhea duration.[16] Based on the severity of output, using an oral rehydration solution may be recommended. The composition of commonly used oral maintenance and rehydration solutions are presented in **TABLE 12.3**.

If commercial ORT are not available and rehydration is critical, home-made oral rehydration-like recipes may be tried. Homemade oral rehydration recipes do not contain the same electrolyte profile as commercial products. Examples of these types of solutions are presented in **TABLE 12.4**.

If oral rehydration fluids are unable to adequately rehydrate a patient, intravenous fluids may be required. Acute diarrhea often does not require **enteral nutrition** (EN) support for management. Chronic diarrhea, specifically malabsorptive and inflammatory, may require EN support when patients are malnourished, have inadequate oral intake or require slow continuous feeds to minimize osmotic load. In a patient with persistent diarrhea requiring long-term nutrition support, **parenteral nutrition** (PN) may be indicated if oral rehydration and enteral feedings are unable to provide adequate nutrition needs.[19]

In addition to rehydration, medical management focuses on treating the underlying gastrointestinal disorder associated with a diarrhea episode. If the diarrhea is infectious, antibiotics may be used as treatment. Anti-diarrheal agents are not recommended in those with infectious diarrhea. Patients without infection cause, specifically those with malabsorptive diarrhea, may benefit from intermittent use of anti-diarrheal agents.

Nutrition Intervention

Nutrition management varies with the degree of dehydration. The AAP has distinguished stages of dehydration using the following physical signs:

- *Mild:* Slightly dry mucous membranes, increased thirst

TABLE 12.3 Oral Rehydrating Solutions (ORS)

Solution	Glucose/ CHO (g/L)	Sodium (mEq/L)	Potassium (mEq/L)	Osmolality (mmol/L)	CHO/Sodium
Pedialyte (Abbott, Columbus, Ohio)	25	45	20	250	3.1
Pediatric Electrolyte (PendoPharm, Montreal, Quebec)	25	45	30	250	3.1
Kaolectrolyte (Pfizer, New York, New York)	20	48	20	240	2.4
Rehydralyte (Abbott, Columbus, Ohio)	25	75	20	310	1.9
WHO, ORS, 2002 (reduced osmolarity)	75	75	30	224	1
WHO, ORS, 1975 (original formulation)	111	90	20	311	1.2
Cola*	126	2	0.1	750	1944
Apple juice*	125	3	32	730	1278
Gatorade* (Gatorade, Chicago, Illinois)	45	20	3	330	62.5
Whole cow's milk	12 (lactose)	40	1226	285	Not available

*Cola, juice, and Gatorade are shown for comparison only; they are not recommended for use.
Abbreviations: CHO, carbohydrate; WHO, World Health Organization.
Data from Oral therapy for acute diarrhea. In: Kleinman RE, ed. *Pediatric Nutrition Handbook*, 6th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2009:651–659.[5]
Roberts J, Shilkofski N. *The Harriet Lane Handbook*, 17th ed. Elsevier Mosby; 2005:559, Table 20–14a.[17]

- *Moderate:* Sunken eyes, sunken fontanelle, loss of skin turgor, dry mucous membranes
- *Severe:* Signs of moderate dehydration plus one or more of the following: rapid thready (scarcely perceptible) pulse, cyanosis, rapid breathing, delayed capillary refill time, lethargy, coma[1]

Mild Diarrhea with No Dehydration

The AAP encourages continuing a normal diet throughout the acute illness, including breastfeeding or full strength infant formula and a regular diet, excluding beverages high in sugar (resulting in high osmolality) such as juices and sodas. Increased fluid intake is necessary to compensate for losses. Infants and children who are not dehydrated can be kept hydrated by using frequent breastfeeding, usual infant formula, and milk. The use of lactose-free formula is no longer recommended in managing acute diarrhea.[1] Infant formulas with added soy fiber have been reported to reduce liquid stools with no change in overall stool output in acute diarrhea and to reduce the length of antibiotic-associated diarrhea.[14,20,21] Using soy formula with added fiber is not a standard of care because continuation of breast feeding or usual formula works to correct dehydration in most cases. Lactose-containing products, especially when given with complex carbohydrates, are no longer thought to increase diarrheal output or prolong the illness unless stool output clearly increases on a lactose-containing diet.[22]

Eating a regular diet during mild diarrhea does not change the volume of stool output. Parents should be educated to continue feeding their child a regular diet and offer adequate hydration.[5] Most infants and children demonstrate hunger and thirst during mild, acute diarrheal illness, and parents can respond to these cues. Historically, using the BRAT (bananas, rice, applesauce, and toast) diet guided the initial food choices for acute diarrhea; however, it is no longer recommended and should be avoided per the AAP nutrition guidelines.[23] However, foods that are high in carbohydrates, such as rice, wheat, peas, and potatoes, may slow diarrheal output.

Diarrhea with Mild to Moderate Dehydration

Increased fluid intake is necessary to compensate for losses and may require the use of ORS in addition

TABLE 12.4 Home-Made "Oral Rehydration-Like" Solutions Using Alternative Beverage Base

Recipe	Glucose/ CHO (g/L)	Sodium (mEq/L)	Potassium (mEq/L)	Calories per L	Osmolality (mmol/L)
Regular Gatorade® Drink ■ 1 ½ cup Gatorade® ■ 2 ½ cups water ■ ½ plus ¼ teaspoon salt	21	70	1.2	84	298
Powerade® ■ 1 ½ cups Powerade® ■ 2 ½ cups water ■ ¾ teaspoon salt	22	79	1.7	88	306
Grape or Cranberry Juice ■ ¾ cups juice ■ 3 ½ cups water ■ ¾ teaspoon salt	26/23	76/76	6.7/0.7	104/92	289/296
Chicken Broth ■ 4 cups water ■ 1 dry chicken broth cube ■ ¼ teaspoon slat ■ 2 tablespoons sugar	25	75	0.5	100	224
Vegetable Broth ■ 2 cubes vegetable bouillon ■ 4 cups water ■ 2 tablespoons sugar	25	77	1.3	100	228
Tomato Juice ■ 2 ½ cups tomato juice ■ 1 ½ cups water	20	74	27.5	122	259

Gatorade® is a registered trademark of PepsiCo
Powerade® is a registered trademark of Coca-Cola
Abbreviations: CHO, carbohydrate
Modified from Home-Made "Oral Rehydration-Like" Solutions Using Alternative Beverage Base In: Pocket Guide for Patients: Managing Short Bowel Syndrome 3rd Edition by Parrish RE, 2016.[18]

to the regular diet.[9] The use of ORS helps to replace fluid and electrolyte losses from diarrhea. Dehydration can be treated at home by giving an ORS solution by syringe at the rate of 1 teaspoon (5 mL) per minute over 4 hours for a child less than 15 kg or 2 teaspoons (5–10 mL) for children 15–20 kg. This method of fluid administration is adequate to replace the fluid deficit within a 4-hour period. After 1–2 hours of this treatment, the infant or child may begin voluntarily accepting the rehydration liquid. If the child or infant is unable to cooperate, a nasogastric (NG) tube may be used at home or in the hospital. After correction of dehydration, age-appropriate feeding should be initiated as described above.[9]

Diarrhea with Severe Dehydration

Severe dehydration in infants and children is a medical emergency and requires immediate hospitalization. Once rehydration is complete, age-appropriate feeding can be initiated.

Mild zinc deficiency may play a role in both acute and chronic diarrhea. Zinc supplementation in developing countries has been associated with a decrease in number of stools per day and decreased number of days with watery diarrhea in acute diarrhea and reduction of the duration of persistent diarrhea.[24] A randomized double-blind placebo-controlled trial of zinc supplementation in breastfed infants in the United States showed no significant difference in diarrhea

frequency in the supplemented and not supplemented groups.[25] In a Cochrane review, zinc supplementation was not found statistically significant in reducing diarrhea. Zinc supplementation may be beneficial in patient populations susceptible to deficiency. The WHO recommends zinc supplementation in those susceptible at 20 mg per day for 7–14 days for children 6 months and older and 10 mg per day for children younger than 6 months.[16]

Prebiotics and Probiotics

Using probiotics and prebiotics in infant and enteral formulas and in foods has been proposed as beneficial for treating acute and chronic diarrhea of infancy and childhood. *Probiotics* are live microorganisms, historically available in fermented foods such as yogurt, that promote health by improving the balance of healthy organisms in the intestinal tract.[14] Additional proposed mechanisms are preventing adhesion of microbes to the gut mucosa, downregulation of inflammatory responses, and stimulation of immunoglobulin A production.[26,27] Technology has allowed beneficial bacteria to be freeze-dried, added to formula or foods, and be reactivated in the gut when consumed.[28] *Prebiotics* are complex carbohydrates, not microorganisms, that promote the growth of healthy microorganisms in the intestinal tract.[14] Human milk contains oligosaccharides (a type of prebiotic) that promote the growth of *lactobacilli* and *Bifidobacterium* in the colon of breastfed infants.[14,29] Higher intake of breast milk has been associated with a lowered incidence of acute diarrhea.[30]

Questions about consuming live probiotic bacteria and prebiotics include whether long-term consumption is safe and whether consumption has positive health effects. A randomized controlled trial has reported that healthy infants consuming a formula supplemented with prebiotic mixtures achieved normal growth and had stools more similar to breastfed infants when compared with infants fed an unsupplemented formula. The prebiotic mixtures included polydextrose and galactooligosaccharides in one group and polydextrose, galactooligosaccharides, and lactulose in the second group.[31] A double-blind randomized placebo-controlled trial evaluated the tolerance and safety of long-term consumption of different levels of cow's milk formula not supplemented and supplemented with different levels of *Bifidobacterium lactis* and *Streptococcus thermophilis* in infants 3–18 months of age. Healthy infants consuming the probiotic-supplemented formula reported a lower frequency of colic or irritability, and reduced severity of antibiotic-induced acute diarrhea.[32]

Manufacture of infant formulas with added prebiotics and probiotics have steadily increased in the past decade. Formula companies claim the benefits for inclusion, promoting digestive health, improving fussiness, and mimicking breast milk. Common prebiotic and probiotic additives in infant formulas include *Lactobacillus rhamnosus* GG and galactooligosaccharides. Pediatric enteral formula for children over 1 year of age with added pre- and probiotics are also on the rise. Examples include Boost Kids Essentials 1.0 and 1.5 (*L. reuteri* inserted in optional straw to use for drinking) by Nestlé,[33] Pediasure enteral formula with fiber (NutraFlora and scFOS) by Abbott;[34] Pediasure Peptide 1.0 and 1.5 (NutraFlora and scFOS), also from Abbott;[34] and Peptamen Jr with fiber (contains insoluble fiber and Prebio, a blend of FOS and inulin) and Peptamen Jr with Prebio (no insoluble fiber) by Nestlé.[33]

In pediatric patients with acute gastroenteritis using probiotics, specifically *Lactobacillus* GG (LGG) and *saccharomyces boulardii (s boulardii)*, may reduce the duration and intensity of symptoms in healthy children.[35] A meta-analysis showed that in children *LGG* reduced the duration of diarrhea, decreased mean stool frequency, and decreased the risk of diarrhea lasting 4 or more days.[35–36] Another meta-analysis study examining *s boulardii* showed similar results in healthy infants and children. Using *s boulardii* decreased duration of diarrhea by 1 day, decreased the risk of diarrhea lasting 4 or more days, and decreased the length of stay in hospitalized children.[37] In both meta-analysis high dose probiotics showed the most benefit in reducing diarrhea suggesting a dose of $\geq 10^{12}$ CFU per day LGG and 250–750 mg per day of *s boulardii* for a duration of 5–7 days.[35] Probiotics that show low efficacy for managing acute enteritis include *Lactobacillus reuteri Strain DSM 17938* and Heat Killed *L. acidophilus LB.*[38] In children, strong recommendations have been made to avoid the probiotic, *enterococcus faecium*, due to safety issues as a possible recipient of the vancomycin-resistant genes.[39]

There is still no consensus concerning the types and amounts of probiotics that are beneficial. More research is needed.

Diarrhea Associated with Enteral Nutrition Support

Diarrhea associated with enteral feeds have been seen in up to 95% of critically ill patients.[10] Causes for diarrhea on enteral feedings include an excessive rate or volume of feeding into the stomach or small intestine, the presence of unabsorbed carbohydrate in the large intestine, and diarrhea secondary to medications, especially those with sugar alcohols.[40] Gastric feedings versus post pyloric feedings have been shown to decrease diarrhea. Most patients are able to tolerate a polymeric formula; however, if diarrhea occurs, switching to a lower osmotic peptide based formula, specifically less

than 300 mOsm, or high **medium-chain triglycerides** (MCT) may improve output.[40,41] Soluble liquid pectin at 1% to 3% mL per total formula volume and microlipids at 1–2 mL per 50 mL formula can be added to enteral formula in order to decrease and bulk stool output.[40] Using fiber containing formula in critically ill patients is discouraged due to the risk of splanchnic perfusion and mesenteric ischemia. ASPEN guidelines recommend 10–20 grams of fiber should be given in 24 hours if diarrhea is present.[42]

Monitoring and Evaluation

It is important to monitor patients closely to see if any adjustments are needed in nutrition intervention. These measurements can help monitor hydration status; intake/output records, stool consistency, laboratory values/electrolytes, and daily weights. As diarrhea improves and diet is advanced, it is important to monitor tolerance/intolerance to foods. Foods that exacerbate diarrhea can be eliminated from the diet temporarily.[19]

Constipation

Constipation is a prevalent functional gastrointestinal disorder of childhood. The vast majority of constipation is termed *functional constipation*, which is idiopathic in nature and "characterized by the passing of infrequent, inconsistent, dry, hard, or painful stools"; whereas *anatomic constipation,* is experienced by individuals born with gastrointestinal and/or genitourinary tract anomalies.[43] Constipation presents with an estimated worldwide prevalence rate of 0.7 to 29.6 percent and it is reported that approximately 17 to 40 percent of children experience constipation within their first year of life. Additionally, constipation accounts for roughly 1 to 3 percent of all annual pediatric primary care visits and results in additional US healthcare costs estimated at $3.9 billion dollars or more precisely $3,362.00 per treated child.[43–47]

Risk factors for constipation have been identified and include intestinal causes such as Hirschsprung's disease, Celiac disease, or anatomic (anorectal) malformations; diseases or consequences of metabolic/endocrine origin (ex. vitamin D intoxication, diabetes mellitus, hypothyroidism), as well as medication/vitamin use (e.g. antidepressants, antihistamines, iron/calcium supplements, opioids). Additionally, those who have been diagnosed with anorexia nervosa, cystic fibrosis, or have experienced physical and psychological trauma, as well as emotional stress are also at an increased risk for constipation. Lastly, low consumption of dietary fiber, physical inactivity, genetic predisposition, and prematurity of birth also increase one's risk of experiencing constipation.[43,45] Whether or not infants and children diagnosed with a cow's milk protein allergy are at greater risk for constipation remains debatable since scientific evidence is inconclusive.[46]

Anatomy, Physiology, Pathology

The physiology and pathophysiology of pediatric constipation is multi-factorial. In some instances, constipation occurs secondary to innate defects in colonic function or a malfunction of the defecation process. Conversely, a fully functioning large intestine allows chyme (which enters the large intestine from the small intestine via the ileocecal sphincter) to move within the colon, promoting adequate absorption of fluid, electrolytes, and nutrients, all while converting digested food into waste (feces), which is then transported from the sigmoid colon to the rectum and then expelled. Any interruption experienced during this process can result in constipation. Lastly, the reabsorption of fluid and electrolytes that occurs during prolonged colonic transit time may result in constipation.[43,45]

When stooling is chronically difficult or painful, children may withhold stool, aggravating the existing problem. Encopresis may result due to the stretched rectal wall, allowing a softer stool to leak out involuntarily. A bowel program to treat encopresis, after a thorough clean-out, often includes a high-fiber diet, adequate fluid, and increased physical activity.[45]

Nutrition Screening

Nutrition screening in pediatric patients with constipation should be consistent with clinical malnutrition (undernutrition) risk screening as defined by the AND and ASPEN.[13]

Nutrition Assessment

Patients with constipation should be referred to a pediatric RDN with expertise in functional gastrointestinal disorders in order to receive individualized nutrition assessment. Nutrition assessment for patients with constipation consists of obtaining a thorough diet history with emphasis placed on food, fluid, and breastmilk/formula intake, as well as the amount of caffeine, psyllium, and dietary fiber detailed in the diet history. Inadequate fluid intake, excessive cow's milk intake, and suboptimal dietary fiber consumption contribute to constipation. Besides nutritional intake, one should also evaluate changes in feeding behaviors, food acceptance, appetite, nausea/vomiting/retching, as well signs/symptoms of **gastroesophageal reflux disease** GERD given the co-occurrence of GERD and constipation in the pediatric population.[43,45–46] Close attention should also be paid to anthropometric assessment, growth trends, and

the need for multivitamin and mineral supplementation to help prevent and/or correct nutrient deficiency.[43,45]

Diagnosis

Presently, the diagnosis and classification of functional constipation is based on the Rome IV criteria (expert consensus criteria for diagnosing functional gastrointestinal disorders), history obtained, and physical examination.[43,46–47] Scientific evidence does not support the routine use of colonic transit studies, digital rectal examination, rectal ultrasound, or abdominal radiography to diagnose functional constipation in the pediatric population.[46]

Nutrition Diagnostic Statements (PES)

- Inadequate fiber intake (NI-5.8.5) related to limited oral consumption of fruits, vegetables, and whole grains as evidenced by a three-day diet history revealing an average fiber intake meeting less than the **Recommended Daily Allowance** (RDA) for age.
- Food and nutrition knowledge deficit (NB-1.1) related to patient and family presenting without prior knowledge of fiber-containing foods as evidenced by patient/family disclosure during clinic visit.

Management

Vital components for managing childhood constipation include medical education, nutrition intervention, treatment of fecal impaction to achieve disimpaction, maintenance therapy via osmotic or stimulant laxatives, and close follow-up.[45–46] Behavioral therapies aimed at decreasing defecation-related anxiety, discouraging the withholding of stools, and both improving and establishing toileting routines may serve as a beneficial non-invasive therapeutic intervention.[43,45] Lastly, surgical options may be considered in severe constipation cases when medical and nutrition therapy interventions have failed.[45]

To date, **polyethylene glycol** (PEG), an osmotic laxative not digested by colonic bacteria, is the primary laxative used for fecal disimpaction and maintenance treatment in the prevention of re-impaction.[45,48–49] PEG should not be used in children who are allergic, have a history of known or suspected bowel obstruction, and should be used prudently in children with swallowing difficulties secondary to an increased risk of aspiration occurrence.[48] The use of PEG has grown over recent years due to its efficacy, safety profile, and non-invasive administration route. Minor adverse reactions have been reported in the literature, which include bloating, increased flatulence, abdominal pain, nausea/vomiting, etc.[48] Recently, concerns have been raised regarding the lack of **Food and Drug Administration** (FDA) approval for the use of PEG for functional constipation in the pediatric population as well as parental fears related to laxative addiction.[48–49] Neuropsychiatric events, such as tics, tremors, and obsessive compulsive behavior have been reported to the FDA; prompting an ongoing research study investigating the overall safety of PEG in children.[48–49]

Nutrition Intervention

The specific nutrition diagnosis established from the nutrition assessment helps determine the appropriate medical nutrition therapy intervention. Treatment in infants may include juices that contain natural sorbitol such as apple, pear, or prune (0.5 g/100 g, 2.1 g/100 g, and 12.7 g/100 g, respectively), increasing fluids, and verifying that infant formula is mixed correctly.[50–51] Rice cereal, a common first food for infants, as well as commercially available jarred baby foods, contain minimal amounts of fiber and may lead to constipation. Replacing rice cereal with oatmeal cereal and preparing homemade infant purees from fresh fruits and vegetables with the skin left on should be considered and may help resolve symptoms.[43] Higher fiber diets, alongside adequate fluid intake and physical activity is recommended as the first line of therapy in children with constipation.[43,45–46] Recommended fiber intake for individuals between the ages of 1 and 18 years is listed in **TABLE 12.5**. The fiber content of common foods is presented in **TABLE 12.6**. Fiber supplements may be considered when dietary intake is

TABLE 12.5 Adequate Intake of Fiber for Children

Age (in years)	Total Fiber Intake (g/day)
1–3	19
4–8	25
Boys	
9–13	31
14–18	38
Girls	
9–13	26
14–18	26

Data from National Academy of Sciences. Institute of Medicine. Food and Nutrition Board. Dietary Reference Intakes for Energy, Carbohydrate, Fiber, Fat, Fatty Acids, Cholesterol, Protein, and Amino Acids (Macronutrients) 2005. Available at: https://www.nap.edu/read/10490/chapter/32#1324. Accessed: October 18, 2017.[52]

TABLE 12.6 Good Sources of Dietary Fiber

	Grams of Fiber	Serving Size
Fruit		
Apple	2.2	1 med. w/skin
Apple	2	1 med. w/o skin
Apricot	7.8	dried, 3 oz
Blueberries	4.4	1 cup raw
Dates, dried	4.2	10
Kiwi	3.4	3 oz
Pear	4.1	1 med. raw
Prunes, dried	7.2	3 oz
Prunes, stewed	6.6	3 oz
Raisins	5.3	3 oz
Raspberries	5.8	1 cup
Strawberries, raw	2.8	1 cup
Vegetables and Legumes		
Avocado, California, raw	3	1 med.
Beans, black, boiled	7.2	1 cup
Beans, great northern, boiled	6	1 cup
Beans, kidney, boiled	6.4	1 cup
Beans, lima, boiled	6.2	1 cup
Beans, baby lima, boiled	7.8	1 cup
Beans, navy, boiled	6.6	1 cup
Beans, green, canned	6.8	½ cup
Broccoli, boiled	2.2	½ cup
Chickpeas (garbanzo beans)	5.7	1 cup
Cowpeas (black-eyed peas)	4.4	1 cup
Lentils, boiled	7.9	1 cup
Ready-to-Eat Cereal		
FiberOne (General Mills)	14	½ cup
All-Bran (Kellogg's)	10	½ cup
100% Bran (Post)	9	⅓ cup
Shredded Wheat and Bran (Post)	8	1¼ cup
Raisin Bran (Post)	8	1 cup
Grape-Nuts (Post)	7	½ cup
Multi Bran Chex (General Mills)	6	¾ cup
Cracklin' Oat Bran (Kellogg's)	6	¾ cup
Mini Wheats (Kellogg's)	6	1 cup (30 biscuits)
Frosted Mini Wheats with raisins (Kellogg's)	6	¾ cup (24 biscuits)
Shredded Wheat (Post)	6	1 cup
Mini Wheats with strawberries (Kellogg's)	5	¾ cup (24 biscuits)
Wheat Chex (General Mills)	5	¾ cup
Bran Flakes (Post)	5	¾ cup
Banana Nut Crunch (Post)	4	1 cup
Raisin Bran Pecan Date Crunch (Post)	4	½ cup
Cranberry Almond Crunch	3	¾ cup
Low Fat Granola (Kellogg's)	3	½ cup
Grape-Nut Flakes (Post)	3	¾ cup

Data from Data for sections on fruits and vegetables/legumes from: Pennington JAT. *Bowes and Church's Food Values of Portions Commonly Used*, 15th ed. Philadelphia: JB Lippincott; 1989[53]
U.S. Department of Agriculture. *USDA Provisional Table on the Dietary Fiber Content of Selected Foods*; 1988. HNIS/PT-106.[54]
Data for section on ready-to-eat cereals from a survey of manufacturer's Websites as of November 2009.

inadequate. Presently, insufficient evidence exists that supports the use of prebiotics and probiotics in the treatment of pediatric constipation.[43,46]

Monitoring and Evaluation

Ongoing monitoring and evaluation is essential for treating, maintaining, and preventing pediatric constipation. Following the initiation of treatment, 60 percent of children with functional constipation experience resolution of symptoms between 6 and 12 months, with the remaining 40 percent of children still reporting symptoms.[45–46] Additionally, approximately 30 percent of children continue to experience symptoms beyond puberty.[45] It is very important for parents to understand that the duration of maintenance therapy ranges from 6 to 24 months.[45] Therefore, individualized medical and nutrition follow up is paramount in order to allow for appropriate adjustments in the treatment plan and receive ongoing support to promote compliance. Focus should be placed on medical nutrition therapy components previously discussed, along with determining dietary compliance, goal achievement, and the current status of the patient's nutrition diagnosis (e.g. "resolved, improved, no change, worsened").[43] One should also assess the comprehension level of the patient and family, along with addressing individual questions/concerns and providing continued support in an effort to achieve improved quality of life.

Gastroesophageal Reflux

Gastroesophageal reflux (GER) is the passage of gastric contents into the esophagus. GER is a normal physiologic process that occurs in healthy infants. GER may cause involuntary regurgitation or vomiting. GER is common in infants with more than 60% of infants regurgitating daily and over 25% regurgitating four or more times per day.[55] GER usually peaks at 4 months of age with tapering by 6 months and improvement of symptoms by 12–15 months when the child achieves a more upright posture.[55,56]

Anatomy, Physiology, Pathology

The primary physiologic cause of GER in infants is the transient relaxation of the lower esophageal sphincter allowing gastric contents to flow into the esophagus. Often, relaxation of the sphincter is spontaneous, however, increased abdominal pressure can cause reflux symptoms. Crawling, sitting, and coughing are common infant milestones that increase abdominal pressure and may worsen reflux.[57] When gastroesophageal reflux causes troublesome symptoms or complications of persistent GER such as recurrent regurgitation, weight loss, poor weight gain, dysphagia, or strider, it is classified as GERD.[58]

Nutrition Screening

Nutrition screening in pediatric patients with GERD should be consistent with clinical malnutrition (undernutrition) risk screening as defined by the AND and ASPEN.[13]

Nutrition Assessment

Patients with GER or GERD that present with growth related and intake related issues should be referred to a pediatric RDN with expertise in gastrointestinal disorders in order to receive an individualized nutrition assessment. A nutrition assessment for infants with GER/GERD consists of obtaining a thorough diet history with emphasis placed on breastmilk/formula intake, feeding position, frequency of feedings as well as using formula thickening agents. For children and adolescents, dietary recall, meal portion size, meal times, and frequency of meals should be assessed. Close attention should also be paid to anthropometric assessment and growth trends.[43]

Diagnosis

In most infants and children, a detailed history including feeding behavior, regurgitation occurrences, and a physical exam can diagnose uncomplicated GER.[46] To date, no set of symptoms or diagnostic test has been established to diagnosis GERD or identify what patients would respond to therapeutic interventions.[47] Diagnostic questionnaires related to reflux symptoms can be a useful tool in monitoring and detecting GERD in infants and children.[48] In certain cases a pH probe or impedance probe may be used to detect the number of reflux episodes and quantify GER, however, these test are unable to diagnosis GERD. An upper GI tract series or barium swallow test may be used in patients with troublesome GER/GERD symptoms in order to detect if there is a structural abnormality or motility disorder[58–61]; however, sensitivity and specificity of these test are low in the infant population.[62] In adolescent patients, there is a high reliability of diagnosing GER with the most common reported symptom being heartburn.[63]

Nutrition Diagnostic Statements (PES)

- Inadequate oral intake (NI-2.1) related to vomiting after feeding as evidenced by insufficient growth velocity
- Inadequate energy intake (NI-1.4) related to altered gastrointestinal function as evidence by weight for length z-score

Management

Thickening of feeds, formula modification, positioning changes, and changes in meal frequency are primary nonpharmacological management techniques for

infants with GER. Pharmacological acid suppression medications like histamine2 (H_2) antagonists and proton pump inhibitors are used and have been shown to reduce gastric acidity and heal esophagitis in children.[64] Currently, there is minimal evidence of efficacy of using acid suppression in infants.[65,66] The **North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition** (NASPGHAN) guidelines do not recommend using acid suppression therapy in healthy infants with regurgitation.[58] Shortterm use of acid-suppression therapy for infants with intractable or severe symptoms should only be considered after a hypoallergenic formula is trialed.[58] For patients with intractable GER, a surgical fundoplication in which the fundus of the stomach is wrapped around the lower end of the esophagus to prevent reflux, may be indicated.

Nutrition Intervention

In 2009, a joint committee of NASPGHAN and the **European Society for Pediatric Gastroenterology, Hepatology, and Nutrition** (ESPGHAN) revised clinical practice guidelines on pediatric GER and GERD.[58] The "happy spitter" is an infant who has frequent episodes of regurgitation but continues to grow well. Parental education concerning the expected improvement as the infant develops is all that is necessary. If the infant is not growing well or has increased irritability, changes in feeding are recommended. For recurrent regurgitation and poor weight gain, NASPGHAN guidelines suggest a 2 to 4-week trial of a hydrolyzed amino acid based formula in order to decrease reflux symptoms. If the infant is breastfed, eliminating cow's milk and/or soy from the mother's diet may be suggested. Discontinuing breastfeeding is rarely recommended. The exact mechanism for reducing reflux with a hypoallergenic formula is unknown. A common symptom of cow's milk protein allergy is reflux, however, milk protein allergy and GER are not mutually exclusive. No data exist to support the idea of an allergy to soy protein in infants that causes regurgitation and vomiting. Decreasing the volume of each feeding and offering more frequent feedings may also improve GER symptoms; however, total intake may decrease with this change.[58] Thickened formula may decrease the number of vomiting episodes and esophageal pain but does not reduce the number of reflux episodes.[58,67] Using infant rice or oatmeal cereal to thicken formula is the most common thickening technique in North America.[58] It is important to note that The Academy of Pediatrics does not recommend the introduction of infant cereal into a child's diet before 4 months of age and the early introduction through formula thickening has been linked to increase incidents of **necrotizing enterocolitis** (NEC) in preterm infants.[68,69] Recommendations on adding infant cereal per formula ounce varies with the minimum addition of 1 teaspoon per ounce to a maximum addition of 1 tablespoon of infant cereal to 1 ounce of formula.[61] One tablespoon of infant cereal addition increases a 20 calorie per ounce formula to 34 calories per ounce. Formula with added infant cereal may require an enlarged nipple to allow flow, which may cause episodes of coughing.[58] Adding infant cereal can result in too rapid a weight gain and decrease the percentage of calories provided by protein and fat. Using antiregurgitant formulas such as Enfamil AR (Mead Johnson) may similarly reduce the number of vomiting episodes but not decrease the number of reflux episodes. If the infant is diagnosed with failure to thrive, increasing the caloric density of formula is recommended, especially if extensively hydrolyzed or amino acid formulas have improved symptoms. Although GER symptoms do improve in the flat, prone position, prone positioning is not recommended in infants because of its association with SIDS unless the infant is observed and awake, particularly in the postprandial phase.[70] The left-lateral feeding position has also been shown to decrease reflux symptoms and is considered safe.[71,72] The semi-supine position, such as in a car seat, does not offer any benefit and may worsen symptoms.[65]

Monitoring and Evaluation

Simple GER usually resolves around 18 months of age. GERD usually requires medical management including using acid suppression medications.[58] Using transpyloric feeds have been shown to decrease reflux episodes, but they may not completely eliminate the symptoms of GERD. When feeding modifications and use of medications do not improve GERD, surgical management such as a fundoplication, may be warranted. In older children and adolescents, there is no evidence that changes in diet improve symptoms, although in adults late night eating and obesity are associated with GER. Expert opinion suggests that children and adolescents eliminate caffeine, chocolate, and spicy foods. Alcohol use may also increase symptoms.[58] Elevating the head of the bed during sleeping in children and adolescents also may improve symptoms.

Lactose Intolerance

Lactose intolerance is defined as the inability to metabolize and digest lactose, which is the sugar most commonly found in milk and milk products, due to lactase deficiency. It is a clinical syndrome characterized by abdominal cramping, bloating, flatulence, and diarrhea that occurs approximately 30 minutes to 12 hours following the ingestion of lactose containing food and beverage. Individual tolerance varies per individual and is based on one's degree of enzyme (lactase) deficiency, which directly influences how much lactose an individual can

tolerate.[73,74] Throughout this section, four types of lactase deficiency will be discussed: primary lactase deficiency, secondary lactase deficiency, developmental lactase deficiency, and congenital lactase deficiency.

Anatomy, Physiology, and Pathology of Lactose Intolerance/Malabsorption

Lactose intolerance is caused by a shortage of the enzyme lactase, which is produced by the cells that line the small intestine. People who lack this enzyme are unable to metabolize and completely digest lactose (a disaccharide) into its simpler forms—glucose and galactose (monosaccharides)—following the ingestion of a lactose-containing food or beverage.[74–76] This results in digestive discomfort, such as abdominal cramping, bloating, flatulence, and diarrhea. The aforementioned symptoms result from bacterial fermentation of malabsorbed carbohydrate (undigested lactose) in the colon causing excess gas and increasing gut motility/osmotic load, which results in loose stools and/or diarrhea.[76]

Diagnosis

Lactose malabsorption is commonly diagnosed noninvasively via a breath hydrogen test. Following the ingestion of a lactose-containing beverage (typically 1–2 g lactose/kg body weight, up to a maximum of 25 grams lactose), the patient blows into a balloon-like bag at intervals for a specified amount of time (typically 2–3 hours).[74] Intermittent samples are taken and analyzed. Carbohydrate malabsorbed in the small intestine (undigested lactose) is fermented by colonic bacteria and released as hydrogen gas, allowing for the measurement of hydrogen in the breath. A raised hydrogen breath level greater than 20 ppm signifies the inability to digest lactose.[74–77] It is important to note that additional diagnostic tests are available such as the fecal pH test that measures the acidity of stool, small bowel biopsy that can determine lactase activity, and genotyping; however, the breath hydrogen test is considered the gold standard.[74]

Primary Lactase Deficiency

Primary lactase deficiency (PLD), also referred to as adult-type hypolactasia, lactase nonpersistence, and hereditary lactase deficiency occurs secondary to "relative or absolute absence" of the enzyme lactase.[74,77] PLD presents with a worldwide prevalence rate of 70 percent and is the most common enzyme deficiency known to cause lactose intolerance and/or malabsorption.[74,76,77] The prevalence and age of onset vary secondary to ethnicity and the overall use of dairy products in the diet.[74,76] Populations known to consume greater lactose containing diets present with lower prevalence rates (e.g. Europeans), whereas, populations with lower lactose containing diets present with higher prevalence rates (e.g. Asia, Africa, American Indians, Ashkenazi Jewish people).[74–77] Individuals with PLD present with varying degrees of enzyme deficiency because **lactase-phlorizin hydrolase** (LPH), the enzyme responsible for digesting lactose, begins to decrease at 2 years of age, resulting in varying levels of lactose tolerance.[77] Nutrition management must be flexible and tailored to the individual (e.g. the complete removal of lactose from the diet vs. limited lactose dietary consumption).[74]

Secondary Lactase Deficiency

Secondary lactase deficiency may occur if the lining of the small intestine that houses lactase-containing epithelial cells is destroyed as a direct result of underlying disease (e.g. celiac disease), small intestinal resection, gastrectomy, chemotherapy treatments, viruses (e.g. rotavirus), bacteria, parasitic infections (e.g. giardiasis), or acute diarrheal disease.[74,75] Lactase deficient, underdeveloped epithelial cells replace those destroyed by disease, resulting in secondary lactose deficiency and lactose malabsorption.[74] According to the American Academy of Pediatrics Committee on Nutrition, eliminating lactose from the diet is generally not required for treating secondary lactase deficiency and lactose malabsorption.[74] Treating the underlying condition is key. Once the primary issue is resolved, lactose-containing products can be reintroduced according to individual tolerance.[74]

Developmental (Neonatal) Lactase Deficiency

Developmental or neonatal lactase deficiency occurs in premature infants as lactase is the last of the major intestinal disaccharidases to develop.[74,75] Therefore, the infant is born before the optimal development of the lactase enzyme.[75] Lactase activity is estimated at 30 percent (deficient) between 26 and 34 weeks of gestation and increases to an estimated 70 percent by gestation age range 35–38 weeks.[75] As a direct result, premature infants have lower levels of lactase activity and may be unable to digest and absorb lactose as well as their term counterparts.[75] Carlson and colleagues demonstrated that the "addition of lactase to preterm formula reduces the amount of lactose by 70% after a two hour incubation period at room temperature."[78] Therefore, using lactase to hydrolyze lactose in preterm formulas and maternal breast milk may aid in decreasing lactose malabsorption in preterm infants and further lead to enhanced weight gain and improved feeding tolerance.[74,79]

Congenital Lactase Deficiency

Congenital lactase deficiency is an extremely rare autosomal recessive disorder caused by coding mutations in

the LPH gene, which results in the reduction or complete absence of LPH.[75,77,80] Primarily affecting those of Finnish decent, affected newborn infants present with severe diarrheal disease, metiorism (tympanites), and malnutrition immediately to a few days following the introduction of maternal breast milk or lactose-containing formula.[47,75,77,80] Treatment includes immediate and complete removal of lactose from the diet, followed by the replacement/consumption of a lactose-free formula during infancy and lactose-free milk products after infancy. If left untreated, congenital lactase deficiency can be life threatening as a result of dehydration and loss of electrolytes.[74,77,80]

Nutrition Screening

Nutrition screening in pediatric patients with lactose intolerance should be consistent with clinical malnutrition (undernutrition) risk screening as defined by the AND and ASPEN.[13]

Nutrition Assessment

Patients with lactose intolerance should be referred to a pediatric RDN in order to receive individualized nutrition assessment. Nutrition assessment consists of obtaining a thorough diet history with emphasis placed on both energy and nutrient intake, specifically calcium and vitamin D; two nutrients required for optimal bone health. Prolonged inadequate calcium and vitamin D intake are associated with reduced bone mass, osteoporosis, decreased bone mineral density, and osteomalacia. Close attention should also be paid to anthropometric assessment, growth trends, clinical data, and the need for multivitamin and mineral supplementation to help prevent and/or correct nutrient deficiency. The diet, clinical history, and additional data obtained by the pediatric dietitian may reveal a relationship between lactose ingestion and reported symptoms. Therefore, the nutrition assessment should also seek to identify potential hidden sources of lactose that may be continuing to cause reported discomfort, as well as assessing the patient/client and caregiver's readiness to make dietary changes and any barriers to learning or compliance.[73,74]

Nutrition Diagnostic Statements (PES)

Nutrition diagnosis is determined from information gathered during the nutrition assessment.

- Altered gastrointestinal function (NC-1.4) related to the inability to digest lactose as evidenced by hydrogen breath tests results greater than 20 ppm.
- Inadequate mineral intake (NI-5.10.1) related to eliminating calcium rich food and beverages from the diet as evidenced by 24-hour dietary recall of calcium rich foods meeting less than 50% RDA for age.

Nutrition Intervention

The specific nutrition diagnosis established from the nutrition assessment aids in determining the appropriate medical nutrition therapy intervention is nutrition education. The goal of nutrition education provided in the dietary management of lactose intolerance is to achieve resolution of symptoms and also prevent the onset and/or re-occurrence of symptoms. Clients and their caregiver(s) should receive nutrition education and resources related to reading food labels, identifying the lactose content of foods/beverages, meal preparation/planning, recipe modification, as well as how to gradually re-introduce lactose back into the diet & establish an individual threshold (when appropriate). Nutrition counseling should also focus on hidden sources of lactose, strategies for eating outside of the home, and identified age-related social situations that may require special attention in order to avoid feelings of depression, distress, or anxiety. Additionally, multivitamin with mineral supplementation may be recommended in addition to an increase in oral nutrient intake to aid in the prevention and/or correction of nutrient insufficiency vs. deficiency.[73,74,76] Lastly, one should always assess the comprehension level of the patient and caregiver, along with addressing individual questions/concerns. Please refer to the Pediatric Nutrition Care Manual for details on a Lactose Controlled Diet.[81]

Nutrition Monitoring and Evaluation

Ongoing monitoring and evaluation is essential to ensure that the nutrition intervention provided was effective and is sustainable. Feedback provided by the client and his/her caregiver(s) allows for appropriate adjustments in the dietary management plan to be made. Follow-up visits include the re-evaluation of diagnostic clinical markers, such as gastrointestinal symptoms/discomfort, growth trends, and review of food-symptom diary (if maintained) to reveal potential adverse relationships and to aid in establishing individual lactose tolerance/threshold. Nutrition-related vitamin/mineral lab results (ex. vitamin D) should also be monitored and addressed, with supplementation recommended and/or adjusted as appropriate. It is paramount that the caregiver and pediatric client receive continued support in an effort to achieve improved quality of life and for each to feel successful in their efforts.[73,74]

Celiac Disease

Celiac disease (CD) is a multisystem, T-cell-mediated chronic autoimmune intestinal disorder that occurs in genetically predisposed individuals carrying human leukocyte antigen (HLA)-DQ2 and/or DQ8 haplotypes.

Both the incidence and prevalence of CD have risen in recent years due to increased awareness and it is now estimated to affect approximately 1% of the world's population with the exception of South East Asia, in which risk alleles of the populace are uncommon.[82–87] First degree relatives of individuals diagnosed with CD are at greatest risk with an estimated prevalence rate 10 times greater than that of the general population.[84] Additional at-risk populations include those previously diagnosed with type 1 diabetes mellitus, autoimmune thyroid disease, selective IgA deficiency, Trisomy 21, Turner Syndrome, and Williams Syndrome.[82,83,85,86]

Early Infant Feeding Practices and Celiac Disease

It was once believed that environmental factors, such as breastfeeding and the time in which gluten is introduced into an infant's diet were influential in the development and presentation of CD.[88,89] Over the last few years, research has revealed that the early introduction of gluten and breastfeeding have no effect on the risk of developing CD during childhood. However, whether or not delayed introduction of gluten into a genetically predisposed infant's diet increases versus decreases one's risk of developing CD during childhood remains highly debatable.[88–90] These new findings prompted ESPGHAN to update their 2008 recommendations and release a revised position paper in 2016. ESPGHAN recommends the safe introduction of gluten in all infants between the ages of 4 and 12 months and recommends avoiding consuming large amounts of gluten (quantity undefined) during the first few weeks that follow initial gluten introduction.[88,90] For a complete listing of position statements and recommendations, please refer to *Gluten Introduction and the Risk of Coeliac Disease: A Position Paper by the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition*.[90]

Clinical Manifestations

The clinical presentation of CD is multi-systemic and ever changing. Classic gastrointestinal-related symptoms are most prominent in younger children and include diarrhea/steatorrhea, constipation, chronic abdominal pain, abdominal distention, and vomiting with associated malnutrition and growth failure.[83,91] A mild elevation of serum liver enzymes is also commonly seen within the pediatric population.[83] Non-gastrointestinal or extra-intestinal symptoms include short stature, inadequate weight gain, weight loss, delayed puberty, dental enamel defects, aphthous ulcers in the mouth, dermatitis herpetaformis, and reduced bone mineral density.[82,86,91] Although CD is traditionally associated with inadequate weight gain or weight loss, some children may initially present as overweight or obese.[82,83] Additionally, iron deficient anemia that is resistant to oral iron supplementation, fatigue, migraines, and joint pain may also present as symptoms.[82,83,86,92] Lastly, anxiety, depression, hallucinations, panic attacks, and suicidal behavior have also been reported amongst adolescents.[83] Experts believe that CD remains underdiagnosed because of the great variability in clinical presentation, which often results in misdiagnoses of irritable bowel syndrome or lactose intolerance.[82,83]

Diagnosis

Those who present with symptoms suspicious of CD must first receive serology testing, which includes **immunoglobulin A** (IgA) antibody and **tissue transglutaminase** (TTG), along with **tissue transglutaminase immunoglobulin G** (TTG IgG) if IgA deficiency is present. For younger children than 2 years of age, research supports combining the tTG-IgA with a DGP-IgG to improve the accuracy of the result.[83] It is important that gluten remain in the diet to ensure reliable serology testing at a quantity of greater than 10 grams per day, which is the equivalent to 2 slices of bread, for no less than 8 weeks.[83] Formal diagnosis must be further confirmed via intestinal biopsy. In some cases, diagnosis may be uncertain. If this is the case, **human leukocyte antigen** (HLA) typing and repeat biopsy can be performed. A trial of a gluten-free diet may also be considered. After a trial of the gluten-free diet, repeat serology testing and intestinal biopsy are recommended. If once positive serology tests become negative after the trial of the gluten-free diet, this is tangible and supportive evidence for the diagnosis of CD.

While the diagnostic gold standard for CD is duodenal biopsy, ESPGHAN published evidence-based guidelines in 2012 that suggest that a formal diagnosis of CD may be made without performing a duodenal biopsy within pediatric patients who meet specified criteria. This specific subpopulation includes symptomatic individuals with a tTg-IgA greater than 10 times the upper normal limit for their age, present with a positive EMA-IgA antibody result, possess the HLA-DQ2 and/or HLA-DQ8 halpotypes, and experienced relief of symptoms following trial of a gluten-free diet.[82,94–96] However, discretion is advised even though a non-biopsy confirmed diagnosis of CD within the pediatric population is desirable with respect to avoiding sedation and endoscopy related risks, as well as decreasing overall diagnostic costs. Standardization of serological tests for CD in the United States does not exist and "variation in antibody levels between commercial assays when the same serum samples are tested" has been well documented; thus directly affecting the interpretation of results and leading to diagnostic inconsistencies.[83,94] Secondly, gastrointestinal disorders that may occur as comorbidities in those with CD, such as Eosinophilic Esophagitis, Helicobacter Pylori Gastritis, and Peptic Esophagitis, will remain undiagnosed in the absence of a biopsy.[83,97]

Nutrition Screening

Nutrition screening in pediatric patients with CD should be consistent with clinical malnutrition (undernutrition) risk screening as defined by the AND and ASPEN.[13]

Nutrition Assessment

All patients with CD should be referred to a RDN with expertise in celiac disease and the gluten free diet to receive individualized nutrition assessment. Nutrition assessment consists of obtaining a thorough diet history with emphasis placed on total energy intake, as well as both macronutrient and micronutrient intake. Close attention should also be paid to biochemical and clinical data results, anthropometric assessment, growth history/trends, and the need for multivitamin/mineral supplementation to aid in preventing and/or correcting nutrient deficiency. Lastly, the nutrition assessment should also seek to identify potential hidden sources of gluten that may be continuing to cause reported gastrointestinal discomfort, as well as assessing the patient/client and caregiver's readiness to make dietary changes and any barriers to learning or compliance, such as financial constraints.[82,88]

Nutrition Diagnostic Statements (PES)

Nutrition diagnosis is determined from information gathered during the nutrition assessment.

- Altered GI function (NC-1.4) related to damage to the intestinal villi as evidenced by elevated celiac antibody levels and positive duodenal biopsy.
- Food and nutrition related knowledge deficit (NB-1.1) related to lack of education in regard to the gluten free diet and its associated lifestyle as evidenced by new diagnosis of celiac disease.
- Limited adherence to nutrition related recommendations (NB-1.6) related to consumption of gluten-containing foods as evidenced by elevated celiac antibody tests, complaints of intermittent abdominal pain, and patient report of routinely eating gluten containing food items.

Nutrition Intervention

Immediately following diagnosis, affected individuals should be referred to a registered dietitian/nutritionist with expertise in CD in order to receive comprehensive nutrition education with emphasis placed on the importance of adhering to a gluten-free diet and the gluten-free lifestyle. When nutritive sources of gluten (e.g., wheat, rye, barley, and its derivatives) and nonnutritive sources of gluten (e.g., toothpaste) are completely eliminated from the diet, gastrointestinal and often extra-intestinal symptoms, serologic test results, histology, and growth and development should normalize as celiac symptoms improve and move into a state of remission.[83,88,91]

Attendance of all primary caregivers should be encouraged during the initial nutrition consultation. Nutrition counseling should focus on nutritive and non-nutritive sources of gluten, gluten-free alternatives, hidden sources of gluten, various aspects of cross-contamination, where one can purchase gluten-free products; credible resources and support groups, eating outside of the home, and identified age-related social situations that may require special attention to avoid feelings of depression, distress, or anxiety. Close attention should also be paid to anthropometric assessment, growth trends, nutritional intake, and the need for multivitamin and mineral supplementation to aid in preventing and/or correcting nutrient deficiency.[88]

Inclusion of Oats in a Celiac Diet

Whether or not an individual with CD can safely consume oats remains controversial and is under scientific investigation. Oats may become contaminated by gluten during the harvesting and milling process, have been reported to cause gastrointestinal symptoms, induce an immunological response as confirmed via detection of serum anti-avenin antibodies, and cause small intestinal damage in some individuals with CD.[82,98,99] Clinical research supports the safe introduction and consumption of 20–25 grams per day (¼ cup gluten-free oats) in most children with CD following the resolution of symptoms and normalization of celiac antibody levels.[82] The patient should discuss including gluten-free oats with his or her gastroenterologist and/or registered dietitian/nutritionist before ingestion because individual tolerance varies and monitoring of antibody levels is required.[82]

Gluten Free Diet and Nutrient Deficiencies

Gluten is composed of two proteins, gliadin and glutenin. Gluten is the general name for storage proteins, referred to as prolamins that are found in wheat, rye, barley, and oats. Gliadin is the specific prolamin found in wheat, secalin is found in rye, and hordein is found in barley. When ingested, it is these specific prolamins that cause villous atrophy, which may further result in nutrient malabsorption and/or deficiency.[82,99]

Iron, calcium, and folate are key nutrients that are often affected in those with celiac disease, because these nutrients are absorbed in the proximal small bowel.[82,88,100] If the disease progresses further down the small intestinal tract, malabsorption of carbohydrates, fat, fat-soluble vitamins, and protein may also occur. The most common causes of anemia in those with celiac disease are iron, folate, or vitamin B_{12} deficiency. Calcium, phosphorus, and vitamin D deficiencies may also occur secondary to malabsorption or decreased intake of dairy products if

lactose intolerance is present. Secondary lactose intolerance is commonly observed as the enterocytes at the tips of damaged villi are absent, and therefore unable to produce the enzyme lactase.[82,88,100]

Those diagnosed with celiac disease are at higher risk for developing metabolic bone disease secondary to "malabsorption, hypogonadism, and inflammation" and should receive dual energy x-ray absorpiometry (DXA scan), quantitative CT scan, or computerized bone age estimation; however, no guidelines exist regarding when or how often to assess bone mineral density via DXA in newly diagnosed pediatric patients.[101,102] Strict adherence to a gluten-free diet that contains sufficient nutrients required to achieve optimal bone health during childhood will result in improved bone mineral density in adulthood.[82,102]

Nutrition Monitoring and Evaluation

It is understood that individuals with CD have a permanent intolerance to gluten and must adhere to a gluten-free diet since this is the only known treatment. Ongoing monitoring and evaluation is essential in treating pediatric CD in order to achieve optimal health outcomes. Frequent medical and nutrition visits are recommended during the first one to two years following the initial diagnosis; however, no formal guideline exists.[83] Subsequent nutrition visits should encompass a complete review of new and pertinent data (e.g. repeat serology tests, DXA scan results), medical nutrition therapy components previously mentioned, along with determining dietary compliance, as well as addressing barriers to compliance (if applicable). One should also continue to assess the comprehension level of the patient and family, along with addressing individual questions/concerns and providing continued support in an effort to achieve improved quality of life.[82,83]

Special Considerations

The only known treatment for CD is strict adherence to a gluten-free diet. If left untreated, CD can result in nutritional deficiencies, decreased bone mineral density, and neurological disorders. Scientific research further suggests that if left untreated, affected individuals are at an increased risk for developing intestinal lymphoma, infertility, spontaneous abortion, and the delivery of low-birth-weight infants.[82,88,103,104] A gluten-free diet will allow for normal growth and development as well as relief from symptoms; however, some may continue to experience persistent symptoms despite adherence.[88] Alternative non-dietary therapeutic treatments, such as oral glutenase supplements (enzyme therapy), therapeutic vaccine, and zonulin inhibitors, for example, are currently in varying stages of development.[105,106] It is important to note that commercially available enzyme supplements are neither approved for use nor proven effective within the pediatric celiac population and therefore should not be recommended for use.[82]

Inflammatory Bowel Disease

The two major types of **inflammatory bowel disease** (IBD) are **Crohn's disease** and **ulcerative colitis** (UC). Crohn's disease may occur in any portion of the gastrointestinal tract. Ulcerative colitis is by definition confined to the colon with minimal involvement of the terminal ileum.[107] Patients who develop nonspecific IBD-like symptoms are often temporarily termed as having indeterminate colitis. The two diseases have many features in common: diarrhea, gastrointestinal blood and protein loss, abdominal pain, weight loss, anemia, and growth failure. Children with IBD may experience growth failure because of inadequate intake, malabsorption, excessive nutrient losses, drug-nutrient interaction, and increased nutrient needs. Inadequate intake may be due to abdominal pain, gastritis, personal effort to decrease the incidence of diarrhea, and taste changes with zinc deficiency. The timing and referral to a gastroenterologist to endoscopically diagnose a patient with IBD is crucial as the symptoms can lead to growth impairment and malnutrition.

Condition Specific Nutrition Screening

Nutrition screening in pediatric patients with IBD should be consistent with clinical malnutrition (undernutrition) risk screening as defined by the AND and ASPEN.[13]

Nutrition Assessment

Nutrient deficits have been noted in 30–40% of adolescents and children with IBD.[108] At the time of diagnosis, about 85% of pediatric patients with Crohn's disease and 65% with UC present with weight loss.[109] A cohort study by Ricciuto et al, found that the median time of diagnosis is 4.5 months and that there is an independent association between delayed diagnosis and linear growth. Bone mass deficits in IBD children range between 10–40%.[110] Plateau linear growth may be the only presenting sign of IBD. Therefore, it is important to have early diagnosis in children in order to maximize growth potential.[111]

Malnutrition occurs in 32% of children at the time of diagnosis of IBD.[112] Patients who present in a flare, or active disease state, often have multiple macro- and micronutrient losses. Inflammation of the mucosa and bowel resections can lead to general malabsorption. Depending on the location and length of bowel resection, specific nutrients may no longer be absorbed and will need to be supplemented. Bacterial overgrowth due to altered motility or strictures can also lead to malabsorption. All these

components can and do lead to malnutrition which is the key to identifying malnutrition and inflammation parameters in order to have improved patient outcomes.[113]

There has been a lack of consensus for estimated caloric needs in pediatric patients with IBD. In the Azcue, et al study, the authors looked at the **resting energy expenditure** (REE) and body composition of pediatric Crohn's patients. It was noted that Crohn's patients were similar to that of the control group per kilogram of lean body mass. The REE per kilogram of body mass was not regulated since it is in the starvation state.[114] A more recent study by Hill et al in 2011 compared four commonly used predicted equations for REE in children with IBD. The study concluded that the Schofield equation was more accurate at measuring and predicting REE for IBD patients.[115] In the adult population, it has been documented that total energy expenditure is not significantly elevated in active disease compared with remission or inactive disease.[116]

Protein requirements are likely elevated due to protein losses as well as increased needs for healing, especially in post-op patients. In patients with IBD who present with protein-losing enteropathy, the body cannot synthesize new proteins as fast as they are lost through the gastrointestinal tract.[117] The resulting hypoalbuminemia cannot be corrected by adding protein to the diet.

Bile malabsorption leads to decreased absorption of long-chain fatty acids but not medium chain fatty acids, because bile is not required for transporting medium-chain fatty acids through the mucosa. Fat malabsorption contributes to the malabsorption of fat-soluble vitamins.

All patients should be assessed for micronutrient deficiencies and repleted as necessary. Blood losses via the stool can lead to iron deficiency. Folate deficiency can result from drug-nutrient interactions as well as decreased intake of folate-rich food sources such as green leafy vegetables. The fat-soluble vitamins, A, D, E, and K as well as zinc, magnesium, and calcium are lost when steatorrhea is present. Zinc deficiency may be related to diarrheal, high-output fistula losses and inadequate dietary intake. Magnesium and potassium are also lost via diarrhea. Patients with resections of the stomach or ileum, or severe disease of the terminal ileum may not be able to absorb Vitamin B_{12}.[118,119] Patients should be on a daily multivitamin unless they are receiving enteral formula, which provides the equivalent of a multivitamin.[120] A study of 54 adults with Crohn's disease revealed that although those patients in clinical remission for longer than 3 months were able to meet macronutrient needs via food intake, micronutrient deficiencies remained. Low plasma levels of vitamin C, copper, niacin, and zinc were found in greater than 50% of the patients.[121]

Although bone disease in IBD is multifactorial, vitamin D and calcium are important nutrients to monitor. Pediatric patients with IBD are known to be at increased risk of osteopenia and osteoporosis.[122] Several factors along with medical treatment are thought to be the reason for impaired bone mineralization in children with IBD. Steroids are known to affect bone mineral density,[123] and new research shows that inflammatory cytokines lead to lower bone mineral density.[124] Even though calcium and vitamin D are not the sole answer in IBD-related low bone mineral density, blood levels should be kept within the normal range. Fat malabsorption can lead to low vitamin D levels, and vitamin D plays many roles beyond bone health in the body. A review article recommends 50,000 units of vitamin D_2 every week up to every day depending on the degree of fat malabsorption, or up to 10,000 units of vitamin D_3 every day for up to 5 months. This is followed by a maintenance dose of 50,000 units of vitamin D_2 every week. For children and adults without fat malabsorption, the recommendation ranges from 400 to 1000 units of vitamin D_3 every day.[125]

Nutrition Diagnostic Statements (PES)

Nutrition diagnosis is determined from information gathered during the nutrition assessment:

- Inadequate oral intake (NI-2.1) related to symptoms of IBD as evidenced by recording of about 50% or less of normal food intake per meals.
- Unintended weight loss (NC-3.2) related to increased needs resulting from active disease of IBD as evidenced by 10% weight loss.

Nutrition Intervention

Drug-nutrient interaction from the pharmacotherapy used in treating IBD may negatively affect nutrition. Sulfasalazine (azulfidine) interferes with folate absorption.[126] Methotrexate is a folic acid antagonist, and supplementation of folic acid helps reduce methotrexate's hepatic side effects.[127,128] Corticosteroid therapy interferes with absorption of calcium, phosphate and zinc.

Controlled studies have not supported using low-residue, high-fiber, or low-refined-sugar diet in order to maintain remission of Crohn's disease.[129,130] Dairy products need not be restricted in patients with IBD; however, lactose malabsorption is more common in patients with small bowel Crohn's disease than in patients with disease involving the colon or UC. Lactose intolerance is often temporary during times of active disease. Advice concerning the intake of dairy products should be individualized in order to avoid unnecessary dietary restrictions.[131]

Nutrition support has both primary and an adjunctive role in treating IBD.[108] Primary nutrition therapy appears to be more effective in treating Crohn's disease than ulcerative colitis.[107,132] In a pediatric study, patients with active Crohn's disease were treated with exclusive

EN therapy (multiple formulas) versus corticosteroids. The duration of clinical remission, degree of mucosal healing and improvement in mucosal inflammation, and improvements to linear growth were greater with any of the enteral formulas than with steroids.[113,133] These results are similar to another pediatric study that showed exclusive enteral feeds with intact proteins were just as effective as corticosteroids in inducing remission, and had a lower rate of relapse.[134] Borelli also looked at a polymeric diet versus corticosteroid therapy, and found that using polymeric formulas led to increased mucosal healing at the end of the 10-week study.[135] A randomized controlled trial of polymeric enteral formula and an elemental formula found similar results in inducing remission in children with Crohn's disease. Children given a polymeric diet, however, achieved better weight gain.[136]

Despite these positive studies in the pediatric population, a Cochrane review, including both pediatric and adult studies, reported that corticosteroids were more effective in inducing remission. The protein composition of the formulas did not reveal any difference in the effectiveness of the EN therapy,[137] however, for the maintenance of remission in Crohn's disease, enteral feeds were found to be beneficial in all age groups with no known side effects.[138] Due to lifestyle changes that accompany exclusive EN therapy, there is interest in the effectiveness of partial EN therapy. A pediatric study using elemental formula provided as either 50% or 100% of calorie needs showed higher remission rates and improved hemoglobin, albumin, and **erythrocyte sedimentation rate** (ESR) in those receiving 100% of their calorie needs from formula.[139] However, in a study of patients with steroid-dependent Crohn's disease in state of remission, adding enteral formula (either elemental or polymeric) taken orally along with a regular diet allowed 43% of patients to discontinue steroids and remain in remission at the 12-month follow-up.[140]

Bamba looked at elemental diets containing different amounts of **long-chain triglycerides** (LCT). The high LCT diet had a remission rate of 25% at 4 weeks, whereas the low LCT diet had a remission rate of 80%.[141] Other studies have found that formulas containing higher amounts of **medium-chain triglycerides** (MCT) have resulted in remission rates comparable to low-fat formulas or steroid therapy.[132–143] Per a Cochrane Review, there is inadequate evidence to support the use of omega-3 fatty acids for inducing or maintaining remission of ulcerative colitis[144,145] nor do they appear to be effective in maintaining remission in Crohn's disease.[146] Probiotics have not been shown to be effective for inducing and maintaining remission of Crohn's disease,[147,148] but when added to standard therapy for UC, they may lead to a reduction of disease activity.[149]

Enteral feeding has been shown to reduce inflammation and improve well-being, nutrition, and growth, but the exact mechanism is unknown. A study on the use of enteral feeding showed that inflammatory markers were significantly improved over the first 7 days of treatment; significant improvement in growth-related proteins and nutritional markers were not seen until day 14 or later.[150]

In patients not treated with EN therapy during periods of active disease and/or weight loss, oral nutritional beverages in addition to food can be useful. Because protein composition does not affect the rates of remission or disease improvement, formula choice should be based on palatability and patient tolerance. If voluntary intake is insufficient, nasogastric enteral supplementation may be considered. PN should be reserved for patients who have bowel obstructions or short bowel syndrome, or are unable to tolerate sufficient quantity of EN because of active disease. PN comes with greater risk as well as higher cost.

Most recently there has been more nutritional modifications done to the overall oral diet. Some of these new "diets" are gaining attention in treating symptoms of IBD. **Specific Carbohydrate diet** (SCD) and Cow's milk protein elimination diet have been used for inducing remission in pediatric IBD.[151] However, the available evidence is not strong enough to recommend this kind of nutrition intervention in the pediatric population.[151]

Nutrition Monitoring and Evaluation

Ongoing monitoring and evaluation is essential to ensure that the nutrition intervention provided was effective and is sustainable. Feedback provided by the client and his/her caregiver(s) will allow for appropriate adjustments in the dietary management plan. Follow-up visits include re-evaluating diagnostic clinical markers, such as gastrointestinal symptoms/discomfort, growth trends, and a review of the food-symptom diary. Nutrition related vitamin/mineral lab results should also be monitored and addressed, with supplementation recommended and/or adjusted as appropriate.

Pancreatitis

Pancreatitis can be acute or chronic and divided into three groups: mild, moderate, and severe. Approximately 80% of all cases are mild and will often resolve within 5–7 days with bowel rest, fluid, and analgesic support.[152,153] The remaining cases are moderate to severe and require medical and nutritional therapy. Moderate and severe pancreatitis creates a state of hypermetabolism and catabolism with significant nitrogen losses, and results in an elevated systemic inflammatory response. It is often associated with infectious complications and in severe cases, multiple organ failure and pancreatic necrosis.[152–154]

Anatomy, Physiology, Pathology

Pancreatitis is the inflammation of the pancreas that occurs when pancreatic enzyme secretion accumulates in the pancreas, causing the organ to digest itself. In most cases, pancreatitis is an acute episode lasting a few days to a week, however, in rare circumstances, it can be a progressive chronic condition.

Acute pancreatitis occurs suddenly with rapid onset. Common causes of acute pancreatitis in pediatrics include physical injury, certain medications, gallstones, or problems in the anatomy of the ducts in the liver or pancreas. In 35% of children, acute pancreatitis is idiopathic.[155] Medications that are associated with pancreatitis include anti-seizure medications, chemotherapy agents, and certain antibiotics.

Chronic pancreatitis is a life-long condition with multiple episodes occurring over time. About 10% of children will have a recurrent episode of pancreatitis.[155] After the second episode of pancreatitis, additional genetic testing is required in order to evaluate the cause of recurrence. Children with genetic, metabolic, or anatomic abnormalities are at greatest risk of developing chronic pancreatitis.[156]

Pancreatitis can be categorized as mild, moderate, or severe. Based on laboratory markers, clinical findings, and CT-imaging severity of pancreatitis is classified.[157]

Nutrition Screening

Nutrition screening in pediatric patients with pancreatitis should be consistent with clinical malnutrition (undernutrition) risk screening as defined by the AND and ASPEN.[13]

Nutrition Assessment

Patients with pancreatitis should be referred to a pediatric RDN with expertise in gastrointestinal disorders in order to receive an individualized nutrition assessment. Nutrition assessment for patients with pancreatitis consists of obtaining a thorough diet history with emphasis placed on food and fluid intake both before and after onset of pancreatitis. Due to the acute nature of pancreatic episodes, decrease in oral intake may occur suddenly at the onset of abdominal discomfort. A thorough review of gastrointestinal systems including nausea, vomiting, and GERD should be evaluated. Close attention should also be paid to anthropometric assessment, growth trends, and feed tolerance. In patients with chronic pancreatitis, serum levels of fat soluble vitamins, vitamin B_{12}, calcium, folate, zinc, copper, and magnesium should be monitored and oral supplementation may be required.[156]

Diagnosis

The diagnosis of pancreatitis is based on multiple factors and includes clinical symptoms such as sudden abdominal or back pain, nausea, vomiting, loss of appetite, or sweating; abnormal blood tests, commonly elevated amylase and lipase levels; or radiographic images like ultrasound and CT scans showing inflammation in the pancreas. A diagnosis of acute pancreatitis can be made if two or more of these criteria are present.[155] Chronic pancreatitis is progressive and includes multiple episodes of pancreatitis throughout a lifetime. Diagnosis of chronic pancreatitis includes exocrine and/or endocrine insufficiency resulting in steatorrhea.[156]

Nutrition Diagnostic Statements (PES)

- Inadequate oral intake (NI-1.1) related to compromised pancreatic function as evidenced by need for enteral feedings to meet nutritional needs.
- Inadequate enteral intake (NI-2.3) related altered GI function as evidence by intolerance on enteral feeds.
- Impaired nutrient utilization (NC-2.1) related to compromised pancreatic function as evidenced by fat soluble vitamin deficiencies.

Management

Treating acute pancreatitis includes fluid resuscitation with intravenous hydration at up to 150% maintenance fluid requirement within the first 24 hours of hospitalization.[158] Antibiotics may also be used to manage inflammation as well as pain medication for discomfort.[154] Chronic pancreatitis may require daily enzyme supplementation and the possible need for surgery or islet cell transplant.[156]

Nutrition Intervention, Monitoring, and Evaluation for Mild Acute Pancreatitis

Traditionally in clinical practice, patients were often initially made NPO, and then advanced to a clear liquid diet as pain resolved. In a randomized controlled trial, patients with mild pancreatitis who resumed oral intake of liquids and solid foods before the resolution of pain were able to advance to a solid food diet sooner than those on bowel rest, thereby decreasing length of stay.[159] A study compared the initiation of a clear liquid diet versus soft solid food diet and found no significant difference between the two groups in cessation of feeds due to pain. Also, patients started on the soft solid diet took in significantly more calories and protein overall, with a decreased length of stay.[160] This research supports the initiation of oral feeds, both liquids and solids, during the first few days with the benefit of improved nutrient intake.

Nutrition Intervention, Monitoring, and Evaluation for Moderate to Severe Acute Pancreatitis

Early initiation of enteral feedings is highly recommended in patients with moderate to severe pancreatitis.[154] Studies have shown that by stimulating the gastrointestinal tract, early feeding has the potential to reverse organ failure with improvement in peristalsis and the return of gut function in those with ileus.[161] More so, jejunal feeds have been found most beneficial in resolution of ileus and gas stasis over PN and bowel rest.[162,163] EN provides adequate calories, protein, and vitamins to support organ recovery and decreases the risk of bacterial overgrowth and maintains gut integrity.[164] Traditionally, PN was used as first line therapy in patients with severe pancreatitis to minimize pancreatic stimulation and provide 'pancreatic rest.' Multiple studies now show that PN increases the proinflammatory response and contributes to gut atrophy, likely leading to bacterial translocation and increased infection rates.[165,166] A systematic review of 11 randomized controlled trials showed a statistically significant reduction in the risk of infectious complications and a statistically nonsignificant reduction in the risk of death with enteral feedings compared with PN.[167] The benefits of using EN over PN include a decrease in systemic inflammation markers[161] and a decrease in expense.[162,163] If PN must be used, it should not be initiated within the first 5 days of admission due to the higher inflammatory response that is present in the beginning of the disease process.[168]

Initiation of early enteral feeding should occur between 24 to 48 hours of admission to the hospital, especially in patients at risk for organ failure and pancreatic necrosis.[169] It is, however, recommended that patients are not fed within the first 24 hours of admission during the initial fluid resuscitation phase.[158] Feeding before fluid resuscitation can increase risk of an ileus and could result in feeding intolerance.[170] The route of enteral feeding is determined by the severity of disease. For patients with severe pancreatitis, a nasojejunal tube is recommended in an attempt to minimize or eliminate pancreatic secretion. A study of patients with severe pancreatitis found that patients on an elemental formula with a feeding tube 20 to 120 cm post the ligament of Treitz loss the stimulatory effect of the pancreas vs. a low-fat formula into the duodenum, which decreased secretory response but did not eliminate.[171] Additionally, patients with severe pancreatitis have a higher probability of gastroparesis due to decreased intake before hospitalization, increasing gastric feed intolerance.[154] Patients with peripancreatic collections also benefit from nasojejunal feeds as it offers a mechanism to splint the compressed stomach and duodenum. This splint allows feeds to flow openly into the bowel without obstruction, unlike gastric feeds.[154,158] Patients with moderate pancreatitis have a greater probability of tolerating nasogastric feedings; however, in patients with gastric feeding intolerance a nasojejunal tube is preferred. The benefits of nasogastric feedings include ease of placement, quickness of initiation, and cost effectiveness.[158] Another review of 20 randomized controlled trials reported that using polymeric vs. semi-elemental formula showed no statistically significant difference in feeding tolerance, infectious complications, and mortality.[172]

As the patient improves clinically, low-fat oral feeds are initiated and slowly advanced, as enteral feeds are weaned to a normal diet.

Nutrition Intervention, Monitoring, and Evaluation for Chronic Pancreatitis

More than 80% of patients with chronic pancreatitis can follow a regular diet when macro and micro nutrients are met in addition to pancreatic enzyme supplementation.[156] Six to 8 small meals a day is recommended, especially in patients with early satiety. Patients with chronic pancreatitis have a 30–50% higher resting energy expenditure than healthy individuals and may require 35 kcal/kg/day to meet needs.[173] Adding a MCT like coconut oil and palm kernel oil to the diet will maximize fat absorption. In cases where oral nutrition cannot meet caloric demand, enteral feeding tubes are required. Less than 1% of patients with chronic pancreatitis will require long term PN.[174] Additionally, patients with chronic pancreatitis are at risk of nutritional deficiencies, most commonly vitamin K, vitamin D, vitamin E, and vitamin A, respectively.[175] Vitamin B_{12}, calcium, folate, zinc, copper, and magnesium are nutrients that may also be deficient in patients with chronic pancreatitis. These nutrients should be monitored with the addition of vitamin supplementation, if needed.[156]

Cholestatic Liver Disease

Introduction

Biliary atresia is one of the more common chronic cholestatic liver diseases that presents in infancy and requires surgical intervention. Alagille syndrome, Byler syndrome, primary biliary cirrhosis, and primary sclerosing cholangitis are cholestatic liver diseases seen in childhood through adulthood. Long-term use of PN can lead to cholestasis; a complete discussion of PN–induced cholestasis is found in Chapter 11. In cholestatic disease, a decrease in biliary bile acids results in fat and fat-soluble vitamin malabsorption. As the liver damage progresses, leading to cirrhosis, there is decreased synthesis of

albumin and transport proteins, causing laboratory protein markers to become low. Nutritional concerns progress from fat malabsorption, protein-energy malnutrition, and excessive catabolism.[176]

Anatomy, Physiology, Pathology

Biliary atresia occurs only in infants. It is characterized by fibrotic obliteration or discontinuity of the extrahepatic biliary system, resulting in obstruction to bile flow. Bile ducts within the liver are initially patent during the first few weeks of life, but are progressively destroyed thereafter. This process occurs within the first 3 months of life.[177] If left untreated, biliary atresia progresses to cirrhosis and eventual death within 18 to 24 months of age.[178] The pathogenesis of biliary atresia is poorly understood.

Nutrition Screening

Nutrition screening in pediatric patients with cholestatic liver disease should be consistent with clinical malnutrition (undernutrition) risk screening as defined by the AND and ASPEN.[13]

Nutrition Assessment

Patients with cholestatic liver disease should be referred to a pediatric RDN with expertise in gastrointestinal disorders in order to receive individualized nutrition assessment. Nutrition assessment for patients with biliary atresia consists of obtaining a thorough diet history with emphasis placed on breastmilk/formula intake. Fat-soluble vitamins are malabsorbed in patients with biliary atresia and should be checked at the initial diagnosis and 1–3 months afterwards depending on the level. Patients with chronic cholestasis/cirrhosis can present with ascites and/or organomegaly, which will falsely elevate the patient's weight. Using this elevated weight would skew the results of a calculated BMI or the weight-for-length or weight/height assessment on a growth chart. These measurements can underestimate the degree of malnutrition. Using the mid-upper arm circumference and tricep skin folds to assess subcutaneous fat and skeletal muscle mass is the most accurate measurement of predicting malnutrition for patients with liver disease.[179] Mid-upper arm circumference z-scores can be used to assess the degree of malnutrition for infants greater than 3 months of age and children until age 5. For infants less than 3 months, serial MUAC measurements can be used to trend changes in nutrition status. For children over 5 years of age, MUAC percentile ranges can be used to predict BMI. Length/height-for-age and head circumference should be evaluated and monitored when appropriate. Infants with cholestatic liver disease are at risk for both stunting and microcephaly.[180]

Diagnosis

Infants with biliary atresia initially present to the doctor's office with jaundice, yellowing of the eyes, and acholic stools. Lab results are significant for elevated bilirubin and elevated **gamma-glutamyl transferase** (GGT).[181] Due to a range of cholestatic liver diseases, additional testing including ultrasound examination, X-rays, and liver biopsy are needed. Most commonly, a liver biopsy confirms diagnosis. Hallmark findings of biliary atresia found on biopsy include bile duct proliferation, portal tract expansion, and canalicular plugging.[182]

Nutrition Diagnostic Statements (PES)

- Malabsorption of fat related to altered gastrointestinal dysfunction as evidenced by need for modified diet high in medium chain triglycerides.
- Inadequate oral intake (NI-1.1) related to gastrointestinal dysfunction as evidenced by need for enteral nutrition to meet needs.

Management

According to the American Liver Foundation, there is no non-invasive treatment for biliary atresia. Patients identified to have biliary atresia will undergo a surgical procedure called a hepatoportoenterostomy, also known as a Kasai procedure, within days of diagnosis. This procedure reestablished bile flow in more than 80% of patients if performed within 60 days of birth. Endpoints include jaundice clearance, survival with native liver, and liver transplant. With a Kasai procedure, 35–50% of patients will survive with native liver at 3 years of age. This procedure improves outcomes and overall survival, however, about half of children will have a failed Kasai and require a liver transplant by 2 years of age.[178] Up to 85% of all patients with biliary atresia will require a liver transplant in their lifetime.

Nutrition Intervention

Chronic cholestasis in infancy and early childhood leads to increased calorie needs due to excessive catabolism. Calorie needs have been suggested to be 110–160% of the RDA for ideal body weight in children. Infants with biliary atresia require calories in the range of 120–200 kcal/kg/day.[183] Protein requirements should focus on providing equal to or greater than the RDA for protein in the nonencephalopathic patient. Recommendations for protein in infants have been made at 2–3 g/kg/day,[184] With encephalopathy, protein should be temporarily restricted below the RDA or a formula predominate in branch chain amino acids should be utilized until mental status returns to normal.[185]

Cholestatic liver disease leads to decreased biliary bile acids, resulting in malabsorption of LCT. Bile is not

required for solubilization of MCT, which are absorbed unmodified into the portal circulation. Formulas containing a high percentage of MCT oil, in addition to emulsifiable MCT oil to formula, or supplementing foods with MCT oil is often recommended to provide fat and calories, while minimizing steatorrhea.[186] MCTs do not, however, provide essential fatty acids or help absorp fat-soluble vitamins;[187] thus, a source of linoleic acid is essential. In a study of infants on a formula containing 3% of calories from linoleic acid, they developed essential fatty acid deficiency. These results show that due to malabsorption with hepatobiliary disease, infants should receive well above 3% of calories from linoleic acid.[188] For infants with biliary atresia receiving formulas, those containing 40–60% of the fat from MCT are recommended. For premature infants, the formula Similac Special Care (Abbott) provides 50% of its fat source as MCT. For term infants, the formula Pregestimil (Mead Johnson) contains 55% of its fat source as MCT. For infants with milk protein allergy, Alfamino Infant (Nestle) a free amino acid based formula, contains 43% MCT. Liquigen (Nutricia) is an emulsifiable MCT oil that can be added to formula to increase caloric concentration and MCT content. Typically, 1 ml of Liquigen (0.5 gm fat, 4.5 calories) is added to 1 or 2 ounces of formula. Liquigen can be added to both oral and enteral formula regimens. For children over 1 year of age and older, Peptamen Junior (Nestlé) contains 60% of its fat as MCT. Additionally, oils naturally high in MCT like coconut oil and palm kernel oil can be added to soft foods. MCT oil given as an oral medication can be added in addition to formula in order to increase caloric intake of absorbable fats. Additional sources of MCT should be added gradually. Excessive intake of MCT oils and Liquigen can cause diarrhea.

To optimize nutrition after hepatoportoenterostomy or before liver transplant, using an enteral feeding tube may be warranted. A randomized study evaluating the use of a predominate MCT based formula showed a significant improvement in length and head circumference z-scores in patients who were enterally fed vs. PO ad libitum.[180] When initiating enteral feeds, continuous nocturnal feeds with PO ad libitum during the day is preferred to maintain oral skills. In cases of severe liver disease, continuous EN feeds may be required to maintain glucose levels by administering a constant **glucose infusion rate** (GIR).

Monitoring and Evaluation

In liver disease, the prevalence of fat-soluble vitamin deficiencies correlates with increasing bilirubin levels.[185] Infants with a total bilirubin greater than 2 mg/dL are at the highest risk for fat soluble vitamin malabsorption.[189] There is a wide array of supplemental and deficiency treatment doses throughout the literature. It is important to remember that the degree of fat malabsorption is unique to each patient's disease state and can change as treatments or medications are changed.

Prophylactically, a liquid water-soluble, fat-soluble multi-vitamin like aquADEKS can be started at the time of diagnosis for presumed fat-soluble malabsorption. Based on the child's age and weight, dosing can vary. When greater than 1 ml is given per day it is best to split the dose into two or three daily dose intervals in order to maximize absorption.

Vitamin A is stored in the liver and requires retinol-binding protein to circulate throughout the body. As liver disease progresses, there is impaired synthesis of retinol-binding protein, which leads to low vitamin A levels without a true deficiency. Clinical judgment needs to be used when reviewing both serum retinol and retinol-binding protein laboratory results. Of note, vitamin A is highly hepatotoxic and the risk benefit of under vs. oversupplementing should be considered before starting. Standard supplementation recommendations for Vitamin A range from 5000–15,000 International Units (IU)/day orally. For severe deficiency, intramuscular injections may be used.[176,186]

Vitamin E is stored in the liver and transported via lipoproteins. In patients with high triglycerides and cholesterol levels, the vitamin E level may be falsely elevated. It is best to check serum vitamin E levels in conjunction with total serum lipids to assess for deficiency. In children less than 12 years of age, a ratio of less than 0.6 mg/g and in children over 12 years of age, a ratio less than 0.8 mg/g is indicative of a deficiency. Supplementation recommendations in pediatrics range from 20–25 IU/kg/day of vitamin E or 10–200 IU/kg/day of alpha-tocopherol for intramuscular injections. Recommendations in adults range from 400–800 IU/day of alpha-tocopherol. The preferred source of vitamin E is D-alpha-tocopherol polyethylene glycol 1000 succinate TPGS vitamin E.[190,191] This suspension precludes the necessity of bile acid micellar solublization and is well absorbed in patients with severe cholestasis.[190]

In cholestatic and noncholestatic liver disease, metabolic bone disease has a multifactorial etiology that includes a decrease in insulin-like growth factor 1, hypogonadism, malnutrition, low BMI, loss of muscle mass, low calcium, and vitamin D deficiency. Vitamin D is hydroxylated in the liver, as one of the steps to forming its active state. Increased vitamin D levels lead to increased calcium and phosphorus absorption in the intestine. In cholestatic patients with fat malabsorption, less vitamin D is absorbed, which decreases calcium and phosphorus absorption. The nonabsorbed fatty acids bind to calcium in the intestine, further decreasing calcium absorption. Absorption of calcium is also reduced by corticosteroid therapy.[192,194] A 25-OH vitamin D level shows the pool of both dietary and endogenous vitamin D. One study recommends checking vitamin D levels

every 1 to 2 months until it is within an appropriate range.[186] DEXA scans are the gold standard for assessing bone density, therefore all patients at risk should be screened.

Recommendations for supplementation of vitamin D differ significantly in light of the emerging research on vitamin D. In patients with biliary atresia, supplementation doses can range from 1200–8000 units per day for levels of Vitamin D less than 15 ng/mL.[189] In adults with primary biliary cirrhosis and primary sclerosing cholangitis, 400–800 IU/day of vitamin D in conjunction with TPGS vitamin E to enhance absorption has been recommended. In an adult-based review article, Holick, recommends 50,000 units of vitamin D_2 every week up to every day depending on the degree of fat malabsorption, or up to 10,000 units of vitamin D_3 every day for up to 5 months, then a maintenance dose of 50,000 units of vitamin D_2 every week.[105] Recent pediatric practice has shown wide use of 10,000–50,000 unit Vitamin D_2 per week in populations most susceptible to deficiency. The vast differences in these recommendations point to the importance of trending laboratory values and making dosage adjustments based on each patient's clinical response to supplementation.

Vitamin K deficiency is due to fat malabsorption and decreased absorption from gut flora alterations. This deficiency, as well as impaired hepatic synthesis of clotting factors, leads to prolonged **prothrombin time** (PT). If the abnormal PT is greater than 1.5, it is thought to be related to a vitamin K deficiency, the medical team can prescribe oral or intravenous supplementation. In a study of cholestatic children taking vitamin K supplementation with normal or near-normal PT, 54% were found to be vitamin K deficient per measure of a PIVKA-II level (protein induced by vitamin K absence).[194] Although following a PIVKA-II level is not widely done in clinical practice, it should be considered in order to assess for subclinical signs of vitamin K deficiency.

Liver Transplant

Introduction

The most common disease requiring liver transplantation in childhood is biliary atresia (over 50% of cases). Other less prominent causes are inherited metabolic disorders (e.g. alpha-1-antitrypsin deficiency, tyrosinemia, and urea cycle defects), intra-hepatic cholestasis syndromes (e.g. Alagille syndrome, Byler syndrome), chronic hepatitis with cirrhosis, and all forms of acute liver failure. Indications for transplantation include hepatic failure, complications of portal hypertension (e.g. variceal bleeding, ascites), specific metabolic disorders, and malignancy.

The patient with end-stage liver disease awaiting transplantation presents a formidable challenge to the medical and nutritional team. The particular liver disease involved, the magnitude of liver dysfunction, the presence of complications, and the transplantation procedure itself combine to present a complex treatment process including meeting nutritional needs for healing and growth.

Anatomy, Physiology, Pathology

Cirrhosis is a chronic disease of the liver marked by degeneration of cells, inflammation, and fibrous thickening of tissue. Cirrhosis may be caused by a degree of pediatric diseases discussed (See the discussion under "Cholestatic Liver Disease."). Without a liver transplant, cirrhosis left untreated results in death.

Nutrition Screening

Nutrition screening in pediatric patients requiring a liver transplant should be consistent with clinical malnutrition (undernutrition) risk screening as defined by the AND and ASPEN.[13]

Nutrition Assessment

Patients listed for liver transplant should be referred to a pediatric RDN who is a certified organ transplant provider in order to receive an individualized nutrition assessment. In most institutions, an RDN evaluation is required for a patient to be listed for liver transplant regardless of nutrition status. During the evaluation, the RDN determines if the patient is an appropriate candidate for organ transplant based on nutrition parameters. Nutrition assessment during liver transplant evaluation should consist of obtaining a thorough diet history with emphasis placed on food, fluid, and breastmilk/formula intake as well as the use of vitamin supplementation. Fat-soluble vitamins may need be checked at the initial diagnosis based on the pathophysiology of liver failure and in patients with cholestasis.[195]

Patients with chronic cholestasis/cirrhosis can present with ascites and/or organomegaly, which will falsely elevate the patient's weight as occurs in patients with cholestatic liver disease. (See the discussion under **Cholestatic Liver Disease**). In listing a patient for liver transplant, MUAC, length/height for age and head circumference are the preferred anthropometrics in evaluating nutrition status for determining PELD score.

Diagnosis

The need for a liver transplant is based on multiple factors. Institutions commonly have a liver transplant team composed of a transplant surgeon, hematologist, nurse practitioner, social work, psychiatrist, and dietitian that

meet to discuss appropriate candidates for transplant based on the following criteria:

- Irreversible cirrhosis with at least two signs of liver insufficiency
- Fulminant hepatic failure: coma Grade 2
- Unresectable hepatic malignancy confined to the liver that is less than 5 cm in diameter
- Metabolic liver disease that would benefit from liver replacement

Factors that are listed below are often the precipitating reason for proceeding with liver transplantation:

- Severe fatigue
- Unacceptable quality of life
- Recurrent variceal bleeding
- Intractable ascites
- Recurrent or severe hepatic encephalopathy
- Spontaneous bacterial peritonitis
- Hepatorenal syndrome
- Development of small hepatocellular carcinoma on hepatic imaging

Nutrition Diagnostic Statements (PES)

- Inadequate oral intake related to cirrhosis as evidenced by EN to meet needs.
- Altered gastrointestinal function related to cirrhosis of the liver as evidence by fat-soluble vitamin supplementation.

Management

Once a patient is identified as a liver transplant candidate, they are listed on the national United Network for Organ Sharing (UNOS) roster.[196] Organ allocation for children is based on the Pediatric End-Stage Liver Disease (PELD) score.[196] The child in most urgent need of a transplant are placed highest on the status list and are given first priority.

As soon as a liver is available it is surgically placed into the recipient candidate. Post-liver transplant management requires a prolonged hospital stay with the use of multiple medications in order to decrease chances of rejection including using immunosuppressants and steroids.

Nutrition Intervention: Pre-transplant – End-stage liver disease creates a hypermetabolic state that elevates the REE.[197] In the hypermetabolic state, once the body's glucose stores are exhausted, lean body mass and proteins will be broken down for gluconeogenesis resulting in protein-energy malnutrition. In a study evaluating the use of predicted energy equations in the pediatric patient with liver disease, commonly used estimation methods, like Food and Agriculture Organization/World Health Organization/United Nations University (FAO/WHO/UNU) and Schofield were shown to underestimate calorie needs based on indirect calorimetry.[197] The study suggested that indirect calorimetry should be used when available in order to determine estimated needs in patients with liver disease who are malnourished.

Children with liver-disease have been found to have inadequate oral intake due to decreased appetite secondary to the anorexic effects of elevated serotonin levels[198] and increased serum leptin levels.[199] Long periods of suboptimal intake and fasting are not recommended in the pediatric population. For older children, small more frequent meals with a nighttime snack, specifically high in carbohydrates and branched-chain amino acids, can optimize nutrition intake and utilization.[200,201] Using oral nutrition supplements and/or enteral feedings may be required to provide adequate nutrition.[200] Protein should not be restricted in infants and children with end-stage liver disease and is often provided at levels above the RDA, unless encephalopathy is present.[183,185] **Branched-chain amino acid** requirements (BCAAs; leucine, isoleucine, valine) are increased in pediatrics patients with cholestatic liver disease at up to 142% of the recommended dietary allowance for the healthy child.[202] Additional supplementation with BCAA may be beneficial, however, dosing recommendations have not been established in children. Formulas with additional BCAA added has been shown to improve height, weight, and muscle mass in addition to decreased albumin infusions.[203]

If the end-stage liver disease is related to a cholestatic liver disease, then the decreased pool of bile acids will result in malabsorption of long-chain triglycerides. MCTs should be used to provide calories and minimize steatorrhea, but MCTs do not provide essential fatty acids or help absorp fat-soluble vitamins.[186] For more specific macro- and micronutrient information, please refer to the "Cholestatic Liver Disease" section of this chapter.

Multiple studies have shown that pre-operative malnutrition negatively affects the outcome of a liver transplant. Height z-score and head circumference is a good indicator of pre-transplant malnutrition. Severe growth retardation is linked to an increased length of stay and increased hospital costs after transplant.[204] In a study of infants with biliary atresia listed for liver transplant, researchers found the post-transplant mortality and graft failure risks include weight/height that is below two standard deviations at the time of listing.[205] Close nutrition monitoring in end-stage liver disease and optimizing nutritional intake will maximize growth potential and nutritional status going into transplant.

Monitoring and Evaluation: Post-Transplant – In many cases, the underlying disease is corrected by transplant; therefore, calorie and protein needs should be individualized to the patient's specific postoperative needs and nutritional status.

Immediately after transplant, early nutritional support should be implemented due to preoperative malnutrition, surgical stress, and postoperative catabolism.[206] Nutrition support should be initiated and slowly advanced to meet calorie and protein needs. The enteral formula should be chosen based on each patient's protein and fat composition needs. The continuation of a high MCT formula post-transplant is often not required. In a study evaluating tube feedings, nasointestinal tube feedings that were started post-transplant vs. maintenance intravenous fluids were maintained until an oral diet could be initiated and showed improved nitrogen balance by the fourth postoperative day and less overall infections.[207]

The nutritional goal is catch-up growth to achieve an age-appropriate weight and height. Post-transplant studies show that weight recovery is rapid and often normalizes within one year of transplant regardless of malnutrition status before transplant.[208] Long-term data evaluating height potential in patients from transplant to 15 years of age found that after transplant a growth spurt occurred with growth potential reaching its max by 10 years of age.[209,203] Deceleration of growth occurred by age 15 and was mostly related to graft dysfunction.[209] Catch-up growth potential was greatest in patients who had a transplant after 2 years of age and had the most growth retardation before transplant, although they remained significantly shorter and lighter at age 15 than age standards.[210] Overall, linear growth achievements should be analyzed by the underlying disease that necessitated transplant. Patients with biliary atresia and alpha-1-antitrypsin achieve better linear growth post-transplant than patients with fulminant liver failure, Alagille syndrome, or chronic hepatitis.[211]

Drug–nutrient interactions are key considerations post-transplant. Steroids are known to affect linear growth and bone health, and to contribute to hyperglycemia and diabetes mellitus.[212] Immunosuppressive medications also contribute to electrolyte and magnesium losses.

Concern regarding metabolic bone disease continues after liver transplant. Risk factors include continued corticosteroid use, malnutrition, muscle wasting, preexisting osteopenia/osteoporosis, and immunosuppressive agents. Bone loss is greatest in the first year after transplant.[168] Patients with cholestatic liver disease who undergo liver transplant improve their bone mineral density.[213] In a prospective pediatric study an increase in bone mineral density was noted at 3 months post-transplant along with improvements in 25-OH vitamin D levels.[214]

Using immune-suppressants after an organ transplant increases a patient's susceptibility to food-borne illness. Caregivers should be informed of safe food practices and receive information regarding cross-contamination and temperature danger-zones.

Short Bowel Syndrome

Introduction

Short bowel syndrome (SBS) is a condition in which the patient has an anatomic or functional loss of more than 50% of the intestine.[215] It has been defined as the reduction of functional gut mass below the minimal amount necessary for digestion and absorption adequate to satisfy the nutrient and fluid requirements for maintenance in adults or growth in children.[216,217] Typically, SBS results from NEC, volvulus, intestinal atresia, gastroschisis, ruptured omphalocele, or vascular infarct.[215,216,218,219] The result of the injury and/or resection is a decreased small intestinal surface area, which leads to malabsorption and large volume watery diarrhea.[220] Although most patients have had a significant bowel resection, some patients may have had a rather small amount of bowel removed, but have poor motility and absorption by the remaining intestine.[220,221] The degree of malabsorption is dependent upon the extent of missing or injured bowel; most affected individuals have difficulty sustaining appropriate growth and development without nutritional support. Growth is a major concern in this population, affected not only by adequate nutrient provision but also by absorption.

Intestinal adaptation is the process by which the remaining intestine grows, dilates, and changes via cellular hyperplasia and villous hypertrophy in order to compensate for its loss of surface area or function. Adaptation is the ultimate goal of treatment and it is dependent on small bowel length, intact **ileocecal valve** (ICV), intestinal continuity, and preservation of the colon.[222] Adaptation usually occurs within the first 3 years but it may take as long as 13.5 years.[222] A retrospective analysis of children with short bowel syndrome that required long-term PN by Quiros-Tejeira et al., found that patients with ICV even with USBS and patients with >15 cm of SBL without ICV have a chance of intestinal adaptation. The ability of the intestine to adapt is largely dependent on how much intestine remains, the health of the remaining intestine, the presence or absence of the ICV, and whether the colon is in continuity with the small bowel.[222]

Anatomy, Physiology, Pathology of Condition

Absorption of fluids and nutrients occurs throughout the small intestine and colon. Half of the mucosal surface is contained within the proximal one-fourth of the small intestine.[220] **TABLE 12.7** illustrates sites of nutrient absorption in the small intestine and colon, as well as the possible implications of intestinal resections of these areas.

The duodenum and jejunum are the primary sites of digestion and absorption of proteins, carbohydrates, lipids, and most vitamins and minerals. Resections in

TABLE 12.7 Intestinal Resection: Function Adaptation and Complications of Resection

Bowel Segment	Function	Adaptation	Complications of Resection
Duodenum	Absorption of CHO, FAT, Pro, Iron Calcium Phos, Mg, Folic Acid	Cannot adapt for the loss of other segments	Macronutrient malabsorption. Acidosis Anemia Osteopenia
Jejunum	Primary site of CHO & PRO absorption Water soluble vitamin absorption	Linger villi and large surface area can adapt for loss of the ileum and/or duodenum. Cannot absorb B_{12} or bile salts	Fluid/electrolyte losses Macronutrient and water soluble vitamin malabsorption
Ileum	Vitamin B_{12} & bile salt absorption Fat soluble vitamin absorption	Increased amount of water and CHO absorbed Does adapt for the Jejunum	Unable to absorb B_{12} Loss of fat soluble vitamins Increased risk of renal oxalate stones
Ileocecal Valve	Limits the reflux of colonic contents into the ileum Limits the rate of food passage into the cecum	Bowel cannot adapt for loss of the valve	Bacterial overgrowth causing inflammation and leading to further malabsorption Rapid transit time
Colon	Fluid absorption Na, Cl, K and fatty acid absorption	Can help in water absorption and enhance carbohydrate absorption (from short chain fatty acids)	Dehydration Electrolyte abnormalities Reduced ability to absorb bile salts Decrease in transit time

Beer S, Bunting KD, Canada N, Rich S, Spoede E, Turybury, K, eds. *Texas Children's Hospital Pediatric Nutrition Reference Guide.* Houston, Tx: Texas Children's Hospital; 2016:110.[223]

these areas result in a decreased surface area for absorption and cause increased osmotic diarrhea and loss of water-soluble vitamins.[224] Loss of calcium, iron, and magnesium can occur, as well as losses of trace minerals such as copper, chromium, and manganese. Decreased secretin, cholecystokinin, and pancreatic/biliary secretions result in decreased digestion and absorption of fats, proteins and fat-soluble vitamins. Decreased disaccharidase secretion allows increased substrate for bacterial overgrowth because carbohydrates are not fully metabolized when these enzymes are decreased.

Decreased surface area from ileal resection results in decreased vitamin B_{12} absorption. Decreased bile salt reabsorption in the ileum increases bile acid in the stool and decreases enterohepatic circulation.[224] This causes bile acid pools and decreased micelle formation. Decreased absorption of long chain fats results, as well as decreased absorption of fat-soluble vitamins. There is increased steatorrhea, and the potential for cholelithiasis and renal oxalate stones. Many gut hormones that affect GI motility are produced in the ileum, including entero-glucagon and peptide YY. Resection of the ileum can impair the nutrient-regulated gut motility.[216,225]

With jejunal resection, the ileum can assume the place of the jejunum in some ways to absorb less site-specific nutrients such as electrolytes and fluid, which the jejunum is responsible for when the bowel is intact.[226] However, due to some of the specific roles of the ileum, the jejunum cannot assume the role of the ileum. For example, loss of the terminal ileum requires B_{12} provision via intramuscular or nasal route for absorption because there are no other sites for B_{12} absorption in the proximal ileum, duodenum, or jejunum. With the loss of the jejunum there are no absolute requirements for supplementation, because absorption will vary depending on the individual child.

The ICV slows transit time and acts as a barrier to bacteria moving into the small bowel from the colon.[224] Resection of the ICV leads to vitamin B_{12} and possibly folate deficiency because the sites for absorption are often lost with adjacent ileum resections. Combined ICV and ileum resections lead to decreased transit time and large influx of nutrients into the large intestine, which can result in malabsorption. Bacterial overgrowth in the small bowel can be a major problem and lead to increased diarrhea and malabsorption.[215] Some centers

routinely cycle patients on either a single antibiotic such as metronidazole for 1 to 2 weeks every month or a combination of two alternating antibiotics such as metronidazole and neomycin for overgrowth treatment. Length of treatment for overgrowth varies depending on the response to treatment; some children will continue treatment for only a few weeks whereas others can continue for several years.

The colon absorbs water, salvages malabsorbed carbohydrates and absorbs sodium. Colonic resection decreases water and sodium reabsorption and increases risk for dehydration.[224]

Nutrition Assessment

Growth is the ultimate measurement of nutritional adequacy. Caloric and protein needs are dependent on age and state of growth, as well as the degree of overall malabsorption. Caloric needs vary from child to child, and can be as low as normal for age or quite increased. No good studies or data exist for recommendations on specific calorie levels, and clinical judgment along with known intake levels and growth outcomes for the specific child should be used. Adequate carbohydrate, fat, micronutrients, and fluid provision are also dependent on age and state of growth; however, they are more influenced by resection site. Micronutrients should be monitored routinely depending upon the specific anatomy of the individual and should be replaced as needed in available forms (enteral, intramuscular, or parenteral.) The most common deficiencies are the fat-soluble vitamins, Vitamin B_{12}, zinc, and iron.

Nutritional management of the pediatric patient with short bowel syndrome can be divided into four phases. The first phase involves fluid, electrolyte, and hemodynamic stability, and PN initiation. The second phase is the initiation of EN. The third phase is weaning of PN and advancing oral nutrition and EN. The fourth phase is long-term nutrient provision and growth monitoring.

The first phase in treating a new patient with SBS is PN while the GI tract is recovering postoperatively. PN should be started as soon as possible via a central line.[221] Initial PN macronutrient needs vary depending on the infant and the clinical situation. Initiation of dextrose should be at 5–7 mg/kg/min and advancement by 1–3 mg/kg/min to endpoint goal of 12–14 mg/kg/min.[221] Various institutions are reporting anecdotal success with pushing the endpoint goal to as much as 16 mg/kg/min in order to use minimal lipids while providing adequate calories. Initiation of protein at 1–2 g/kg/day and advancement by 1–2 g/kg/day to endpoint of 3–3.5 g/kg/day is recommended.[221]

Initiation of lipid at 1 g/kg/day and advancing by 1 g/kg/day to an endpoint of no more than 3 g/kg/day is recommended.[221] Soy-based intravenous lipids have been associated with the development of liver disease.[216] Due to an increased risk of developing **intestinal failure-associated liver disease** (IFALD) many centers restrict lipids to 1 g/kg/day while adjusting other macronutrients within PN to meet calorie goals. Due to decrease/restriction of IV lipids, the clinician needs to be aware of essential fatty acid deficiency and monitor for those patients who are receiving less 1 g/kg/day.[216] Presently, there are newer lipid IV emulsions derived from fish oils that contain predominantly n-3 fatty acids, as opposed to the n-6 fatty acids that have been approved for adults.[216] These lipid emulsions are currently in clinical trials and long-term studies are needed to compare fat emulsions and examine the risk of hepatic cirrhosis in this patient population.[216]

Initial stool output, whether per rectum or ileostomy, typically increases the needs for sodium, magnesium, and zinc to compensate for losses.[221,227] Serum zinc is not always reflective of zinc status, but monitoring trends can be helpful. An infant not responding to adequate caloric and protein provision, but with high output stool volume may benefit from additional sodium and zinc supplementation despite normal serum values. PN should be monitored carefully to minimize complications associated with PN therapy. **Parenteral nutrition associated liver disease** (PNALD) is a major cause of death in patients with SBS.[228] See Chapter 11 on PN management details for maximizing calcium and phosphorus and for preventing associated complications with use of PN.

EN is the second phase of nutrition therapy and should be started as soon as the patient is stable and gastrointestinal motility has returned as a necessary component to promote intestinal adaptation.[219,229] A slow continuous infusion is thought to bathe the lumen and allow for better nutrient absorption.[219] In some conditions, if infants have a proximal high ostomy and a mucous fistula connected to a substantial portion of the bowel, refeeding the mucous fistula with the proximal bowel content can be done to prevent diffuse atrophy.[230]

If available from the mother or via donor, breast milk should be used in infants with SBS to initiate EN. The abundance of growth factor and nucleotides available in breast milk vs. formula makes it the ideal choice for these children.[219,221] If breast milk is unavailable, however, controversy exists as to the ideal formula for infants with SBS. Each infant is different, and a formula should be selected to start EN depending on his or her clinical situation.

Polymeric formulas have been shown to be better for mucosal adaptation in adults and in animal studies.[219,231,232] In pediatrics, hydrolyzed and even amino acid formulas are often preferred. In this population, mucosal breakdown, bowel dilation, and bacterial overgrowth all

predispose the infants to higher rates of food allergies; thus, the provision of an amino acid formulation is a considerable advantage.[233] A 2001 prospective, randomized, crossover trial involving 10 children with SBS and lasting 60 days found no difference in energy and nitrogen balance when using a hydrolyzed protein formula versus an intact protein formula. This study is often cited in pediatric SBS guides.[234] It should be noted, however, that the study did not measure adaptation, diarrhea, or perceived tolerance to formulas. The study concluded that it is common practice that a semi-elemental diet is used to optimize absorption of the remaining bowel and that there was a difference in intestinal permeability, weight gain, and nitrogen balance in children with SBS whether they were receiving hydrolyzed or non-hydrolyzed protein formulas.[234] Also, some centers advocate amino acid formulations that were better tolerated and helpful in weaning from PN.[234]

If fat malabsorption exits from bile acid hypersecretion, PNALD, or pancreatic insufficiency, using a formula with a higher percentage of fat from MCTs is preferred.[233] Even though MCTs increase the osmotic load slightly and offer fewer calories per gram than LCTs, the lack of micelle needed for absorption makes them the ideal fats in this situation.[233] If bilirubin levels are elevated, using a very high MCT formula is recommended.[233,235] In rate studies, the use of LCTs have facilitated better adaptation of the remaining bowel than the use of MCTs.[233,235] Many have extrapolated this to indicate that LCTs are better than MCTs for infants with SBS and facilitate better intestinal adaption.[219,229,232] Interesting work has been done recently in adults with SBS indicating that the use of oleic acid (LCT) supplementation actually did not cause a delay in transit time (which is thought to stimulate adaption better) and actually decreased energy absorption by 14% vs. placebo overall.[236] The fact that this is an adult study with a small sample size (n=57) poses limitation for its use in pediatrics. However, it does make an argument in the setting of a human model with LCTs. Several institutions advocate using a blend of both MCTs and LCTs.[233,237] Most typically in practice, amino acid formulations with higher MCT content such as Elecare (33% MCT, Abbott Labs) and Neocate Infant DHA/ARA (33% MCT, Nutricia) are preferred formulas used for this patient population.

The third stage of nutritional management should be started while the infant is still hospitalized, if possible. PN should be weaned as EN and oral tolerance increases, stool output decreases, and growth is achieved. The ultimate goal is eliminating PN because cholestasis occurs in 30% to 60% of children with SBS; liver failure develops in 3% to 19% of children who acquired SBS in the neonatal period.[238–240] Gradual advances in EN/oral intake are dependent upon gastric tolerance, stool output (frequency and consistency), and rate of growth.[1] This stage can be quite lengthy, depending on setbacks and the rate of advancement. Stool should be monitored for frequency, acidosis, and reducing substances as EN and oral intake advances. PN should be cycled when 35–50% of EN goal has been achieved and if blood sugar levels are stable. Two to 6 hours off of PN each day allow for GI hormone release.[221,233] As PN is weaned, vitamin and mineral status should be closely monitored and supplemented as needed because the absorption route has changed and the potential for malabsorption has increased.

Oral feedings, even small amounts of volumes, should be started as soon as clinically stable and feasible, to prevent oral aversion.[1,219,229,233] Feeding aversions are common in this population for many reasons. First and foremost, prematurity and severe illness delay oral attempts and result in immature feeding skills.[224] Furthermore, frequent vomiting and diarrhea prevent pleasurable associations with food.[219] Oral feeds can be introduced as developmentally appropriate, and general precautions regarding monitoring for food allergies should be practiced. If the infant is unable to take any oral feedings, and oral motor stimulation program should be in place to help develop feeding skills. Feeding aversion in patients with SBS is notoriously difficult to treat, and the focus is on prevention of aversion as feasible.[1,232,241]

The fourth phase of management is long-term advancement of EN and maximizing oral intake. Once EN is at goal, it may be beneficial to start fiber therapy if the colon (or most of it) is present. The conversion of complex carbohydrate to **short-chain fatty acids** (SCFAs; acetate, propionate, and butyrate) in the colon can cause significant caloric reuptake by colonic cells.[1,232,240] Additionally, SCFAs also stimulate sodium and water reabsorption, which aids in fluid management. Adding a 1–3% pectin solution can decrease reducing substances and improve pH to allow for more fermentation of carbohydrates.[221] In orally fed patients, using guar gum fiber such as Nestlé's Resource Benefiber (hospital grade) at 0.5 g/kg/day may be used as a more palatable replacement for pectin.

As the intestine adapts, the goal is to normalize the EN schedule while promoting oral intake and growth. If possible, EN should be cycled overnight to maximize oral intake in the daytime hours. It is necessary that children find palatable beverages providing complete nutrition to help lessen EN dependence and increase oral intake. Higher calorie formulas typically have higher osmolarity and may interfere with desired weight gain. However, some nutritional products provide a high percentage of calories from fat, with lower osmolarity. Overly strict guidelines for oral intake should be discouraged. Each child needs to be carefully assessed because nutrient

needs differ due to amount and location of the resections. General guidelines include:

- Limiting simple sugars (juice, candy) in order to minimize osmotic diarrhea.[1,219,221,232,233,235,242]
- Fat and protein should be adequate to support growth.
- Soluble fiber of at least 5–10 g/day as pectin/guar gum is beneficial to slow the gut transit time.[219,229,235] Those with a colon have an additional bonus of caloric reuptake from provision of soluble fiber in the diet.
- Lactose should be avoided only if symptoms are reported with ingestion.[1,235]
- Liberal salt intake at meals helps make the meal bolus isotonic in the gut.[235]

Much has been reported on the guidelines for adults with SBS, and many of these have been adapted to the child.[235] In adults with a colon, 50–60% of kcal from CHO and 20–30% of kcal from fat are recommended as a balance of LCT and MCT. Those without a colon are encouraged to consume 40–50% kcal from carbohydrates and 30–40% for kcal from fat with more LCTs than MCTs. Isotonic fluids are always encouraged, regardless of the presence or absence of a colon; however, hypoosmolar fluids that are higher sodium-containing fluids are encouraged for those without a colon. Sometimes oral rehydration solutions (ORS) are recommended for those without a colon (Table 12.3).

Increased renal oxalate stones occur with intestinal resections because of increased enteric absorption of oxalate (enteric hyperoxaluria).[243] Renal oxalate stones are well documented in the literature for adults with SBS, and up to 30% of this population develop stones.[1,235] However, the incidence is relatively rare in children.[243,244] A retrospective 5-year review at a tertiary pediatric medical center by Chang-Kit reported 72 causes of urolithiasis. Of note, seven patients had Crohn's disease, seven had cystic fibrosis, and four had SBS. It is not routine to restrict oxalates in patients with Crohn's disease or cystic fibrosis. Thus, empiric restrictions, especially in the setting of a history of food aversions, are not necessary. Allowing oxalated foods in diet or provision of extra calcium is recommended if 24-hour urinary oxalate levels are elevated.[235] Urinary oxalate levels should be monitored annually in this population.

Nutrition Diagnostic Statement (PES)

Nutrition diagnosis is determined from information gathered during the nutrition assessment.

- Altered GI function (NC 1.4) related to short bowel syndrome as evidenced by need for parenteral nutrition to support growth
- Inadequate oral intake (NC 2.1) related to oral aversion and oral diet modification as evidenced by need for enteral nutrition to maintain growth as advancing to oral autonomy

Nutrition Intervention

The overall nutrition goal is to achieve adaptation and autonomy and wean the patient off of PN. This includes careful consideration of managing the oral and enteral feeding regimen while weaning the patient off of PN and monitoring laboratory values and macro- and micronutrients.

Nutrition Monitoring and Evaluation

Ongoing monitoring and evaluation is essential to ensure that the nutrition intervention provided was effective and is sustainable. Feedback provided by the client and his/her caregiver(s) will allow for appropriate adjustments in the dietary management plan. Follow up visits include the re-evaluation of diagnostic clinical markers, such as gastrointestinal symptoms/discomfort, growth trends, and review of food-symptom diary. Nutrition-related vitamin/mineral lab results should also be monitored and addressed, with supplementation recommended and/or adjusted as appropriate.

Special Considerations

Although growth hormone and glutamine have been used in some adult studies,[242–246] more research is needed and there are no current recommendations for their use in pediatrics.[235,242–247] Alternative surgical procedures such as bowel lengthening, tapering enteroplasty, or intestinal transplant may be considered for those patients who are not making progress.[237,248–250]

Intestinal Transplantation

Introduction

An intestinal transplant is typically reserved for those pediatric patients who have developed PNALD and failed other nutrition modalities to wean off of PN or have had life-threatening line infections. Intestinal transplant is not limited to an isolated intestine, it can also be a modified multivisceral transplant, which includes the stomach and pancreas or a multivisceral transplant that includes stomach, pancreas, and liver.[251] Nutrition management both pre/post-transplant is varied from center to center. Managing children with intestinal failure has improved at centers that use a multidisciplinary team approach. Intestinal transplant is reserved for those who obtain life-threatening episodes of line sepsis, loss of venous access, and

parenteral nutrition induced liver failure.[251] The overall postoperative management includes a complex treatment of immunosuppression and management of possible infectious complications as well as ongoing nutrition management that benefit from a multidisciplinary team approach. Grant et al reported that transplants since the year 2000 have a survival rate of 77% at year 1 and 55% at year 5. Sepsis remained the leading cause of graft loss.[252] The Intestinal Transplant Registry Report also noted that by 6 months after transplantation most recipients resume an oral diet and improved quality of life.[252]

Anatomy, Physiology, Pathology of Condition

Indications for intestinal transplant has not changed over time.[252] Intestinal transplant is not acceptable for all pediatric patients who have a diagnosis related to having intestinal failure. Transplantation should be considered when other modalities have not been successful. Recent advancements have been made in treating patients with intestinal failure such as restricting intralipids or even using other intravenous fat sources, rotating antibiotic therapy and medical advances to central line care as well as surgical procedures to help patients meet a more autonomy of enteral feeds and use less PN.[253] The primary indications for transplant of the intestine are not limited to: life-threatening episodes of line sepsis, loss of venous access, PNALD, or frequent life-threatening dehydration or electrolyte imbalances.[251] Intestinal transplant is not limited to an isolated intestine transplant, but may also be modified multivisceral and or a multivisceral transplant.[251] Overall intestinal transplant may include all of the following or selected organs with the intestine: stomach, liver, and pancreas.

Nutrition Assessment

Nutrition assessment is a complex assessment that entails growth history, feeding regimen, laboratory values, and nutrition-focus exam in developing and adjusting to meet the patient's nutritional requirements and aid in continued support of optimizing growth. These patients also typically have significant losses from output and need this volume and constituents replaced.

Nutrition Diagnostic Statement (PES)

Nutrition diagnosis is determined from information gathered during the nutrition assessment:

- Altered GI function (NC 1.4) related to short bowel syndrome as evidenced by the need for intestinal transplant due to PNALD.

Nutrition Intervention

Formalizing an individual nutrition regimen is based on findings from a patient's laboratory values, output losses, and growth. An early nutrition onset post-transplant complication seen is chylous ascites. Chylous ascites occur when lymphatic disruption is present in the abdominal cavity with resultant accumulation of chyle in the abdomen.[254] Post-transplant patients are started on a fat-free diet for the first 2–4 weeks after transplant and then the introduction of fat is adjusted after this time. Most patients receive gastrostomy or jejunostomy feedings to help achieve EN while weaning from PN.[255] The overall nutrition goal is to achieve autonomy and wean off of PN. Careful consideration of managing the oral and enteral feeding regimen while weaning PN is needed. Close monitoring of laboratory values and macro- and micronutrients is also required.

Nutrition Monitoring and Evaluation

Following a patient's individual growth, labs, and output is required as is adjusting these to continue to achieve appropriate growth. As immunosuppression is common, careful consideration in assessing vitamins and mineral levels and replenishment is needed. Since these patients are advanced on an EN regimen, feeds should be adjusted to prevent growth failure or the development of malnutrition.

Conclusion

The gastrointestinal tract is a complex organ system designed to ingest, digest, and absorb nutrients before waste expulsion. Its functions are essential to optimal health; however, if disrupted, alterations in nutritional requirements and status result. The clinical presentation of gastrointestinal disorders is multi-systemic and formal diagnoses are physician made and based on testing results. Nutrition screening in pediatric patients should be consistent with clinical malnutrition risk screening as defined by the AND and ASPEN. Additionally, pediatric patients benefit from receiving an individualized nutrition assessment from an experienced pediatric RDN, which often consists of obtaining a thorough diet history with emphasis placed on total energy intake, as well as both macronutrient and micronutrient intake, biochemical and clinical data results, anthropometric assessment, growth history/trends, and the need for multivitamin/mineral supplementation to aid in preventing and/or correcting nutrient deficiency. Information gathered during the nutrition assessment process allows for the determination of the nutrition diagnosis and appropriate intervention. Ongoing monitoring and evaluation is essential to achieve optimal health outcomes and improved quality of life within the pediatric gastrointestinal population.

CASE STUDY

LC is a 14-year 2 month-old female who was diagnosed with celiac disease one month ago.
This is her first visit to the celiac clinic post diagnosis. Recent lab values reflect vitamin D of 28.1. DEXA revealed z-score of -0.8. LC has begun eliminating gluten from her diet. Weight at today's visit is 35.1 kg, height is 151.5 cm.
24 Hour Diet recall reveals:
Breakfast: 1 packet of Quaker oatmeal mixed with 1 cup skim milk, apple
Lunch: Dietz and Watson turkey and cheese on Udi's gluten-free bread, Snyder's gluten-free pretzels, carrots, water
Snack: Smartfood popcorn or Tostitos tortilla chips and salsa
Dinner: chicken, rice, green beans, apple juice
Snack: Schar gluten-free cookies

Please calculate the following:

Wt/age% tile:
IBW (kg):
% IBW:
BMI:
BMI %tile:

Ht/age %tile:
Stnd. Ht/age (cm):
% Stnd Ht/age:

Please classify LC's nutritional status:

☐ Normal ☐ Wasted (mild/moderate/severe) ☐ Stunted (mild/moderate/severe)

Please estimate needs:

Calories:	per day	(REE x ___ used)
Protein (gm):	per kg	
Fluid (ml):	per day	
Calories:	per day	(REE x ___ used)
Protein (gm):	per kg	
Fluid (ml):	per day	

Review Questions

1. Can you identify any sources of gluten that LC is currently receiving in her diet?
2. Comment on DEXA results and vitamin D level
3. Would you recommend vitamin/mineral supplementation? If so, please specify.
4. What are your MNT goals for LC?

Answer Key:

Wt/age %tile: <5th
IBW (kg): 42
% IBW: 83
BMI: 15.2 KG/M^2
BMI %tile: <5th

Ht/age %tile: 5–10th
Stnd. Ht/age (cm): 161
% Stnd Ht/age: 94

Mild wasting
Mild stunting
1995 kcal/day (REE x 1.7) 1 gm protein/kg/day, 1802 ml/day (REE = 1174)

1. Quaker oats should be avoided due to cross contamination risk during milling and processing.
2. Introduction of certified gluten-free oats into diet (such as Bob's Red Mill brand) should be discussed with healthcare provider
3. DEXA normal and vitamin D insufficiency

4. Research reports that adherence to the gluten-free dietary pattern may result in a diet that is low in iron, folate, niacin, vitamin B_{12}, calcium, phosphorus and zinc. Therefore, a daily complete multivitamin is recommended. LC's dietary intake of calcium is also low. DRI for age indicates that LC should be taking in 1300 mg of calcium per day. In light of low vitamin D and suboptimal calcium intake, a calcium with vitamin D supplement is recommended. Ideally, a supplement that provides 1000 mg of calcium and 1000 IU of vitamin D.

Goals:

- Replete vitamin deficiencies and promote weight gain with diet and supplementation.
- Identify need for additional calorie boosting such as use of Ensure or Boost beverage.
- LC will verbalize good understanding of gluten-free diet.

Websites

Websites that provide additional information on the various GI conditions are listed below.

General Gastrointestinal

https://www.gikids.org/http://www.naspghan.org

Celiac Disease

Foundations/Associations
Beyond Celiac
https://www.beyondceliac.org/
Celiac Disease Foundation
https://celiac.org/
National Celiac Association
https://www.nationalceliac.org/
Online resources for gluten-free food/recipes
https://simplygluten-free.com/
http://www.glutenfreemall.com
http://www.glutenfreeda.com
https://www.glutenfreeandmore.com/

Magazines for People with Celiac Disease & Other Food Allergies/Sensitivities

Gluten Free Living
http://www.glutenfreeliving.com
Simply Gluten Free
http://simplyglutenfreemag.com/

Crohn's and Colitis Foundation of America

http://www.ccfa.org

Review Questions

1. The gastrointestinal tract is a large muscular tube designed to ingest, digest, and absorb nutrients. Functions of the gastrointestinal tract may be disrupted by which of the following?
   A. Disease
   B. Antibiotics
   C. Environmental Toxins
   D. Bacterial Overgrowth
   E. All of the above

2. Approximately 95% of critically ill patients experience diarrhea associated with enteral feeds. Which feeding method has been shown to aid in decreasing diarrhea?
   A. Gastric feedings
   B. Post-pyloric feedings

3. The specific nutrition diagnosis established from the nutrition assessment aids in determining the appropriate medical nutrition therapy intervention. Which one of the following is NOT recommended as a first line of therapy in children with constipation?
   A. High Fiber Diet
   B. Use of Prebiotics and Probiotics
   C. Increased fluid intake
   D. Increased physical activity

4. The only known treatment for celiac disease is adherence to a gluten-free diet. Which of the following foods listed below **may** contain gluten?
   A. Baked Beans
   B. Hot chocolate mix
   C. French Fries
   D. Miso Soup
   E. All of the above

5. Crohn's disease may occur in any portion of the gastrointestinal tract; whereas, Ulcerative Colitis is confined to the colon with minimal involvement of the terminal ileum.
   A. True
   B. False

6. Early initiation of enteral feedings **is not** recommended in patients with moderate to severe pancreatitis.
   A. True
   B. False

7. Nutrition interventions in an infant/child with cholestatic liver disease includes all of the following **except**:
   A. Recommend infant formulas that contain 40–60% of the fat from medium chain triglycerides (MCT).
   B. Provide nutrition education regarding foods high in linoleic acid.
   C. Ensure that infants receive less than 3% of calories from linoleic acid.
   D. Ensure that additional sources of MCT are added gradually.

8. What nutrient deficiencies may be found in a child with Short Bowel Syndrome. Why?

9. Which of the following statements are true?
   A. Nutrition screening in pediatric patients should be consistent with clinical malnutrition risk screening as defined by the Academy of Nutrition and Dietetics and American Society for Parenteral and Enteral Nutrition.
   B. Information gathered during the nutrition assessment process allows for the determination of the nutrition diagnosis and appropriate intervention.
   C. Ongoing monitoring and evaluation is essential to achieve optimal health outcomes.
   D. All of the above

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