This article develops an approach to nutrition and gastrointestinal (GI) problems in children who have disabilities that is geared to the primary care practitioner. Practical day-to-day issues of the patient and caregiver are emphasized. The general considerations of this article are the requirements and assessments for nutritional well-being, how to meet these needs, and delineate factors responsible for malnutrition and poor growth in children who have disabilities. The specific considerations are the GI problems common in children who have disabilities, with emphasis on management and some recent developments.

GENERAL CONSIDERATIONS

All children and adolescents have caloric requirements necessary for metabolism, growth, activity, fecal losses, and specific dynamic action of protein. The basic building blocks that provide these calories are protein, including essential amino acids (approximately 4 cal/g); fat, including essential fatty acids (approximately 9 cal/g); and carbohydrate (approximately 4 cal/g). In addition, it is important to consider water, trace element, and vitamin needs. To attain normal growth, both height and weight, and health, it is necessary to meet these needs. All nutrients must be ingested, retained, processed, absorbed, metabolized, and excreted, the net result of which should produce nutritional sufficiency. Ingestion and retention are dependent upon proper mechanics

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a Michigan State University College of Human Medicine, Kalamazoo Center for Medical Studies, 1000 Oakland Drive, Kalamazoo, MI 49008, USA
b Cleveland Clinic, 9500 Euclid Avenue, Cleveland, OH 44195, USA
* Corresponding author.
E-mail address: feinberg@kcms.msu.edu (A.N. Feinberg).
of sucking and swallowing and normal esophageal peristalsis and valve function with input from the autonomic nervous system to the enteric nervous system.

As children and adolescents are all works in progress, there are changes over time in metabolic, activity, and growth needs in all of them, disabled or not. It is impossible to establish a single simple formula or equation to calculate all dietary needs. This article presents a few rules of thumb, mainly to establish a starting point for achieving nutritional goals with the understanding that monitoring is crucial and that the prescribing physician or dietician should be prepared to make several modifications to suit the needs of the individual patient. In individuals without disabilities, a rule of thumb for caloric needs is: 1000 kcal at 10 kg, from 10 to 20 kg; add 50 cal/kg up to 20 kg. After 20 kg, add 20 kcal/kg. Box 1 outlines basic nutritional needs.

The Oxford Feeding Study\(^1,2\) notes that children with neurologic impairment have inadequate caloric intake even with the change in mind-set that those who have disabilities should not be obese. Hogan\(^3\) discusses energy requirements of children and adults who have cerebral palsy. Certain conditions such as tachypnea, fever, seizures, and spasticity and ambulation will alter caloric needs. Studies assessing total energy expenditure in children who have cerebral palsy vary considerably. Krick and colleagues\(^4\) proposed a formula for energy needs of children who have cerebral palsy taking the above into account. In spite of many suggested formulas, however, the variation among studies, ranging from 1.1 to 1.6 times resting energy expenditure, precludes establishing standards for nutritional needs of handicapped patients.\(^5\) Thus it is necessary to estimate initially, not only the patient’s needs, but also how to prescribe a diet that meets these needs. Some disabled individuals will be able to thrive with a normal diet for age. Others, because of oro–motor fatigue or GI problems, may require pureed foods. In many instances, it may not be possible for the patient to meet all needs orally without some modification. It is possible to increase the caloric density of standard formula feedings. This is of importance in certain conditions such as cardiac or renal failure, where it may be necessary to limit fluid intake, yet provide adequate calories. If a patient cannot meet these needs orally with formula modification or supplements, it may be necessary to provide feedings through a naso or oro-gastric tube or a gastro- or ileostomy tube. In addition, certain patients may require specific types of protein, fat, or carbohydrate. Special modular formulas provide these needs. Table 1 presents guidelines for increasing caloric density of standard formulas, Table 2 reviews modular formulas, and Table 3 contains some formulas used for special nutrition needs. Table 4 lists a few standard nutritional products used to enhance caloric intake.\(^6\)

It is important to take measurements at every visit and follow the length and weight percentile trends. Although body mass indices (BMI) weight (in kilograms)/height (in meters squared) are helpful for normal children and adolescents, there have been no anthropometric standards established for many handicapping conditions. There are growth charts for conditions such as Down syndrome, prematurity, meningomyelocele, sickle cell anemia, Turner, and other genetic syndromes.\(^7\) In patients who have contractures or scoliosis, some investigators have proposed formulas to calculate ideal length based on tibial length. An example of such is:

\[
\text{Tibial length (from medial popliteal line to bottom of medial malleolus)} \times 3.26 + 30.8 \text{ cm} = \text{ideal stature.}
\]

Similarly, it is difficult to assess true height in a patient who has spina bifida (lower extremities may be disproportionately short), and upper arm length can be a basis for growth standards.\(^7\)

To assess nutritional status, one may use another rule of thumb that defines mild malnutrition as less than 90% of ideal weight, moderate malnutrition as less than
### Box 1
**Vitamin and mineral needs**

#### Kilocalories (water)\(^a\)
- 0 to 10 kg—100 cc/kg/day Based on 1 kcal metabolized produces 1 cc
- 11 to 20 kg—1000 cc +50 cc/kg>10 kg H\(_2\)O
- 20+ kg—1500 cc+ 20 cc/kg>20 kg

#### Protein—10% to 15% of calories—breast milk 10%, formulas 20%; premature infants and disabled children approximately 20%; need essential amino acids

#### Fat\(^b\)
- 50% of calories age 0 to 2
- 30% of calories after age 2

#### Carbohydrates—50% of calories—major energy source

#### Vitamins\(^c,d\)
- A: 400 \(\mu\)g/d infants & children, up to 900 \(\mu\)g/d teens
- D: 5 \(\mu\)g (200 IU)/d
- E: 5 mg/d infants, 7 mg/d children, up to 15 mg/d teens
- K: 2.5 \(\mu\)g/d infants, 30-55 \(\mu\)g/d children, up to 75 \(\mu\)g/d teens
- B\(_1\) (Thiamine) 0.3 mg/d infants, 0.6 mg/day children, up to 1.2 mg/day teens
- B\(_2\) (riboflavin) 0.4 mg/d infants, 0.6 mg/d children, up to 1.3 mg/d teens
- B\(_6\) (Pyridoxine) 0.3 mg/d infants, 0.6 mg/d children, up to 1.3 mg/d teens
- B\(_{12}\) 0.5 \(\mu\)g/d infants, 1.2 \(\mu\)g/d children, up to 2.4 \(\mu\)g/d teens
- Niacin: 4 mg/d infants, 8 mg/d children, up to 16 mg/d teens
- Folate: 80 \(\mu\)g/d infants, 200 \(\mu\)g/d children, up to 400 \(\mu\)g/d teens
- Pantothentic acid: 1.8 mg/d infants, 3 mg/d children, 5 mg/d teens

#### Trace elements\(^e\)
- Na\(^+\): 2 to 4 mEq/kg/d, K\(^+\) 2 to 3 mEq/kg/d
- Calcium: 270 mg/d infants, up to 800 mg/d children, up to 1300 mg/d teens
- Fe: 11 mg/d infants, 10 mg/d children, 11 mg/d \(\geq\) teens, 15 mg/d \(\geq\) teens
- Mg\(^2+\) 75 mg/d infants, up to 130 mg/d children, up to 400 mg/d teens
- PO\(_4\)\(^-\) 275 mg/d infants, 500 mg/d children, 1250 mg/d teens
- Others include: chromium, copper, fluoride, iodine, manganese, molybdenum, selenium, zinc

**Abbreviations:** LC-PUFA, long chain polyunsaturated fatty acids; MCT, medium chain triglycerides; RDA, recommended dietary allowance.

\(^a\) This is a starting point but must be modified to individual needs. See text for varying needs.

\(^b\) Need essential LC-PUFA (linolenic, \(\alpha\) linoleic). MCT absorbed directly.

\(^c\) Vitamins A, D, E, and K are fat-soluble.

\(^d\) B vitamins are water-soluble.

\(^e\) RDA for all elements.

80% of ideal weight, and severe malnutrition as less than 70% of ideal weight, but as with BMI, there are no established anthropometric standards for many handicapping conditions. Supplemental measurements of midarm diameter or triceps or subscapular skin fold thickness are helpful to assess nutrition, taking into account both adipose and muscle tissue. Although there are no standards developed for bone density in

| Table 1 |
| Formulas: how to increase caloric content of existing formulas |

<table>
<thead>
<tr>
<th>Powder add water to make</th>
<th>4 scoops (tablespoons) 8 oz.</th>
<th>4.5 scoops 8 oz</th>
<th>4.75 scoops 8 oz</th>
<th>5.25 scoops 8 oz</th>
</tr>
</thead>
<tbody>
<tr>
<td>Powder add water</td>
<td>1 cup (level) 29 oz</td>
<td>1 cup 26 oz</td>
<td>1 cup 24 oz</td>
<td>1 cup 21.5 oz</td>
</tr>
<tr>
<td>Concentrate add water</td>
<td>13 oz</td>
<td>13 oz</td>
<td>13 oz</td>
<td>13 oz</td>
</tr>
<tr>
<td></td>
<td>13 oz</td>
<td>10½ oz</td>
<td>8½ oz</td>
<td>7 oz</td>
</tr>
<tr>
<td>End result</td>
<td>20 kcal/oz</td>
<td>22 kcal/oz</td>
<td>24 kcal/oz</td>
<td>26 kcal/oz</td>
</tr>
</tbody>
</table>

For each column, refer to the bottom entry in the same column for the caloric content.


| Table 2 |
| Modular components of formulas |

<table>
<thead>
<tr>
<th>Ingredient</th>
<th>Source</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Carbohydrate (cal/g)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lactose (4)</td>
<td>Cow milk</td>
<td>Standard formulas</td>
</tr>
<tr>
<td>Dextrose (3.4)</td>
<td>Corn sugar</td>
<td>Increases osmolality</td>
</tr>
<tr>
<td>Fru (4)</td>
<td>Fruit sugar</td>
<td>Increases osmolality</td>
</tr>
<tr>
<td>Fru + Glu (3)</td>
<td>Honey</td>
<td>Botulism in children &lt; 1 year old</td>
</tr>
<tr>
<td>Corn syrup</td>
<td>Corn</td>
<td>Patient may be allergic to corn products</td>
</tr>
<tr>
<td>Karo syrup (2)</td>
<td>Light corn syrup</td>
<td>Also contains vanilla and salt</td>
</tr>
<tr>
<td>Glu polymer (3.8)</td>
<td>Corn</td>
<td>Lowers osmolality</td>
</tr>
<tr>
<td>Sucrose (4)</td>
<td>Cane or beet sugar</td>
<td>Increases osmolality when broken down</td>
</tr>
<tr>
<td>Corn starch</td>
<td>Corn and maltodextrin</td>
<td>Food thickener; useful in patients with glycogen storage disease</td>
</tr>
<tr>
<td>Fat</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Corn oil (8.8)</td>
<td>Corn</td>
<td>High in linoleic and linolenic acid</td>
</tr>
<tr>
<td>MCT oil (8.3)</td>
<td>Coconut oil</td>
<td>Absorbed directly in portal system; mainly triglycerides of C8-C10 fatty acids</td>
</tr>
<tr>
<td>Safflower oil (8.8)</td>
<td>Safflower</td>
<td>68% linoleic acid; increases total caloric content of feeding</td>
</tr>
<tr>
<td>Protein</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Casein + whey (3.8)</td>
<td>Cow milk</td>
<td>Standard formulas</td>
</tr>
<tr>
<td>Casec Ca caseinate</td>
<td></td>
<td>Protein supplement</td>
</tr>
<tr>
<td>Resource Beneprotein</td>
<td>Whey protein isolate</td>
<td>Protein supplement, easier-to-digest whey; soy proteins cross-react in 50% of patients with milk allergy</td>
</tr>
</tbody>
</table>

Abbreviations: Fru, Fructose; Glu, Glucose; MCT, Medium chain triglycerides.

<table>
<thead>
<tr>
<th>Product</th>
<th>(Osmolality) Content</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Soy formulas (lactose-free)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Good Start Supreme Soy DHA and ARA</td>
<td>(180) CHO: sucrose, corn, maltodextrins; protein: hydrolyzed soy; fat: soy, palm olein, coconut, safflower, and sunflower</td>
<td>Cross-reaction with cow’s milk in 50% of patients with milk allergy</td>
</tr>
<tr>
<td>Isomil</td>
<td>(200) CHO: corn syrup, sucrose; protein: soy; fat: palm olein, soy, coconut, sunflower</td>
<td>Carnitine added + see above</td>
</tr>
<tr>
<td>Prosobee</td>
<td>(200) CHO: corn syrup; protein: soy; fat: soy oil, coconut, sunflower</td>
<td>See above + sucrose and lactose-free</td>
</tr>
<tr>
<td>Other formulas for special nutritional needs</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Alimentum Advance</td>
<td>(370) CHO: sucrose, tapioca; protein: casein hydrolysate; fat: MCT, soy oils, DHA, and ARA</td>
<td>Hypoallergenic, enhances fat absorption</td>
</tr>
<tr>
<td>Neocate Infant</td>
<td>(375) CHO: corn syrup; protein: L amino acids; fat: safflower, coconut and soy oils, 5% MCT</td>
<td>Hypoallergenic</td>
</tr>
<tr>
<td>Nutramigen LIPIL</td>
<td>(375) CHO: corn syrup, corn starch; protein: L amino acids, casein hydrolysate; fat: palm olein, soy coconut and safflower oils (DHA &amp; ARA)</td>
<td>Hypoallergenic</td>
</tr>
<tr>
<td>Pregestamil LIPIL</td>
<td>(330) CHO: corn syrup, corn starch, dextrose; protein: casein hydrolysate, L amino acids; fat: MCT, soy, safflower, and corn oil</td>
<td>Hypoallergenic and for fat malabsorption</td>
</tr>
<tr>
<td>Peptamin Junior</td>
<td>(260, unflavored, 380-400 flavored) CHO: maltodextrin, corn starch; protein: hydrolyzed whey; fat: MCT (60%), soy, and canola oils</td>
<td>Elemental protein formula, for milk allergy and impaired gastrointestinal function</td>
</tr>
<tr>
<td>Similac PM 60/40</td>
<td>(280) CHO: lactose; protein: whey, Na caseinate; fat: corn, coconut, and soy oils</td>
<td>For hypercalcemic infants, low renal solute load, Ca:PO₄ like human milk; Whey:casein ratio is 60:40</td>
</tr>
<tr>
<td>Vivonex</td>
<td>(360) CHO: maltodextrin, modified starch; protein: L amino acids; fat: MCT (68%) and soy oils</td>
<td>MCTs good for fat malabsorption</td>
</tr>
</tbody>
</table>

*Abbreviations: ARA, Arachidonic acid; CHO, Carbohydrate; DHA, Decahexanoic acid; MCT, Medium chain triglycerides.
children or adolescents, Chad and colleagues\textsuperscript{8} determined that osteopenia is worse in nonambulatory children who have spastic cerebral palsy. Duncan and colleagues\textsuperscript{9} determined that poor intake played a significant role in low bone density.

Box 2 lists laboratory and imaging studies helpful to assess appropriate nutritional status.

Cohen and Navathe\textsuperscript{10} in 1999 delineated several factors why children and adults may not meet their nutritional needs (Table 5). The following discussion incorporates the concepts in Table 5 as an aid to diagnosis in handicapped children and adolescents. It is important to recognize that all disabled patients are subject to the same medical conditions as their normally developing peers. Discussing all diagnoses goes well beyond the scope of this article. The following is a framework to classify conditions and provide some scenarios that affect nutritional status of those with disabilities. It is important to approach diagnosis in the context of disability, but, be prepared to consider other diagnoses, some of which may not be of gastrointestinal origin.

**Congenital/Genetic/Metabolic Derangements**

These will lead to malnutrition a priori. For a few examples, chromosomal disorders such as trisomies and gene deletions are associated with short stature.
regardless of the patient’s weight. Congenital heart disease with failure may lead to caloric deprivation for many reasons, including fatigue from feeding and increased metabolic demand secondary to tachypnea and tachycardia. Inborn errors of metabolism will cause vomiting, with diminished caloric intake, acidosis, and hence poor growth. Cystic fibrosis and celiac disease will cause length and weight problems due to malabsorption. Endocrine problems such as hypothyroidism, Cushing’s disease, hypopituitarism, and rickets are associated with

<table>
<thead>
<tr>
<th>Box 2</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Laboratory and imaging parameters to assess nutritional status</strong></td>
</tr>
</tbody>
</table>

**Laboratory**
- Hemoglobin/indices—assess iron sufficiency
- White blood cell count, diff, immunoglobulins—assess immune status
- Isohemagglutinins, *Candida* skin test
- Serum urea nitrogen, creatinine—assess renal function, protein sufficiency, hydration
- Electrolytes—assess hydration, acid-base balance
- Ca⁺⁺, PO₄⁻³, alkaline phosphatase—assess calcium sufficiency and bone health
- Albumin, prealbumin—assess protein sufficiency
- Drug levels (eg, anticonvulsant)—assess for adverse effects, toxicity, or compliance

**Imaging**
- Radiograph of long bone—assess for osteomalacia, osteoporosis
- Radiograph of wrist for bone age—assess for endocrine disorders and progress
- Dexascans—no established standards for children

<table>
<thead>
<tr>
<th>Table 5</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Etiologies of feeding disorders and undernutrition for children and adults with developmental disabilities</strong></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Historical perspectives</th>
<th>Genetic endowment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Underlying illness</td>
<td>neurodevelopmental status</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Current state</th>
<th>Oral motor competence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intercurrent illness</td>
<td>Behavioral resistance</td>
</tr>
<tr>
<td>food–medication interaction</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Nutriture</th>
<th>Caregiver involvement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Food availability</td>
<td>Nutrient adequacy</td>
</tr>
<tr>
<td>Appetite/alternative sources</td>
<td>Energy requirements</td>
</tr>
<tr>
<td>Activities, expenditures, losses</td>
<td>iatrogenic</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Metabolic adaptation</th>
<th>Digestion, absorption</th>
</tr>
</thead>
<tbody>
<tr>
<td>Uptake, use</td>
<td>Storage, excretion</td>
</tr>
<tr>
<td>hormonal homeostasis</td>
<td></td>
</tr>
</tbody>
</table>

short stature irrespective of weight. Hyperthyroidism, on the other hand, leads to poor weight gain from a hypermetabolic state. Anatomic malformations of the GI system including oral, such as cleft palate, esophageal webs, fistulae, bowel atresias, malrotations, and many others will limit intake because of vomiting. Even after corrective surgery for these conditions, there may be residual symptoms. Congenital central nervous system conditions will affect oral intake throughout life. Spasticity consistently will increase the metabolic rate, thus raising maintenance caloric needs.

**Traumatic/Hypoxic**

Oral motor dysfunction occurs in patients who have traumatic brain injuries whether occurring prenatally, at birth, or later. Injury to the GI tract, acquired accidentally or iatrogenically (surgery, radiation) will affect nutritional adequacy. In these patients, there is significant vomiting, choking, and regurgitation.

**Infection, Inflammation, Malignancy**

Intercurrent illness plays a significant role when growth and weight gain are marginal in the baseline state. Specifically, intercurrent infection may cause poor intake because of lethargy and vomiting, or may produce a hypermetabolic state because of fever. The immunocompromised are more subject to any GI infection, and it is important to consider opportunistic infections such as gram-negative, staphylococcal, and fungal infections, and parasitic infestations. Patients who have disabilities may convalesce more slowly because of their hypostatic state and abilities to rid themselves of respiratory secretions. They also may be more subject to urinary tract infection because of a neurogenic bladder associated with central nervous system (CNS) and spinal conditions, or because of congenital urologic anomalies. Seizures also will increase caloric requirements. Other intercurrent conditions include inflammatory conditions such as collagen vascular disease, inflammatory bowel disease, malignancy, and others.

**Psychosocial**

Good nutrition depends upon the presence and availability of proper food and the ability of the patient to ingest and retain it with or without external help. Patients may develop secondary food aversions, because they associate the unpleasant sensations of pain, nausea, vomiting, or instrumental manipulation with feeding. Feeding evokes some of the deepest emotions between parent and child, and variations from the norm may promote significant dysfunction in this extremely sensitive parent–child dyad. Feeding often is prolonged and fraught with frustration when there is associated vomiting, crying, and food refusal. Hunt discusses several psychosocial aspects of changing from oral to enteral feeding.

**Iatrogenic**

The composition of nutrients should meet the patient’s individual metabolic and growth needs. This is contingent upon proper content, monitoring that the caregiver is following the regimen correctly. It is important to consider that medications may have a profound effect on nutritional status. Some examples are:

- Anticonvulsants, which may decrease vitamin D levels
- Diuretics, which may cause increased excretion of electrolytes
- Mineral oil, which may interfere with absorption of fat-soluble vitamins
- Stimulants, which may cause decreased appetite and growth
Corticosteroids, which may cause increased appetite, fluid retention, and hyperglycemia. Commonly-used interventions such as total parenteral nutrition (TPN) are not without significant complication. It is important to monitor these patients for infection and specific metabolic derangements such as abnormal glucose, electrolyte, calcium, phosphorus, magnesium, and trace element levels. TPN over time is associated with hepatotoxicity and cholestasis requiring close monitoring. Increased fat content of parenteral feedings may be associated with emboli to the lungs. Complications and malfunctions from procedures such as TPN or percutaneous gastrostomy (PEG) tubes may prevent nutrients from reaching the patient.

**SPECIFIC CONSIDERATIONS**

This section examines GI problems common in children who have disabilities. Sullivan and colleagues and Sullivan\(^1\)\(^-\)\(^12\) determined that GI problems in children who had neurologic impairment were very high. Fifty-nine percent were constipated; 22% had problems with vomiting, and 31% had respiratory complications of gastroesophageal reflux (GER). Eighty-nine percent of these children had some feeding problem. This section is limited to problems more likely to occur in special needs children either a priori, or as a complication of disability and its treatment.

**Vomiting and Regurgitation**

GER is a normal physiologic phenomenon in people. Gastroesophageal reflux disease (GERD) is by definition GER with pathologic consequences such as mucosal damage or decline in the individual’s quality of life. In the pediatric population, up to 50% of infants younger than 3 months of age display regurgitation on a daily basis. This is predominantly caused by transient lower esophageal sphincter relaxation (TLESR). This physiologic issue becomes more pronounced as 66% of infants at 4 months of age demonstrate regurgitation. By 1 year of age, only 5% have symptoms. Two basic theories exist. Increased inspiratory and expiratory effort in children who have chronic lung conditions (ie, cystic fibrosis) leads to increased negative intrathoracic pressure, which may in turn predispose the child to GER. Because the embryonic origins of the respiratory tract and esophagus are the same, they share autonomic innervation by means of the vagus nerve. Esophageal irritation may lead to airway constriction because of this common nerve conduction pathway.

Other disorders of the proximal esophagus confirmed on manometric studies occur with various neurodevelopmental disorders. An intricate sequence of muscle relaxation is necessary for effective swallowing of a bolus of food. With disruption of this sequence, vomiting and dysphagia occur. Secondary causes of disorders affecting the proximal esophagus may include meningocele, Arnold Chiari malformation, cerebrovascular accidents, polymyositis, dermatomyositis, and muscular dystrophy. Disorders of the distal esophagus include achalasia, dysmotility of the esophagus, and inability of the lower esophageal sphincter (LES) to relax. This constellation of findings can occur with several other disorders, including Hirschsprung’s disease (aganglionosis elsewhere), pyloric stenosis, Down syndrome, intestinal pseudo-obstruction, diabetes mellitus, and sarcoidosis. Triple A, or Allgrove syndrome, is associated with adrenocorticotropic hormone insensitivity, alacrima, and achalasia. Gastroparesis is associated with conditions such as type 1 diabetes mellitus, celiac disease, cystic fibrosis, thyroid hormone abnormality, and anorexia nervosa. Medication may be associated with feeding intolerance. Hyperglycemia found in type 1 diabetics may lead to
gastric hypomotility early on in the disease course. Prematurity and postviral gastroenteritis are common causes of temporary lactase deficiency and delayed gastric emptying time.

Aspiration of gastric contents is a complication found in children who have recurrent pneumonia with a history of GERD or other esophageal disorders. Bronchoscopy demonstrates lipid laden macrophages and contributes to the diagnosis of recurrent aspiration. Impedance probe testing is the diagnostic test of choice for evaluating GERD. This technology allows for testing of acidic and nonacidic fluids and gas refluxed from the stomach into the esophagus. It also provides a means to correlate symptoms with reflux based on probe detection and symptom diary on the probe device. Limitations of this study exist in the pediatric population, as there is yet to be an established standard reference range. Upper GI series are useful to evaluate for GI obstruction or malrotation. They lack adequate sensitivity and specificity for a definitive diagnosis of GERD, however. Gastric emptying studies can provide insight into gastroparesis, although they also do not provide the clinician with a definitive diagnosis regarding the etiology. Gastroenterologists employ esophageal and antral–duodenal manometry as a means to confirm dysmotility. Although endoscopic and histologic evaluations are the gold standard for esophagitis, patients still may present with symptoms of GERD with unremarkable findings.

Treatment generally consists of managing the underlying systemic ailment. Dietary modification such as reduction in high fat-containing foods and fiber are useful interventions. In severe cases, jejunostomy feedings may be necessary to bypass the stomach altogether. A gastrostomy tube may be beneficial in alleviating gastric distention by allowing venting. Medical management of GERD involves several approaches. Prokinetics are a class of drugs that helps facilitate GI motility. Metoclopramide, a dopamine receptor blocker, is a commonly used prokinetic agent. Unfortunately, CNS effects are documented complications. These adverse effects include extrapyramidal signs such as dystonic reactions. To date, there has been no definitive proof of metoclopramide’s effectiveness in treating GERD. Domperidone (not available in the United States) is a more tolerable prokinetic with less frequent dystonic reactions. Possible adverse effects include cardiac arrhythmias caused by prolonged QT interval and subsequent ventricular fibrillation. Erythromycin demonstrates some effect on gastric motility, but has not been a proven treatment for GERD. There is a reported risk for development of hypertrophic pyloric stenosis in patients taking erythromycin. Cisapride is the only prokinetic drug with statistically significant effectiveness to reduce esophageal acid exposure. The United States has banned its use because of cardiac arrhythmia and prolonged QT interval, especially in infants younger than 3 months of age. Cytochrome P4503A4 is responsible for the metabolism of cisapride. Its levels are lower in infancy, contributing to the age bias of reported adverse effects. Grapefruit juice can affect the metabolism of cisapride. Histamine-2 blockers such as famotidine and ranitidine are common treatments for GERD. Tachyphylaxis may occur after several weeks of use. It is not as effective in healing esophagitis as proton pump inhibitors (PPIs), lansoprazole, omeprazole and esomeprazole. These are prodrugs as the active component is the result of an acidic environment. They bind irreversibly to H+/K+ ATPase enzyme, which inhibits acid production. Children who have adequate acid suppression may demonstrate healing of underlying esophagitis after 4 to 8 weeks of therapy. Prolonged hypochlorhydria may lead to gastric bacterial overgrowth. Vitamin B₁₂ deficiency is another complication to acid suppression because of impaired absorption of B₁₂ from food in an acid-suppressed environment. Surgical intervention for GERD primarily consists of Nissen fundoplication (NF), which is a gastric wrap around the fundus and distal esophagus. This procedure prevents complete
relaxation of LES and reduces TLESRs. One retrospective study of more than 7000 children found good-to-excellent results in 95% of normal children and in 84.6% of children who had underlying neurologic medical issues. There is 0.07% mortality in normal children and 0.8% mortality in those children who have neurologic problems. Negative outcomes included GERD reoccurrence in 7.1% of the study population, gas bloat syndrome in 3.6% of the cases, and obstructive symptoms in 2.6% of the study cases.13

Feeding Tube Problems

Feeding Tube Problems

Malnutrition is a major medical issue for children who have neurodevelopmental delays or diseases causing increased caloric demand such as chronic cardiac or respiratory medical issues. In up to 60% of the cases, poor feeding in infancy precedes the diagnosis of cerebral palsy.14 Nasogastric tubes may supplement short-term nutritional needs (less than 2 months), or act as a trial for gastric feedings before gastrostomy tube placement. PEG is the placement of a more stable and long-term means of enteral nutritional support. The initial gastrostomy tube is replaced after approximately 8 weeks with a button device consisting of an internal balloon to maintain tube placement. Gastrostomy tubes have several risks, including infection and perforation. Perforation rates have been reported as high as 15%.15 Studies show that children who have neurologic disorders and underlying aspiration do not have a lower risk of aspiration with gastrostomy tube feeds, and that GT feedings in combination with NF only reduces the chances of aspiration.16 In general, a patient’s tolerance of enteral feedings will determine the dietary regimen. Bolus feedings over 15 to 20 minutes provide nutrition throughout the day. Night time feedings are usually continuous over several hours. Patients who have neurologic disabilities may require either transpyloric feedings (by means of gastrostomy–jejunostomy tube or direct jejunostomy tube) or an NF procedure. Patients who undergo NF may require simultaneous pyloroplasty if they have underlying delayed gastric emptying. A trocar pierces into the stomach with endoscopic observation of internal placement. There is, however, an area between the stomach and abdominal surface that is not visible during the procedure. Immediately after the PEG placement procedure, there is a critical time necessitating close observation.

Complications include perforation into an organ overlying the stomach such as the colon. Another potential for perforation is during difficult GT changes at home or in a medical office. If there is concern about proper placement of the GT, a CT scan may be helpful. In patients who have gastrostomy tubes that present with diarrhea of unknown etiology, it is important to consider GT complications. In this setting, it is possible that the GT migrated into the colon and seemingly is causing diarrhea, but rather is infusing formula directly into the large bowel. A GT injection study under fluoroscopy can make this diagnosis. Another major concern is dislodgment of the GT either by rupture of the internal balloon or by trauma. It is imperative to maintain stoma patency, as it takes relatively very little time for onset of closure (hours). In a medical setting, it is appropriate to insert a catheter of comparable or slightly smaller size into the stoma until performing definitive replacement of the GT. In cases where the stoma already has begun to close, serial dilatation with caution may be helpful. GT fluoroscopy is then necessary to confirm appropriate placement of the new GT. The occurrence of granulation tissue is common in post-GT placement. Chemical cauterization is useful to keep surrounding skin dry, so as to not cause burns outside of the granulation tissue. Topical steroids are also helpful. In refractory situations, surgical consultation for resection of the granulation tissue may be necessary. GT occlusion can make enteral feedings difficult to impossible depending on the degree. Stagnant
formula or undissolved medications are the usual sources of occlusion. A common
treatment of GT occlusion involves water flushes (up to 30 cc). Occasionally, Viokase
mixed with bicarbonate may help to dissolve the obstructing agent. As a rule, infuse no
more than 5 cc of water into the balloon port of the button gastrostomy tube. More
than that volume can cause rupture of the balloon, or ischemia of the stomach mucosa
because of the pressure of the balloon against the lining of the gastric tissue.

**Constipation**

Constipation is an exceedingly common problem in healthy children and is even more
prevalent in those who have neurologic impairments. It is unclear why this subgroup of
patients has this problem. A large percentage of these patients is not mobile, and this
lack of ability to ambulate may contribute to delayed transit through the GI tract. De-
creased truncal tone and inability to effectively generate adequate intra-abdominal
pressures can play a role in ineffective evacuation also. Oral motor incoordination
leads to inadequate intake and thus decreases stool bulk. Inadequate fluid intake
also may cause harder stools with difficult passage. Additionally, the diet of these
patients is often lacking in fiber.17

When first seeing a patient for constipation, a complete history is paramount. This
includes size, frequency, and consistency of stools. Ask the patient or caregiver about
pain with passage of stools, which may lead to withholding behaviors. Also, address
behaviors and postures during defecation. Typical withholding behaviors include
arching, scissoring/straightening of the legs, standing on tiptoes, or hiding. Care
givers should know what the patient’s normal stool pattern was, and at what age
the patient first developed constipation. Constipation during infancy is more indicative
of underlying pathology. Delayed passage of meconium specifically raises concerns
about Hirschsprung’s disease and cystic fibrosis. Review dietary history also, with
an emphasis on fiber and fluid intake. Growth charts should be available, looking for
failure to thrive and decreased growth velocity. Note prior treatments and whether
they were effective. A comprehensive list of medications in the history is important,
as several medications (particularly antiepileptic drugs and medications used to treat
spasticity) cause constipation. The presence of bright red blood with bowel move-
ments that are large and hard is indicative of a fissure, and this may appear on physical
examination.

The physical examination of the constipated patient begins with the general appear-
ance of the patient. Is he or she well nourished, and is his or her size appropriate to
age? Is there evidence of abdominal distention? Document height and weight and
plot on the appropriate growth chart. Special charts for specific syndromes are avail-
able and helpful. Inspect the patient’s abdomen again looking for distention, poor
muscle tone, or compromised abdominal musculature that could compromise the
ability to generate increased intra-abdominal pressures. Auscultate bowel sounds.
Hypoactive bowel sounds may be indicative of slow transit constipation. High pitched
or tinkling bowel sounds can be heard in the context of a small bowel obstruction.
Carefully palpate the abdomen assessing for masses (fecal or otherwise). A rectal ex-
amination is essential. Inspect the external anus visually, looking specifically at place-
ment. There may be evidence of encopresis on the examination in the toilet trained
child. Fissures may be present in the chronically constipated patient, as well as skin
tags. Assess anal wink by lightly stroking the perianal area in all four quadrants. The
digital examination will help to evaluate the tone of the anal sphincter and reliably de-
termine whether the patient has a fecal impaction. Visualize the spine and palpate with
attention to the sacral region. The presence of midline pigment lesions or hair tufts
points to potential spinal abnormalities. Sacral dimples can be associated with
tethered cord. Deep tendon reflexes in the lower extremities and sensation should be assessed if possible. In the ambulatory patient, always evaluate gait.

Laboratory evaluation of the constipated patient is fairly minimal. Screening for hypothyroidism is appropriate. If the patient has a history that includes failure to thrive, celiac titers may be in order. Consider other laboratory tests including sweat chloride testing. If there is a high index of suspicion for Hirschsprung’s disease in a constipated child who is not growing well, rectal suction biopsy looking for absence of ganglion cells is necessary for definitive diagnosis. History and physical examination should guide imaging choices. A plain radiograph of kidney, ureter and bladder (KUB) with upright may be useful in assessing the degree of fecal retention, especially if there is a limited physical examination because of the patient’s body habitus or inability to cooperate. Barium enema in infants may help in the evaluation of Hirschsprung’s disease, although this is not as sensitive or specific as rectal suction biopsy. If an abdominal mass is present and not thought to be caused by feces, ultrasound or CT may help to clarify its etiology.

Treatment of constipation in the child who has medical disabilities is not significantly different than treatment in the otherwise healthy patient. If a fecal impaction is present, then address this first, either with enema or with high-dose polyethylene glycol; in some patients, maintenance with polyethylene glycol is adequate. Other medications that may be used include lactulose, magnesium hydroxide, mineral oil, and stimulants such as senna or bisacodyl. A guideline was published recently in the *Journal of Pediatric Gastroenterology and Nutrition*, outlining the different medications and their appropriate dosing. Of note, patients who have spinal cord injury or spinal cord defects often fail medical management alone. In this subset of patients, enema regimens or transanal irrigation systems may be beneficial in maintaining regular stooling habits and daytime continence. Other options for treatment include surgical placement of a cecostomy or colostomy for anterograde enemas, typically done if medical management fails to provide adequate relief of a patient’s constipation.

**Hepatobiliary System**

Jaundice and hepatitis in the patient who has underlying disabilities may occur for a myriad of reasons. Often the cause is difficult to determine, in spite of careful history taking. It is important to consider the patient’s underlying medical conditions, as often they involve the liver. Family history is vital and should not be limited to liver disease. Many syndromes, such as Alagille’s syndrome, have variable presentation, and affected family members may have no hepatic involvement whatsoever. Review travel history with emphasis on travel to endemic areas for infectious etiologies.

Physical examination should include general survey of the patient. Note dysmorphic features associated with specific syndromes. If family members are available, examine them also. Monitor vital signs closely, as jaundice and hepatitis may be the presenting complaints of sepsis. Obtain neurologic examinations at baseline and follow-up, particularly if fulminant hepatitis is a concern. Mental status changes are often difficult to determine in the developmentally delayed patient, and careful monitoring for deterioration is the best means of assessing for hepatic encephalopathy. Abdominal examination may show hepatosplenomegaly or ascites. Examine the abdominal wall looking for dilated venous patterns that are concomitant with portal hypertension. When examining the skin, the presence of excoriation indicates pruritus secondary to cholestasis. Rashes and their distribution may help in determining viral etiologies for liver disease.

Consider infectious causes of hepatitis. Hepatitis A is more common in residential centers for the disabled. Hepatitis B is decreasing in incidence, due predominantly
to early vaccination. Hepatitis C still was transmitted via the blood supply as late as 1989, when serologic screening tools became available.\textsuperscript{22} Other viruses that may cause hepatitis include Epstein Barr virus, cytomegalovirus, herpes simplex virus, coxsackievirus, adenovirus, enterovirus, and varicella zoster. History of outbreaks, either in the community or in the environment in which the patient resides, may help in diagnosing infectious etiologies. Symptoms of systemic illness including rashes typical of certain viral illnesses may provide clues to specific causes. Serologic testing may be helpful in confirming diagnosis. Treatment for acute viral hepatitis consists of supportive care. Treatment of chronic viral hepatitis requires subspecialist management.

Some hereditary/metabolic conditions present with liver disease as part of the underlying medical problem itself. These include, but are not limited to: Crigler-Najjar syndrome, cystic fibrosis, alpha 1-antitrypsin deficiency, disorders of fatty acid oxidation, tyrosinemia, galactosemia, urea cycle defects, and glycogen storage diseases. Patients who have sickle cell disease or other hemoglobinopathies will present with hyperbilirubinemia caused by hemolysis. Other disorders that may present as hepatic dysfunction include hemachromatosis, Wilson’s disease, and hereditary fructose intolerance. Treatment is highly specific to each underlying disorder and is not within the scope of this article.

Laboratory evaluation of patients who have liver disease should include transaminases, a marker for hepatocellular damage. It is important to recall that damage to other tissues including muscle and gut mucosa also may cause elevated aspartate aminotransferase (AST) and alanine aminotransferase (ALT). Other markers of hepatobiliary damage include alkaline phosphatase, which is elevated in cholestatic liver disease. Bone is another source of this marker, and it is the heat-stable fraction that is biliary in origin. Gamma glutamyl transferase is a more specific marker for bile duct injury. Obtain fractionated bilirubin and follow it as a marker of hepatic function. It is vital to determine whether it is conjugated or unconjugated bilirubin that is elevated, as there are significantly different diagnoses that each of these will lead to. Other laboratory values that are useful in assessing synthetic function of the liver include coagulation studies and specific clotting factor levels such as II, V, VII, and XI.\textsuperscript{23} Low albumin is also a marker of impaired liver function, but as the half life is much longer, it is a more accurate reflection of chronic, rather than acute disease.

Viral antibody titers may be helpful in determining the etiology of infectious hepatitis. Serum ceruloplasmin and 24 hour urine copper may aid in diagnosing Wilson’s disease. Normal iron studies help to rule out hemochromatosis. Hereditary fructose intolerance will yield abnormal tissue enzyme assays, specifically deficiency of aldolase B. Autoimmune markers such as ANA, anti-LKM, and anti-SM are part of the evaluation for autoimmune hepatitis. Obtain complete blood count with differential, blood culture, and urine culture to rule out sepsis, and if the index of suspicion is high, start antibiotics immediately. Liver biopsy is also useful in determining etiology of hepatobiliary disease, and always requires subspecialist consultation for indication, procedure, and interpretation.

The physician should not forget iatrogenic causes of liver disease. Medications are a common cause of cholestasis and hepatitis. Drug toxicity may be dose-related, as in acetaminophen, or may be idiosyncratic. Many herbal remedies or supplements that are allegedly safe can have deleterious effects on the liver. Review carefully the patient’s medication list, including any over-the-counter medications or supplements. Pay particular attention to dosing and the timing of any new or old medications relative to the onset of symptoms. Stopping the causative agent typically leads to resolution of symptoms. It is important to consult the physician who prescribed the medication, as
abrupt withdrawal of some medications may have serious consequences. In fulminant hepatic failure, supportive care is essential. Discontinue all identified hepatotoxins. In some instances, antidotes are appropriate. Specifically, if acetaminophen toxicity is the cause, start N-acetyl cysteamine and continue it until drug levels are no longer in the toxic range.

Parenteral nutrition (PN) is the intravenous administration of nutritional support. Common indications for chronic PN include gut failure, short gut, necrotizing enterocolitis, severe GI dysmotility, and refractory inflammatory bowel disease. PN also may be helpful in the short term, if a patient is not able to take nutrition through the gut. Such instances include postoperative care or insults to the mucosa that should heal or resolve relatively quickly. Hepatobiliary complications are associated with PN. The onset may be as early as 2 weeks after the start of therapy. The exact incidence is unclear, but at-risk populations include premature infants who have necrotizing enterocolitis, short gut, those with and prolonged periods without enteral feeds. Children on chronic PN develop cholestasis, cirrhosis, and portal hypertension. These patients are at risk for gallstones or biliary sludge formation. Ursodeoxycholic acid may mitigate the cholestasis, but there is little literature to support its use. Prevention of PN-associated liver disease includes avoiding overfeeding, windowing the infusions, and instituting enteral feeds as able.

REFERENCES