Question: 1

You are evaluating a 6-month-old child who has a ventricular septal defect and is scheduled for cardiac surgery. The child's weight is 6 kg (3rd percentile), length is at the 30th percentile, and head circumference is at the 50th percentile. His mother states she prepares the formula by adding 1 scoop of powder to 2 oz of water. She estimates that he drinks 24 oz of formula per day. You estimate the baby's intake is approximately 500 kcal per day of cow milk formula, which is the recommended dietary allowance (RDA) for his age. According to his mother, he spits up three times a day and passes two soft stools daily. On physical examination, you hear a 3/6 holosystolic murmur and palpate the liver 1 cm below the right costal margin.

Of the following, the BEST explanation for the child's malnutrition is

A. caloric requirements exceeding the RDA

B. cow milk protein intolerance

C. incorrect preparation of the formula

D. pathologic gastroesophageal reflux

E. undiagnosed pancreatic insufficiency

Critique: 1 Preferred Response: A

Children who have large ventricular septal defects, such as described for the child in the vignette, have increased pulmonary blood flow and may have ventricular hypertrophy and heart failure. Because their hearts have to work harder, their caloric needs often are increased above the recommended dietary allowance (RDA) for healthy children. The absence of significant vomiting, diarrhea, or rectal bleeding suggests that the patient does not have significant gastroesophageal reflux, cow milk protein intolerance, or pancreatic insufficiency. Incorrect preparation of the formula always should be considered, but the mother's reported preparation is correct.

The RDA is defined as "a nutrient intake level that is ... sufficient to meet the nutrient requirements of 97% of healthy individuals" in a given group, categorized by sex and age. RDAs are developed by the Food and Nutrition Board of the Institute of Medicine in collaboration with Health Canada. A full list of RDAs may be found in many reference textbooks and at the Food and Nutrition Information Center on the United States Department of Agriculture web site (http://fnic.nal.usda.gov). RDAs estimate energy requirements of healthy children and do not adjust for chronic illness states that increase caloric needs, such as cystic fibrosis or congenital heart disease. Children who have chronic illnesses often require more calories than the RDA to grow and develop.

The estimated daily caloric requirement of a patient is the aggregate of the patient's basal metabolic rate and physical activity. Various mathematic equations can be used to estimate the daily caloric intake of children and adults of different ages. Such equations usually take into account the individual's age, sex, physical activity level, and either the weight and height or, preferably, the body surface area. For children who have chronic illnesses, more accurate estimates of daily caloric requirements can be obtained by using a laboratory-based technique, such as indirect calorimetry.

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Question: 3

You are seeing a 1-month-old girl for follow-up after a hospitalization for acute gastroenteritis caused by rotavirus. Her diarrhea had decreased in the hospital while taking oral rehydration solution, but when her mother resumed her usual cow milk formula, the girl began to have an increased number of very watery stools. She appears well hydrated, and findings on her abdominal examination are normal.

Of the following, the MOST appropriate approach to managing this infant's diarrhea is to

A. change to a lactose-free formula for the next few days

B. dilute the cow milk formula with oral rehydration solution for the next few days

C. give her only oral rehydration solution until the diarrhea resolves

D. readmit her to the hospital for administration of intravenous fluids

E. repeat her stool studies to confirm the diagnosis of rotavirus infection

Critique: 3 Preferred Response: A

The infant described in the vignette most likely has lactase deficiency due to rotavirus infection. Lactase is an enzyme found in the most superficial villous portion of the intestinal brush border, which hydrolyzes lactose to glucose and galactose. Lactase deficiency may have several causes in children and adults. Primary lactase deficiency, the most common type, is a genetically determined condition that affects children and adults at different ages but is unusual before 5 years. Symptoms include abdominal distention, bloating, flatulence, or nausea after the ingestion of lactose, with the amount of lactose needed to cause such symptoms varying from person to person. The diagnosis is made by breath hydrogen testing, and management consists of removing some or all lactose from the diet. Congenital lactase deficiency is extremely rare.

Secondary lactase deficiency may develop after an infectious gastroenteritis, such as rotavirus, giardiasis, or cryptosporidiosis. Other causes include celiac disease and enteropathy related to immunodeficiency. Secondary lactase deficiency is suggested when a child who has a recent diarrheal illness experiences worsening diarrhea or bloating after the reintroduction of lactose into the diet, as described for the girl in the vignette. Most children who have gastroenteritis do not develop lactase deficiency. For this reason, most infants can tolerate and should continue taking human milk or standard lactose-containing formula throughout a diarrheal illness. For very young infants (eg, <3 months old), such as the one described in the vignette, or those who have significant fluid losses, a lactose-free formula may be attempted until the diarrhea resolves. Infants who are breastfed should be encouraged to continue breastfeeding, even if secondary lactase deficiency is suspected.

Giving full-strength formula or human milk is recommended to supply the child with sufficient calories during the recovery phase of a diarrheal illness; therefore, diluting the formula or providing only oral rehydration solution is inappropriate. If the child is not vomiting, oral hydration is optimal, and intravenous hydration is not necessary. There is no need to confirm the diagnosis of rotavirus infection; doing so would not alter management plans.

References:

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Question: 4

During a prenatal visit with expectant parents, they report that they are strict vegans. They ask you to advise them on a healthy diet and any required supplements. The mother plans to breastfeed the newborn exclusively for the first 6 months.

Of the following, you are MOST likely to tell them that their newborn may require supplemental

A. calcium

B. folate

C. iron

D. vitamin B6

E. vitamin B12

Critique: 4 Preferred Response: E

A vegan diet, by definition, excludes all foods derived from animal products. A lacto-ovo-vegetarian diet may include milk and eggs. Although a vegan diet may be healthy, there is a risk for vitamin B12 deficiency because vitamin B12 is only found in foods of animal origin. Breastfeeding vegan mothers may produce milk that is deficient in this vitamin and require supplementation that generally is achieved by continuing the consumption of prenatal vitamins containing vitamin B12.

The recommended supplementation for breastfed vegan infants to prevent vitamin B12 deficiency is 0.4 mcg/day during the first 6 postnatal months and 0.5 mcg/day from 6 months to 1 year of age. Vegan infants who are not breastfed should receive iron-fortified soy infant formula until 1 year of age to avoid deficiencies in iron. Vegan infants require no other mineral or vitamin supplementation.

Vegan diets in older children and adolescents may be low in calcium (similar to the typical American "teenage diet" that contains less than the recommended intake of dairy products), and the zinc consumption may be relatively low due to the absence of phytate, which renders zinc more bioavailable. Children who follow vegan diets may have relatively diminished overall energy intake because such diets commonly are low in fat and high in fiber.

Review of nutrient intake and energy intake in conjunction with growth curves of children eating vegan diets in both the United States and the United Kingdom demonstrate no significant health issues. Height and weight measured in vegan populations may be slightly lower than average but not in the range of failure to thrive or short stature. Adolescents eating vegan diets are more likely than adolescents eating a typical American diet to meet nutritional goals, including recommended intake of fruits and vegetables. Vegan adolescents are less likely to be obese because they consume fewer foods high in fat. However, they remain at risk for vitamin B12 deficiency and should consume at least a daily multivitamin. They are less likely to have anemia but just as likely to have low calcium intake as their non-vegan peers. Adolescents who follow a lacto-ovo-vegetarian diet are less likely to have deficiencies in vitamin B12, calcium, and iron.

Folate and vitamin B6 are not likely to be deficient in persons who consume vegan diets because those nutrients are found in many legumes, fruits, and vegetables that are the mainstays of the diet.

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Question: 14

A 3-month-old infant who has a history of renal dysplasia associated with obstructive uropathy has marked polyuria. He is breastfeeding and receiving supplemental cow milk-based formula. In an effort to reduce the high urine output, you consider reducing the renal solute load by changing feedings from the milk-based formula currently being used.

Of the following, the MOST appropriate change is to

A. a hydrolyzed formula containing medium-chain triglycerides

B. a more concentrated (24-kcal) milk-based formula

C. human milk exclusively

D. soy milk-based formula

E. whole cow milk

Critique: 14 Preferred Response: C

The infant described in the vignette has polyuria caused by a urinary concentrating defect. The concentrating defect is the result of tubular damage due to the obstructive uropathy. The inability to concentrate the urine causes the kidneys to create an "excessive" volume of urine to excrete the solute load presented to them.

One strategy to reduce polyuria is to reduce the solute burden placed on the kidneys. Potential renal solute load is affected by intake of protein, sodium, potassium, chloride, and phosphorus. The protein and phosphorus content are the most important variables when comparing infant feeding regimens.

Human milk possesses a lower potential renal solute load than cow milk or cow milk-based formulas. Accordingly, the most appropriate change in feeding for the infant in the vignette is to recommend that the mother stop cow milk formula supplementation and exclusively breastfeed. If human milk is not available, a "low-solute" cow milk-based formula can be used. A low calcium-phosphorus formula has the next lowest potential renal solute load compared with human milk. Cow milk, soy milk-based formula, hydrolyzed formula with medium-chain triglycerides, and 24-kcal milk-based formula all have greater renal solute loads than human milk.

Renal solute load should also be considered in nephrogenic diabetes insipidus.

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Question: 15

A mother brings in her 13-month-old daughter for evaluation because her girl developed a perioral rash and "hives" on two occasions last week. One episode occurred while eating yogurt and another happened immediately after eating a bagel with cream cheese. She states that her daughter has eaten other foods such as eggs and bread without problems but is breastfeeding and never has been given milk-based formulas or cow milk. The infant has been given rice milk, but she became fussy and seems to prefer breastfeeding. The mother is concerned that her daughter may be allergic to milk but would like to stop breastfeeding.

Of the following, the BEST advice is to recommend

A. a cow milk food challenge in the clinic

B. avoidance of milk, egg, soy, and wheat products

C. breastfeeding until the child is 3 years old

D. switching to an amino acid-based formula

E. switching to soy milk

Critique: 15 Preferred Response: E

Milk protein allergy is an immunoglobulin (Ig) E-mediated food reaction that affects 2% to 3% of infants within the first postnatal year. Typical symptoms include urticaria, angioedema, atopic dermatitis, and anaphylaxis. With IgE-mediated reactions, the quantity of milk required to result in a reaction often is minimal (eg, milk touching the face, a taste of ice cream). Taking a detailed history about the specific food(s) involved, timing of the onset of symptoms, and type of symptoms is important to distinguish IgE-mediated reactions, as described for the child in the vignette, from other adverse milk reactions, such as milk protein enterocolitis and lactose intolerance. Once an IgE-mediated food allergy is suspected, the clinician should consider allergy skin testing or serum IgE testing for the suspected food.

While awaiting results from either blood testing or allergy consultation for skin testing, the first reasonable action is to switch to a soy-based formula. Approximately 10% to 15% of infants and children who have IgE-mediated milk protein allergies may not tolerate soy formula, but this risk applies to infants younger than 6 months of age. For infants older than 6 months, the risk is closer to 5%. Nonetheless, the initial soy formula trial should be performed in the clinic. Other acceptable formula options in this scenario include an extensively hydrolyzed or an amino acid-based formula, although the unpleasant taste and significantly higher cost can be limiting for many families. Also, because almost all affected infants can be fed successfully with a soy or extensively hydrolyzed formula, switching initially to an amino acid-based formula is not required.

Food challenges often are used to assess adverse food reactions, but they generally are reserved for foods that are unlikely allergens or if the clinical history is inconsistent or vague (eg, a patient who eats a particular food and does not always have a reaction). Food challenges may result in anaphylaxis and generally are avoided when the history and testing results support an IgE-mediated reaction.

Infants who have a specific food allergy sometimes are placed incorrectly on restricted diets that avoid multiple foods. Without a specific history of other adverse food reactions, avoidance of other foods such as egg or wheat is not recommended. However, parents should be counseled that children can develop other food allergies and should monitor their children during ingestion of other common food allergens.

Breastfeeding until age 3 years old is an option that is not preferred by the mother in the vignette. Although most IgE-mediated cow milk allergies resolve by 3 years of age, milk protein allergy can persist past 5 years of age in up to 20% of affected children.

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Question: 17

You are following a 3-month-old infant who was born at 30 weeks' gestation, underwent a distal ileal resection for necrotizing enterocolitis at 2 weeks of age, and subsequently was placed on parenteral nutrition for 2 months. The baby has residual cholestasis from the parenteral nutrition (total bilirubin, 5.0 mg/dL [85.5 mcmol/L]; direct bilirubin, 3.0 mg/dL [51.3 mcmol/L]). Currently, she is receiving a cow milk protein hydrolysate formula concentrated to 24 kcal/oz (0.8 kcal/mL). You are considering adding a dietary supplement to increase the caloric density of the formula.

Of the following, the supplement that is the MOST likely to be tolerated and cause less diarrhea in this infant is

A. flaxseed oil

B. medium-chain triglyceride oil

C. olive oil

D. omega-3 polyunsaturated fatty acid (fish oil)

E. soybean oil

Critique: 17 Preferred Response: B

Infants who have chronic illnesses may have specialized nutritional requirements and often do not tolerate the standard 20-kcal/oz formula given to healthy term infants. For example, children who have some forms of congenital heart disease or renal disease may require a more concentrated formula because the standard formula may lead to volume overload. Children who have intestinal disease or malabsorption, such as the child described in the vignette, also may require a more concentrated formula to decrease the likelihood of feeding intolerance or diarrhea.

Formula may be concentrated by increasing the concentration of protein, carbohydrate, or fat. Perhaps the easiest method of increasing the caloric density of a formula is to mix more powder with the same amount of water. For example, four scoops of most commercially available formulas mixed with 8 oz of water yields standard 0.67-kcal/mL (20-kcal/oz) formula, but mixing five scoops in 8 oz results in 0.83-kcal/mL (25-kcal/oz) formula. However, exceeding 25-kcal/oz formula by increasing the amount of powder may yield too high a concentration of protein, which could result in an excessive renal solute load. For this reason, carbohydrate and fat supplements are available to concentrate infant and toddler feedings further. The most common adverse effect of carbohydrate supplements is diarrhea, and lipid supplements may cause either diarrhea or delayed gastric emptying. For these reasons, caution is recommended when increasing the caloric density of a formula, especially when the caloric density is increased to greater than 1 kcal/mL (30 kcal/oz).

Because the patient in the vignette has had an ileal resection and cholestasis, the best fat supplement for him is oil composed of medium-chain triglycerides (MCT oil). MCT oil can be absorbed directly across the enterocyte and does not require intraluminal digestion by bile acids. In contrast, soy, olive, flaxseed, and fish oils are long-chain fatty acids that require bile acids for digestion and might cause diarrhea in a child who has cholestasis and ileal resection.

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Question: 33

A 7-month-old child presents for a follow-up office visit after undergoing a Kasai procedure for biliary atresia at 6 weeks of age. The mother states that the boy is irritable when his right arm is moved. On physical examination, the infant is jaundiced. You detect tenderness in the anterior radial head. Radiography of the affected region demonstrates metaphyseal fraying (Item Q33) and a fracture.

Of the following, the MOST appropriate laboratory studies to obtain next are

A. calcium and phosphorus measurement and bone densitometry (DEXA scan)

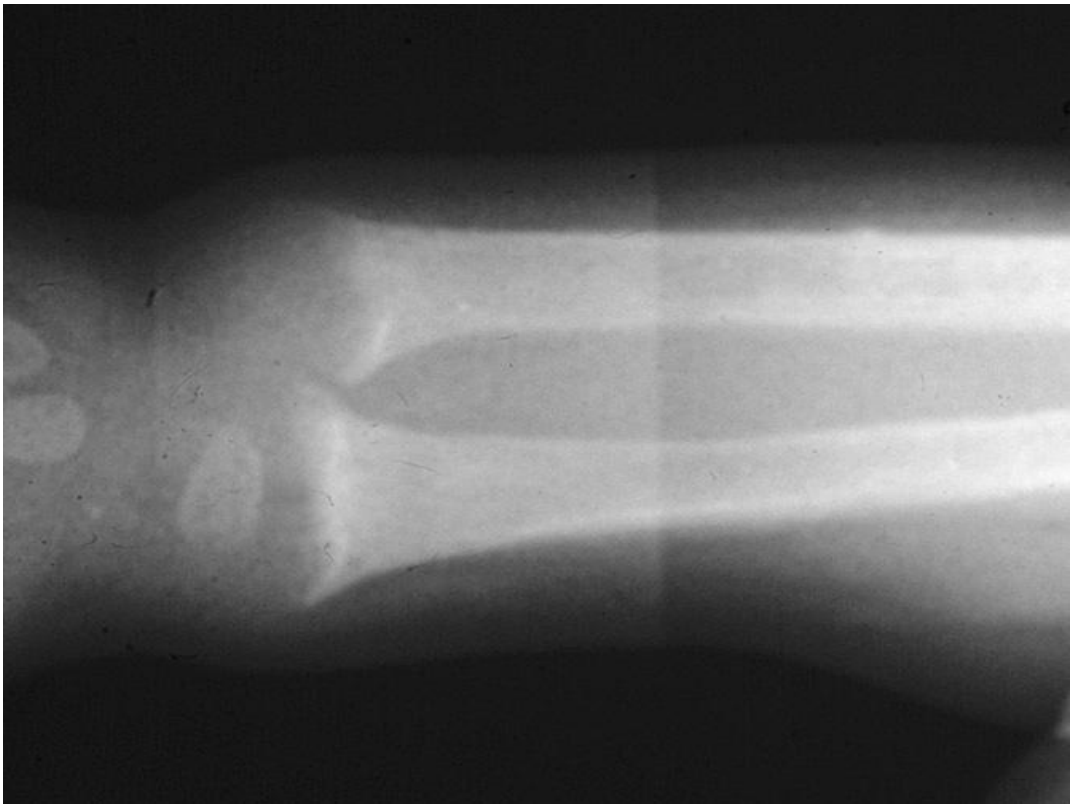
B. calcium and phosphorus measurement and urinary calcium-to-creatinine ratio

C. calcium, phosphorus, and 25-hydroxyvitamin D measurement

D. calcium, phosphorus, and magnesium measurement

E. magnesium, phosphorus, and parathyroid hormone measurement

Question: 33



Metaphyseal fraying, cupping, and widening, as described for the infant in the vignette. (Couretsy of R. Schwartz)

Critique: 33 Preferred Response: C

Chronic cholestasis due to biliary atresia results in decreased bile flow into the intestine. The absence of intraluminal bile acids, in turn, causes decreased digestion of lipids, leading to fat malabsorption. In addition, absorption of fat-soluble vitamins (A, D, E, and K) is impaired, which may lead to clinical sequelae of fat-soluble vitamin deficiency. Finally, steatorrhea may impair calcium absorption because intraluminal free fatty acids may bind calcium.

The clinical presentation of the patient in the vignette strongly suggests the presence of rickets from vitamin D deficiency. Therefore, the most helpful initial laboratory testing is determination of calcium, phosphorus, and 25-hydroxyvitamin D concentrations. The 25-hydroxyvitamin D assay is the best measure of hepatic stores of vitamin D and is a better marker of vitamin D status than either serum vitamin D or 1,25-dihydroxyvitamin D. Although bone density testing, measurement of serum magnesium and parathyroid hormone, and determination of the urinary calcium-to-creatinine ratio may provide useful additional information, they will not help establish the diagnosis of vitamin D-deficient rickets.

Rickets is a potentially preventable complication of biliary atresia, but requires monitoring of calcium, phosphorus, and 25-hydroxyvitamin D concentrations two to four times a year. Infants who have biliary atresia routinely receive supplementation with approximately 8,000 IU of ergocalciferol (vitamin D2) daily. This dose of vitamin D is approximately 20 times the recommended dietary allowance for a healthy toddler. If rickets develops or the vitamin D concentration cannot be maintained within the normal range, the patient should receive either calcitriol (1,25-dihydroxyvitamin D3) or intramuscular vitamin D.

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Question: 49

A 12-year-old boy has had cholestasis since infancy from Alagille syndrome. He has been lost to medical follow-up for the last several years. He now presents to your office with pain in his right upper thigh after a fall. His thigh is intensely tender, and ultrasonography demonstrates a large hematoma in his quadriceps. The parents state that he has tended to bruise easily in the past few months.

Of the following, the condition MOST likely to account for this patient's symptoms is

A. factor VIII deficiency

B. idiopathic thrombocytopenic purpura

C. vitamin C deficiency

D. vitamin K deficiency

E. von Willebrand disease

Critique: 49 Preferred Response: D

Alagille syndrome is characterized by cardiac disease (especially peripheral pulmonary stenosis), vertebral anomalies, ocular anomalies (posterior embryotoxon), facial dysmorphism (triangular facies, macrocephaly, large ears) (Item C49A), and paucity of the intrahepatic bile ducts. The hepatic manifestations of this syndrome account for much of the medical morbidity. Specifically, impaired bile flow results in chronic cholestasis, which leads to severe pruritus, jaundice, malabsorption of nutrients, and malabsorption of fat-soluble vitamins (Item C49B). Although most affected children have their jaundice improve as they grow older, a subset progresses to cirrhosis and requires liver transplantation.

Patients who have hepatic disease must have their nutritional status monitored carefully. Chronic anorexia, recurrent illnesses, and fat malabsorption may result in caloric deficiency and growth failure. Caloric supplementation by nasogastric tube or gastrostomy may be necessary to ensure adequate caloric intake. In addition, patients who have cholestasis are at risk for fat-soluble vitamin deficiency. Vitamin D deficiency typically causes osteopenia and rickets, vitamin E deficiency causes peripheral neuropathy and ataxia, and vitamin A deficiency may cause night blindness or corneal lesions. The bruising described for the patient in the vignette most likely is due to vitamin K deficiency. Vitamin K is a cofactor essential in posttranscriptional carboxylation of the clotting factors II, VII, IX, and X. Thus, vitamin K deficiency leads to prolonged prothrombin and partial thromboplastin time, which predisposes to bruising. Although factor VIII deficiency, vitamin C deficiency, von Willebrand disease, and idiopathic thrombocytopenia purpura also may cause bruising, the patient who has Alagille syndrome is not at increased risk for developing these conditions.

In addition to supplementing patients who have chronic liver disease with fat-soluble vitamins, the clinician caring for these patients also must supply adequate calories. Patients who have advanced chronic liver disease may have both anorexia and increased caloric requirements. In addition, patients who have portal hypertension and ascites may need to have total fluid intake restricted, which, in turn, means that they may require a more concentrated and less palatable formula. For these reasons, nasogastric or gastrostomy feedings sometimes are necessary to achieve optimal growth, especially when preparing a patient for liver transplantation.

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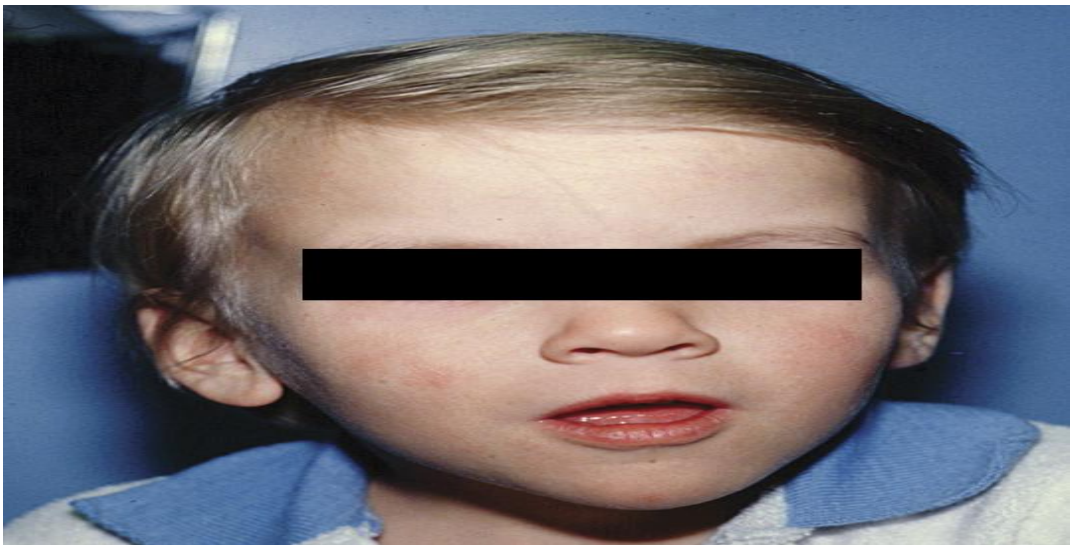
Critique: 49



The broad forehead and pointed chin characteristic of Alagille syndrome. (Courtesy of M. Rimsza)

Critique: 49

Consequences and Medical Management of Prolonged Cholestasis

Consequence of Cholestasis	Management
Malabsorption	
Long-chain triglycerides (malnutrition)	• Supplement formula with medium-chain triglycerides or change to special formula • Supply adequate calories (125% to 150% RDA) • Supply adequate protein
Fat-soluble vitamin deficiency	
Vitamin A deficiency (night blindness)	• Vitamin A 10,000 to 25,000 IU/d
Vitamin D deficiency (metabolic bone disease)	• Vitamin D_2 5,000 to 8,000 IU/d or 25-hydroxycholecaliciferol 3 to 5 mcg/kg per day
Vitamin E deficiency (degenerative neurologic syndrome)	• Vitamin E 50 to 400 IU/d or d-alpha-tocopherol polyethylene glucol-1000 succinate 15 to 25 IU/kg per day
Vitamin K deficiency (hypoprothrombinemia)	• Vitamin K (water-soluble derivative of menadione) 2.5 to 5 mg/d
Water-soluble vitamin deficiency	• Supplement with twice RDA
Mineral deficiencies	
Calcium deficiency (bone disease)	• Elemental calcium 25 to 100 mg/kg per day
Phosphate deficiency (bone disease)	• Phosphate 25 to 50 mg/kg per day
Zinc deficiency	• Zinc 1 mg/kg per day
Retention of Constituents of Bile	
Cholesterol and bile acids (pruritus and xanthoma)	• Bile acid-binding agents: cholestyramine 0.25 to 0.5 g/kg per day in three divided doses • Ursodeoxycholic acid (presently undergoing clinical trials)
Copper	• Low-copper diet
Fibrosis or Cirrhosis	
Portal hypertension (esophageal and gastric variceal hemorrhage)	• Resuscitation, appropriate blood products • Esophageal sclerotherapy • Balloon tamponade, vasopressin infusion • Portovenous shunting • Prophylactic role of propranolol?
Ascites	• Sodium restriction 1 to 2 mEq/kg per day • Aldosterone antagonist (spironolactone 3 to 5 mg/kg per day in three or four divided doses) • Loop diuretic (furosemide 1 to 2 mg/kg per day in two to three divided doses)
Respiratory ascites	• Albumin infusion 1 g/kg and furosemide infusion 0.5 to 1 mg/kg • Large-volume paracentesis and intravenous infusion of albumin 1 g/kg
End-stage Liver Disease	• Liver transplantation

RDA=recommended dietary allowance
Reprinted with permission from Mews C, Sinatra FR. Cholestasis in infancy. *Pediatr Rev*. 1994;15:233-240

Question: 81

A 4-month-old infant who has gastroschisis underwent surgical repair on the first day after birth, but continues to require support with parenteral nutrition and lipids. He now has developed poor feeding, irritability, and progressive diarrhea. Radiography demonstrates metaphyseal fraying, but calcium, phosphorus, and 25-hydroxyvitamin D concentrations are normal. When you review his prior laboratory studies, you note he has had neutropenia for the past 4 weeks.

Of the following, this child's symptoms are MOST consistent with

A. copper deficiency

B. magnesium deficiency

C. vitamin A deficiency

D. vitamin B6 deficiency

E. zinc deficiency

Critique: 81 Preferred Response: A

The poor feeding, irritability, metaphyseal irregularity, normal vitamin D value, and neutropenia described for the infant in the vignette are consistent with copper deficiency, a rare disorder that develops when children receive parenteral nutrition without trace element supplementation. Copper is a trace element that has many important biochemical functions. It is an important component of respiratory chain enzymes (eg, cytochrome C) and lysyl oxidase (an enzyme critical to collagen production and bone formation). Menkes disease, a rare syndrome involving a defect in copper transport and profoundly low serum copper concentrations, is characterized by hypotonia, developmental delay, seizures, and "steely hair." Copper deficiency also may be seen in preterm infants who do not receive appropriate trace element supplementation. The clinical presentation of copper deficiency in the preterm infant is more subtle, but can include pallor, poor feeding, hypochromic anemia, neutropenia, and skeletal changes (including metaphyseal fraying and osteoporosis). The patient's symptoms are more consistent with copper deficiency than with magnesium, vitamin B6, zinc, or vitamin A deficiency. Specifically, magnesium deficiency can cause hypotonia and apnea, vitamin B6 deficiency can cause hypotonia and seizures, zinc deficiency can cause diarrhea and skin rashes, and vitamin A deficiency can lead to corneal lesions and impaired vision.

To prevent copper deficiency, most parenteral nutrition is supplemented with 200 mcg/L of copper. Infants who have cholestasis may have impaired copper excretion into the bile, necessitating a decrease in the concentration of copper in the parenteral nutrition to prevent copper overload.

References:

Collier S, Gura KM, Richardson D, Duggan C. Parenteral nutrition. In: Hendricks KM, Duggan C. *Manual of Pediatric Nutrition*. 4th ed. Hamilton Ontario, Canada: BC Decker; 2005:317-375

Giles E, Doyle LW. Copper in extremely low-birthweight or very preterm infants. *NeoReviews*. 2007;8:e159-e164. Available for subscription at: http://neoreviews.aappublications.org/cgi/content/full/8/4/e159

Question: 94

An 8-month-old girl who has a history of cardiomyopathy following viral myocarditis presents with poor weight gain. She is receiving a 20-kcal/oz milk-based formula and has no history of vomiting or diarrhea. Her only medication is furosemide. Physical examination findings include a heart rate of 130 beats/min, respiratory rate of 60 breaths/min, and blood pressure of 88/44 mm Hg.

Of the following, the MOST appropriate initial strategy to increase weight gain for this girl is to

A. change to a 24-kcal/oz formula

B. discontinue furosemide therapy

C. increase the volume of 20-kcal/oz formula

D. place a gastrostomy feeding tube

E. start parenteral nutrition

Critique: 94 Preferred Response: A

Young infants who have cardiac dysfunction, such as the girl described in the vignette, often have difficulty ingesting sufficient calories for growth because they frequently have increased caloric expenditure with feeding. Increased caloric intake may place the infants at risk of fluid overload, necessitating the use of diuretics. Because an increased volume of feedings with a 20-kcal/oz formula may place the infant at risk of fluid overload, use of a more concentrated formula is required. Accordingly, the infant described in the vignette should be changed to a 24-kcal/oz formula. Increasing the caloric density of feedings can meet the goals of increased calories and "relative" fluid restriction.

Discontinuation of furosemide likely would lead to weight gain from fluid retention, not a true weight gain. A gastrostomy tube provides a conduit for feeding and probably plays a role in a child incapable of taking in sufficient calories (eg, chronic renal failure or severe developmental delay), but the child in the vignette deserves a trial of high-calorie feedings before subjecting her to this surgical procedure. Finally, parenteral nutrition is not a suitable option because this child has a functional gastrointestinal tract and can tolerate enteral nutrition.

References:

Kelleher DK, Laussen P, Teixeira-Pinto A, Duggan C. Growth and correlates of nutritional status among infants with hypoplastic left heart syndrome (HLHS) after stage 1 Norwood procedure. *Nutrition*. 2006; 22:237-244. Abstract available at: http://www.ncbi.nlm.nih.gov/pubmed/16500550

Pillo-Blocka F, Adatia I, Sharieff W, McCrindle BW, Zlotkin S. Rapid advancement to more concentrated formula in infants after surgery for congenital heart disease reduces duration of hospital stay: a randomized clinical trial. *J Pediatr*. 2004;145:761-766. Abstract available at: http://www.ncbi.nlm.nih.gov/pubmed/15580197

Yahav J, Avigad S, Frand M, et al. Assessment of intestinal and cardiorespiratory function in children with congenital heart disease on high-caloric formulas. *J Pediatr Gastroenterol Nutr*. 1985;4:778-785. Abstract available at: http://www.ncbi.nlm.nih.gov/pubmed/4045636

Question: 97

A 16-year-old boy in your practice has cystic fibrosis. As a complication of his illness, he has developed cirrhosis and cholestasis. He now complains of shaky hands. Neurologic examination demonstrates hyporeflexia and tremor with hands outstretched.

Of the following, the patient's symptoms are MOST consistent with deficiency of

A. vitamin A

B. vitamin B1 (thiamine)

C. vitamin C

D. vitamin D

E. vitamin E

Critique: 97 Preferred Response: E

Because the young man described in the vignette has chronic cholestasis, he is at risk for developing deficiency of any of the fat-soluble vitamins, including vitamins A, D, E, and K. His neurologic symptoms of tremor and hyporeflexia most strongly suggest vitamin E deficiency.

Vitamin E (tocopherol) is an important factor in stabilizing the lipid membrane of the red blood cell and the lipids in the myelin sheath of neurons. Therefore, the most common presenting features of hypovitaminosis E are hemolysis (primarily reported in preterm infants) and peripheral neuropathy (identified in infants and children who have chronic cholestasis, pancreatic insufficiency, or malabsorption).

Supplementation of formulas and parenteral nutrition with vitamin E has reduced substantially the incidence of hemolysis in the vitamin E-deficient preterm infant. However, patients who have cystic fibrosis or cholestatic liver disease require both monitoring of vitamin E concentrations and supplementation with vitamin E. Because vitamin E is a fat-soluble vitamin, those who have cholestasis may have difficulty absorbing alpha-tocopherol, the form of vitamin E available in most dietary supplements. For this reason, d-alpha-tocopheryl polyethylene glycol 1,000 succinate, a water-soluble form of vitamin E, should be given to patients who have significant cholestatic liver disease. The recommended dose for a patient who has cholestatic liver disease is 15 to 25 IU/kg per day.

Deficiency of vitamin A, B1, C, or D would not be expected to cause such a clinical presentation. Vitamin A deficiency causes impaired vision ("night blindness") and corneal ulcers; vitamin B1 deficiency can cause myopathy and heart failure ("beriberi"); vitamin C deficiency causes irritability, bone lesions, and bruising (scurvy); and vitamin D deficiency causes osteopenia or rickets.

References:

Harmatz P, Burensky E, Lubin B. Nutritional anemias. In: Walker WA, Watkins JB, Duggan C, eds. *Nutrition in Pediatrics*. 3rd ed. Hamilton, Ontario, Canada: BC Decker; 2003:830-847

Spinozzi NS. Hepatobiliary diseases. In: Hendricks KM, Duggan C. *Manual of Pediatric Nutrition*. 4th ed. Hamilton, Ontario, Canada: BC Decker; 2005:586-592

Question: 116

A 10-year-old boy who recently emigrated from Central America is referred by the school nurse for evaluation of obesity.Physical examination reveals an obese but generally healthy boy who has acanthosis nigricans (Item Q116).He has had limited access to medical care in the past.

Of the following, the physical finding MOST likely to suggest an underlying cause for the child's obesity is

A. a normal blood pressure

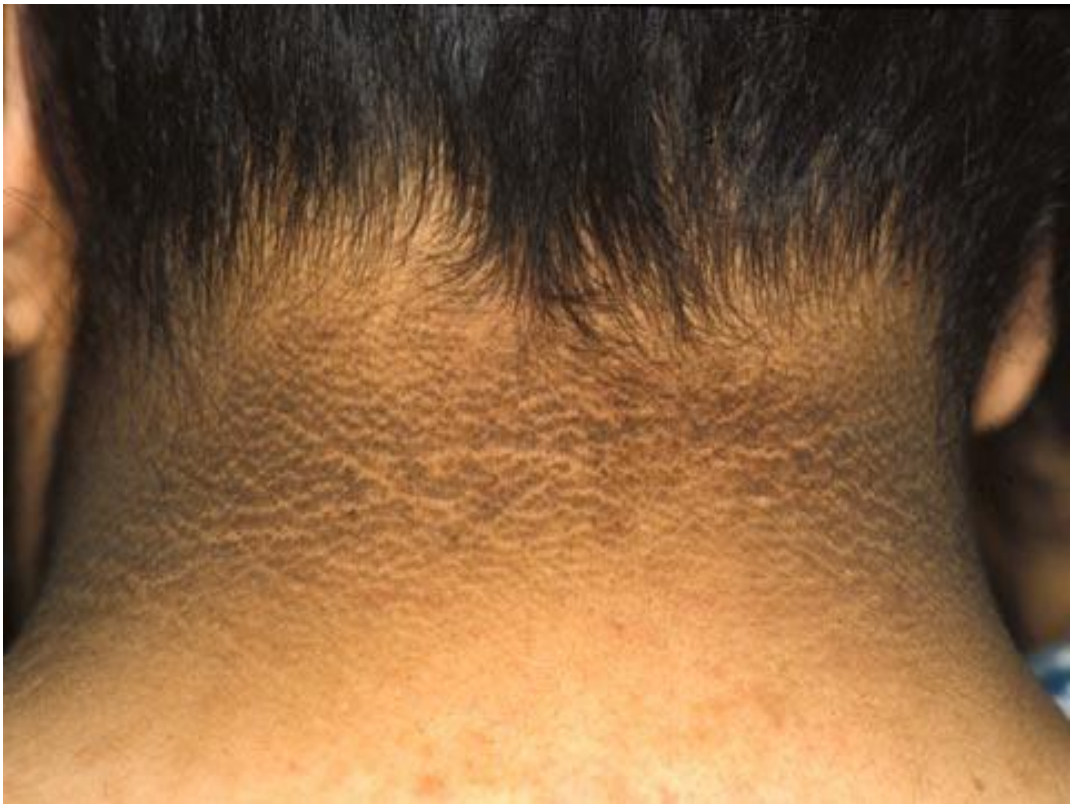
B. abdominal striae

C. penile length at 1 standard deviation below the mean

D. small hands and feet

E. stature greater than the 95th percentile

Question: 116



(Courtesy of M. Rimsza)

Critique: 116 Preferred Response: D

The incidence of obesity continues to increase among children in industrialized nations. In general, the causes in most children relate to increased caloric intake and decreased physical activity. However, the pediatrician must exclude genetic, metabolic, or other underlying causes of obesity in children for two reasons: 1) some underlying causes may require treatment and 2) the focus on changing the child's lifestyle must involve parents who are reassured that hormonal or other abnormalities are not the cause. A child who exhibits normal linear growth, has normal developmental milestones, and has normal findings on physical examination is unlikely to have an underlying cause for his or her obesity.

The most common metabolic cause of obesity is hypothyroidism, but routine testing for thyroid function in the overweight child who has normal linear growth velocity for age and no clinical signs of hypothyroidism is unwarranted. A stature greater than the 95th percentile should be interpreted with caution because one measurement is not sufficient to determine growth velocity. However, this finding is reassuring because normal to above-average height is unlikely in a child who has hypothyroidism and common in children who have exogenous obesity.

Children who have small hands and feet, hypogonadism, learning disabilities, or mental retardation should be evaluated for Prader- Willi and Bardet-Biedl syndromes. These syndromes often present in infancy or early childhood with hypotonia and developmental delay. Small hands and feet are common and strongly suggest an underlying anomaly for the child described in the vignette (Item C116A). Genetic testing for these conditions is available.

The normal blood pressure reported for the boy in the vignette is reassuring because hypertension often complicates obesity. Penile length is normal at 1 standard deviation below the mean, but it is important to remember that measurement of penile length can be a challenge in obese children due to interference from the pubic fat pad.

Striae may be seen in many overweight children simply due to rapid weight gain. Although this finding also occurs with cortisol excess (Item C116B), conditions associated with hypercortisolism usually are associated with other signs and symptoms. Thus, the finding of striae in an obese child is not an indication that additional testing is necessary.

Treatment of childhood obesity is difficult, although there is some evidence for a variety of interventions that may prove successful in some populations. Numerous diets, including low-carbohydrate diets, appear to have some impact on obesity in children. For morbidly obese adolescents who have comorbidities unresponsive to diet and exercise, some centers are moving toward bariatric surgery.

Barriers to healthy lifestyles (lack of availability in some communities of safe outdoor play areas, specific exercise programs geared to very obese children, and school lunch programs) remain topics of scientific and political investigation.

References:

Arterburn DE. Obesity in children. *BMJ Clinical Evidence*. 2007. Available for subscription at: http://clinicalevidence.bmj.com/ceweb/conditions/chd/0325/0325.jsp

Rodearmel SJ Wyatt HR, Stroebele N, Smith SM, Ogden LG, Hill JO. Small changes in dietary sugar and physical activity as an approach to preventing excessive weight gain: the America on the Move Family Study. *Pediatrics*. 2007;120:e869-e879 Available at: http://pediatrics.aappublications.org/cgi/content/full/120/4/e869

Schneider MB, Brill SR. Obesity in children and adolescents. *Pediatr Rev*. 2005;26:155-162. Available at: http://pedsinreview.aappublications.org/cgi/content/full/26/5/155

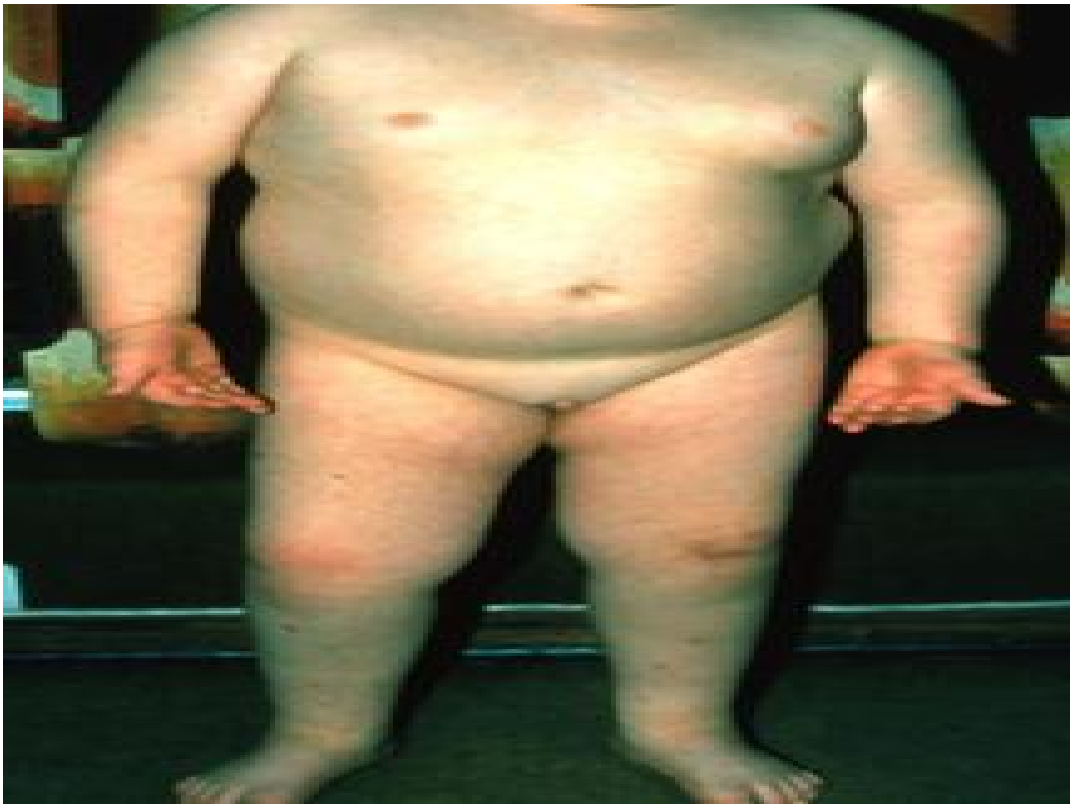
Shaw K, Gennat H, O'Rourke P, Del Mar C. Exercise for overweight or obesity. *Cochrane Database Syst Rev*. 2006;4:CD003817. Available at: http://www.mrw.interscience.wiley.com/cochrane/clsysrev/articles/CD003817/frame.html

Summerbell CD, Waters E, Edmunds LD, Kelly S, Brown T, Campbell KJ. Interventions for

preventing obesity in children. *Cochrane Database Syst Rev*. 2005;3:CD001871. Available at: http://www.mrw.interscience.wiley.com/cochrane/clsysrev/articles/CD001871/frame.html

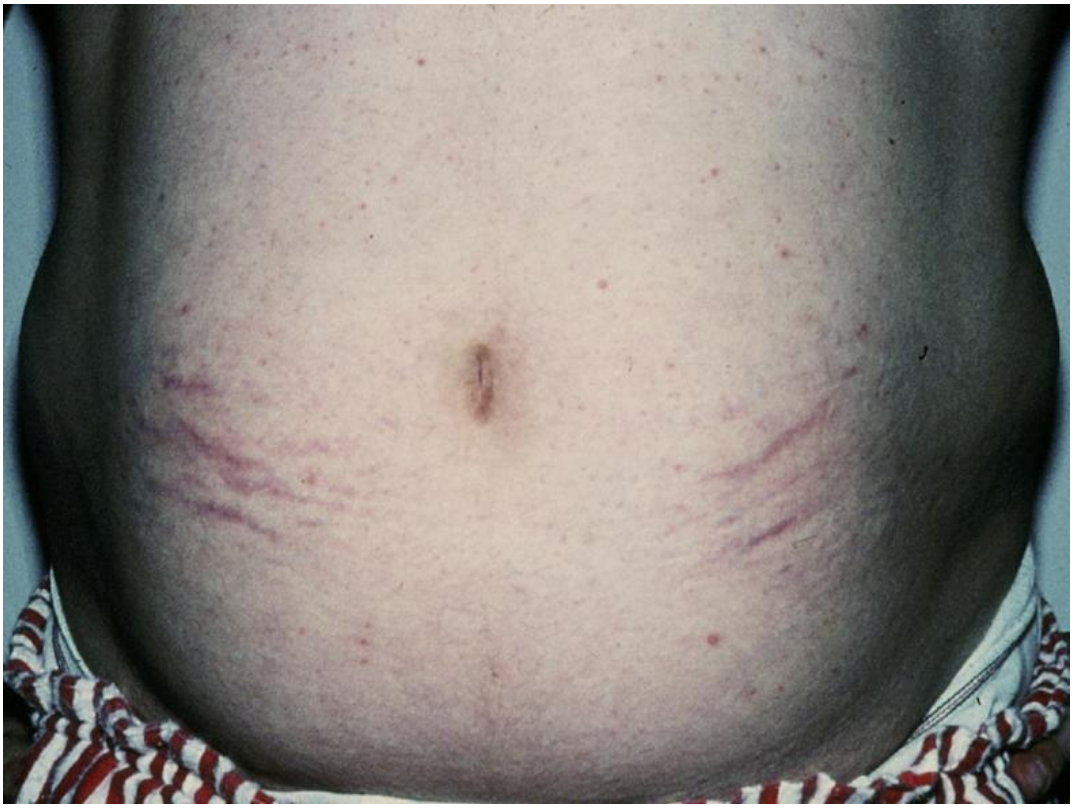
Thomas DE, Elliott EJ, Baur L. Low glycaemic index or low glycaemic load diets for overweight and obesity. *Cochrane Database Syst Rev*. 2007;3:CD005105. Available at: http://www.mrw.interscience.wiley.com/cochrane/clsysrev/articles/CD005105/frame.html

Critique: 116



Prader-Willi syndrome is characterized by short stature, obesity, hypogonadism, and small hands and feet. (Courtesy of Y. Lacassie)

Critique: 116



The striae observed in patients who have Cushing syndrome often have a violaceous color. (Courtesy of M. Rimsza)

Question: 227

You are addressing a group of expectant mothers who are due to deliver their infants in the next few weeks. You discuss the benefits of breastfeeding and explain that it is the best nutrition for most babies. One woman asks you if it is acceptable to breastfeed if she has had hepatitis in the past. You explain that there are only a few infections that would prevent a mother from being able to breastfeed her baby.

Of the following, breastfeeding is MOST likely to be contraindicated if a mother

A. has active untreated pulmonary tuberculosis

B. has genital herpes without breast lesions

C. is a cytomegalovirus carrier

D. is hepatitis B surface antigen-positive

E. is hepatitis C antibody-positive

Critique: 227 Preferred Response: A

Human milk is the optimal nutrition for infants. Benefits include transference of protective maternal antibodies, improved bonding between mother and child, and probable improvement in cognitive and developmental function of the infant. Most mothers can breastfeed successfully, although there are contraindicated conditions for both infants and mothers. Infants who have galactosemia should not receive human milk, and infants who have other forms of metabolic disease, such as urea cycle defects or phenylketonuria, may receive only a limited amount. Mothers infected with human immunodeficiency virus (in the United States) or human T-cell lymphotrophic virus-1 or -2 and those who have active untreated tuberculosis or active herpes lesions on the breast should not breastfeed their infants. However, mothers who are hepatitis B surface antigen-positive or hepatitis C antibody-positive, are cytomegalovirus carriers, or have genital herpes without breast lesions can breastfeed safely. Maternal medications that preclude breastfeeding include antineoplastic agents, immunosuppressants, lithium, and radiopharmaceutical agents.

Several disorders of the breast may make breastfeeding difficult, but they are not contraindications to breastfeeding. Previous breast surgery may cause ineffective lactation, but this varies among mothers. Women who have inverted or flat nipples may experience difficulties with latch-on, but this can be improved with early feedings, use of nipple shields, and lactation consultation. Use of a breast pump also may help. Women who have breast cancer may be able to breastfeed if they are not taking antineoplastic medications. Mastitis, inflammation of the breast usually caused by obstruction of ducts, may make breastfeeding painful, but more frequent nursing is the best recommendation to help resolve this condition. Sore or cracked nipples may develop, especially if the infant has oral-motor dysfunction, and adjusting the infant's latch-on may improve these symptoms. A mother who has *Candida* infection of the breast may continue to breastfeed, but both she and her infant should be treated for the infection to avoid a cycle of reinfection.

References:

American Academy of Pediatrics Section on Breastfeeding. Breastfeeding and the use of human milk. *Pediatrics*. 2005;115:496-506. Available at: http://pediatrics.aappublications.org/cgi/content/full/115/2/496

Chandran L, Gelfer P. Breastfeeding: the essential principles. *Pediatr Rev*. 2006;27:409-417. Available at: http://pedsinreview.aappublications.org/cgi/content/full/27/11/409

Powers NG, Slusser W. Breastfeeding update 2: clinical lactation management. Pediatr Rev. 1997;18:147-161. Available at: http://pedsinreview.aappublications.org/cgi/content/full/18/5/147

Question: 17

A 6-month-old infant presented in the newborn period with intestinal malrotation and mid-gut volvulus. Emergency surgery resulted in resection of his entire small bowel, except for 7 cm of duodenum. He has been maintained on parenteral nutrition since then and is listed for small bowel transplantation at the regional transplant center. His parenteral nutrition regimen provides 120 kcal/kg per day and includes 20% dextrose, 3 g/120 kcal per day amino acids, and 3 g/kg per day lipids. He has had increasing jaundice over the past month. On physical examination, the alert, afebrile, and icteric infant has a firm liver edge palpable 3.5 cm below the right costal margin. Laboratory data include:

- Total bilirubin, 12.5 mg/dL (213.8 mcmol/L)
- Direct bilirubin, 8.0 mg/dL (136.8 mcmol/L)
- Alanine aminotransferase, 200 units/L
- Aspartate aminotransferase, 150 units/L
- Gamma-glutamyl transpeptidase, 180 units/L

Of the following, the MOST appropriate treatment is to

A. add phenobarbital 5 mg/kg per day orally

B. add ursodeoxycholic acid 20 mg/kg per day orally

C. decrease amino acids to 1.5 g/120 kcal per day

D. decrease dextrose to 10%

E. decrease lipids to 1.0 g/kg per day

Critique: 17 Preferred Response: E

Parenteral nutrition (PN) is a life-sustaining therapy for patients whose limited gastrointestinal function does not permit adequate enteral absorption of nutrients. Despite its ability to maintain nutritional status and support growth, long-term PN has been associated with numerous serious and sometimes life-threatening complications, including bloodstream infections due to bacterial and fungal contamination of central venous catheters, metabolic derangements, and PN-associated liver disease (PNALD). The infant described in the vignette has PNALD, an extremely challenging problem that is especially common in patients who have short-bowel syndrome, for reasons that are not completely understood. Although the causes of hepatocellular dysfunction and cholestasis (indicated by direct hyperbilirubinemia and elevated gamma-glutamyl transpeptidase values) in this clinical setting may be multifactorial, current data suggest that excess intravenous lipids or specific elements in lipid emulsions play a major role in the pathogenesis of PNALD. Accordingly, the most reasonable approach for the infant in the vignette is to decrease the amount of infused lipid.

For the patient who has an extremely short bowel, PN serves as a "bridge therapy." Because the infant in the vignette retains only a short length of duodenum, the prognosis is dire for achieving any significant small bowel adaptation that can lead to recovery of intestinal absorptive function. Therefore, long-term survival clearly depends upon a successful small intestinal transplant. For patients in whom residual bowel length is sufficient to undergo adaptive change and support some level of fluid and nutrient absorption, early reintroduction of enteral feedings remains the best option for limiting the occurrence and severity of PNALD.

Careful ongoing clinical assessment is essential for all patients receiving PN to identify and treat potential complications of this therapy. In addition to routine biochemical monitoring (Item C17), scrupulous attention must be paid to strict adherence to aseptic technique, both in parenteral fluid preparation and central line care. Ideally, central venous catheter access, including care required for routine dressing changes, should be carried out only by personnel trained and credentialed in line management. Such precautions should limit PN-associated complications, although PNALD remains a vexing and often unavoidable problem.

All major nutrients infused in PN solutions, as well as other factors (eg, infection, hepatic accumulation of bile acids) have been implicated in the pathogenesis of PNALD. However, advances in the composition and delivery of nutrients to infants receiving PN have resulted in amino acid profiles that mimic those during breastfeeding, and control of carbohydrate intake avoids hyperglycemia and wide swings in blood glucose values. Recently, increased focus has been placed on the role of intravenous lipids in the pathogenesis of PNALD. Both excess lipid administration and phytosterols, present in soybean emulsions used for PN, have been postulated as causative factors. Reducing infused lipids to 1.0 g/kg per day in this infant should ameliorate PNALD at least partially while preventing the onset of essential fatty acid deficiency. One recent study in two infants reported dramatic improvement in PNALD following replacement of the standard omega-6 fatty acid-based lipid infusate with a phytosterol-free omega-3 fatty acid emulsion.

Control of carbohydrate and protein intake, as demonstrated in the vignette, should limit PN-associated metabolic derangements. Accordingly, these major nutrients are much less likely to be implicated in the pathogenesis of PNALD in this infant, and reduction in parenteral glucose or

amino acid intake would not be expected to ameliorate cholestasis. Early studies suggested that phenobarbital enhances bile salt-independent biliary flow. However, its efficacy in this condition has not been demonstrated, and this therapy is not recommended for use in any pediatric cholestatic state. Ursodeoxycholic acid is a bile acid that does not form micelles, undergoes enterohepatic circulation, and increases hepatocellular bile excretion. However, the extreme short bowel with absent ileum (the site of active bile salt reabsorption) described for the infant in the vignette precludes its effectiveness.

Suggested reading:

Clayton PT, Whitfield P, Iyer K. The role of phytosterols in the pathogenesis of liver complications of pediatric parenteral nutrition. *Nutrition.* 1998;14:158-164. Abstract available at: http://www.ncbi.nlm.nih.gov/pubmed/9437703

Colomb V, Jobert-Giraud A, Lacaille F, Goulet O, Fournet JC, Ricour C. The role of lipid emulsions in cholestasis associated with long-term parenteral nutrition in children. *JPEN J Parenter Enteral Nutr.* 2000;24:345-350. Abstract available at: http://www.ncbi.nlm.nih.gov/pubmed/11071594

Gura KM, Duggan CP, Collier SB, et al. Reversal of parenteral nutrition—associated liver disease in two infants with short bowel syndrome using parenteral fish oil: implications for future management. *Pediatrics.* 2006;118:e197-e201. DOI: 10.1542/peds.2005-2662. Available at: http://pediatrics.aappublications.org/cgi/content/full/118/1/e197

Kleinman RE. Parenteral nutrition. In: *Pediatric Nutrition Handbook*. 6th ed. Elk Grove Village, Ill: American Academy of Pediatrics; 2009:519-540

Critique: 17

Variable Monitored	Initial Period*	Later Period†
Serum electrolytes (and carbon dioxide)	3–4 times/wk	Weekly
Serum urea nitrogen	3 times/wk	Weekly
Serum calcium, magnesium, phosphorous	3 times/wk	Weekly
Serum glucose		
Serum protein or albumin		Weekly
Liver function studies	Weekly	Weekly
Hematocrit	Weekly	Weekly
Urine glucose	Daily	Daily
Clinical observations (eg, activity, temperature)	Daily	Daily
Complete blood cell count and differential count	As indicated	As indicated
Cultures	As indicated	As indicated
Serum triglyceride	4 hours after an increase in dose	Weekly

In several institutions, the frequency of monitoring after the initial period has been significantly prolonged. It should be noted that prealbumin/transferrin are short term indicators of nutritional status, whereas albumin has a longer half life and therefore, more indicative of stable nutritional status. The usefulness of weekly liver function tests needs to be reexamined, because direct hyperbilirubinemia, the most specific indicator of cholestasis, takes several weeks to develop.

* Initial period is the period before full glucose, protein, and lipid intake is achieved or any period of metabolic instability.

† Later period is the period during which patient is in a metabolic steady state.

Blood glucose should be monitored closely during a period of glucosuria and for 2 to 3 days after cessation of parenteral nutrition to determine the degree of hypoglycemia. In the latter instance, frequent determination of blood glucose levels in fingertip venous blood constitutes adequate screening. After a month or more of receiving total parenteral nutrition, measurements can be made once a week or less frequently.

Adapted from Kleinman RE, ed. *Pediatric Nutrition Handbook*. 6th ed. Elk Grove Village, Ill: American Academy of Pediatrics; 2009.

ITEM C17: Recommended monitoring schedule during parenteral nutrition therapy in the hospitalized patient. Schedule may vary with age and underlying medical condition of the patient.

Question: 18

During the 1-week health supervision visit, a mother who is exclusively breastfeeding asks about vitamin and iron supplementation for her healthy term infant. She explains that her previous child, who was born at 30 weeks' gestation, was discharged with an oral iron supplement and vitamins.

Of the following, the MOST appropriate oral supplement to initiate for this infant at this visit is

A. calcium

B. folic acid

C. iron

D. vitamin D

E. vitamin K

Critique: 18 Preferred Response: D

The most appropriate oral supplement to initiate at 1 week of age for an exclusively breastfeeding term infant is vitamin D. In 2008, new guidelines from the American Academy of Pediatrics recommended that breastfed and partially breastfed infants be supplemented with 400 IU of vitamin D daily within days of birth. This change in the recommended amount of vitamin D supplementation arose because of continued reports of rickets in breastfed infants. Exclusive breastfeeding without adequate sun exposure or vitamin D supplementation is a risk factor for vitamin D deficiency and rickets.

Term newborns are assumed to have adequate iron stores for the first 4 to 6 months after birth. The adequate intake of iron for the first 6 postnatal months is 0.27 mg/day and is estimated from the content of iron in human milk and its high bioavailability. Recent studies have shown that exclusively breastfed infants who received iron supplementation between 1 and 6 months of age had improved hemoglobin concentrations at 6 months of age as well as better visual acuity and higher Bayley Psychomotor Indices at 13 month of age when compared with unsupplemented peers. The AAP Committee on Nutrition recently recommended that exclusively breastfed term infants receive a supplement of elemental iron at 1 mg/kg per day, starting at 4 months of age and continuing until appropriate iron-containing foods have been introduced. The preterm infant has lower iron content than the term infant and requires initiation of iron supplementation between 2 and 4 weeks of age and extending through 12 months of age.

The term infant has adequate calcium stores, in contrast to the preterm infant, who is born during the period when 80% of calcium, phosphorus, and magnesium are accrued. The term infant also has adequate folate stores. The routine use of a standard dose of intramuscular vitamin K at birth minimizes the risk of hemorrhagic disease of the newborn, which results from vitamin K deficiency.

As a result of reviewing this information, do you intend to make a change in practice to provide better patient care?

Yes No

Suggested reading:

Baker RD, Greer FR, and the Committee on Nutrition. Diagnosis and prevention of iron deficiency and iron-deficiency anemia in infants and young children (0—3 years of age). *Pediatrics*. 2010;126:1040-1050. Available at: http://pediatrics.aappublications.org/cgi/content/abstract/peds.2010-2576v1

Dee DL, Sharma AJ, Cogswell ME, Grummer-Strawn LM, Fein SB, Scanlon KS. Sources of supplemental iron among breastfed infants during the first year of life. *Pediatrics*. 2008;122(suppl 2):S98-S104. DOI: 10.1542/peds.2008-1315m. Available at: http://pediatrics.aappublications.org/cgi/content/full/122/Supplement_2/S98

Kleinman RE. Iron. In: *Pediatric Nutrition Handbook*. 6th ed. Elk Grove Village, Ill: American Academy of Pediatrics; 2009:403-422

Wagner CL, Greer FR, and the Section on Breastfeeding and Committee on Nutrition. Prevention of rickets and vitamin D deficiency in infants, children, and adolescents. *Pediatrics*. 2008;122:1142-1152. DOI: 10.1542/peds.2008-1862. Available at: http://pediatrics.aappublications.org/cgi/content/full/122/5/1142

Question: 34

A mother in your pediatric practice recently delivered a 28 weeks' gestation infant who is in the neonatal intensive care unit. The woman exclusively breastfed her previous child, who was born at 36 weeks' gestation. She is concerned that something is wrong with her milk for this infant because it is being combined with human milk fortifier before being given to her infant. You reassure her by explaining that fortification helps to meet the additional needs of her preterm infant.

Of the following, the MOST important role of such fortification is to

A. augment the immunologic properties of human milk

B. boost the carbohydrate content of human milk

C. decrease the osmolality of human milk

D. enhance the absorption of iron from human milk

E. increase the protein content of human milk

Critique: 34 Preferred Response: E

The protein content of human milk must be increased to meet the requirements of a preterm infant. The estimated protein requirement for a preterm infant is 3.0 to 4.0 mg/kg per day compared to 1.5 to 2.0 mg/kg per day for the term infant. The requirement for the preterm infant does not include additional allowances for catch-up growth that are due to losses of lean body weight prior to the infant surpassing birthweight. If these allowances are included, the recommended protein content increases to 3.4 to 4.2 mg/kg per day.

Milk supplied by the preterm infant's mother is the preferred enteral feeding. Although milk produced by a mother who delivers prematurely may have increased protein content compared to term milk, the protein content declines in the first weeks of lactation to that of term milk. Human milk fortifier supplies additional protein, which has been shown to increase concentrations of blood urea nitrogen and weight gain in the enterally feeding preterm infant.

Human milk fortifier does not augment the immunologic properties of human milk. Human milk contains secretory immunoglobulin A as well as other factors such as lactoferrin and lysozyme that confer immunologic protection upon the infant. The carbohydrate content of preterm human milk may be slightly lower than term human milk, but it is well tolerated in spite of the intestinal lactase functioning at 30% of the level of a term infant. The iron content of human milk is lower than that of iron-fortified formula, but nearly 50% is absorbed. Human milk fortifier does not significantly increase the carbohydrate content or enhance the absorption of iron. Of note, the addition of human milk fortifier does increase the osmolality of enteral human milk feedings.

Suggested reading:

Adamkin DH. Nutrition management of the very-low birthweight infant: II. Optimizing enteral nutrition and postdischarge nutrition. *NeoReviews*. 2006;7:e608-e614. Available at: http://neoreviews.aappublications.org/cgi/content/full/7/12/e608

Kleinman RE. Nutritional needs of the preterm infant. In: *Pediatric Nutrition Handbook*. 6th ed. Elk Grove Village, Ill: American Academy of Pediatrics; 2009:79-112

Kuschel CA, Harding JE. Multicomponent fortified human milk for promoting growth in preterm infants. *Cochrane Database Syst Rev.* 2004;1:CD000343. DOI: 10.1002/14651858.CD000343.pub2. Available at: http://www.mrw.interscience.wiley.com/cochrane/clsysrev/articles/CD000343/frame.html

Premji SS, Fenton TR, Sauve RS. Higher versus lower protein intake in formula-fed low birth weight infants*. Cochrane Database Syst Rev*. 2006;1:CD003959. DOI:10.1002/14651858.CD003959.pub2. Available at: http://www.mrw.interscience.wiley.com/cochrane/clsysrev/articles/CD003959/frame.html

Question: 145

You are caring for an 18-year-old girl who was diagnosed 6 months ago with Crohn disease affecting the terminal ileum and colon. Her medications include prednisone 10 mg/day, sulfasalazine 1 g three times per day, and 6-mercaptopurine 75 mg/day. She has experienced an excellent symptomatic response to treatment and presents today complaining only of occasional streaks of blood in an otherwise normal daily bowel movement. Since diagnosis, she has gained 2.5 kg on a regular diet with only iron supplementation.

Of the following, the girl is MOST at risk of developing a deficiency of

A. copper

B. folic acid

C. selenium

D. vitamin D

E. zinc

Critique: 145 Preferred Response: B

Malnutrition is a common finding in inflammatory bowel disease (IBD), particularly Crohn disease. Although the cause is multifactorial, decreased energy intake is the most frequent contributing factor. Other causes include increased energy requirements, enteric losses, malabsorption, and drug-nutrient interactions. As a result, up to 85% of pediatric patients who have Crohn disease exhibit documented weight loss at the time of diagnosis, and growth failure has been described in 15% to 40% of affected children. Although nutrient deficits are more prevalent in patients who have extensive small bowel involvement and in those who exhibit increased disease activity, nutritional problems are not limited to these subgroups.

In most cases, nutritional status may be improved and catch-up growth realized by enteral alimentation (which also may serve as primary disease therapy), nutritional supplements, and controlling symptoms, which increases appetite.

Treatment for the girl described in the vignette included the use of sulfasalazine, a known competitive inhibitor of folate absorption. Accordingly, she is at risk of developing folic acid deficiency. All patients receiving this drug, many of whom already may manifest reduced folate stores, should receive dietary supplementation with folic acid to prevent a deficiency state. Because, in part, of this property of sulfasalazine, its use in IBD largely has been supplanted by the 5-aminosalicylate analogs, which do not interfere with folate metabolism.

Although one recent study in pediatric patients demonstrated normal folate stores at the time of Crohn disease diagnosis, previous work in adults found both reduced folate concentrations and impaired folate absorption. Low folate stores may result from several factors, including reduced intake of green, leafy vegetables; increased disease activity; and drug-nutrient interactions. The clinical consequences of this micronutrient deficit in IBD have not been fully elucidated, but reduced folate has been associated with elevated homocysteine concentrations in children who have both Crohn disease and ulcerative colitis, suggesting a possible link between folate deficiency and the thrombotic complications of these disorders. Deficiencies of vitamins B6 and B12, the latter a particular concern in patients who have extensive ileal disease, also have been linked to elevated concentrations of homocysteine.

Iron deficiency is the most common mineral deficiency associated with Crohn disease and is usually due to enteric blood loss. Children who have Crohn disease may exhibit reduced serum concentrations of copper, selenium, and zinc. These mineral deficiencies appear to be related directly to disease activity and develop as a result of both enteric loss and oxidative stress. Zinc deficiency also has been associated with impaired metabolism of retinol-binding protein, leading to vitamin A deficiency. However, such nutrient deficits are unlikely to develop in patients who exhibit low disease activity indices.

Oxidative stress may lead directly to impaired vitamin A and vitamin E nutriture. In such cases, concentrations normalize in response to control of disease activity. Vitamins A and E, as well as vitamins D and K, are fat-soluble and require micellar solubilization for absorption. Accordingly, nutritional adequacy may be a concern in the setting of extensive ileal disease leading to bile salt loss and fat malabsorption.

Maintenance of skeletal health is an important objective in the care of patients who have Crohn disease. This is especially critical in children, for whom dysregulation of calcium and vitamin D metabolism may lead to osteopenia and inhibit skeletal growth. Problems related to

calcium homeostasis have been demonstrated in those who have Crohn disease, independent of glucocorticoid use. Studies have shown that a significant percentage of patients exhibit increased 1,25-dihydroxyvitamin D and reduced 25-hydroxyvitamin D concentrations, probably because of the overexpression of 1-alpha-hydroxylase in inflamed small bowel mucosa. Low concentrations of 25-hydroxyvitamin D result in reductions in serum calcium that, in turn, lead to increased parathormone secretion. As a consequence, 1,25-dihydroxyvitamin D concentrations rise, stimulating bone resorption and calcium release.

Suggested reading:

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Question: 216

A 15-year-old, who wishes to join the school track team, decided to test his endurance with a 3-hour run. He drank water before and during the run to maintain hydration, but he fainted and was taken to an emergency department. You advise him about fluids and nutrition and make specific suggestions for days when he plans to run for more than 1 hour.

Of the following, the MOST appropriate advice for these days is for him to ingest

A. caffeine-containing drinks during and after long runs

B. carbohydrate drinks before, during, and after long runs

C. daily amino acid supplements

D. high-protein meals 3 to 6 hours before long runs

E. increased amounts of water after the run

Critique: 216 Preferred Response: B

Optimal exercise performance requires good hydration, with attention given to fluids before, during, and after exercise (Item C216). Fluid requirements for an athlete vary by the type of sport; the duration of activity; the ambient temperature; and individual factors such as body weight, genetics, and sweating rates. In addition to water, electrolyte and carbohydrate intake becomes important when sports participation lasts longer than 1 hour. Accordingly, the boy described in the vignette should ingest carbohydrate drinks before, during, and after long runs.

A 1% loss in body weight with exercise can result in an increased heart rate and impaired heat transfer to the skin and environment. Loss of 1 kg of weight is equivalent to loss of 1 L of fluid. Dehydration or loss of 2% of body weight impairs performance, cognition, short-term memory, attention, and visual motor tracking and increases the risk for heat exhaustion and heat stroke. Water alone is adequate for exercise that lasts 1 hour or less. Excessive water intake puts the athlete at risk for hyponatremia, which may have been the case for the boy in the vignette.

The recommended daily allowance for protein is 0.95 g/kg per day in 4 to 13 year olds, 0.85 g/kg per day in 14 to 18 year olds, and 0.8 g/kg per day in adults. No evidence currently supports the need for additional dietary protein requirements in healthy adolescents who undertake endurance or resistance exercise. Supplements of amino acids and other proteins do not enhance performance, and long-term studies on their safety have not been conducted. A high-carbohydrate, low-fat meal is recommended before an endurance event.

Energy drinks are marketed for their stimulant effect and may contain caffeine, vitamins, and herbal supplements. They have the same amount of calories as standard soft drinks. Ingestion of more than 400 mg of caffeine daily by adults has the potential to induce behavioral, cardiovascular, bone, and reproductive effects, and ingestion of more than 100 mg by adolescents may result in high blood pressure. Sports drinks are manufactured for those undertaking physical activity lasting more than 1 hour at a time and contain carbohydrates and electrolytes. These drinks reduce fatigue by maintaining hydration, electrolyte balance, and blood glucose concentrations during endurance sports. Use should be at a rate consistent with sweat loss. The sodium in these drinks helps with water and glucose absorption from the small intestine and is especially important in maintaining hydration during exercise in the heat.

Suggested reading:

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Critique: 216

3–4 h before exercise	Carbohydrate: 4 g/kg
1–2 h before exercise	Carbohydrate: 0.5–1 g/kg
	Fluid: 90–180 mL if weight <40 kg 180–360 mL if ≥40 kg
During exercise (especially if >1 h duration)	Carbohydrate: 0.7 g/kg per h, divided every 15–20 min
	Fluid: 150 mL every 20 min if weight approx 40 kg 250 mL every 20 min if weight approx 60 kg
After exercise	Carbohydrate: 1–1.5 g/kg
	Fluid: replace losses: 450–680 mL/0.5 kg of body weight lost
	Protein: approx 0.2–0.4 g/kg

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ITEM C216: Dietary recommendations before, during, and after physical exercise.

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Dietary recommendations before, during, and after physical exercise.

Question: 232

You are evaluating a 15-year-old girl who runs track and has completed orthopedic treatment for a stress fracture of her tibia. You note that she is shorter than other family members. She has had a few light periods since menarche 1 year ago. Her mother states that she has always been a picky eater, but the girl has experienced no weight loss.

Of the following, the MOST appropriate anticipatory guidance for the girl is to

A. begin combined oral contraception

B. do stretching exercises after running

C. increase her calcium intake

D. increase her overall nutritional intake

E. start daily vitamin D supplements

Critique: 232 Preferred Response: D

Active individuals need proper diets to meet increased energy and fluid needs and maintain their body weights, replenish their glycogen stores, and provide protein to build and repair tissue. The needs depend on sex; body size; and the activity undertaken, its duration, and its intensity. Dietary components include macronutrients (carbohydrates, protein, and fat) and micronutrients (fluids, electrolytes, vitamins, and minerals). Most athletes increase their intake to accommodate these increased needs. If adequate energy is ingested from a variety of foods to maintain body weight, vitamin and mineral supplements are not needed.

Inadequate intake of energy, protein, and some micronutrients is more common among participants in sports that have weight requirements; encourage leanness; and require clothing that is contour-revealing, such as gymnastics, ballet, diving, cheerleading, distance running, and weight lifting. Female athletes, who are more likely to participate in most of these activities, are at greater risk for nutritional complications than males. Chronic energy deficits, which the girl described in the vignette most likely is experiencing, can result in impaired growth and development in younger athletes, loss of body weight, disruption of endocrine function, loss of strength and endurance, increased risk for injuries (including stress fractures), and compromised immune function. Female athletes are at risk for the female athlete triad that includes the following three interrelated components: disordered eating, amenorrhea, and osteoporosis. Calcium, vitamin D, and weight-bearing exercises are needed to achieve peak bone mass in all adolescents. Calcium requirements do not change with athletic participation. However, if intake of these micronutrients is inadequate and caloric intake is decreased, the risk for stress fractures increases, as for this girl. Supplementation with calcium, vitamin D, or estrogen (combined oral contraception) without increasing caloric intake does not reverse bone changes such as osteopenia or osteoporosis. Stretching after exercise does not affect bone health.

Suggested reading:

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