TABLE 2-4 Potential Complications of a Vegetarian Diet

Calcium absorption may be inhibited as a consequence of the presence of phytates in plant foods; vegetarian diets are a risk for pregnant women, children, and adolescents if calcium intake is not carefully planned.

Iron Deficiency Anemia. Females should be sure to obtain an adequate amount of absorbable iron (Sharma et al, 2003). The iron in dairy, eggs, and plant foods is largely nonheme, of which only about 2–20% is absorbed.

Excess Fiber or Inadequate Energy Intake. In some circumstances, this regimen can restrict energy intake in the first few years of life (Murphy and Allen, 2003). This is also true for adults who consume large amounts of fiber to the extent that other nutrients are not able to be absorbed in the small intestine.

Omega-3 Fatty Acids and essential amino acids **methionine** and **lysine** are found in significantly lower amounts in vegetarian diets (Mezzano et al, 2000). It may be necessary to use a supplemental form.

Protein may be limited. Suggest complementary food combinations to acquire all amino acids.

Vitamin B_{12} Deficiency. An individual following a vegan diet should use supplements to obtain this vitamin (Stabler and Allen, 2004).

Vitamin D Deficiency or Rickets. The human body can synthesize vitamin D from sunlight, but this is only possible when the sun reaches a certain intensity level. For people who live in northern latitudes, for a few months of the year, they will have to seek other sources of vitamin D. Milk is generally fortified with vitamin D; for vegans who do not consume dairy products, supplements are necessary (Outila et al, 2000). A very low-fat vegan diet can be nutritionally adequate with the exception of vitamin D; supplementation is needed (Dunn-Emke et al, 2005).

Zinc intake may be lower in vegetarian diets (Hunt, 2003).

INTERVENTION

OBJECTIVES

- Encourage use of a wide variety of foods in adequate quantity with a mix of nutrients and amino acids throughout the day.

SAMPLE NUTRITION CARE PROCESS STEPS

Excessive Fiber Intake

Assessment Data: Food records; adverse side effects from high fiber intake; low BP; altered nutritional labs for calcium and iron.

Nutrition Diagnoses (PES): Excessive fiber intake related to vegan lifestyle as evidenced by complaints of excessive gas at night and intake of 45+ g of fiber daily, especially with large amounts at dinner.

Interventions: Food and Nutrition Delivery: ND 3.2.1 and ND 3.2.3 provide supplementation of a multivitamin and vitamin B_{12}; ND-1.4 educate and counsel patient on following a healthy vegan diet and teach patient how to plan and monitor diet carefully.

Nutrition Education: E-2.2 Registered dietitian (RD) to provide recommend modifications to patient's diet through instruction and training to lead to a better understanding of the importance of monitoring vegan diet and supplementing to get required nutrients not supplied through diet.

Counseling: C-2.4 Problem Solving: RD will teach and counsel patient on how to get nutrients she needs while continuing to follow a vegan diet. They will discuss solutions to patient's vitamin B_{12} deficiency by discussing vegan foods that are fortified with vitamin B_{12} and foods such as nutritional yeast that will provide her with a source of vitamin B_{12} in her diet.

Coordination of Care: RC-1.2 Refer patient to RD specializing in vegetarianism.

Monitoring and Evaluation: Improved quality of life with reduced symptoms of gas; lab reports for calcium, iron, ferritin; diet history revealing intake of fiber within desired range of 25–35 g/d.

- Provide nutritionally adequate menus with sufficient energy for weight goals. Discourage excessive use of sweets.
- Monitor the vegetarian diet carefully if the client is a pregnant woman, lactating mother, or elderly person. Infants, children, and teens on vegan diets should be monitored even more closely to ensure adequate energy intake and mineral and vitamin intakes (Perry, 2002). High-fiber diets may replace calories and cause some stunting or other growth deficits.
- Monitor fiber intake in general; excesses may interfere with absorption of calcium, zinc, and iron.
- Prevent or correct anemias, which could be either microcytic or macrocytic.
- The limiting amino acids in typical protein foods include wheat (lysine), rice (lysine and threonine), corn (lysine and tryptophan), beans (methionine), and chickpeas (methionine). Vary food mixtures such as using bread with milk, rice with cheese, or pasta with cheese; rice with beans, bread with beans, or corn and beans; garbanzo beans with sesame seeds (as in dips or in roasted snacks). Serve vegetables with nuts, dairy products, rice, sunflower seeds, or wheat germ. Different food combinations provide essential amino acids that produce higher quality proteins (American Dietetic Association, 2003).
- Plant sources of protein can provide adequate amounts of essential amino acids. Using a variety of plant foods is key, and energy needs should readily be met. Although vegetarian diets are lower in total protein, protein intake in both lacto–ovo–vegetarians and vegans appears to be adequate (Messina and Messina, 1996).
- Soy foods can be useful in reducing elevated Chol as part of a healthy vegetarian diet (Rosell et al, 2004).

FOOD AND NUTRITION

The American Dietetic Association recommends that vegetarians consult with a registered dietitian or other qualified nutrition professional, especially during periods of growth, breastfeeding, pregnancy, or recovery from illness.

- For a balanced diet, minimize intake of less nutritious foods such as sweets and fatty foods. Choose whole or

unrefined grain products instead of refined products. Choose a variety of nuts, seeds, legumes, fruits, and vegetables, including good sources of vitamin C to improve iron absorption. Choose low-fat or nonfat varieties of dairy products, if they are included in the diet.
- Vegetarian foods rich in iron include many breakfast cereals, oatmeal, raisins, black beans, cashews, lentils, kidney beans, black-eyed peas, soybeans, hempseed, sunflower seeds, chickpeas, molasses, whole-wheat bread. Follow the food guide for North American vegetarians by including: 6–12 servings from the bread group, 2–3 servings of protein-rich foods such as legumes, nuts and seeds, or eggs (if used), 2–3 servings from the dairy group as tofu, yogurt, or fortified soy milk, 4 or more servings of vegetables, 3 or more servings of fruits, 2–3 servings of fats and oils, including olives and avocado.

Common Drugs Used and Potential Side Effects

- Monitor use of medications that deplete vitamins and minerals, especially iron and B-complex vitamins.

Herbs, Botanicals, and Supplements

- Many cultures use herbs and botanicals as part of their meal patterns, rituals, and celebrations. Identify those that are used and monitor side effects. Counsel about use of herbal teas, especially regarding toxic substances.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Beneficial changes to diet may occur on changing to a self-selected vegetarian diet; for example, it is one way of achieving a better blood lipid profile (Robinson et al, 2002).
- Explain patterns of food intake that provide complementary amino acids. Whole grains, legumes, seeds, nuts, and vegetables contain sufficient essential and nonessential amino acids if taken in the right combinations.
- Emphasize the importance of a balanced diet.
- Describe the role vegetarian diets play in lowering serum Chol, TG, and glucose. These are beneficial changes that can result after starting a vegetarian diet (Phillips et al, 2004).
- Counsel about appropriate products for infants and children, as protein may be the biggest problem. Soy milk should be fortified with calcium and vitamin B_{12}.
- Unless otherwise advised by a doctor, those taking dietary supplements should limit the dose to 100% of the Daily Reference Intakes.

Patient Education—Food Safety

- Discuss food handling, preparation, and storage, especially careful washing of fruits and vegetables. Spinach and sprouts have been contaminated in recent years; wash produce thoroughly. Discuss hand washing.
- Starches such as hot cereals and rice should not be prepared and held in large batches because of the risks of *Bacillus cereus*.

For More Information

- Food and Nutrition Information Center
  http://www.nal.usda.gov/fnic/pubs/bibs/gen/vegetarian.pdf
- Hindu Food Practices
  http://monarch.gsu.edu/WebRoot$/multiculturalhealth/handouts/Hindi/Hindi_food_pyramid.pdf.
- Lacto-Ovo Vegetarian Cuisine
  http://www.nhlbi.nih.gov/health/public/heart/obesity/lose_wt/lacto_ov.htm
- North American Vegetarian Society
  http://www.navs-online.org/
- Oldways Preservation and Trust
  http://www.oldwayspt.org/
- Soy Connection
  http://www.soyconnection.com/newsletters/soy-connection/health-nutrition/index.php
- Seventh-Day Adventist Diet
  http://www.sdada.org/aboutsda.htm
  http://www.andrews.edu/NUFS/veggiediet.html
- UCLA Vegetarian Nutrition
  http://apps.medsch.ucla.edu/nutrition/vegetarianism.htm
- Vegetarian Cuisine and Recipes
  http://vegweb.com/
- Vegetarian Diets for pregnancy
  http://my.clevelandclinic.org/healthy_living/pregnancy/hic_nutrition_during_pregnancy_for_vegetarians.asp
- Vegetarian Network (Victoria, Australia)
  http://www.vnv.org.au/
- Vegetarian Recipes
  http://allrecipes.com/HowTo/Vegetarian-Cuisine/Detail.aspx
- Vegetarian Resource Group
  http://www.vrg.org/
- Vegetarian Recipes for Teens
  http://kidshealth.org/teen/recipes/
- Vegetarian Society of the United Kingdom
  http://www.vegsoc.org/
- World Guide to Vegetarianism
  http://www.veg.org/veg/

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Mezzano D, et al. Cardiovascular risk factors in vegetarians. Normalization of hyperhomocysteinemia with vitamin B_{12} and reduction of platelet aggregation with omega-3 fatty acids. *Thromb Res.* 100:153, 2000.

Murphy SP, Allen LH. Nutritional importance of animal source foods. *J Nutr.* 133:3932S, 2003.

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Perry CL, et al. Adolescent vegetarians: how well do their dietary patterns meet the Healthy People 2010 objectives? *Arch Pediatr Adolesc Med.* 156:431, 2002.

Phillips F, et al. Effect of changing to a self-selected vegetarian diet on anthropometric measurements in UK adults. *J Hum Nutr Diet.* 17:249, 2004.

Robinson F, et al. Changing from a mixed to self-selected vegetarian diet—influence on blood lipids. *J Hum Nutr Diet.* 15:323, 2002.

Rosell MS, et al. Soy intake and blood cholesterol concentrations: a cross-sectional study of 1033 pre- and postmenopausal women in the Oxford arm of the European Prospective Investigation into Cancer and Nutrition. *Am J Clin Nutr.* 80:1391, 2004.
Sharma JB, et al. Effect of dietary habits on prevalence of anemia in pregnant women of Delhi. *J Obstet Gynaecol Res.* 29:73, 2003.
Siener R, Hesse A. The effect of a vegetarian and different omnivorous diets on urinary risk factors for uric acid stone formation. *Eur J Nutr.* 42:332, 2003.
Stabler SP, Allen RH. Vitamin B_{12} deficiency as a worldwide problem. *Annu Rev Nutr.* 24:299, 2004.
Szeto YT, et al. Effects of a long-term vegetarian diet on biomarkers of antioxidant status and cardiovascular disease risk. *Nutrition.* 20:863, 2004.

EASTERN RELIGIOUS DIETARY PRACTICES

NUTRITIONAL ACUITY RANKING: LEVEL 2 (ADVISEMENT/PLANNING)

DEFINITIONS AND BACKGROUND

Hinduism, Jainism, and Sikhism

Hindus may be vegetarian while adhering to *ahimsa*, related to nonviolence as applied to the infliction of pain on animals. Beef is never eaten (the cow is considered sacred), and pork is usually avoided. Foods prohibited may include snails, crab, poultry, cranes, ducks, camels, boars, and some types of fish. The Brahmins, "high caste" folk, have stricter rules and practices, and there are differences between the North Indian Brahmins and the South Indian Brahmins. Some foods promote purity of the body, mind, and spirit. Devout Hindus avoid alcoholic beverages and foods that stimulate the senses, such as garlic and onions. Feast days include Holi, Dusshera, Pongal, and Divali (varying each year according to the lunar calendar). In addition, personal feast days include the anniversaries of birthdays, marriages, and deaths. Fasting depends on a person's social standing (caste), family, age, gender, and degree of orthodoxy. Fasting can be complete, adopting a completely vegetarian diet, or it can be abstaining from favorite foods.

Jainism is a branch of Hinduism that also promotes the nonviolence of ahimsa. Jains are expected to practice nonviolence, including against animals. Devout Jains are complete vegans. They avoid blood-colored foods (tomatoes) and avoid root vegetables, which may result in the death of insects clinging to the vegetable when it is harvested. Jains drink only boiled water. Fasting is a tool for connecting with the inner being during festivals. Fasting is based on three levels of austerity: Uttam, Madhyam, and Jaghanya. When one has finished with the roles of life, he or she willingly gives up food and drink; this can take up to 12 years with a gradual decline in eating.

Sikhs participate in many Hindu practices but differ by their belief in a single God. Sikhs abstain from beef and alcohol, but pork is permitted. Everyone is equal, no matter what color, sex, race, wealth, height, weight or religion; there is only one true race, the human race. Everyone sits on the floor when eating, as equals.

Buddhism

Buddhist dietary customs vary considerably depending on sect (Theravada or Hinayana, Mahayana, Zen) and on country of origin. Most Buddhists also subscribe to the concept of ahimsa, and many are lacto–ovo–vegetarians. Some eat fish, whereas some only abstain from beef. Some believe that unless they personally slaughter an animal, they may eat its meat.

Buddhist monks fast completely on the days of the new moon and full moon each lunar month; they also avoid eating any solid food after the noon hour. Buddhist feasts vary from one region to another. Celebrations include the birth, enlightenment, and death of Buddha in Mahayana Buddhism; the 3 days are unified into the single holiday of Vesak for Theravada Buddhism.

Buddhist vegetarian diets tend to allow more natural insulin sensitivity, so diabetes is less common (Kuo et al, 2004). However, serum tHcy should be monitored because of possibly lower intakes of vitamin B_{12} (Hung et al, 2002).

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Clinical/History

Height
Weight
BMI
Recent weight changes
Diet history
BP

Lab Work

Gluc
Chol, Trig
Serum Na^+, K^+
Ca^{++}, Mg^{++}
Alk phos
H & H, serum Fe

SAMPLE NUTRITION CARE PROCESS STEPS

Inadequate Mineral (Iron) Intake

Assessment Data: Food records showing low intake of heme iron; altered nutritional labs for iron and ferritin; normal folate and B_{12} levels; complaints of easy fatigue and irritability.

Nutrition Diagnoses (PES): Inadequate iron intake related to Hindu (vegan) lifestyle as evidenced by intake of 4–5 g nonheme iron daily and low-serum Fe and ferritin levels.

Intervention: Education about increasing intake of iron-rich foods while decreasing excess of wheat bran. Counseling about using iron-fortified cereals or a supplement that provides 100% DRI for iron.

Monitoring and Evaluation: Improved energy and less fatigue; improved lab reports for iron and ferritin; diet history revealing improved intake of nonheme iron with supplements as needed.

INTERVENTION

OBJECTIVES

- Serve appropriate menu choices, and omit foods or beverages that are not permitted.
- Respect traditions and preferences of the individual and family members.

FOOD AND NUTRITION

- Support dietary practices as followed by the individual and family members.
- Counsel about specific nutritional changes according to the medical diagnosis and current condition.

Common Drugs Used and Potential Side Effects

- During periods of fasting, identify potential interactions from drugs that are dependent on energy sources for their metabolism.

Herbs, Botanicals, and Supplements

- Many cultures use herbs and botanicals as part of their meal patterns, rituals, and celebrations. Identify those that are used and monitor side effects.
- Counsel about use of herbal teas, especially regarding toxic substances.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Show the patient how to prepare foods to reduce Chol, fat, or sodium if heart disease or hypertension is present.
- Various types of cancer may prevail in different parts of the world and in different cultures. Discuss diet in relationship to what is common.
- While medical students and physicians with healthful personal practices (such as vegetarianism) are more likely to encourage such behaviors in their patients, these beliefs do not affect their actual nutrition counseling (Spencer et al, 2007).

Patient Education—Food Safety

- Discuss safe preparation and storage of foods to reduce likelihood of bacterial contamination.

For More Information

- Asian Foods
  http://www.asiafood.org/
- Asian Society
  http://www.asiasociety.org/
- Buddhism
  http://www.buddhanet.net/
- Ethnic Recipes
  http://asiarecipe.com/religion.html
- Hinduism
  http://www.hindunet.org/vegetarian/
- International Studies
  http://www.internationaled.org/
- Jainism
  http://www.diversiton.com/religion/main/jainism/holydays-festivals-rituals.asp
- Sikhism
  http://jainguru.com/diets.html

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Kuo CS, et al. Insulin sensitivity in Chinese ovo-lactovegetarians compared with omnivores. *Eur J Clin Nutr.* 58:312, 2004.

Spencer EH, et al. Personal and professional correlates of US medical students' vegetarianism. *J Am Diet Assoc.* 107:72, 2007.

WESTERN RELIGIOUS DIETARY PRACTICES

NUTRITIONAL ACUITY RANKING: LEVEL 2 (ADVISEMENT/PLANNING)

DEFINITIONS AND BACKGROUND

Judaism (Edited by Rabbi Allan Bernstein)

Jewish congregations in the United States are either identified as Orthodox, Conservative, or Reform. Orthodox Jews believe the laws are the direct commandments of God, to be explicitly followed by the faithful. Reform Jews follow the moral law but believe that the laws are still being interpreted (some are considered dated or currently irrelevant) and may be observed selectively. Conservative Jews fall in between the other congregations in their beliefs and adherence to the laws. About 25–30% of Jews in America keep kosher to one extent or another (http://www.jewfaq.org/kashrut.htm).

Jewish dietary laws are known as *Kashrut* and are among the most complex of all religious food practices. The term *kosher*, or *kasher*, means "fit" and describes all foods that are permitted for consumption. Kosher is loosely used to identify Jewish dietary laws, and to "keep kosher" means that the laws are followed. The dietary laws are complex. Briefly, they include what foods are fit to eat, what foods are prohibited (a lengthy list that includes pork, shellfish, and other foods), how animals must be slaughtered, how they must be prepared, and when they may be consumed (specifically, rules regarding when milk products can be consumed with meat products).

Jewish feast days include Rosh Hashanah, Sukkot, Hanukkah, Purim, Passover, and Shavout (dates vary because Judaism uses a lunar calendar). Specific foods are associated with the feasts but may differ nationally. Complete fast days (no food or water from sunset to sunset) include Yom Kippur and Tisha b'Av. Partial fast days (no food or water from sunrise to sunset) include Tzom Gedaliah, Tenth of Tevet and Seventeenth of Tamuz, Ta'anit Ester, and Ta'anit Bechorim. Special kosher laws are observed during Passover, including the elimination of any products that can be leavened.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Clinical/History

Height
Weight
BMI
Recent weight changes
Diet history
BP

Lab Work

Gluc
Chol, Trig
Serum Na^+, K^+
Ca^{++}, Mg^{++}
Alk phos
H & H, serum Fe

INTERVENTION

OBJECTIVES

- Observe dietary practices as followed by the laws of Judaism: meats are limited to cud-chewing animals with cloven hooves (cows and sheep) that are properly slaughtered. Pork (including ham and all pork products), shellfish, and scavenger fish are forbidden.
- Separate utensils are used for preparation and eating and especially for separating meat and milk foods.
- Monitor the kosher diet, which tends to be high in Chol, saturated fats, and sodium. Encourage application of the DASH diet principles where possible, but reduce lactose and sodium if necessary.

FOOD AND NUTRITION

The Jewish dinner table follows these guidelines (http://www.jewfaq.org/kashrut.htm):

- Certain animals may not be eaten at all. This restriction includes the flesh, organs, eggs, and milk of the forbidden animals. No pork, ham, bacon, pork products, rabbit, shellfish, or eel may be eaten.
- Of the animals that may be eaten, the birds and mammals must be killed in accordance with Jewish law. All blood must be drained from the meat or broiled out of it before it is eaten. Certain parts of permitted animals may not be eaten. Sheep, cattle, goats, and deer are kosher.
- Meat (the flesh of birds and mammals) cannot be eaten with dairy. Fish, eggs, fruits, vegetables, and grains can be eaten with either meat or dairy. According to some views, fish may not be eaten with meat.
- Dairy: Milk may be consumed before a meal, but once meat is eaten, 3–6 hours (depending on individual traditions) must pass before dairy products can be consumed. Omit lactose if not tolerated; provide other sources of calcium and riboflavin.
- Utensils that have come into contact with meat may not be used with dairy and vice versa. Utensils that have come into contact with nonkosher food may not be used with kosher food.
- Fruits, vegetables, and grains can be used, except that breads made with milk products are forbidden with meat meals. Grape products made by non-Jews may not be eaten.
- Leavened (raised) bread is forbidden during Passover. Matzoh bread or crackers may be used. Haroset and fried matzoh are traditional Passover foods. Seder plates and other items appropriate for the Seder dinner are important additions to the menu at this time.
- Common food choices include matzoh ball soup, chicken soup with kreplach, gefilte fish with beet horseradish,

cheese blintz with sour cream, flanken tzimmes, chopped liver, noodle Kugel, and kishka. Frozen kosher meals may be available in some areas.
- Fasting is common during Yom Kippur.
- Traditional Hanukkah foods include latkes and sour cream or applesauce.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Show the patient how to limit foods high in Chol/fat if weight and elevated lipid levels are a problem.
- Discuss sodium and obesity in relationship to hypertension, as appropriate. Recommend other herbs, spices, and cooking methods.
- Low-fat cheeses should be substituted for high-fat cheeses such as cream cheese.
- Note that food labels with a "U" with an "O" encircling it are considered kosher. Many other foods are considered kosher, but an inquiry should be made.
- Discuss holiday preferences and alternatives when needed.

Patient Education—Food Safety

- Discuss safe preparation and storage of foods to reduce likelihood of bacterial contamination.

For More Information

- Determining Kosher
  http://www.ou.org/kosher/primer.html
- Hebrew Food Pyramid
  http://monarch.gsu.edu/WebRoot$/multiculturalhealth/handouts/hebrew//Hebrew_food_pyramid.pdf
- Judaism 101
  http://www.jewfaq.org/kashrut.htm
- Kashrut–Dietary Laws
  http://www.myjewishlearning.com/daily_life/Kashrut.htm
- Kosher certification
  http://www.okkosher.com/
- Kosher recipes
  http://www.okkosher.com/Content.asp?ID=79
- Kosherfest
  http://www.kosherfest.com/
- Union for Traditional Judaism
  http://www.utj.org/

Christianity

There are three major branches of the Christian faith: Roman Catholicism, Eastern Orthodox Christianity, and Protestantism. Dietary practices vary; some are minimal.

(1) Roman Catholicism: Devout Catholics observe several feast and fast days during the year. Feast days include Christmas, Easter, the Annunciation (March 25th), Palm Sunday (the Sunday before Easter), the Ascension (40 days after Easter), and Pentecost Sunday (50 days after Easter). Catholics in each country observe many food traditions. Fasting (one full meal per day permitted; snacking according to local custom) and/or abstinence (meat is prohibited, but eggs, dairy products, and condiments with animal fat are permitted) may be practiced during Lent, on the Fridays of Advent, and Ember Days (at the beginning of the seasons) by some Catholics; some fast or abstain only on Ash Wednesday and Good Friday. Today, Catholics may avoid meat only on the Fridays of Lent (40 days before Easter). Food and beverages (except water) are to be avoided for 1 hour before communion is taken.

(2) Eastern Orthodox Christianity: The 14 self-governing churches that form the Orthodox Church differ from Catholicism in their interpretation of the Biblical theology, including the use of leavened bread instead of unleavened wafers in communion. Numerous feast and fast days are observed (dates vary according to whether the Julian or Gregorian calendar is used). Feast days include Christmas, Theophany, Presentation of the Lord into the Temple, Annunciation, Easter, Ascension, Pentecost Sunday, the Transfiguration, Dormition of the Holy Theotokos, Nativity of the Holy Theotokos, and Presentation of the Holy Theotokos. In addition, Meat Fare Sunday is observed the third Sunday before Easter (all meat in the house is consumed, and none is eaten again until Easter). Cheese Fare Sunday is observed on the Sunday before Easter (all cheese, eggs, and butter are consumed). On the next day, Clean Monday, the Lenten fast begins. Food and drink are avoided before communion.

Meat and all animal products (milk, eggs, butter, and cheese) are prohibited on fast days; fish is avoided, but shellfish is permitted. Some devout followers may avoid olive oil on fast days, too. Fast days include every Wednesday and Friday (except for three fast-free weeks each year), the Eve of Theophany, the Beheading of John the Baptist, and Elevation of the Holy Cross. Fast periods include Advent, Lent, the Fast of the Apostles, and Fast of the Dormition of the Holy Theotokos.

(3) Protestantism: The only feast days common in most Protestant religions are Christmas and Easter. Few practice fasting.

(4) Mormons (Church of Jesus Christ of Latter Day Saints): Mormons avoid alcoholic beverages, hot drinks (coffee and tea), and caffeine-containing drinks. Followers are encouraged to eat mostly grains and to limit meats. Some Mormons fast 1 day a month and donate that food money to the poor.

(5) Seventh-Day Adventists avoid overeating. Most are lacto–ovo–vegetarians, but when meat is consumed, most avoid pork. Tea, coffee, and alcoholic beverages are prohibited. Water is consumed before and after meals. Eating between meals is discouraged. Strong seasonings and condiments, such as pepper and mustard, are avoided.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Clinical/History	Lab Work	
Height	Gluc	H & H, serum Fe
Weight	Chol, Trig	C-reactive protein (CRP)
BMI	Serum Na^+, K^+	
Recent weight changes	Ca^{++}, Mg^{++}	
Diet history	Alk phos	
BP		

INTERVENTION

OBJECTIVES

- Observe dietary practices as followed by the individual. Discuss the role of special meals, fasting, or events and plan menus accordingly.
- Assist guests and immigrants in maintaining their healthy dietary practices and religious traditions, as appropriate (Kaplan et al, 2004).

FOOD AND NUTRITION

- Promote a healthy diet. For example, principles of a Mediterranean diet may be suitable for many individuals (Bilenko et al, 2005).
- Fasting may be common during special holidays. Discuss concerns related to pregnancy, children, the elderly or those in a malnourished state.
- Some individuals avoid caffeine and alcohol as part of their religious preferences; honor those wishes.
- Determine if any foods are avoided on special days of the week and plan alternatives accordingly.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Show the patient how to limit foods high in Chol/fat if weight and elevated lipid levels are a problem.
- Discuss sodium and obesity in relationship to hypertension, as appropriate. Recommend other herbs, spices, and cooking methods.
- Discuss holiday preferences and alternatives where needed.
- There tend to be few specific relationships between religion, fat intake, and physical activity in contemporary U.S. society; religion may play only a small role in the context of how diet and exercise are developed and maintained (Kim and Sobal, 2004).

Patient Education—Food Safety

- Discuss safe preparation and storage of foods to reduce likelihood of bacterial contamination.

For More Information

- Andrews University–Seventh Day Adventist diet http://www.sdada.org/position.html

WESTERN RELIGIOUS DIETARY PRACTICES—CITED REFERENCES

Bilenko N, et al. Mediterranean diet and cardiovascular diseases in an Israeli population. *Prev Med.* 40:299, 2005.

Kaplan MS, et al. The association between length of residence and obesity among Hispanic immigrants. *Am J Prev Med.* 27:323, 2004.

Kim KH, Sobal J. Religion, social support, fat intake and physical activity. *Pub Health Nutr.* 7:773, 2004.

MIDDLE EASTERN RELIGIOUS DIETARY PRACTICES

NUTRITIONAL ACUITY RANKING: LEVEL 2 (ADVISEMENT/PLANNING)

DEFINITIONS AND BACKGROUND

Islam is an Arabic word that means submission, surrender, and obedience; it also means peace, as it is derived from the word "Salam," which means peace. As a religion, Islam stands for complete submission and obedience to God. Followers of the Islamic faith are known as Muslims. Muslims promote the concept of eating to live, not living to eat. They advise sharing food.

Prohibited foods as described in the Koran are called *haram*; those in question are *mashbooh*. Pork and birds of prey are haram; meats must be slaughtered properly. Alcohol is prohibited, and stimulants, such as coffee and tea, are allowed. *Halal* is the term for all permitted foods. The flesh of animals must be slaughtered according to Islamic law or *halal*; kosher items may be used for this reason. Feast days (dates vary according to the lunar calendar) include Eid al-Fitr, Eid al-Azha, Nau-Roz (a Persian holiday), Al-Ghadeer, and Maulud n'Nabi. Fasting is considered an opportunity to earn the approval of Allah, to wipe out sins, and to understand the suffering of the poor. Fasting includes abstention from all food and drink from dawn to sunset. Voluntary fasting is common on Mondays and Thursdays; it is undesirable to fast on certain days of the month and on Fridays. Muslims are required to fast during the entire month of Ramadan and are encouraged to fast 6 days during the month of Shawwal, on the Al-Ghadeer day, and on the 9th day of Zul Hijjah.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Clinical/History

Height
Weight
BMI
Recent weight changes
Diet history
BP

Lab Work

Gluc
Chol, Trig
Serum Na^+, K^+
Ca^{++}, Mg^{++}
Alk phos
H & H, serum Fe

INTERVENTION

OBJECTIVES

- During fasting, eating occurs only before dawn and after sunset. Plan accordingly.
- Monitor dietary patterns, which include fasting 3 days a month. Pregnant and breastfeeding mothers need not fast.
- Monitor need for vitamin D in women if sun exposure is minimal.

FOOD AND NUTRITION

- Pork and pork products are forbidden, including gelatin.
- Alcohol is not used, even in vanilla extract and other preparations.
- Foods such as dates, seafood, honey, sweets, yogurt, milk (goat's milk also), meat, and olive or vegetable oils are encouraged. Beef, chicken, and lamb are commonly used. Couscous, pita bread, rice, millet, and bulgur are frequently included. Eggplant, cucumbers, green peppers, pomegranates, and tomatoes are readily available.
- Typical combination foods include: falafel (grain, fat), hummus (grain, fat), kibbeh (meat, grain, fat), tabouli (vegetable, grain, fat), baba ghanouj (vegetable, fat), pilaf (grain, fat), stuffed grape leaves (meat, grain, fat), and shawarma (meat, grain, fat). Khoresh is a stew with meats (lamb, beef, or veal), poultry, or fish with vegetables; fresh or dried fruits; beans, grains, and even nuts.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- If diet is low in heme iron, anemia may occur. Discuss options if necessary.
- Fasting is not recommended for persons who have diabetes, cancer, or HIV/AIDS. Discuss menu planning for religious occasions.
- Discuss useful dietary changes for managing obesity and diabetes.

Patient Education

- Discuss periods of fasting if there are undesirable side effects, such as hypotension or fainting.

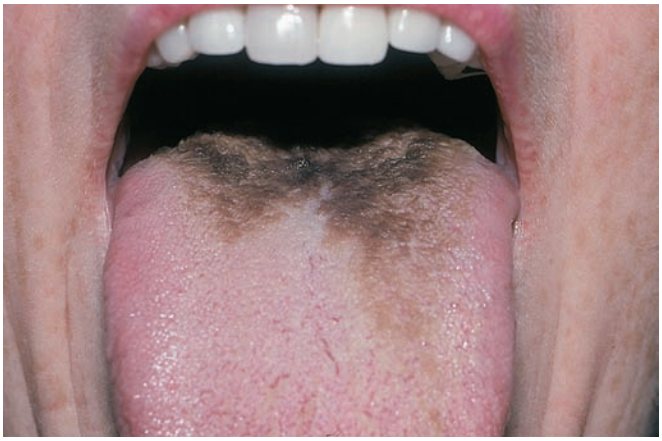
For More Information

- Catering for Muslim Patients
  http://www.med.umich.edu/multicultural/ccp/culture/muslim.htm
- Iranian Cooking
  http://www.asiafood.org/persiancooking/index.cfm
- Islamic Food and Nutrition Council of America
  http://www.ifanca.org/
- Jordanian Food
  http://www.gondol.com/English/food.htm
- Muslim Consumer group
  http://www.muslimconsumergroup.com/hfs.htm

OROFACIAL CONDITIONS

DENTAL DIFFICULTIES AND ORAL DISORDERS

NUTRITIONAL ACUITY RANKING: LEVEL 2–3



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DEFINITIONS AND BACKGROUND

Diet and nutrition affect many oral diseases. Cell turnover is rapid in the tongue and oral mucosa; therefore, the oral cavity is one of the first areas where signs of systemic disease appear. Two oral infectious diseases are diet related: dental caries (tooth decay related to diet composition and frequency), and periodontal disease, associated with malnutrition (Touger-Decker et al, 2003). In dental caries, chronic infectious disease leads to progressive destruction of tooth substances from interactions between bacteria and organic tooth compounds. *Streptococcus mutans* and *Lactobacillus* form acids within 20 seconds to 30 minutes after contact. Erosion of tooth enamel may occur in patients who chronically consume acidic beverages and/or keep such beverages or foods in the mouth for a period of time (e.g., sucking lemons, chewing vitamin C tablets, chewing lemon hard candies). Tooth loss can prevent proper bite and may lessen the ability

to chew foods properly. Dietary advice given when dentures are placed results in increased consumption of fruits and vegetables during stages of change (Moynihan, 2005a).

Health professionals should check the oral and dental health of their patients. Many Americans lack fluoridated water, an effective safeguard against dental cavities. Those who are poor or have no dental insurance are also at risk for caries. Water fluoridation can reduce caries by 20–40% (American Dental Association, 2009).

Some dental problems are age specific. Infants should be monitored for early childhood caries (ECC); dental decay often occurs during the growth spurts of adolescents; and older patients should be monitored for changes in eating habits, inadequate diet, and caries. Elderly persons who wear dentures are more prone to malnutrition (de Oliveria and Frigerio, 2004) and problems with chewing, swallowing, and mouth pain often precede hospitalizations (Bailey et al, 2004).

Poor oral hygiene can increase the likelihood of gingival abnormalities when vitamin C and D intakes have been poor. Some conditions, such as diabetes, can also make individuals prone to dry mouth and dental decay. An increase in water intake, extra care with oral hygiene, chewing sugarless gum, and prevention of periodontal disease are important steps.

With tongue disorders, mastication of food may be affected. The ability to push mashed food with the tongue and anterior hard palate will be affected. Other oral problems may cause pain, problems with chewing, dysphagia, mouth dryness, or infection including aphthous stomatitis, cheilosis, oral cancer, lichen planus, oral herpes, candidiasis, thrush, or xerostomia. Many of these conditions occur because of altered immunity and debility, as in cancer or HIV infection.

Fracture of the lower jaw (mandible) is an injury that requires intermaxillary fixation (wiring). Patients with wired jaws face a whole new lifestyle for up to 6 weeks following surgery. Patients have to eat liquefied meals; proper presurgical patient education is essential.

Proper nutrition is essential for good dental and oral health. Table 2-5 provides a list of the key nutrients needed for healthy oral mucosa and teeth. Vitamins A and C are significant for prevention and treatments of leukoplakias (Scully, 2000; Sheiham et al, 2001). Treatment with beta-carotene or retinoids is often recommended (Lodi et al, 2004). Table 2-6 lists dental problems, treatment, and prevention.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Clinical/History

- Height
- Weight
- BMI
- Recent weight changes
- Diet history
- Mouth, gum or tongue lacerations
- Dental caries
- Missing or loose teeth
- Dentures, missing, loose or ill-fitting
- Taste alterations
- Sore or bleeding gums
- High intake of sugars and sticky starches?

Lab Work

- Alb, transthyretin
- Serum Na^{+}, K^{+}
- Ca^{++}, Mg^{++}
- Alk phos
- H & H, serum Fe
- X-rays (mandible)
- Serum folate
- Serum ascorbate and retinol

TABLE 2-5 Nutrients Needed for Proper Oral Tissue Synthesis and Dental Care

Protein	Needed for healthy tissue growth and maintenance.
Vitamin A	Necessary for epithelial tissue and enamel. Beta-carotene may play a role in oral cancer prevention (Lodi et al, 2004).
Vitamin B-complex	Deficiencies show a bright scarlet tongue and stomatitis in niacin deficiency; magenta tongue, glossitis, and angular cheilitis in riboflavin deficiency; smooth tongue in vitamin B_{12} deficiency.
Folate	Needed for a healthy blood supply.
Vitamin C	Enables connective tissue cells to elaborate intercellular substances. Deficiency can lead to easy bleeding or swelling of gums and gingivitis. Forms collagen; helps to heal wounds and bleeding gums.
Vitamin K	Aids with calcium absorption in bone; adequate blood clotting; helps in healing.
Vitamin D	Protects against chronic inflammation of the gums, which can lead to gingivitis or periodontal disease (Dietrich et al, 2004). Necessary for dentin, bony tissue synthesis; mineralization; and jawbone sufficiency.
Calcium and phosphorus	Necessary for dentin and bony tissue synthesis. Poor mineralization occurs with deficiency. Maintains jawbone sufficiency.
Chromium	Needed for proper glucose metabolism. Controlled intake of carbohydrates helps to maintain healthier gums and overall health status (Moynihan, 2005b).
Copper	Needed for production of blood and nerve fibers.
Fluoride	Consumption of fluoridated water coupled with a reduction in nonmilk sugar intake is an effective means of caries prevention (American Dietetic Association, 2000; Moynihan, 2005a). Keeps bones healthy. Drinking water should contain 1 ppm; toothpaste, mouth rinses, and topical treatments also help.
Iron	Helps produce red blood cells; promotes resistance to disease; improves health of the teeth, skin, and bones. Maintains energy.
Magnesium	Helps in bone development. Enhances use of vitamin C. Deficiency may lead to calcium resorption.
Potassium	Needed for muscle contraction and proper nerve function.
Zinc	Regulates the inflammatory process; aids in wound healing. Deficiencies can lead to poor healing, susceptibility to infection, loss of taste, and altered metabolism.

TABLE 2-6 Dental Problems, Treatment, and Prevention

Symptoms	Likely Cause	Treatment	Prevention
Bad Breath			
Odor from mouth; bad, metallic taste; coated tongue	Food caught around and between teeth; infection in gums; improper brushing; sinusitis; digestive problems, such as preulcerative conditions; diabetes	Practice good oral hygiene, including rinsing with mouthwash; brush tongue often; see dentist to evaluate throat, sinuses, tongue, and possible gum infection, and professionally clean teeth and gums; review diet	Regular dental visits; flossing, brushing, and rinsing; good nutrition
Broken Tooth or Filling			
Tooth feels sharp; tooth sensitivity to temperature and pressure	Accidental trauma; decay; weak tooth from grinding or improper bite	Do not irritate; place piece of soft dental wax from drugstore over cracked or fractured tooth; see a dentist immediately	Regular dental checkups to discover possible weak teeth, decay, or large, unstable fillings
Canker Sores			
Painful red circular area that develops on the tongue, gums, lips, or cheeks; in certain phases, sores have a yellow or white center area; sore to touch; sensitivity to spicy, salty foods	Bacterial or viral infection; trauma from denture in mouth; stress	Use over-the-counter remedies recommended by the dentist; coat lesions after meals; see dentist to make sure there is no infection or for additional medication if pain persists; the dentist will evaluate dentures for weight-bearing points to be certain the problem does not exist there	Avoid irritating the area; avoid spicy, acidic foods while mouth is sore
Dental Abscess (swelling around tooth or cheek)			
Pain, throbbing in gum or tooth; swelling; sensitive bite; loose teeth; sensitivity to heat	Tooth decay; initial eruption of tooth through the gums or fractured tooth; tooth nerve damage	Rinse with salt water solution; use mouthwash; avoid eating on or near tooth; see dentist immediately; may require antibiotics or root canal treatment to prevent spread of infection	Regular dental checkups; good oral hygiene; brushing, flossing, and rinsing
Discolored Teeth			
Teeth have unsightly and discolored appearance; single tooth begins to turn yellow or gray	Surface stain from certain foods, such as tea and coffee; internal staining from tooth nerve damage or from rheumatic fever; stains from tetracycline	Improve oral hygiene; brush frequently; diminish coffee or tea intake; rinse with peroxide; consult dentist to check nerve in darkened tooth; consider supervised tooth bleaching/whitening	Good oral hygiene; avoid foods and liquids that can stain teeth, such as tea and coffee
Gum Disease			
Gum pain; nonthrobbing ache; swelling; gum bleeding; blood in saliva when brushing; metallic taste	Food debris between teeth; tartar beneath gums; infection; poor bite may worsen this condition	Improve oral hygiene by brushing often and flossing; rinse with mouthwash; consult dentist to evaluate extent of condition; treatment by removing plaque and tartar may require surgery and/or bite adjustment	Good oral hygiene; regular dental checkups and cleanings
Red Inflamed Gums			
Color of gums around teeth progresses from pink to red with swelling or puffiness; dry mouth; snoring	Mouth breathing; some medications, such as antihistamines, blood pressure medications, and antidepressants, decrease salivary flow	Use oral salivary rinses and toothpastes for dry mouth; improve oral hygiene; consult dentist because this condition can lead to tooth decay, advanced gum disease, or other mouth infections	Ask physician if medications can be changed; consult dentist about obtaining oral rinses and a snoreguard

(continued)

TABLE 2-6 Dental Problems, Treatment, and Prevention *(continued)*

Symptoms	Likely Cause	Treatment	Prevention
Loose Teeth			
Teeth move; spongy feel to bite; teeth sensitive or even painful when chewing	Gum disease; tooth grinding; orthodontic appliances too tight; cyst, tumor, abscess, or trauma to teeth	See dentist as soon as possible to determine cause; practice good oral hygiene; be aware of tooth grinding or clenching and use appliance to prevent grinding	Regular dental visits; good oral hygiene; have your dentist evaluate your bite; use a bite appliance if your dentist advises
Lumps Under Jaw or Neck Muscle			
Neck sore to touch or movement; swelling in neck; sore throat; difficulty swallowing	Cold/flu; tooth abscess or infection; tumor	Treat cold/flu symptoms; limit neck movement; check temperature; take pain relievers such as aspirin; see a dentist if symptoms persist to evaluate the extent of swelling and infection	Regular dental checkups; patients should pay special attention to any growth or changes in the head or neck
Toothache (tooth pain on biting or chewing)			
Tooth pain related to temperature change or touch or from chewing or biting; dark brown spots on teeth may indicate new decay	Bacterial acids; large filling broken out of tooth; tooth grinding	Rinse mouth often with vanilla extract to soothe discomfort; avoid chewing on tooth; see a dentist as soon as possible to determine cause and further treatment	Regular dental visits for prevention; the sooner examined, the better the chance of success
Tooth Sensitivity to Temperature Change			
Breathing outside in cold air causes pain; waking up with toothache; pain when eating/drinking cold things	Inflamed gums; gum recession that exposes root surfaces; tooth decay; teeth clenching or grinding that has worn away tooth enamel	Use desensitizing toothpaste on a daily basis; use a soft bristle brush; avoid temperature differences; consult dentist for appropriate treatment	Good oral hygiene; apply fluoride gel; use desensitizing toothpaste; avoid food temperature differences; avoid hard bristle toothbrushes; become aware of and avoid tooth grinding or squeezing teeth together; have fillings bonded to seal areas of sensitivity; dentist may recommend a biteguard for grinding

Adapted from Rhode Island Dental Association, 200 Centerville Road, Warwick, RI 02886; Phone: (401) 732-6833. Web site accessed March 28, 2009, at http://www.ridental.com/dentalproblems.cfm. Used with permission.

INTERVENTION

OBJECTIVES

Broken or Wired Jaw

- Provide adequate nourishment to allow healing while reducing jaw movement.
- Decrease complications such as fever, nausea, and vomiting.
- Prevent excessive weight loss; up to 10% is common.
- Maintain a patent airway.

Dental Caries

- Alter dietary habits; deprive bacteria of substrate; reduce acid by keeping pH at 7.0.
- Maintain frequent fluoride contact with tooth surfaces as directed by a dental professional.

Early Childhood Caries (ECC)

- ECC is a preventable dental disease in which enamel erodes, and tooth surfaces are permanently damaged from long exposure to liquid carbohydrate sources.
- Education is the biggest factor. Children with significant risk factors for caries (e.g., inadequate home dental care, poor oral hygiene, a mother with a high number of cavities, a high sugar intake, enamel defects, premature birth, special health care needs, low socioeconomic status) should be referred to a dentist (Douglass et al, 2004).

Edentulism

- Provide proper consistency to allow the patient to eat.
- Monitor for deficiencies in fiber, vitamins A and C if whole grains, fruits, and vegetables are not consumed (Touger-Decker, 2004).

Mouth Ulcers or Pain

- Lessen mouth soreness to increase dietary intake; mouth sprays may be available to lessen pain while eating.
- Promote healing for a return to normal eating patterns.
- Prevent weight loss or other consequences.

Tongue Disorders

- Provide adequate nourishment despite acute or chronic disability.

SAMPLE NUTRITION CARE PROCESS STEPS

Chewing Difficulty

Assessment Data (sources of info): Food records and intake calculations; dental evaluation for loose dentures; weight changes.

Nutrition Diagnosis: Chewing difficulty related to inability to chew foods from poor dentition as evidenced by weight loss of 2 lb in 14 days and ill-fitting dentures.

Interventions: Food and Nutrient Delivery

ND 1.2 Modify current diet to puree diet until otherwise noted from Dentist/MD.

ND 3.1.1 Continue with shakes BID to enhance energy intake.

Recommend dental referral for dentures.

Monitoring and Evaluation: Intake records, reduction in chewing problems, improved weight after fitting of new dentures.

Inadequate Energy Intake—Early Childhood Caries

Assessment Data (sources of info): Food records (high intake of juice, sweetened beverages throughout the day from the bottle); intake calculations; dental evaluation for ECC; weight loss from inability to chew solids and refusal to drink from a cup.

Nutrition Diagnosis: Inadequate energy intake (NI-1.4) related to inability to chew foods as evidenced by early childhood dental caries with poor weight gain.

Interventions

Goals: Wean from bottle completely. Increase solid food intake and decrease fluids, especially sweetened fluids. Follow a weight gain of at least 0.6 oz/wk, 2.7 oz/mo, or 1 lb/6 mo. Educate parents on importance of healthy oral hygiene and not allowing child to carry liquids around during day or fall asleep with liquids.

Food and Nutrient Delivery: ND-1.1 Provide general/healthful diet, provide information on weaning from the bottle, educate on importance of oral hygiene, increase intake of water, decrease intake of juice to a maximum of 4 oz/d diluted with water, eliminate other sweet drinks, alter diet to reduce need to chew (puree, mash, or chop foods).

Nutrition Education: E-1.2 Provide information on weaning, calorie boosters, importance of oral hygiene, limiting sweetened beverages. DDS to perform oral surgery for removal of dental caries to decrease pain and increase intake of solid foods for weight gain. RD to continue to monitor weight gain and food intake.

Counseling: C-2.5 Counsel patient's mother and father on supporting each other through weaning and the importance of good oral hygiene.

Coordination of Care: RC-1.3 Collaborate with MD and DDS on patient's care through oral surgery and monitoring weight status.

Monitoring and Evaluation: Intake records, reduction in chewing problems, improved weight, and health status.

Tube Feeding

- Children on tube feedings often have dental problems; attend to oral hygiene more carefully than for those fed orally.
- Adults will require special attention to oral hygiene and mouth care while on tube feedings.

Xerostomia

- Dry mouth may be more severe after radiation therapy than with other causes, such as diabetes.
- Artificial saliva agents may be useful for some. Reduced saliva affects patient perception of swallowing ability and changes dietary choices (Logemann et al, 2003).
- Good oral hygiene may prevent dental decay.

FOOD AND NUTRITION

Broken or Wired Jaw

- A diet of pureed or strained foods and liquids of high protein/calorie content are necessary. Use high-energy supplemental beverages (perhaps 2 kcal/mL). Double-strength milk may also be used to keep protein intake at a high level.
- Take adequate amounts of vitamin C for healing.
- Monitor food temperatures carefully, as extremes may not be tolerated.
- Six to eight meals are needed.
- Follow meals with salt water rinse.

Dental Caries

- Decrease sucrose and cooked or sticky starches, as well as the frequency of snacking and duration of exposure time. *Streptococcus mutans* is a common bacterial culprit; others include *Lactobacillus casein* and *Streptococcus sanguis* (Touger-Decker, 2004).
- Use a balanced diet, avoid eating sweets or starches with meals.
- Fluoride exposure should be adequate, including from water supplies.
- The sequence of eating foods, the combination of foods, the form of foods and beverages consumed, and nutrient composition of foods/beverages must be evaluated and altered accordingly (Touger-Decker, 2004).

Early Childhood Caries (ECC)

The following are guidelines for prevention (American Academy of Pediatric Dentistry, 2005–2006; Clarke et al, 2006; Wagner, 2006):

- Do not allow a child to fall asleep with a bottle containing milk, formula, fruit juices, or other sweet liquids. Never let a child walk around with a bottle in his/her mouth. Never put an infant or child to bed with a bottle that is filled with sugar-containing beverages, including fruit juice or Kool-Aid.
- Comfort a child who wants a bottle between regular feedings or during naps with a bottle filled with cool water.
- Always make sure a child's pacifier is clean, and never dip a pacifier in a sweet liquid.
- Introduce children to a cup as they approach 1 year of age. Children should stop drinking from a bottle soon after their first birthday.
- Notify the parent of any unusual red or swollen areas in a child's mouth or any dark spot on a child's tooth so that the parent can consult the child's dentist.
- Maintain good oral hygiene.
- Monitor for iron deficiency anemia, which is common (Clarke et al, 2006).

Edentulism

- A chopped, ground, strained, or pureed diet should be followed as required. Use the least restricted diet and progress as tolerated.
- Identify potential solutions such as obtaining new dentures or repairing current dentures.

Mouth Ulcers or Pain

- Foods low in acid and spices should be consumed; avoid citrus juices, vinegar and other similar foods.
- Supplement the diet with vitamin C, protein, and calories to speed healing.
- Small, frequent meals and oral supplements may be beneficial to prevent weight loss.
- Moist or blenderized foods with additional liquid are helpful.
- Soft, cold foods such as canned fruits, ice cream, popsicles, yogurt, cottage cheese, or cold pasta dishes may be used.
- Use of a straw may be helpful.
- Cut or grind meats or vegetables.
- Extra butter, mild sauces, and gravies may be needed.
- Follow meals by brushing teeth to reduce possibility of dental caries.

Tongue Disorders

- If the patient is unable to chew, tube feeding should be considered (Fig. 2-3).
- Liquids may be added to the diet as tolerated. Many foods are tolerated if liquefied and blenderized.

Tube Feeding

- Good oral hygiene and mouth care will be needed, even if a patient is not fed by mouth. Tube feeding should include all key nutrients to meet patient needs; see Section 17.

Xerostomia

- Moisten foods, adding water or milk when possible. Use sauces or gravies if needed.
- Avoid excessive spices.
- Avoid excessively chewy foods such as steak, crumbly foods such as crackers or cake, dry foods such as chips, or sticky foods such as peanut butter (Touger-Decker, 2004).

Common Drugs Used and Potential Side Effects

- Luride is a fluoride supplement for children to strengthen teeth against tooth decay. Avoid use with calcium or dairy products because it may form a nonabsorbable product.
- For patients with cancer, various therapies affect the mouth and gums. Monitor closely.
- Oral side effects of drugs interfere with functioning and increase risks for infection, pain, and possible tooth loss (Spolarich, 2000).

Herbs, Botanicals, and Supplements

- Herbs and botanicals may be used; identify and monitor side effects.
- Counsel about use of herbal teas, especially unsuitable products such as comfrey tea.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- If needed for oral or dental problems, blended foods and/or tube feedings should be prepared. Sometimes, using a bulb syringe to feed may be useful.
- Provide creative ideas for the seasoning and flavoring of foods. Discuss acceptable restaurant options for persons who are at home.
- Ensure that fluoride is provided in some way by the diet, water supply, or dental office.
- Read milk labels to ensure vitamin D fortification.
- Dental status is an especially important part of assessment and care for the elderly (Sahyoun et al, 2003; Sheiham et al, 2001).
- Integrating dietary counseling into the dental setting warrants further investigation (Moynihan, 2005a).

To Prevent Caries

- Encourage good habits in oral hygiene and diet: detergent foods (raw fruits and vegetables) should be recommended rather than sticky or impactant foods (soft cookies, bread, sticky sweets, dried fruits). Cariostatic foods should be encouraged, such as cheese, raw fruits and vegetables, peanuts, and cocoa.
- Avoid cariogenic foods such as dried fruits, candy, cookies, pies, cakes, ice cream, canned fruit, soft drinks, fruit drinks, lemonade, gelatin desserts, snack crackers, pretzels or chips, and muffins. Brush teeth or eat cheese after meals and sugary snacks to normalize pH.
- Regular use of fluoride daily can help reduce the incidence of root caries (Richards, 2009).

Patient Education—Food Safety

- When traveling, avoid ice made from tap water. Airline water may not be free from contamination.
- Use of bottled water is recommended for brushing teeth in countries where water is not safe.

For More Information

- American Academy of Pediatric Dentistry
  http://www.aapd.org/
- American Academy of General Dentistry
  http://www.agd.org/
- American Academy of Periodontology
  http://www.perio.org/
- American Dental Association
  http://www.ada.org/
- Colgate
  http://www.colgate.com/app/Colgate/US/OralCare/HomePage.cvsp
- International Association for Disability and Oral Health
  http://www.iadh.org/
- Medline—dental health
  http://www.nlm.nih.gov/medlineplus/dentalhealth.html
- National Institute of Dental and Craniofacial Research (NIDCR)
  http://www.nidcr.nih.gov/
- Oral Health America
  http://www.oralhealthamerica.org/

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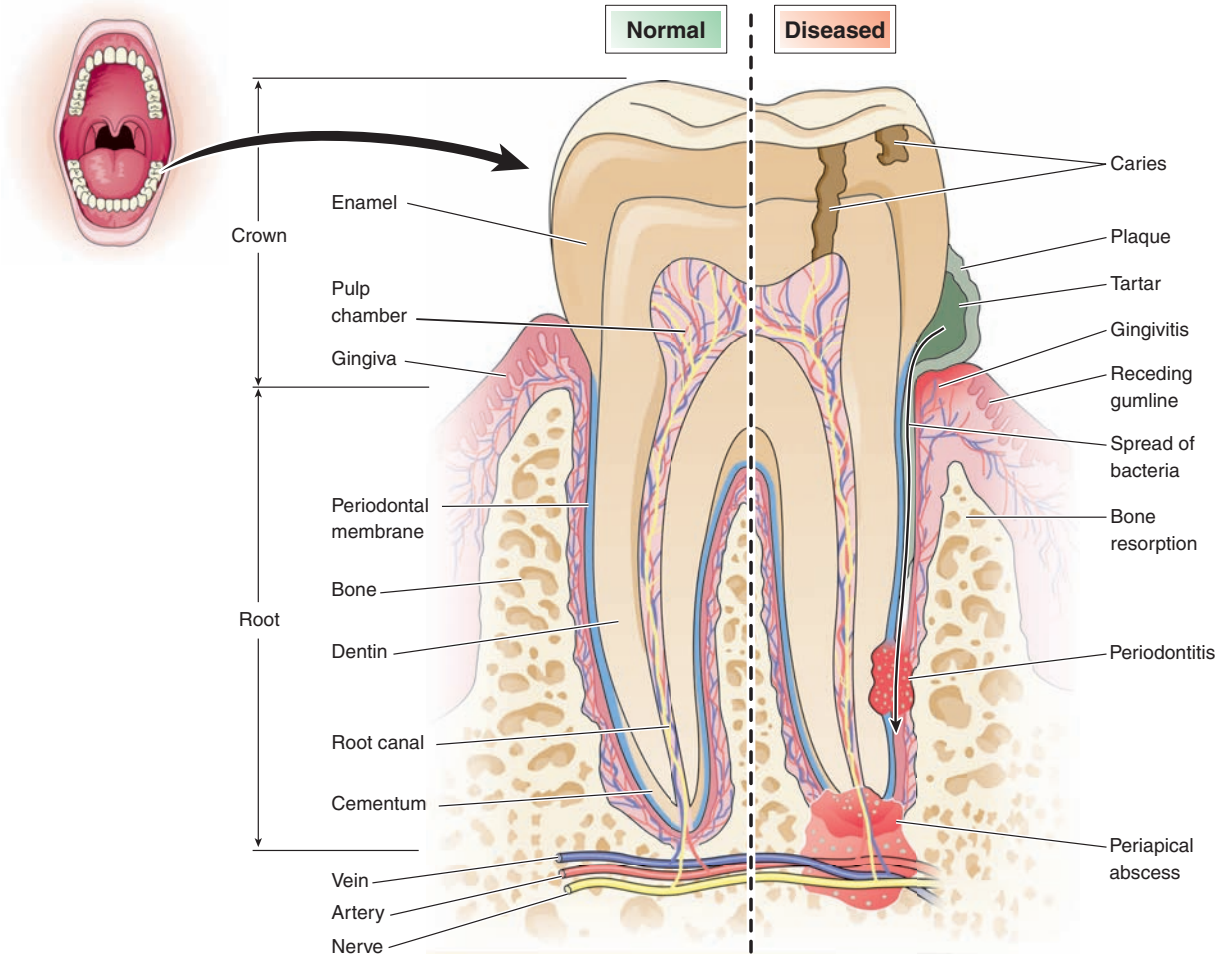
PERIODONTAL DISEASE AND GINGIVITIS

NUTRITIONAL ACUITY RANKING: LEVEL 1–2



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DEFINITIONS AND BACKGROUND

Gingivitis involves minor inflammatory changes in the gums; it may be acute or chronic, local or generalized. Vitamins C and D may reduce gingivitis (Dietrich et al, 2005). *Acute necrotizing ulcerative gingivitis* (Vincent's disease or trench mouth) is an acute ulceration affecting marginal gingiva with inflamed or necrotic interdental papillae. The onset is abrupt and painful with slight fever, malaise, excess salivation, and bad breath. It can be caused by systemic disease.

Tissues that support teeth in the jaws are collectively known as the periodontium (gums, alveolar bone, periodontal membrane). Any abnormality that leads to a visible change or loss of integrity of any component of the supporting tissue is a periodontal disease. Periodontal disease is a painless, chronic inflammatory disease that most commonly manifests as *pyorrhea alveolaris*. It involves a gross breakdown of supporting tissues with progressive loosening and loss of teeth inflammatory disease initiated by oral microbial biofilm (Van Dyke, 2008). It is a major cause of tooth loss in adults.

Periodontoclasia involves destruction of tissues around the teeth. A poor diet and inadequate dental hygiene can cause destruction of the jawbone. Osteoporosis and inflammation-associated bone degradation in periodontitis have a common pathogenesis (Serhan, 2004). Periodontal disease is evident approximately 10 years before osteoporosis.

Periodontal disease can start in the second decade; wisdom teeth are a breeding ground for bacteria that cause problems. Children and teens are also at risk if their oral and dental health needs are not addressed (Cummins, 2006). Nutrient deficiencies are prevalent and smokers are especially vulnerable to vitamin C deficiency (Nishida et al, 2000). Immune-enhancing nutrients for good oral health include protein, zinc, vitamins C and E, calcium, and the B-complex vitamins.

In the United States, periodontal disease affects a large portion of the population. At risk in particular are pregnant women, women after menopause, obese individuals, diabetics, alcoholics, smokers, persons with AIDS or rheumatoid arthritis, persons with respiratory ailments, and persons on medications including heart medicines, antidepressants, and antihistamines. Periodontal disease may precede bacterial pneumonia, so treatment is important. Evidence-based periodontology includes antimicrobial therapy, regenerative periodontal surgery, periodontal plastic surgery, bone regeneration surgery and implant treatment, and advanced soft tissue management at implant sites (Tonetti, 2000).

SAMPLE NUTRITION CARE PROCESS STEPS

Inadequate Oral Food and Beverage Intake—Periodontal Disease

Assessment Data (sources of info): Food records and intake calculations; dental evaluation.

Nutrition Diagnosis: Inadequate oral food and beverage intake related to sore and inflamed gums as evidenced by diet history revealing low use of antioxidant foods and vitamin C from fruits and vegetables; weight loss of 5# in past month; and diagnosis of periodontal disease.

Intervention: Education about the role of diet and oral hygiene in periodontal disease. Recommend nutrient and dietary changes to improve quality of food intake, reduce inflammation, and promote healing.

Monitoring and Evaluation: Intake records, rate of healing of gum disease.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Clinical/History

- Height
- Weight
- BMI
- Diet history
- Gums—color, friability, receding
- Oral examination for tooth mobility, calculus
- Sore mouth
- Overall nutritional status
- History of smoking, diabetes, other chronic diseases

Lab Work

- Serum glucose
- Serum Na^{+}, K^{+}
- Ca^{++}, Mg^{++}
- Serum ascorbic acid
- H & H, serum Fe
- Alb, transthyretin
- CRP

INTERVENTION

OBJECTIVES

- Reduce inflammation and promote healing.
- Correct poor nutritional habits that can lead to chronic subclinical deficiencies in levels of vitamins A, C, D, amino acids, riboflavin, folacin, zinc, and calcium.
- Prevent further decline in status of bones and gums. Protect the jawbone with adequate calcium and vitamin D (Hildebolt et al, 2004), especially in postmenopausal women.
- Review medications and consider alternatives to those causing dry mouth or other problems.
- Pregnant women with this condition are at risk for preterm birth and other adverse obstetric outcomes, such as preeclampsia and low birth weight; they should be closely monitored with prenatal medical and dental care.

FOOD AND NUTRITION

- Ensure adequate intake of calcium, protein, zinc, phosphorus, vitamin C, fluoride, and vitamin A. A multivitamin–mineral supplement and vitamin-D fortified milk should be used.
- Use high-detergent foods (firm, fresh fruits and raw vegetables or those that are lightly cooked). Include cranberries, blueberries, green tea, and other foods rich in antioxidants and polyphenols (Kushiyama et al, 2009).

- Control timing and frequency of meals and snacks to reduce exposure of susceptible gum tissue and teeth to the acids that form plaque. Control blood glucose in diabetes.
- Promote a diet containing foods naturally rich in antioxidants and omega-3 polyunsaturated fatty acids (DHA, EPA), and low in refined carbohydrates (Chapple, 2009).

Common Drugs Used and Potential Side Effects

- Sodium bicarbonate may be used as a mouthwash. Patients with high BP should not swallow this wash.
- Peridex is an oral rinse to control bleeding gums. Taste changes may occur with its use.
- Triclosan-containing dentifrices may slow periodontal disease progression (Niederman, 2004).
- Antibiotic treatment of periodontitis includes amoxicillin/clavulanic acid, metronidazole, and clindamycin.
- Nonsteroidal anti-inflammatory agents may be used, such as Ibuprofen.

Herbs, Botanicals, and Supplements

- Herbs and botanicals may be used; identify and monitor side effects. Counsel about use of herbal teas, especially those containing toxic substances.
- For gingivitis, bloodroot, Echinacea, purslane, chamomile, licorice, and sage have been recommended but not confirmed for efficacy.
- A "Connective Tissue Nutrient Formula" that contains vitamins A, C, and D, glucosamine sulfate, magnesium, oligoproanthocyanidins, copper, zinc, manganese, boron, silicon, and calcium may be prescribed to enhance the integrity of key connective tissue elements.
- Naturopathic physicians may prescribe *Panax ginseng, Withania somnifera,* and *Eleutherococcus senticosus* to reverse the impact of bacterial and psychosocial stressors.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Encourage a proper diet, especially sources of omega-3 fatty acids, calcium and vitamins C and D.
- Recommend meticulous oral hygiene and regular dental examinations to maintain dental hygiene. Brush often and floss after eating sticky foods such as candy, sticky buns, and fruit rolls. Drink lots of water.
- Encourage pregnant women and persons with dentures, diabetes, cancer, HIV/AIDS, rheumatoid arthritis, or leukemia to pay special attention to oral hygiene.
- Encourage intake of polyphenols and antioxidant-rich foods, including green tea (Kushiyama et al, 2009).

Patient Education—Food Safety

- Periodontitis involves host-mediated inflammation, with modulation of inflammation at a cellular and molecular level. Avoidance of infection with added inflammatory response will be needed, especially related to food handling and sanitation.

For More Information

- American Academy of Periodontology
  http://www.perio.org/index.html
- Dental Societies
  http://www.perio.org/links/links.html#dental
- Periodontal Societies
  http://www.perio.org/links/links.html#perio

PERIODONTAL DISEASE AND GINGIVITIS—CITED REFERENCES

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Tonetti M. Advances in periodontology. *Prim Dent Care.* 7:149, 2000.

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TEMPOROMANDIBULAR JOINT DYSFUNCTION

NUTRITIONAL ACUITY RANKING: LEVEL 1

DEFINITIONS AND BACKGROUND

Temporomandibular joint (TMJ) disorders result from local or systemic causes, such as rheumatoid or osteoarthritis and connective tissue disorders. The TMJ is a diarthrodial joint with moving elements (mandible) and fixed elements (temporal bone). With this dysfunction, overuse or abuse of any part of normal action affects the mastication process. Patients with TMJ disorder pain dysfunction have toothaches, facial pains, and food-intake problems. The National Institute of Dental and Craniofacial Research (2009) indicates that over 10 million people in the United States suffer from TMJ

problems at any given time. Osteoarthrosis and internal derangement may coexist in the same joint in about 33% of cases; pathological tissue changes should be examined in patients with TMJ (Dimitroulos, 2005).

Women between the ages of 30 and 60 years account for 75% of all cases. Mandibular deviation may occur from repetitive overloading (stress or habit such as gum chewing, grinding), from functional masseter muscle coordination problems, or from incorrect occlusion (as with missing teeth). Structural problems are treated by surgery (e.g., fusion can be treated by removing the area of fused bone and replacing it with silicon rubber). Sometimes an artificial joint is the answer; but surgery is recommended for only a few patients. Undue muscle tension causes most TMJ, with some other problems stemming from inadequate bite (as from a high filling or a malocclusion). People with TMJ benefit from a visit to their dentist or ear, nose, and throat specialist.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Clinical/History

Height
Weight
BMI
Diet history
Stiff neck, face, or shoulders
Locking of affected joint
Trismus
Gum status
Mouth/jaw pain or clicking noise
Headaches
Shoulder or neck pain

Lab Work

Gluc
Serum Na^+, K^+
Ca^{++}, Mg^{++}
H & H, serum Fe
Chol, Trig
Alb
CRP

INTERVENTION

OBJECTIVES

- Reduce repetitive overloading by use of a splint or by breaking bad habits such as grinding (bruxism).
- Reduce stress with relaxation techniques. Relieve pain and muscle spasms.
- Prevent or correct malnutrition or weight loss.
- Ensure adequate intake of soft, nonchewy sources of fiber.
- Reduce any existing inflammation and prevent complications such as mitral valve prolapse.

FOOD AND NUTRITION

- Use a normal diet with soft foods to prevent pain while chewing.
- Cut food into small, bite-sized pieces. Avoid chewy foods such as caramel, nuts, toffee, chewy candies, and gummy bread and rolls.

SAMPLE NUTRITION CARE PROCESS STEPS

Chewing Problems

Assessment Data: Food records and intake calculations; dental evaluation; pain when chewing.

Nutrition Diagnosis: Chewing problems related to TMJ and pain when eating as evidenced by diet history

Intervention: Education about soft foods and liquids of high nutrient density. Recommend individualized dietary changes to improve intake and prevent weight loss.

Monitoring and Evaluation: Intake records, reduction in TMJ pain, improved intake from nutrient-dense foods.

- Avoid opening mouth widely, as for large and thick sandwiches. Grate vegetables (e.g., carrots) to reduce need for chewing.
- Use adequate sources of vitamin C for adequate gingival health.
- Suggest foods rich in antioxidants, such as green tea, to promote health.

Common Drugs Used and Potential Side Effects

- Pain medicines may be needed when the condition is active. Monitor side effects for the specific drugs used.

Herbs, Botanicals, and Supplements

- Herbs and botanicals may be used; identify and monitor side effects. Counsel about use of herbal teas, especially avoiding toxic ingredients.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Discuss the role of dental care in maintaining adequate health.
- Monitor for any tooth or gum soreness; advise the dentist as necessary. Regular oral hygiene must be continued despite mouth pain.
- Physical therapy may be needed to correct functioning of muscles and joints.
- Nail biting, gum chewing, use of teeth to cut thread, or similar habits should be stopped.
- Smoking is often a cause of bruxism, and programs to stop smoking should be considered if needed.

Patient Education—Food Safety

- Use general safe food handling measures.

For More Information

- Jaw Joints and Allied Musculoskeletal Disorders Foundation http://www.tmjoints.org/
- TMJ Disorder http://www.tmj.org/

TEMPOROMANDIBULAR JOINT DYSFUNCTION—CITED REFERENCES

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SELF-FEEDING PROBLEMS: VISION, COORDINATION, CHEWING, HEARING LOSS

NUTRITIONAL ACUITY RANKING: LEVEL 1–2 (VARIES BY SEVERITY)

DEFINITIONS AND BACKGROUND

Self-feeding ability, one of the activities of daily living (ADLs), can be limited by low vision or blindness, lack of coordination, and chewing problems. Where appropriate, these factors are mentioned in other sections. Dysphagia is a fourth problem, described in Section 7. Assessment for vision changes, self-feeding difficulty, hearing, continence, gait and balance, and cognition can reveal a great deal about an individual's ability to function independently (Rao et al, 2004).

Low vision or blindness can affect any age; children with developmental disabilities and older persons may have cataracts or macular degeneration. The World Health Organization defines "low vision" as visual acuity between 20/70 and 20/400 with the best possible correction, or a visual field of 20 degrees or less. "Blindness" is defined as a visual acuity worse than 20/400 with the best possible correction. Someone with a visual acuity of 20/70 can see at 20 feet what someone with normal sight can see at 70 feet. Someone with a visual acuity of 20/400 can see at 20 feet what someone with normal sight can see at 400 feet. Normal visual field is about 160–170 degrees horizontally. Age-related macular degeneration and cataract are the leading causes of visual impairment and blindness in the United States; both diseases increase dramatically after age 60.

Age-related macular degeneration (AMD) is a vascular condition that damages the retina and affects 25–30 million people worldwide. The Age-Related Eye Disease Study (AREDS), sponsored by the National Eye Institute, found that taking high levels of antioxidants and zinc can reduce the risk of developing advanced AMD by about 25%. The specific daily amounts are 500 mg vitamin C; 400 IU vitamin E; 15 mg beta-carotene; 80 mg zinc oxide; and 2 mg copper as cupric oxide to prevent copper deficiency anemia. Protective foods include nuts, fish (Miljanovic et al, 2005; Seddon et al, 2003) lycopene, lutein and zeanthin. Lutein and zeaxanthin are carotenoids that have a role in filtering destructive blue light as photosensitive antioxidants. Smokers should not take beta-carotene supplements (Age-Related Eye Disease Research Group, 2001). In addition to the AREDS supplement, a lower dietary glycemic index (dGI) with higher intakes of DHA and EPA reduces progression to advanced AMD (Chiu et al, 2009). Diets high in saturated fat, animal fat, linoleic acid, and trans-fatty acids promote higher risk of AMD (Seddon et al, 2003). Abdominal obesity, smoking, and diets high in glycemic index should be avoided. Women tend to have a higher risk than men.

Cataract causes clouding in the crystalline lens of the eye, causing opacity and less passage of light. Blindness occurs if not treated. Regular intake of antioxidant foods, including rich sources of vitamins A, C, E and selenium, can be protective.

Diabetic retinopathy is a major cause of vision loss. All individuals who have diabetes should have a dilated eye exam annually. Diabetic retinopathy affects as many as 80% of individuals who have had diabetes for 10 years or longer. Careful control of blood glucose and hypertension are important measures to protect the small vessels of the eye. Treatment often involves laser surgery.

Glaucoma also has nutritional implications. Risk factors for chronic glaucoma include age over 40; a family history of glaucoma, diabetes, myopia; or being African American. Acute glaucoma may occur in persons with a family history of acute glaucoma, older age, presbyopia, or use of systemic anticholinergic medications. Prostagladins regulate inner eye pressure; glaucoma may be related to a higher intake ratio of omega-3 fatty acids to omega-6 fatty acids (Kang et al, 2004). Soybean, safflower, and sunflower oils may be protective.

Hemianopia yields loss in half of the field of vision; quadrantanopia affects a quarter of the visual field. Vision loss may range from slight to severe. Patients may need guidance about their meals, as they may not see the full plate or tray of food. Vision restoration therapy and the brain's ability to repair itself have made advances for these patients.

Coordination problems may occur at any age. Conditions that can cause coordination problems include Alzheimer's disease, alcohol abuse, attention deficit disorder, brain cancer, chorea, Down syndrome, encephalitis, fetal alcohol syndrome, advanced HIV infection, hydrocephaly, multiple sclerosis, Parkinson's disease, Rett syndrome, stroke, and Wilson's disease. Hand–eye coordination is needed for self-feeding, and when this is not working properly, assistance is needed. Other problems affecting meal intake may include falling forward, feet not touching the floor, leaning to one

side, poor balance while sitting, and poor neck control. Sometimes, it is possible to adjust table height, offer pillows or other positioning equipment, offer a footstool, or adjust pedals on a wheelchair. It is recommended to work with an occupational therapist for the proper types of adjustments to allow for better mealtime food intake.

Chewing problems may cause inability to consume enough food or foods of varying texture. Total edentulism without dentures may contribute to deterioration in health. Without chewing, there is less production of saliva and food is not properly mixed before swallowing. **Dry mouth (xerostomia)**, from a variety of causes, may interfere with chewing and swallowing; it should be corrected where possible.

Hearing loss, while not always affecting food intake, may be related to underlying cardiovascular disease, hypertension, or diabetes. Cochlear vulnerability from microvascular changes may occur (Agrawal et al, 2009). Hearing loss is more common than previously believed and preventive measures, such as lifestyle changes, may be needed as early as adolescence (Helzner et al, 2005).

SAMPLE NUTRITION CARE PROCESS STEPS

Self-Feeding Problems—Low Vision

Assessment Data: Food records and intake calculations; vision and self-feeding problems.

Nutrition Diagnosis: Self-feeding difficulty related to blindness and no adaptive equipment as evidenced by limited intake at meals and long period required for eating.

Intervention: Education about adaptive equipment with careful orientation to items served at meals. Reading menu choices aloud.

Refer to Occupational Therapy for proper equipment, techniques for cueing, positioning at mealtime, and encouragement tips.

Counsel about dietary changes to improve intake and prevent weight loss, such as finger foods and easy to handle foods.

Monitoring and Evaluation: Intake records showing better intake when using adaptive equipment; weight records; improved quality of life with greater independent functioning at mealtimes.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers

Clinical/History

- Height
- Weight
- BMI
- Recent weight changes
- Diet history
- Mouth or tongue lacerations
- Sore or bleeding gums
- Missing or loose teeth, edentulism
- Dentures, especially poorly fitting
- Blindness, cataracts, AMD
- Hemianopia, low vision
- Chewing problems
- Dysphagia (see Section 7)
- Signs of dehydration or edema
- Coordination problems
- Dry mouth (xerostomia)
- Needs feeding assistance
- Needs adaptive feeding equipment

Lab Work

- Gluc
- Alb, transthyretin
- Serum Na^{+}, K^{+}
- Ca^{++}, Mg^{++}
- I & O
- H & H, serum Fe
- X-rays (such as mandible)

INTERVENTION

OBJECTIVES

- Promote independence in self-feeding, when possible.
- Address all nutritional deficiencies and complications individually. Select nutrient-dense foods.
- Promote overall wellness and health.
- Increase interest in eating. Increase pleasure associated with mealtimes.
- Prevent malnutrition and weight loss.
- Decrease instances in which constipation, anorexia, or other problems affect nutritional status.
- Educate the caregiver about adaptive equipment, utensils, and special food modifications. Patients with hemianopia may require special training to be able to see and eat all of the meals served to them.

FOOD AND NUTRITION

Low Vision, Blindness, AMD, Cataracts

- Provide special plate guards, utensils, double handles, and compartmentalized plates with foods placed in similar locations at each meal. Place all foods within 18″ reach at mealtime. Explain placement of foods. Open packets if needed.
- Work with occupational therapist (OT) or family to practice kitchen safety and to determine ability to be independent at mealtimes. Allow sufficient time to complete meals; refrigerate or reheat items as needed.
- Create a feeling of usefulness by delegating appropriate tasks related to mealtime, such as drying dishes and assisting with simple meal tasks that are safe for the individual.
- Support companionship during meals, especially if problems occur or if anything else is needed.
- Use straws for beverages unless there is dysphagia.
- Include a balance of fatty acids (omega 3 and omega 6). Use rich sources of DGA and EPA, such as salmon, sardines, herring, tuna, or a fish oil supplement as needed. Use more oils such as olive and canola. Linoleic acid from vegetable oils reduces the positive effects and should be used less often.
- For a diet lower in dGI, include whole grains, soybeans and lentils more often and exclude desserts, candy, sweetened beverages, potatoes and white bread.
- Include more lycopene from pink grapefruit, tomato sauce, tomato juice, and watermelon. Lutein and zeaxanthin are found in broccoli, spinach, other leafy greens, egg yolk. Antioxidant-rich foods that include good sources of

TABLE 2-7 Nutrients for Healthy Vision[a]

Protein and Amino Acids	Protein undernutrition is associated with increased risk of cataract. Low protein intake may induce deficiencies of specific amino acids that are needed to maintain the health of the lens, or other nutritional deficiencies, particularly niacin, thiamin, and riboflavin (Delcourt et al, 2005).
Vitamin A, Lutein and Zeaxanthin	Vitamin A is needed for healthy cornea and conjunctiva. Many studies have shown that lutein and zeaxanthin reduce the risk of chronic eye diseases, including age-related macular degeneration and cataracts. Cataract and AMD patients tend to be deficient in vitamin A and the carotenes, lutein, and zeaxanthin (Head, 2001; Krinsky et al, 2003; Seddon et al, 2003). Lutein and zeaxanthin are found in green leafy vegetables (kale, collards, spinach, turnip greens, broccoli) as well as eggs, yellow corn, peas, tangerine, and orange bell peppers. Eggland's Best eggs contain 185 mg of lutein. Lutein is facilitated with ascorbic acid supplementation (Tanumihardjo et al, 2005).
Thiamin	For normal retinal and optic nerve functioning. Protective against cataracts (Jacques et al, 2005).
Riboflavin	For corneal vascularization. Protective against cataracts (Jacques et al, 2005). Riboflavin appears to play an essential role as a precursor to flavin adenine dinucleotide (FAD), a cofactor for glutathione reductase activity (Head, 2001).
Niacin	For healthy vision. Avoid excesses, which can cause nicotinic acid maculopathy (Spirn et al, 2003).
Folate	A strong protective influence on cortical cataract from use of folate or vitamin B_{12} supplements is a recent finding (Kuzniarz et al, 2001).
Vitamin B_6	For healthy conjuntiva. Untreated homocystinuria is known to cause ocular changes; vitamin B_6 can help to lower homocysteine levels.
Vitamin B_{12}	For retinal and nerve fibers. Protective against cataract (Kuzniarz et al, 2001). Found only in animal foods such as meat and milk.
Vitamin C	For healthy conjuntiva and vitreous humor. Long-term use of adequate vitamin C may delay or prevent early age-related lens opacity (Ferrigno et al, 2005; Valero et al, 2002). Orange and grapefruit juices, cantaloupe, oranges, green peppers, tomato juice, broccoli, kiwifruit, and strawberries are good sources. Vitamin C (ascorbic acid) is an antioxidant that lowers the risk of developing cataracts, and when taken in combination with other essential nutrients, can slow the progression of age-related macular degeneration and visual acuity loss (American Optometric Association, 2009).
Vitamin D	Helps prevent cancer, heart disease, diabetes, and age-related macular degeneration; it is the most potent steroid hormone in the human body, and is the only vitamin formed with the help of sunlight (American Optometric Association, 2009).
Vitamin E	Important for antioxidant properties. Protects the eyes from free radical damage and may slow the onset of cataracts. Long-term use of supplements may be beneficial (Jacques et al, 2005). Vitamin E is found in almonds, peanuts, peanut butter, sunflower seeds, safflower oil, margarines, fortified cereals, sweet potatoes, and creamy salad dressings.
Omega-3 Fatty Acids	Omega-3 fatty acids and fish are protective against AMD (Cho et al, 2001). Eating fish (sardines, salmon, herring, tuna, fortified eggs) weekly and cutting back on saturated fatty acids are important (Smith et al, 2000). Infants need a supply of DHA for up to a year for healthy visual development (Hoffman et al, 2004). Avoid use of large doses of alpha linolenic acid.
Omega-6 Fatty Acids	Omega-6 fatty acids in soybean, safflower, and sunflower oils may be protective against glaucoma (Kang et al, 2004) but not against cataracts. High doses of canola, flaxseed, and soybean oils may actually increase the risk of cataracts (Jacques et al, 2005).
Selenium	Pathophysiological mechanisms of cataract formation include deficient glutathione levels contributing to a faulty antioxidant defense system within the lens of the eye; nutrients that increase glutathione levels and activity include selenium (Head, 2001; Flohe, 2005).
Sodium	Sodium-restricted diets may protect against cataracts (Cumming et al, 2000).
Zinc	For healthy retina, choroid, and optic nerve. Found in beef, chicken, oysters, mixed nuts, and milk. Zinc is an essential trace mineral, bringing vitamin A from the liver to the retina in order to produce melanin for a protective pigment in the eyes (American Optometric Association, 2009). Zinc is highly concentrated in the eye.

American Optometric Association. Web site accessed March 29, 2009, at http://www.aoa.org/x11813.xml.
[a]Long-term use of multivitamin, B group, and vitamin A supplements is associated with reduced prevalence of either nuclear or cortical cataract (Kuzniarz et al, 2001).

vitamins C and E and zinc should also be consumed daily. Snacking on nuts is an excellent choice.

Coordination Problems

- Self-feeding requires the ability to suck, to sit with head and neck balanced, to bring hand to mouth, to grasp cup and utensil, to drink from a cup, to take food from a spoon, to bite, to chew, and to swallow. Each person should be assessed individually to determine which, if any, aspects of coordination have been affected by his or her condition. Adjust self-feeding accordingly.
- Use clothing protectors at mealtime to maintain dignity.
- Assist with feeding if needed; use adaptive feeding equipment as needed (such as weighted utensils, large-handled cups, larger or smaller silverware than standard). Adjust table or chair height.
- Place all foods within 18″ reach at mealtime.

Chewing Problems

- Dentures should fit well and be adjusted or replaced as needed, such as after weight loss.
- Decrease texture as necessary; use a mechanical soft, pureed, or liquid diet. Season as desired for individual taste. Try to progress in textures if possible because chewing

is important for saliva production and for proper digestion of foods.
- Liquid or blenderized foods may be beneficial. If needed, use a tube feeding.
- For some persons, a straw may be helpful; for others, it is not. Speech therapists can assess this ability.
- Protein foods such as tofu, cottage cheese, peanut butter, eggs, cheese, and milk products can be used when meats or nuts cannot be chewed.
- If fresh fruits and vegetables cannot be consumed, use cooked or canned sources and juices. Use pureed foods when needed. If whole grain breads and cereals are not tolerated, use cooked cereals. Avoid rice or foods with small particles in dysphagia (see Section 7).

Hearing Loss

- Researchers and international scientists have found a gene that causes deafness in humans: *LRTOMT*.
- Alter diets as needed if diabetes, cardiovascular disease, or hypertension are present. A controlled carbohydrate diet, therapeutic lifestyle diet (low saturated fat), or the DASH diet may be appropriate.

Common Drugs Used and Potential Side Effects

- For glaucoma, a combination of medications is used to reduce elevated intraocular pressure and prevent damage to the optic nerve. Some may cause dry mouth or fatigue; monitor individually.
- For AMD, Chol-lowering medications (statins) may be protective. Clinical trials are in order.

Herbs, Botanicals, and Supplements

- Nutrients and botanicals that may prevent cataracts include folic acid, melatonin, and bilberry (Head, 2001). Flavonoids, particularly quercetin, and ginkgo biloba may increase circulation to the optic nerve (Head, 2001). Curcumin is also under study.
- Lutein and zeaxanthin, in whole food or supplemental form, have an impact on retinal function with the potential to preserve vision and prevent degeneration in AMD; further research is needed to determine an effective dose (Carpentier et al, 2009).
- Herbs and botanicals may be used; identify and monitor side effects. For glaucoma, oregano, jaborandi, kaffir potato, and pansy have been recommended but not confirmed as effective. For cataract, rosemary, catnip, and capers have not been found to be effective.

Counsel about use of herbal teas to avoid intake of toxic substances.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Discuss the importance of using the various therapies and medications.
- Discuss the role of nutrition in health, weight control, recovery and repair processes.
- For healthy eyes, nutrition plays an essential role; see Table 2-7. Consume 10 mg of lutein weekly, the equivalent of one cup of cooked spinach four times a week. Orange juice is a good choice for vitamin C. The DASH diet is a good plan.
- Provide instruction regarding simplified meal planning and preparation. Refer to agencies such as Meals-on-Wheels if needed.
- Discuss the tips appropriate for the individual (texture, finger foods, ease of placement at meals).

Patient Education—Food Safety

- Discuss simple hand washing or use of hand sanitizers before meals.

For More Information

- Age-Related Macular Degeneration Alliance
  http://www.amdalliance.org/
- American Association of Ophthalmology
  http://www.eyenet.org/
- American Academy of Ophthalmology
  http://www.aao.org/
- American Council for the Blind
  http://www.acb.org/siteindex.html
- American Occupational Therapy Association
  http://www.aota.org/
- American Optometric Association
  http://www.aoanet.org/conditions/eye_coordination.asp
- Coordination Problems: National Center for Education in Maternal and Child Health
  http://www.brightfutures.org/physicalactivity/issues_concerns/10.html
- Hearing Loss Association
  http://www.hearingloss.org/
- Hearing Disorders and Deafness
  http://www.nlm.nih.gov/medlineplus/hearingdisordersanddeafness.html
- Help for Vision Loss
  http://www.visionaware.org/getstarted_professionals
- Low Vision
  http://www.lowvision.org/
- National Library Service for the Blind and Physically Handicapped (NLS)
  http://www.loc.gov/nls
- National Eye Institute, NIH
  http://www.nei.nih.gov/health/
- Prevent Blindness America
  http://www.preventblindness.org/
- Save your vision
  http://www.aoa.org/documents/nutrition/Save-Your-Vision-Month-Release.pdf
- Vision Loss for Seniors
  http://www.afb.org/seniorsitehome.asp

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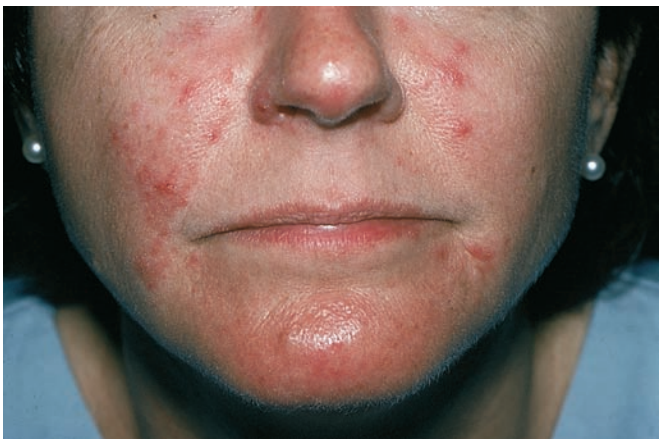
Skin Conditions, Pressure Ulcers, and Vitamin Deficiencies

SKIN DISORDERS

NUTRITIONAL ACUITY RANKING: LEVEL 1



Adapted from: *Goodheart HP. Goodheart's Photguide of Common Skin Disorders*. 2nd ed. Philadelphia: Lippincott Williams & Wilkins, 2003.

DEFINITIONS AND BACKGROUND

Human skin is the largest, independent peripheral endocrine organ of the body (Zouboulis, 2000). Skin is affected by both internal and external influences, which may lead to photo aging, inflammation, immune dysfunction, imbalanced epidermal homeostasis, and other disorders (Boelsma et al, 2001). The skin often reflects internal problems such as GI disturbances, alcoholism, or general malnutrition.

Nutritional factors impact hydration status, sebum production, elasticity, and skin cancer (Greenwald, 2001). A low-fat diet and foods rich in vitamin D and carotenoids may protect against some forms of skin cancer and actinic keratoses (Millen et al, 2004).

Many carotenoids found in nature are also found in the human body. Lutein and zeaxanthin are present in the skin and have shown significant efficacy against ultraviolet light-induced skin damage (Roberts et al, 2009). Retinoids have vitamin A biological activity; they have benefits for skin diseases and reversal of photo aging. Supplementation with vitamins, carotenoids, and polyunsaturated fatty acids has been shown to provide protection against ultraviolet light; however, sunscreen is still important (Boelsma et al, 2001).

Enzymes of the cytochrome P450 (P450 or CYP) family are drug-metabolizing enzymes that are induced in skin in response to xenobiotic exposure; they also play important roles in metabolism of fatty acids, eicosanoids, sterols, steroids, vitamins A and D (Ahmad and Mukhtar, 2004). In psoriasis, for example, many CYP enzymes are elevated; some relationship with celiac disease has been noted.

Acne affects many young people and may cause psychological distress. Iodine can aggravate acne; milk and dairy products contain a high level of iodine, but more studies are needed (Arbesman, 2005). Changes in diet are not the primary solution. Benzoyl peroxide is efficacious in mild-to-moderate acne, whereas adapalene and tretinoin are better for greater severity. Green tea polyphenols have anti-inflammatory effects, both taken internally and in topical creams.

Atopic dermatitis (AD), or eczema, causes itchy, inflamed skin. It usually affects the insides of the elbows, backs of the knees, and the face, but can cover much of the body.

AD often affects people who either suffer from asthma and/or hay fever or have family members who do (the "atopic triad"). AD flares when the person is exposed to trigger factors, such as dry skin, irritants, allergens, emotional stress, heat and sweating, and infections; avoiding triggers is the key. Because essential fatty acids (EFAs) form an important component of cell membranes, are eicosanoid precursors, and are required for both the structure and function of every cell, EFAs may benefit persons who have AD or psoriasis (Das, 1999).

Dermatitis herpetiformis (DH or Durhing's disease) is related to celiac disease (Karpati, 2004). There is the presence of villous atrophy and endomysial antibodies (EMAs) as markers (Kumar et al, 2001). A gluten-restricted diet is needed.

Epidermolysis bullosa is a hereditary condition in which blistering of the skin occurs with even slight trauma. It affects two of every 100,000 live births, occurring in both sexes and all ethnic groups. Nail dystrophy can occur, with rough, thickened, or absent fingernails or toenails. There may be blisters and problems with the soft tissue inside the mouth; protein–energy malnutrition, stunting, anemia, vitamin and mineral deficiencies are common. Treatment involves careful skin and wound care, and prevention of infections.

Nickel dermatitis affects 8–15% of women and 1% of men. Sensitization to nickel is often associated with ear piercing (Garner, 2004). In severe cases, reduce exposure to nickel from foods (Antico and Soana, 1999), and other sources.

Nummular eczematous dermatitis occurs with a rash; etiology is unknown. The rash is coin shaped and worsens in very hot or cold weather. Wool, soaps, frequent bathing (more than once a day), detergents, and rough clothing may be irritants. No special diet is needed unless food allergies are identified.

Psoriasis has patches of scaly red skin that burn, itch, or bleed. Calcitriol and vitamin D analogs are useful, along with controlled exposure to sunlight (Lehmann et al, 2004; May et al, 2004). Omega-3 fatty acids (specifically EPA) with a drug regimen of etretinate and topical corticosteroids may improve symptoms. Enbrel (etanercept), a tumor necrosis factor alpha (TNFα) inhibitor, relieves the clinical symptoms and may also clear up related depression and fatigue. There may be a relationship with celiac disease; if tests are positive, a gluten-free lifestyle will be needed.

Rosacea is a disorder of the central portion of the facial skin with onset in the third decade and peak in the fourth or fifth decade. A chronic and progressive condition of flare-ups and remissions, rosacea can be disfiguring if left untreated. Rosacea resembles other dermatological conditions, especially acne vulgaris. It affects one in 20 people, or 13 million people in the United States. Members of the same family tend to be affected; fair-skinned individuals of Northern and Eastern European descent (English, Scottish, Welsh, or Scandinavian) are most commonly affected (Litt, 1997). Green tea extract in creams may have benefits.

SAMPLE NUTRITION CARE PROCESS STEPS

Psoriasis

Assessment Data: Food records and intake calculations; many skin rashes, with psoriasis diagnosed 15 years ago.

Nutrition Diagnosis: Inadequate vitamin and fatty acid intake related to chronic history of psoriasis and poor diet as evidenced by diet history revealing high intake of beer, low intake of vitamin D and omega 3 fatty acids, and low serum vitamin D.

Intervention: Education about careful exposure to sunshine and use of vitamin D-fortified foods, fatty fish such as salmon and mackerel. Counsel about dietary changes to improve intake of omega-3 fatty acids and vitamin D_3. Advise cutting beer intake down or out. Coordinate care with physician to evaluate for celiac disease.

Monitoring and Evaluation: If positive for celiac, discuss gluten-free lifestyle. Intake records showing better food intake and lower intake of beer; improved quality of life with fewer outbreaks of psoriasis.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: Single genes control mediators such as enzymes, neuroendocrine transmitters, and cytokines that promote rosacea and probably other skin disorders.

Clinical/History	Lab Work	
Height	Alb (decreased in exfoliative dermatitis)	Thyroid-stimulating hormone (TSH), T4 level
Weight	Transthyretin	H & H, serum Fe
BMI	Serum zinc	Gluc
Growth pattern in children	Serum histamine (may be elevated)	Chol, Trig
Diet history	Skin tests for allergies	Serum Na^+, K^+
Family history of skin disorders, allergies	Anti-tTG, AGA, and/or EMA tests (for celiac)	Ca^{++}, Mg^{++}
Psoriasis?		Serum carotene
Rashes, blisters, pustules?		Retinol-binding protein (RBP)
Dermatitis?		CRP

INTERVENTION

OBJECTIVES

- Reduce inflammation, redness, or edema where present. Prevent further exacerbations of the condition.
- Apply nutritional principles according to the particular condition (such as vitamin D, omega-3 fatty acids, carotenoids, zinc).
- Identify any offending foods; omit from the diet any food allergens or intolerances, such as gluten.

FOOD AND NUTRITION

- **Acne.** Encourage intake of adequate zinc, carotenoids, and vitamin A foods. This condition is hormone dependent. It is less common in non-Western societies, so a high-fat diet that is low in fruit and vegetable intake should be avoided (Cordain et al, 2002). Drinking green tea can be highly recommended.
- **Acrodermatitis enteropathica.** Supplement with zinc because absorption is often impaired. Use protein of high biological value. Decrease excess fiber, if necessary, to normalize bowel function.
- **Atopic dermatitis.** Eczema is a general term for any type of dermatitis; AD is the most severe and chronic form. AD is a disease that causes itchy, inflamed skin. It typically affects the insides of the elbows, backs of the knees, and the face, but can cover most of the body. Diet therapies do not work well; do not automatically eliminate important foods such as milk and wheat. A trial without salicylates (berries and dried fruits), aspirin, penicillin, food molds, some herbs and spices, and FD&C yellow 5 may be useful. Infants may have hypersensitivity to milk, egg albumin, wheat, or linoleic acid but tend to outgrow it. Control energy excess in obese infants. Avoid herbal products, such as chamomile tea, for which allergy is possible. Green tea is safe and may be recommended.
- **Dermatitis herpetiformis.** A gluten-free diet is quite successful in treating this condition; see Section 7.
- **Epidermolysis bullosa (EB).** A balanced diet that includes extra protein, calories, and a multivitamin–mineral supplement will be useful. Highlight the nutrients that are beneficial, including omega-3 fatty acids, vitamin A, and zinc. Dysphagia is common, and may even lead to esophageal strictures. Anemia, contractures, gastroesophageal reflux (GERD), or scarring of the tongue may occur. Gastrostomy feeding may be needed.
- **Nickel dermatitis.** Avoid canned foods, such as tuna fish, tomatoes, corn, spinach, and other canned vegetables. Do not cook with stainless steel utensils. Chocolate, nuts, and beans may have slightly higher naturally occurring nickel than other foods; avoid large quantities (Christensen et al, 1999).
- **Psoriasis.** Psoriasis may precede arthritis by months or years; both are inflammatory processes. Because of their antiproliferative effects, calcitriol and other vitamin D analogs are highly efficient in the treatment of psoriasis (Lehmann et al, 2004; May et al, 2004). The therapeutic effect of UVB light therapy may be related to its skin synthesis of calcitriol. If celiac sensitive, omit gluten from diet, beverages such as beer, and medications.
- **Rosacea.** Alcoholic beverages (especially red wine), spicy foods, hot beverages, some fruits and vegetables, marinated meats, and dairy products may trigger flare-ups; avoid as needed. Limit use of all forms of pepper, paprika, chili powder, and curry. Substitute with cumin, oregano, sage, thyme, marjoram, turmeric, cinnamon, basil, and milder spices. Recommend drinking green tea.

Common Drugs Used and Potential Side Effects

When using cortisone ointments, use just a little and massage in well. Application once daily does as much good as using it more often. The potential for long-term use to suppress the adrenal gland exists (Matsuda et al, 2000; Woo and McKenna, 2003).

Acne

- Isotretinoin (Accutane) may be used for acne. Watch for a decrease in high-density lipoprotein (HDL) and an increase in TG. An increase in depression or suicide attempts seem to be related. Avoid taking with vitamin A supplements. Dry mouth can occur. Do not use during pregnancy.
- Retin A (retinoic acid) is useful for moderate cases of acne; side effects are mild.
- Tetracycline should not be taken within 2 hours of use of milk products or calcium supplements. Excesses of vitamin A can cause headaches or hypertension. Use more riboflavin, vitamin C, and calcium in the diet. Protein and iron malabsorption may result from prolonged use. Diarrhea is the major GI effect. Minocycline causes less GI distress and does not affect calcium metabolism as dramatically. Antibiotics are used their anti-inflammatory effect.

Atopic Dermatitis

- Topical cortisone (steroid) creams, such as Aclovate, usually have a mild effect on the nutritional status of the patient. Stronger brands or dosages may act like oral steroids and can suppress the adrenal system if taken for prolonged periods.

Epidermolysis bullosa

- Wound care products are used for tissue regeneration. Fluid replacement and protein loss may be associated with blistering.

Psoriasis

- Calcitriol and other vitamin D analogs are often highly effective. Topical products such as tazarotene (Tazorac) and calcipotriene, a form of vitamin D, have been available for years.
- Enbrel (enteracept), Humira (adalimumab), or Remicade (infliximab) are used for severe chronic plaque psoriasis. TNF inhibitors such as efalizumab (Raptiva) yield less-frequent itching and better quality of life (Ricardo et al, 2004).

Rosacea

- Antibiotic creams such as metronidazole (MetroCream) and azelaic acid (Finacea) are commonly prescribed. Tetracycline may also be prescribed; avoid taking within an hour of dairy or calcium-related supplements.
- Taking an antihistamine about 2 hours before a meal may counter the effects of histamine.
- Aspirin may reduce the effects of niacin-containing foods in sufferers affected by the flushing effect of these foods. Monitor multivitamin supplements and intakes carefully.

Herbs, Botanicals, and Supplements

- Counsel about use of herbal teas, especially related to potentially toxic ingredients. Herbs and botanicals may be used; identify and monitor side effects.
- For **acne**, salicylic acid helps break down blackheads and whiteheads and helps cut down the shedding of cells lining the hair follicles. Tea tree oil topical solutions may be beneficial for **acne** because of anti-inflammatory effects (Koh et al, 2002; Liao, 2001). Ointment containing tea leaf extract is effective for in impetigo, acne, and methicillin-resistant *Staphylococcus aureus*; results equal conventional treatments (Martin and Ernst, 2003).
- For prevention of **AD**, Chinese herbs, dietary restrictions, homeopathy, house dust mite reduction, massage therapy, hypnotherapy, evening primrose oil, emollients, topical coal tar, and topical doxepin are not proven effective (Hoare et al, 2000). Extracts of arnica (*Arnica montana*), chamomile (*Chamomilla recutita*), tansy (*Tanacetum vulgare*), and feverfew (*Tanacetum parthenium*) may cause allergic reactions. Calendula is often recommended but not proven effective.
- For **psoriasis**: bishop's weed, avocado, licorice, red pepper, Brazil nut, and purslane are not confirmed. Red clover is sometimes used for dermatitis or psoriasis; avoid use with warfarin or hormone replacement therapy.
- For **scabies**: evening primrose, onion, neem, and mountain mint have been proposed. No confirming studies are available.
- For **sunburn**: eggplant, plantain, and calendula have been proposed. Topical aloe is used for sunburn and mild burns; it causes GI cramping and hypokalemia if ingested.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Encourage the patient to read food, medication, and supplement labels. A symptom and food diary may be quite useful to identify any relationship between diet, allergies, and skin flare-ups.
- Encourage adequate fluid intake. Hydration of the skin is important. Drinking green tea can be highly recommended.
- Help the patient modify his or her diet as specifically indicated by the condition.
- Discuss the roles of nutrients in skin care. Sunscreens may prevent vitamin D from penetrating the skin, especially formulas with higher protective factors. Therefore, if dietary intakes are low in vitamin D, a supplement may be needed. Protein, vitamin A, and zinc are also important nutrients for healthy skin; describe good sources.
- Discuss the effect of EFAs on membrane function and how to include them in the diet. Omega-3 fatty acids reduce inflammation for some skin conditions. Eskimo skin care products, containing natural stable fish oil, is quite beneficial for improving skin elasticity (Segger et al, 2008). Otherwise, avoid topical products, except as recommended by the doctor.
- Flavones in medicinal plants, some vegetables and spices, have natural anti-oxidant with cytoprotective properties. Carrots, peppers, celery, olive oil, peppermint, rosemary and thyme contain natural levels of luteolin; more research is needed to identify its usefulness in medicine.

Patient Education—Food Safety

- The usual food safety habits are needed for good skin health. With open sores or exudative lesions on hands, a bandaid might be needed when preparing or serving food.

For More Information

- Acne Hotline
  http://www.niams.nih.gov/hi/topics/acne/acne.htm
- Acne Resources
  http://www.acne-resource.org/
- American Academy of Dermatology
  http://www.aad.org/
- Dystrophic Epidermolysis Bullosa Research Association of America
  http://www.debra.org/
- National Eczema Association
  http://www.nationaleczema.org/home.html
- National Psoriasis Foundation
  http://www.psoriasis.org/
- National Quality Measures Clearinghouse
  http://www.qualitymeasures.ahrq.gov/
- National Rosacea Society
  http://www.rosacea.org/
- NIH—Dermatitis
  http://www.niams.nih.gov/hi/topics/dermatitis/
- NIH—Eczema
  http://www.nlm.nih.gov/medlineplus/eczema.html

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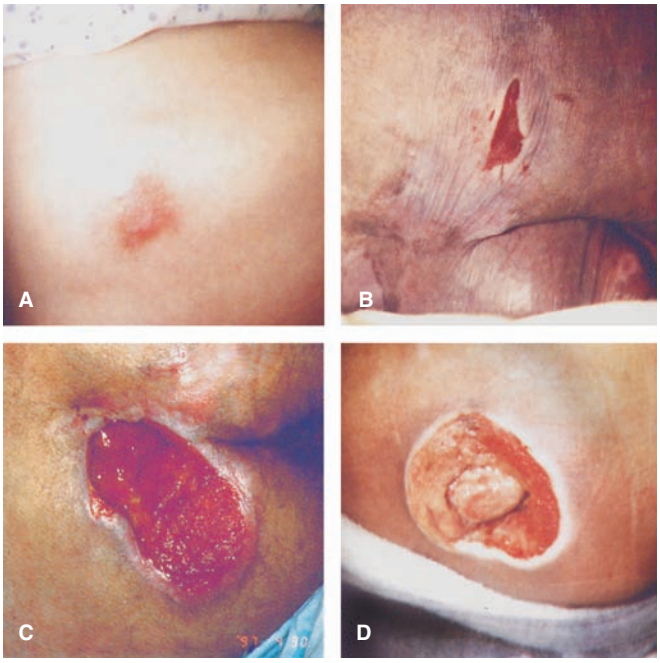
PRESSURE ULCERS

NUTRITIONAL ACUITY RANKING: LEVEL 2–3 (STAGES 1–2), LEVEL 4 (STAGES 3–4)



A = stage 1 B = stage 2 C = stage 3 D = stage 4
Adapted from: Nettina, Sandra M., MSN, RN, CS, ANP, *The Lippincott Manual of Nursing Practice,* 7th ed. Lippincott, Williams & Wilkins, 2001.

DEFINITIONS AND BACKGROUND

Pressure, friction, or shear and a lack of oxygen and nutrition to an affected area are associated with the development of pressure ulcers over bony or cartilaginous prominences (hip, sacrum, elbow, heels, back of the head). Pressure ulcers are common among patients with protein-energy malnutrition in HIV infection, pulmonary and cardiac cachexia, rheumatological cachexia, cancers, renal diseases, and among bedridden or paralyzed patients. With immobility, loss of lean body mass in muscle and skin, and lowered immunity, the risk of pressure ulcers increases by 74% (Harris and Fraser, 2004).

Many patients with pressure ulcers are below their usual body weight, have a low transthyretin level, and are not taking in enough nutrition to meet their needs (Guenter et al, 2000). Poor nutritional status and decreased oxygen perfusion are predictors of pressure ulcers; nutritional status and length of stay are predictors of ulcer severity in institutions (Williams et al, 2000). Patients with malnutrition are at risk for many complications, including incidence, progression, and severity of pressure ulcers. Risk factors should be assessed frequently: unintentional weight loss, incontinence, immobility, poor circulation (as in diabetes, peripheral vascular disease, or anemia), infection, prolonged pressure, multiple medications, serum albumin <3.4 g/dL, reduced functional ability, poor oral intake (<50% of meals over 3 days or longer), chewing or swallowing problems, and Chol levels below 160 mg/dL.

The mini-nutritional assessment (MNA) is a useful tool, with better results than using albumin levels or visceral proteins alone (Langkamp-Henken et al, 2005). Causes of malnutrition that should be carefully monitored include (Harris and Fraser, 2004): decreased appetite, requirement of assistance with meal intake, impaired cognition and/or communication, poor positioning, frequent acute illnesses with GI losses, medications that decrease appetite or increase nutrient losses, polypharmacy, decreased thirst response, decreased ability to concentrate urine, intentional fluid restriction because of fear of incontinence, fear of choking with dysphagia, isolation or depression, monotony of diet, and higher nutrient requirements. Reversible protein-energy malnutrition should be treated. A "MEALS ON WHEELS" mnemonic helps in identifying individuals at risk (Morley and Thomas, 2004): **M**edications—**E**motional problems—**A**norexia/alcoholism—**L**ate-life paranoia—**S**wallowing disorders—**O**ral problems—**N**osocomial infections—**W**andering/other dementia behaviors—**H**yperthyroidism or hypercalcemia—**E**nteric problems—**E**ating problems—**L**ow-salt, low-Chol diets—**S**tones (cholelithiasis).

Wound healing is complex and has three distinct phases. In each phase of wound healing, energy and macronutrients are required. Studies have established specific roles for nutrients including arginine; vitamins A, B, and C; selenium, manganese, zinc, and copper (Mathus-Vliegen, 2004). Administration of nutrition is an important part of wound healing.

Medicare costs attributable to pressure ulcer treatment are over $2 billion annually. The cost of caring for these preventable pressure ulcers is over $60,000 per patient (Edlich et al, 2004) and affects 10% of hospital admissions (Harris and Fraser, 2004). Pain, amputations, and osteomyelitis may result; mortality is often secondary to sepsis (Brem et al, 2003). In most cases, it is difficult to determine whether a pressure ulcer led to the terminal event, or whether the process of dying (decreased cardiac output, severe catabolic state) led to an unpreventable pressure ulcer (Braden, 1996).

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Clinical/History

- Height
- Weight
- Weight changes
- BMI
- BP
- Diet history
- Number, size and stage of ulcer(s)—see Table 2-8
- Exudate, infection or sepsis, fever
- Pain
- Abnormal motor coordination
- Changes in appetite, anorexia, indigestion
- Nausea/vomiting
- Diarrhea, bowel incontinence
- Urinary tract infections or incontinence
- Recent or frequent surgeries
- Prognostic Inflammatory and Nutritional Index (PINI)
- Braden scale: intense or prolonged pressure (activity, mobility, sensory perception) and tissue tolerance for pressure (nutrition, moisture, friction, and shear); scores range from 6 to 23, lower scores suggest higher risk
- Norton scale: physical condition, mental status, activity, mobility, and incontinence; rating >16 suggests high risk

Lab Work

- Gluc
- C-reactive protein
- Serum Chol
- BUN, Creat
- Serum Na^+, K^+ Ca^{++}, Mg^{++}
- H & H, serum Fe, serum ferritin
- Alb, transthyretin, RBP (usually decreased)
- N balance, transferrin
- Total lymphocyte count (TLC)
- Protime or INR
- Serum zinc
- Serum B_{12}

TABLE 2-8 Skin Changes with Aging and Pressure Ulcer Stages

Skin Change	Consequences
Thinning of epidermis	Increased vulnerability to trauma and skin tears
Decreased epidermal proliferation	Slower production of new skin cells
Atrophy of dermis	Underlying tissue more vulnerable to injury; decreased wound contraction
Decreased vascularity of dermis	Easy bruising and injury; decreased wound capillary growth
Compromised vascular response	Impaired immune and inflammatory responses
Fragility	Easy bruising and tearing
Suspected deep tissue injury	Purple or maroon localized area of discolored intact skin or blood-filled blister due to damage of underlying soft tissue from pressure and/or shear. The area may be preceded by tissue that is painful, firm, mushy, boggy, warmer or cooler as compared to adjacent tissue

*Staging**

Stage I	Intact skin with nonblanchable redness of a localized area usually over a bony prominence. Darkly pigmented skin may not have visible blanching; its color may differ from the surrounding area. The area may be painful, firm, soft, warmer or cooler as compared to adjacent tissue. Stage I may be difficult to detect in individuals with dark skin tones. May indicate "at risk" persons (a heralding sign of risk)
Stage II	Partial thickness loss of dermis presenting as a shallow open ulcer with a red pink wound bed, without slough. May also present as an intact or open/ruptured serum-filled blister. Presents as a shiny or dry shallow ulcer without slough or bruising. This stage should not be used to describe skin tears, tape burns, perineal dermatitis, maceration or excoriation. Bruising indicates suspected deep tissue injury
Stage III	Full thickness tissue loss. Subcutaneous fat may be visible but bone, tendon or muscle are not exposed. Slough may be present but does not obscure the depth of tissue loss. May include undermining and tunneling The depth of a stage III pressure ulcer varies by anatomical location. The bridge of the nose, ear, occiput and malleolus do not have subcutaneous tissue and stage III ulcers can be shallow. In contrast, areas of significant adiposity can develop extremely deep stage III pressure ulcers. Bone/tendon is not visible or directly palpable
Stage IV	Full thickness tissue loss with exposed bone, tendon or muscle. Slough or eschar may be present on some parts of the wound bed. Often include undermining and tunneling. The depth of a stage IV pressure ulcer varies by anatomical location. The bridge of the nose, ear, occiput and malleolus do not have subcutaneous tissue and these ulcers can be shallow. Stage IV ulcers can extend into muscle and/or supporting structures (e.g., fascia, tendon or joint capsule) making osteomyelitis possible. Exposed bone/tendon is visible or directly palpable
Unstageable	Full thickness tissue loss in which the base of the ulcer is covered by slough (yellow, tan, gray, green or brown) and/or eschar (tan, brown or black) in the wound bed. Until enough slough and/or eschar is removed to expose the base of the wound, the true depth, and therefore stage, cannot be determined. Stable (dry, adherent, intact without erythema or fluctuance) eschar on the heels serves as "the body's natural (biological) cover" and should not be removed

*Adapted from: National Pressure Ulcer Advisory Panel, 2007 staging. Web site accessed April 1, 2009, at http://www.npuap.org/.

SAMPLE NUTRITION CARE PROCESS STEPS

Pressure Ulcer

Assessment Data: Food intake records, weight records, need for enteral nutrition or feeding assistance, pressure ulcer team reports, other nutritional evaluations and risk measures. Check that Nursing is turning and repositioning every 2 hours and offering fluids.

Nutrition Diagnosis: Inadequate protein intake related to poor appetite and intake as evidenced by new stage 2 and stage 4 sacral pressure ulcers this past month with intake records showing limited protein consumption from milk, eggs, cheese and entrees; poor nutritional lab values (H & H, transthyretin) and elevated CRP.

Intervention: Education of patient, staff, or family members about the role of nutrition in wound healing. Counseling about acceptable sources of protein and enhancing menu items with protein powders or liquid supplements. Encourage use of oral supplements with medication passes; adequate fluid intake calculated as 30 mL/kg. Careful calculation of fluid, protein, and energy requirements according to stage of ulcers; recalculate as needed if healing does not occur. Micronutrient provision with vitamin–mineral supplement meeting 100% DRIs and RDA levels for zinc, copper, vitamins A and C.

Monitoring and Evaluation: Healing of pressure ulcers by 14 days after initiation of treatment; improved intake of protein foods to meet higher needs. Greater understanding by patient, family, or staff about the importance of nutrient-dense foods or formulas. Intake and output records indicating sufficient protein and energy intake. Labs improving for H & H, albumin, CRP.

INTERVENTION

OBJECTIVES

- Restore to a healthy nutritional status. Correct protein-energy malnutrition; this is of paramount importance.
- Monitor risk assessments. Nutrition risks evaluate the person's usual food intake pattern; intake of 3–5 days can be useful.
- Improve low-grade infections, fever, diarrhea, and vomiting. Support the patient's immune system to prevent sepsis.
- Heal the pressure ulcer(s) and prevent further tissue breakdown. Assess healing using an appropriate scale, such as the PUSH scale from the National Pressure Ulcer Advisory Panel at http://www.npuap.org/PDF/push3.pdf.

FOOD AND NUTRITION

- Provide a high-quality protein diet (Mathus-Vliegen, 2004). The guidelines for the treatment of pressure ulcers (2006) recommend 1.0–1.5 g protein/kg body weight. A deep ulcer or multiple sites may require 1.5–2 g/kg. It may be necessary to add protein powders to beverages, casseroles, tube feedings, and liquid supplements to get the adequate amount. Intake of protein greater than 2 g/kg of body weight may not be metabolized to increase protein synthesis.
- Recommended calorie levels for wound healing vary from 25 to 35 kcal/kg body weight; use lower levels for obese patients and higher levels for underweight patients. Whether or not pressure ulcers are preventable is controversial, but removing nutritional deficits is essential (Thomas, 2006).
- Provide small, frequent feedings if oral intake is poor, four to six times daily.
- Supplement diet with a general multivitamin–mineral supplement to supply adequate B vitamins, vitamin A, vitamin C, zinc, and copper; excesses are wasteful, do not necessarily speed the healing process, and may harm the immune system.
- A concentrated, fortified, collagen protein hydrolysate supplement may be of benefit to residents of long-term care facilities who have pressure ulcers (Lee et al, 2006).
- Feed by tube if necessary. With a large sacral pressure ulcer, central parenteral nutrition (CPN) may be the only way to feed if bowel incontinence is present; avoid. EFA deficiencies by including at least 2% of calories from lipid.
- Follow the algorithm given on page 117.

Common Drugs Used and Potential Side Effects

- Monitor the drug profile for potential side effects, especially for depletion of serum proteins or blood-forming nutrients. Drugs that can affect skin include antibacterials, antihypertensives, analgesics, tricyclic antidepressants, antihistamines, antineoplastic agents, antipsychotic agents, corticosteroids, diuretics, and hypoglycemic agents. Antibiotics may be needed in bacterial sepsis.
- Recommend, if needed, an appetite stimulant. Unintentional weight loss may be corrected by using dronabinol or cannabinoids (Marinol); megestrol acetate (Megace); oxymetholone.

Herbs, Botanicals, and Supplements

- Herbs and botanicals may be used; identify and monitor side effects and potential drug interactions.
- Counsel about use of herbal teas, especially regarding ingredients that may be toxic or ineffective.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Instruct nursing personnel and patient's family about the importance of adequate nutrition for healing of tissues.
- Discuss importance of maintaining healthy, intact skin. Skin should be kept clean and dry; avoid massage over bony prominences.
- Provide information about high-protein diets, and appropriate calorie and fluid levels.
- Where possible, improve ambulation and circulation to all tissues. Physical activity can help improve appetite.
- Discuss the role of nutrition in wound healing: collagen and fibroblasts require protein, zinc, and vitamin C for proper formation; adequate vitamins A and K, B-complex vitamins for healthy nerves and muscles.
- Discuss degree of assistance needed at mealtimes and provide ideas for self-help devices to increase overall intake (wet cloth under plate, curved flatware, two-handled cup, other adaptive equipment if needed.

INTERDISCIPLINARY NUTRITION CARE PLAN
Pressure Ulcer

Client Name: _____ **#:** _____ **Initiated by:** _____ **Date:** _____

SCREEN
Nutrition Screen diagnosis: Pressure Ulcer
Signed: _____ Date: _____

ASSESS *(Check any/all)*
❑ **Multiple pressure ulcers**
❑ **Stage III or IV pressure ulcer**
❑ **Receiving enteral or parenteral nutrition support**
Weight/BMI
- ❑ Weight change >3 lb/wk, >5%/mo, or >10%/6 mo
- ❑ BMI <20
- ❑ BMI >27

❑ **Infection (UTI, URI)**
Poor Oral Intake Symptoms
- ❑ Complex diet order
- ❑ Nausea/vomiting
- ❑ Poor appetite/early satiety
- ❑ Problems chewing/swallowing
- ❑ Depression/anxiety
- ❑ GI distress

Signed: _____ Date: _____

None

1 or more

HIGH-RISK INTERVENTIONS *(Check any/all)*
❑ **Power packing your diet** provided and explained
❑ **Food record** provided and explained
❑ **Fluid intake stressed**
Obtain Dr. orders as needed:
- ❑ RD referral for home visit(s)
- ❑ Monitor weight q:_____
- ❑ Monitor I & O q: _____
- ❑ Multiple vitamin/mineral supplement
- ❑ BID/TID supplements
- ❑ Tube feeding

❑ **Other:**_____
(See notes for documention.)

Signed: _____ Date: _____

Next visit

ASSESS RESPONSE *(Check any/all)*
❑ Stage III or IV pressure ulcer
❑ Multiple pressure ulcers
❑ Weight change not appropriate per goal
❑ Infection worsened
❑ Continued dehydration
❑ Continued Poor Oral Intake Symptoms
❑ **Other:**_____
(See notes for documention.)

Signed: _____ Date: _____

None

1 or more

OUTCOMES NOT ACHIEVED
Reassess/evaluate need for EN/PN (refer to Tube Feeding Nutrition Care Plan). Document on Nutrition Variance Tracking form.

GOALS (Check any/all):

❑ Maintain or improve nutritional status in ____ (goal time).

❑ Correct causes of involuntary weight loss where possible in ____ (goal time).

❑ Maintain or improve hydration status to prevent dehydration in ____ (goal time).

Weight ❑ maintained, or ❑ loss / ❑ gain of _____lb in ____ (goal time).

❑ Support pressure ulcer healing in ____ (goal time).

MODERATE RISK INTERVENTIONS *(Check any/all)*
❑ **Power packing your diet** provided and explained
❑ **Food record** provided and explained
❑ **Fluid intake encouraged**
❑ **Wound care explained**
Obtain Dr. orders as needed:
- ❑ RD chart consult
- ❑ Monitor weight q:_____
- ❑ Multiple vitamin/mineral supplement
- ❑ BID/TID supplements

❑ **Other:**_____
(See notes for documention.)

Signed: _____ Date: _____

Next visit

ASSESS RESPONSE *(Check any/all)*
❑ Weight change not appropriate per goal
❑ Dehydration
❑ Development of Stage III or IV pressure ulcer
❑ Development of multiple pressure ulcers
❑ Onset of new infection
❑ Exhibiting poor oral intake symptoms
❑ **Other:**_____
(See notes for documention.)

Signed: _____ Date: _____

None

OUTCOMES ACHIEVED
❑ Pressure ulcers improved
❑ Weight Maintained or improved
❑ Hydration status maintained or improved
❑ Development of multiple pressure ulcers
❑ **Other:**_____
(See notes for documention.)
❑ Repeat Nutrition Risk Screen in ____ days

Signed: _____ Date: _____

OUTCOMES ACHIEVED
❑ Pressure ulcers improved
❑ Weight Maintained or improved
❑ Hydration status maintained or improved
❑ Development of multiple pressure ulcers
❑ **Other:**_____
(See notes for documention.)
❑ Repeat Nutrition Risk Screen in ____ days

Signed: _____ Date: _____

Patient Education—Food Safety

- Hand washing will be important for patients, caretakers, and nurses before and after meals.

For More Information

- Agency for Healthcare Research and Quality
  http://www.ahrq.gov/consumer/bodysys/edbody6.htm
- Centers for Medicare and Medicaid Services
  http://www.cms.hhs.gov/
- European Pressure Ulcer Advisory PanelNutrition Guidelines
  http://www.epuap.org/guidelines/english1.html
- National Pressure Ulcer Advisory Panel
  http://www.npuap.org/
- Wound Care Network
  http://www.woundcarenet.com/index.html

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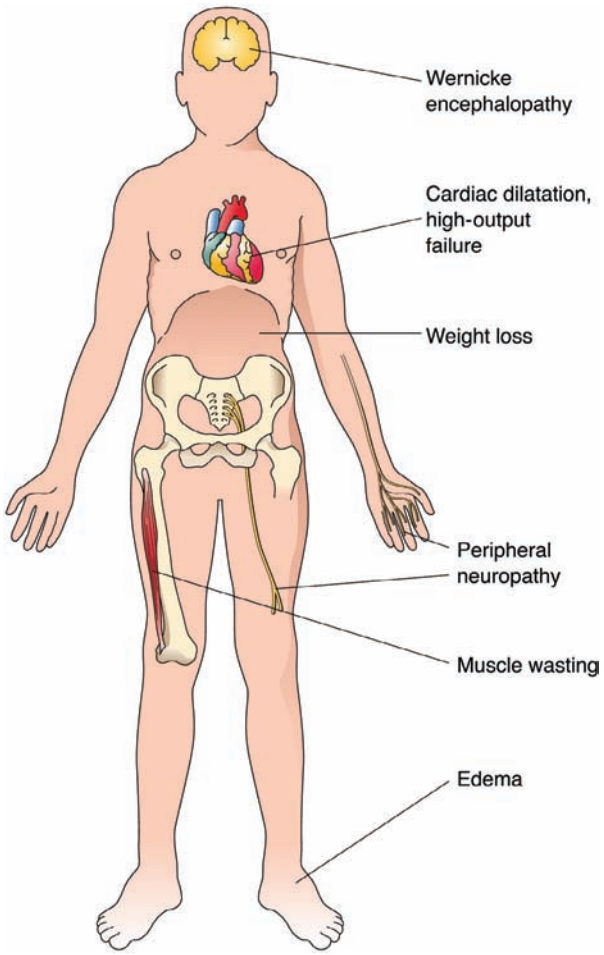
VITAMIN DEFICIENCIES

NUTRITIONAL ACUITY RANKING: LEVEL 3



Adapted from: Rubin E MD and Farber JL MD. *Pathology,* 3rd ed. Philadelphia: Lippincott Williams & Wilkins, 1999.

DEFINITIONS AND BACKGROUND

Vitamins are a part of a healthy diet. If a person eats a variety of foods, deficiency is less likely. However, people who follow restricted diets may not get enough of one or more particular nutrients. Deficiencies may be primary (self-induced by inadequate diet) or secondary to disease process. They are especially common in alcoholics, people who live alone and eat poorly, and among those who follow restrictive food fads. Vegetarians are also susceptible, especially for vitamin B_{12} deficiency. Appendix A provides greater detail about the vitamins, their sources, toxicities, and deficiencies. Table 2-9 summarizes concerns and physical signs of deficiency.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Clinical/History

Height
Weight
BMI
Diet history
Neurological, hepatic or renal changes
Physical signs of malnutrition (see Table 2-9 and Appendix A)

Lab Work

Vitamin A—serum retinol <0.35 mmol/L
Vitamin C—plasma concentrations <0.2 mg/dL
Vitamin D—25-OHD values <25 nmol/L
Vitamin E—plasma alpha-tocopherol <18 μmol/g

TABLE 2-9 Vitamin Deficiency Summary

Vitamin	Issues	Physical Signs
Vitamin A	Common in children. Night blindness and eye changes are often early signs. Many infections (such as measles) may cause deficiency. Vitamin A helps form and maintain healthy teeth, mucous membranes, skeletal and soft tissues, and skin. Retinol generates the pigments in the retina. Vitamin A promotes good vision (especially in dim light) and is required for healthy reproduction and lactation. Because beta-carotene is a precursor for vitamin A and because so many carotenoids play a role in maintaining good health, deficiencies of these phytochemicals may play an even larger role in maintaining vitamin A adequacy in the body.	Night blindness, Bitot's spots, xerophthalmia, follicular hyperkeratosis Reduced growth, changes in epithelial tissue, failure of tooth enamel and/or degeneration, loss of taste and smell.
Thiamin	Common in alcoholics, patients with heart failure, and persons with poor-quality diets. Thiamin helps convert carbohydrate (CHO) into energy; a high-CHO diet can deplete thiamin. It is also important for proper functioning of the heart, nervous system, and muscles.	Impairment of cardiovascular, nervous, and gastrointestinal systems.
Niacin and riboflavin	Often in conjunction with other B-complex vitamin deficiencies. Riboflavin (B_2) is important for growth, red cell production, and releasing energy from carbohydrates. Niacin assists in the functioning of the digestive system, skin, and nerves; it is important in the conversion of food to energy. A deficient diet or failure of the body to absorb niacin or tryptophan can cause signs of deficiency or pellagra. It is common in certain parts of the world where people consume large quantities of corn and is characterized by dermatitis, diarrhea, and schizophrenia-like dementia. It sometimes develops after gastrointestinal diseases or among alcoholics.	Niacin: Symmetrical, pigmented rash on areas exposed to sunlight, bright red tongue, Dermatitis, diarrhea, depression, and (sometimes) death (the 4 Ds). Riboflavin: Sore throat, hyperemia, edema of pharyngeal and oral mucous membranes, cheilosis, angular stomatitis, glossitis, seborrheic dermatitis, and normochromic, normocytic anemia. Magenta tongue.
Vitamin B_6	Can occur after surgery or as a result of poor diet. Vitamin B_6 deficiency has role in cardiac disorders (atrial fibrillation, hyperhomocysteinemia) and inflammation (Friso et al, 2004) and in dopamine release in the brain. Because vitamin B_6 plays a role in the synthesis of antibodies and red blood cells, a healthy immune system and circulatory system depend on it. The higher the protein intake, the more need there is for vitamin B_6; a high protein–low CHO diet may deplete vitamin B_6.	Seborrheic dermatitis, stomatitis, cheilosis, glossitis, confusion, depression. Convulsions or intractable seizures in infants and young children (Gospe, 2006); anemias; nerve and skin disorders.
Folic acid	May result in a megaloblastic anemia; supplementation is needed (see Section 12). Folic acid acts as a coenzyme with vitamins C and B_{12} in the metabolism and synthesis of proteins. It is needed to make red blood cells, to synthesize DNA, and to support tissue growth and cell function. There are roles for folic acid in disease prevention (e.g., neural tube defects, cancers, heart disease).	Depapillation of the tongue, rarely. Pregnancy-induced anemias; neural tube defects (Tamura and Picciano, 2006). Cardiovascular disease with elevated homocysteine levels.
Vitamin B_{12}	May also result in megaloblastic anemia (see Section 12 on anemias). Peripheral neuropathy and a positive Schilling test are needed to indicate B_{12} deficiency. Folic acid supplementation may mask a B_{12} deficiency; both should be given.	Tingling and numbness in extremities, diminished vibratory and position sense, motor disturbances including gait disturbances. Pernicious anemia and other anemias; poor vision; some psychiatric symptoms.
Pantothenic acid and biotin	Not common. Pantothenic acid is essential for metabolism and in the synthesis of hormones and cholesterol. Biotin is essential for metabolism of proteins and carbohydrate and the synthesis of hormones and cholesterol.	*Pantothenic acid:* No visible physical signs of note *Biotin*. Inflammation of the lips and skin.
Choline	May occur in long-term TPN use without lipid replacement. Plays a role in preventing neural tube defects along with folic acid.	No visible physical signs of note. Liver damage and altered DNA function.
Vitamin C	Occurs overtly with scurvy after 3 months without intake from inadequate consumption of fresh fruits and vegetables. Hypovitaminosis C can occur in the elderly and the homeless, among those who live alone or have psychiatric diseases, and in those who follow food fads. It is more common than realized in the general population. Long-term deficiency can be a concern for people with cancer or cataracts.	Follicular hyperkeratosis, petechiae, ecchymosis, coiled hairs, inflamed and bleeding gums, perifollicular hemorrhages, joint effusions, arthralgia, delayed wound healing. Weakness, myalgia, vascular purpura, loss of teeth. Biological signs include anemia, hypocholesterolemia, and hypoalbuminemia.

(continued)

TABLE 2-9 Vitamin Deficiency Summary *(continued)*

Vitamin	Issues	Physical Signs
Vitamin D	Insufficiency is a low threshold value for plasma 25-OHD (50 nmol/L). Secondary hyperparathyroidism, increased bone turnover, bone mineral loss, and seasonal variations in plasma PTH can occur with insufficiency. Deficiency is defined as 25-OHD values below 25 nmol/L; common among community-dwelling elderly who live in higher latitudes and among institutionalized elderly and patients with hip fractures. Vitamin D is produced in the skin by exposure to the sun and is found in fortified milk and other foods. For individuals who are not getting enough vitamin D in the diet, supplements may be helpful. The average adult under 50 needs 200 IU of vitamin D a day; 1 cup of vitamin D-fortified milk provides 50 IU of vitamin D (Surgeon General's Report, 2004). Recent studies suggest a role for vitamin D in autoimmune disorders, including multiple sclerosis (Mark and Carson, 2006); ovarian cancer (Zhang et al, 2006); type 1 diabetes or hypertension (Holick, 2006).	Widening at ends of long bones, rachitic rosary in children, rickets. Abnormal bone growth and repair; osteomalacia in adults; muscle spasms. Decreased immunity (Villamor, 2006).
Vitamin E	It is an antioxidant, protects body tissue from the damage of oxidation, helps form red blood cells, and supports the use of vitamin K. Abetalipoproteinemia is the most severe deficiency and occurs mainly in premature and sick children. Fat malabsorption occurs in deficiency, especially in children.	Rupture of red blood cells; nerve damage.
Vitamin K	Rare except in intestinal problems and short gut syndromes because intestinal bacteria in the healthy gut can make vitamin K. Healthy bones require sufficient vitamin K.	Poor wound healing or blood clotting. Osteopenia (Duggan et al, 2004).

Vitamin K—elevated prothrombin time, altered INR
Thiamin—erythrocyte transketolase activity >1.20 μg/mL/h; AST is often decreased
Riboflavin—erythrocyte glutathione reductase >1.2 IU/mg hemoglobin
Niacin—*N*-methylnicotinamide excretion <5.8 μmol/d
Vitamin B_6—plasma pyridoxal 5′phosphate <20 nmol/L
Vitamin B_{12}—serum concentration <180 pmol/L; elevated tHcy
Folic acid—serum concentration <7 nmol/L; red cell folate <315 nmol/L
Choline—plasma choline and phosphatidylcholine concentrations fall when humans are fed a choline-deficient diet, but otherwise there are no definitive tests; abnormal liver function tests may occur

INTERVENTION

OBJECTIVES

- Replenish the deficient nutrient and restore normal serum levels.
- Prevent or correct signs, symptoms, and effects of nutrient deficiency. For example, reduced immunity and high maternal and child mortality occur in populations with poor intakes of vitamin A (Cox et al, 2006). Vitamin D deficiency can lead to increased incidence of hip fracture (Cauley et al, 2008).

FOOD AND NUTRITION

- **Vitamin A deficiency.** Use a diet including foods high in vitamin A and carotene: carrots, sweet potatoes, squash, apricots, collards, broccoli, cabbage, dark leafy greens, liver, kidney, cream, butter, and egg yolk.
- **B-Complex vitamins:**
- **Thiamin deficiency** (beri-beri). Use a diet including foods high in thiamin: pork, whole grains, enriched

SAMPLE NUTRITION CARE PROCESS STEPS

Vitamin A Deficiency

Assessment Data: Food intake records, physical signs of deficiency, frequent bouts of infectious illnesses.

Nutrition Diagnosis: Inadequate vitamin A intake related to poor appetite and intake as evidenced by prolonged recovery after measles, recent onset of night blindness, serum retinol <0.35 mmol/L, complaints of lethargy and frequent illnesses.

Intervention: Education of patient, staff, or family members about the role of vitamin A for healthy immunity and vision. Counseling about good sources of vitamin A and carotenoids. Micronutrient provision meeting 100% DRI for vitamin A.

Monitoring and Evaluation: Total recovery after measles. Improvement in vision. Labs improving for retinol. Fewer infectious illnesses, colds, flu, etc. Improved quality of life.

cereal grains, nuts, potatoes, legumes, green vegetables, fish, meat, fruit, and milk in quantity. A high-protein/high-carbohydrate intake should be included.

- **Riboflavin deficiency.** Use a diet including foods high in riboflavin: milk, eggs, liver, kidney, and heart. Caution against losses resulting from cooking and exposure to sunlight.
- **Niacin deficiency or pellagra.** Use a diet including foods high in niacin and other B vitamins: yeast, milk, meat, peanuts, cereal bran, and wheat germ.
- **Folic acid deficiency.** Use fresh, leafy green vegetables, oranges and orange juice, liver, other organ meats, and dried yeast.
- **Vitamin B_6 deficiency.** Use dried yeast, liver, organ meats, whole-grain cereals, fish, and legumes.
- **Vitamin B_{12} deficiency.** Use liver, beef, pork, organ meats, eggs, milk, and dairy products.
- **Biotin deficiency.** Use liver, kidney, egg yolks, yeast, cauliflower, nuts, and legumes.
- **Pantothenic acid deficiency.** Use live yeast and vegetables.
- **Choline.** Include eggs, liver, beef, milk, oatmeal, soybeans, peanuts, and iceberg lettuce.
- **Vitamin C deficiency or scurvy.** Use a diet high in citrus fruits, tomatoes, strawberries, green peppers, cantaloupe, and baked potatoes.
- **Vitamin D deficiency.** Use fortified milk, fish liver oils, and egg yolks. Expose skin to sunlight if possible.
- **Vitamin E deficiency.** Use vegetable oil, wheat germ, leafy vegetables, egg yolks, margarine, and legumes.
- **Vitamin K deficiency.** Use a diet high in leafy vegetables, pork, liver, and vegetable oils.

Common Drugs Used and Potential Side Effects or Toxicity

Note: The DRI "tolerable upper intake levels" (UL) address the toxic side.

- **Vitamin A.** Absorption of vitamin A depends on bile salts in the intestinal tract. Controlled high doses may be prescribed for a short period of time. Beware of doses greater than the recommended upper limit per day for a long time, especially for children.
- **Thiamin.** A common dose is 5–10 mg/d of thiamin; anorexia and nausea may be common at the beginning of treatment. Intravenous therapy may be better tolerated.
- **Riboflavin.** Achlorhydria may precipitate a deficiency and may preclude successful correction. Alkaline substances destroy riboflavin.
- **Niacin.** Treatment with niacin may cause flushing. Niacinamide is a better choice; 200–400 mg of niacin or niacin equivalents may be used for a short time. Nicotinic acid can cause nausea, vomiting, and diarrhea.
- **Vitamin B_6.** Pyridoxine hydrochloride is the common content.
- **Pantothenic acid.** Pantholin is a drug that is prescribed as needed.
- **Choline.** Choline hydrochloride salt may be degraded by intestinal bacteria and cause a fishy body odor. This does not occur when lecithin is eaten in the diet.
- **Vitamin C.** Excesses can cause false-positive glucosuria tests. Cevalin or Cevita are drug sources; 50–300 mg/d may be given to correct scurvy. Excesses may have an antihistamine effect or cause diarrhea.
- **Vitamin D.** Calderol, Rocaltrol, Hytakerol, and Calciferol are common drug sources. Be sure to use vitamin D_3 [25-hydroxyvitamin D (25-OHD)] for greater effectiveness.
- **Vitamin E.** Aquasol E has no adverse side effects if used within measured dosage for age and daily requirements.
- **Vitamin K.** Vitamin K is usually injected to correct deficiency rather than using diet alone. Synkayvite, Mephyton, and Konakion are trade names.

Herbs, Botanicals, and Supplements

- Herbs and botanicals may be used by many individuals; identify and monitor side effects.
- Counsel about use of herbal teas, especially regarding ingredients that may be toxic.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Explain where sources of the specific nutrient may be found.
- Demonstrate methods of cooking, storage, etc., that prevent losses.
- Help the patient plan a menu incorporating his or her preferences.
- Discuss the use of vitamin and mineral supplements. Although they may be appropriate to correct a deficiency state, they may not be warranted for continuous or long-term use.

For More Information

- American Cancer Society—Vitamins
  http://www.cancer.org/docroot/eto/eto_5_2_5.asp
- Food and Nutrition Information Center
  http://www.nal.usda.gov/fnic/
- Medline Plus
  http://www.nlm.nih.gov/medlineplus/vitamins.html
- Merck Manual
  http://www.merck.com/mmpe/sec01/ch004/ch004a.html
- NIH Office of Dietary Supplements
  http://www.cc.nih.gov/ccc/supplements/intro.html
- Nutrient Data Laboratory
  http://www.ars.usda.gov/main/site_main.htm?modecode=12354500
- Nutrition Information
  http://www.nutrition.org/
- U.S. Department of Health and Human Services
  http://www.dhhs.gov/
- Vitamin Information Service
  http://www.vitamins-nutrition.org/

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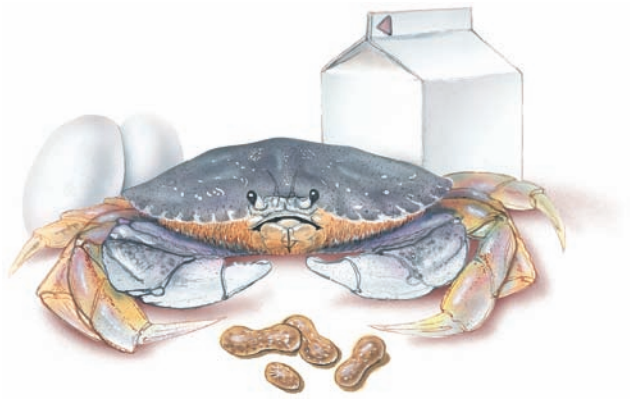
Food Allergy and Ménière's Syndrome

FOOD ALLERGY AND INTOLERANCES

NUTRITIONAL ACUITY RANKING: LEVEL 2–3 (SIMPLE); LEVEL 3–4 (COMPLEX)



DEFINITIONS AND BACKGROUND

It is important to distinguish food allergies from intolerances caused by toxins or drugs and metabolic disorders such as lactase deficiency or celiac disease. People who have a tendency toward allergy may develop sensitivity to new foods at any time. The manifestations of allergy are caused by the release of histamine and serotonin.

Prevalence of food allergies in the United States is about 6 million people; worldwide, they affect 3.5–4% of all individuals (Taylor, 2008). Allergic tendencies are inherited but not necessarily to a specific antigen (i.e., a parent with a genetic predisposition to severe bee sting reactions could have a child with a bee sting allergy, food allergy, or other allergy). Immune deviation and disturbed gut motility occurs in children with multiple food allergies, along with a maternal history of autoimmunity (Latcham et al, 2003). It is now recognized that children who have a fever before age 1 are less likely to develop allergies by age 6–7 years, and exposure to pets such as dogs at an early age builds immunity.

The most common symptoms of food allergies affect the GI tract (GIT): diarrhea, nausea, vomiting, cramping, abdominal distention, and pain. GI allergic manifestations can be classified as immunoglobulin E-mediated; "mixed" GI allergy syndromes, involving some IgE components and some non-IgE or T cell–mediated components; or eosinophilic gastroenteritis (Garcia-Careaga and Kerner, 2005; Lee and Burks, 2009). **GI food allergies** include a spectrum of disorders that result from adverse immunologic responses to dietary antigens, as defined by the National Institute of Allergy and Infectious Diseases (NIAID) in 2010:

- **Immediate GI hypersensitivity** refers to an IgE-mediated FA in which upper GI symptoms may occur within minutes and lower GI symptoms may occur either immediately or with a delay of up to several hours. This is commonly seen as a manifestation of anaphylaxis. Among the GI conditions, acute immediate vomiting is the most common reaction and perhaps the one best documented as immunologic and IgE mediated.
  - **Eosinophilic esophagitis** (EoE) involves localized eosinophilic inflammation of the esophagus. While EoE is commonly associated with the presence of food-specific IgE, the precise causal role of FA in its etiology is not well defined. Both IgE- and non-IgE-mediated mechanisms seem to be involved.
  - **Eosinophilic gastroenteritis (EG)** also is both IgE- and non-IgE-mediated, and commonly linked to food allergies. EG symptoms vary depending on the portion of the GIT involved and a pathologic infiltration of the GIT by eosinophils that may be quite localized or very widespread.
  - **Dietary protein-induced proctitis/proctocolitis** typically presents in infants who seem generally healthy but have visible specks or streaks of blood mixed with mucus in the stool. [5]IgE to specific foods is generally absent.
  - **Food protein-induced enterocolitis syndrome** (FPIES) is another non-IgE-mediated disorder presenting in infancy with vomiting and diarrhea severe enough to cause dehydration and shock. Cow's milk and soy protein are the most common causes. It typically manifests in the first few months of life, with severe projectile vomiting, diarrhea, and failure to thrive. A similar

condition has also been reported in adults, most often related to crustacean shellfish ingestion. Children will often outgrow an allergy to milk and soy by age 5 or 6. Recent research suggests that many children tolerate products containing egg or milk if extensively heated (Sicherer and Leung, 2009).

- **Oral allergy syndrome** (OAS), or pollen-associated FA syndrome, is a form of localized IgE-mediated allergy. Reaction is usually from direct contact with fresh fruits or vegetables, confined to the lips, mouth, and throat. OAS most commonly affects patients who are allergic to pollens. Symptoms include itching of the lips, tongue, roof of the mouth, and throat, with or without swelling, and/or tingling of the lips, tongue, roof of the mouth, and throat. There is a rapid onset of symptoms, but it is rarely progressive. Cross-reactivity occurs between certain pollen and food allergens, such as ragweed allergy with ingestion of bananas or melons; birch pollen allergy with ingestion of raw carrots, celery, potato, apple, hazelnut, or kiwi; or latex-fruit allergy with apples, avocado, banana, bell pepper, cherries, chestnut, kiwi, nectarines, peach, plums, potato and tomato.

Over 170 different foods have been known to produce an allergy or intolerance. Yet, over 90% of food allergies are caused by eight foods: eggs, milk, wheat, soy, fish, shellfish, peanuts, and tree nuts (TNs). Peanut and TN allergies are the leading cause of fatal or near-fatal food allergic reactions, including respiratory arrestand shock (Sicherer, 2003). The greatest danger comes from anaphylaxis, a violent allergic reaction that involves many parts of the body. Signs include itchy lips, tongue, or palate; metallic taste; flushing and itching or urticaria of skin; angioedema and edema of lips and tongue; nausea, vomiting, or diarrhea; tightness in chest or throat; dysphagia; hoarseness; dry cough; shortness of breath or wheezing; rhinorrhea or congestion; bronchospasm; syncope; chest pain; and hypotension.

Food allergy reactions usually occur within 2 hours. Immediate (1 minute–2 hours) or delayed reactions (2–48 hours) may also occur. **Food-induced anaphylaxis** is an IgE-mediated, rapid-onset, potentially life-threatening systemic reaction in which the affected individual may experience cardiovascular shock and/or serious respiratory compromise due to airway obstruction or bronchoconstriction (NIAID, 2010). Anaphylaxis occurs when a person is exposed to an allergen after being sensitized by at least one previous exposure. Peanuts, TNs, shellfish, milk, eggs, and fish are the most problematic. Tropomyosin is the protein that causes allergic reactions in shellfish (Taylor, 2008). Peanut allergy is quite serious, where even miniscule amounts have caused deaths (Lee and Burks, 2009; Scurlock and Burks, 2004).

Histamine mediates anaphylaxis by triggering a cascade of inflammatory mediators (Winbery and Lieberman, 2002). Histamine occurs naturally in foods such as cheese, red wines, spinach, eggplant, and yeast extract; it may elicit a response, including hives, urticaria, GI irritability, nausea, and flushing (Maher, 2002). A nonallergic reaction may occur from eating spoiled (scombroid) fish, which tends to be high in histamine; it may cause a reaction similar to anaphylaxis.

The GIT is highly involved in nutritional, physiological and immunological health. Gut-associated lymphoid tissue (GALT) is developed after birth with bacterial colonization, supporting development of protective IgA. An imbalance in T cells (type th2 greater than type th1) promotes autoimmune disease, Crohn's disease, or gut disease such as NEC (Walker, 2008). Persons with chronic urticaria may have impaired small bowel enterocyte function, with higher sensitivity to histamine-producing foods (Guida et al, 2000). Probiotics are being widely promoted for a role in reducing undesirable GIT responses to pathogens and allergens. In addition, retinoids regulate immunosuppressive factors within the mucosa; this role is just becoming understood (Strober, 2008).

Exercise and aspirin may facilitate allergen absorption from the GI tract (Matsuo et al, 2005). Food-dependent exercise-induced anaphylaxis (FDEIA) is an allergic reaction induced by intense exercise with the ingestion of a causative food (Matsuo et al, 2005). FDEIA is often associated with celery, chicken, shrimp, oyster, peaches, and wheat.

Alcoholic beverages may also contribute to hypersensitivity reactions: flushing syndrome, anaphylactoid reactions of urticaria/angioedema, asthma, food allergy, or exercise-induced anaphylaxis in susceptible subjects (Gonzalez-Quintela et al, 2004). Alcohol may promote development of IgE-mediated hypersensitivity to different allergens; alcohol abuse and even moderate alcohol consumption are associated with increased total serum IgE levels (Gonzalez-Quintela et al, 2004).

The Food Allergen Labeling and Consumer Protection Act (FALCPA) became effective January 1, 2006. Food ingredient labeling is the first line of defense for those with food allergies and their caregivers. Food ingredient labels should be read every time a food is purchased and used. Under FALCPA, food labels are to provide clear, consistent, and reliable ingredient labeling information by including "common English" names of the top eight major food allergens in food labeling. Legislation requires one of two options for food labeling with these common terms. The first is to list the food allergen in parentheses following the required ingredient term; for example, "whey (milk)" or "semolina (wheat)." The second option is to follow the ingredient declaration with a statement such as "contains flounder, pecan, wheat, and soy." In addition, all spices, flavors, and incidental additives that contain or are derived from a major food allergen will be labeled with the name of an allergen under either ingredient labeling option. For example, a flavor that contains an ingredient derived from milk might say "natural butter flavor (milk)."

Genetically modified (GM) foods are the product of biotechnology. Genetic bioengineering may, eventually, be able to reduce the level of allergens in the food supply (Lehrer, 2004), a common concern among members of the public (Celec et al, 2005). For GM foods, possible allergenicity of proteins is evaluated by comparison of their amino acid sequence with that of known allergens and determination of their stability during processing. GM crops that have been grown commercially are regularly evaluated for allergenic properties (Goodman et al, 2005). Overall, biotechnology can enhance the safety, nutritional value, and variety of foods without promoting allergies (American Dietetic Association, 2006).

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: Both genetics and environment play a role in promoting food-specific IgE responses (Tsai et al, 2009). Major food allergens are water-soluble glycoproteins. In normal individuals, allergens cause an IgA response along with suppressor CD8+ lymphocyte production. Antigen uptake is altered in children with cow's milk allergy; this affects about 3% of children in the U.S. Soy allergy affects about 1% of children. Peanuts have an Ara h 1 component that promotes IgE-mediated mast cell degranulation; tree nuts have similar effects (Khodoun et al, 2009). Gliadin is often found to be a factor in wheat allergy. Latex contains Hev b protein, to which some people are hypersensitive.

Clinical/History	Lab Work	
Height	H & H, serum Fe	Double-blind food challenge
Weight	Serum tryptase (elevated)	Patch tests for delayed hypersensitivity reactions
BMI	Serum histamine	CRP
Recent weight changes	Allergen microarray test for IgE profiling	Alb, transthyretin
Food diaries and symptoms history	Radioallergosorbent test (RAST)	BUN, Creat
BP	Allergen-specific IgE levels: skin prick tests (SPT) give 50% false-positives but are very reliable if negative	
Temperature		
Chronic GI distress, diarrhea		
Asthma or rhinitis		
Angioedema, urticaria		
Double-blind food challenge test		

SAMPLE NUTRITION CARE PROCESS STEPS

Intake of Unsafe Foods—Food Allergies

Assessment: Food diaries, food history, history of previous anaphylaxis and known food allergens.

Nutrition Diagnosis: Intake of unsafe food related to knowledge deficit as evidenced by anaphylaxis reaction after consuming peanuts.

Intervention: Education and counseling about identified food allergies (peanuts,) food labeling, recipes, and ingredients; evaluation of nutritional adequacy. Teach how to keep a food diary, keeping a chronological record of all foods eaten and any associated adverse symptoms. If needed, implement an elimination diet.

Monitoring and Evaluation: Review of food diaries; reports of no further problems with anaphylactic reactions to foods.

INTERVENTION

OBJECTIVES

- Careful clinical history, diagnostic studies, endoscopy, or double-blind food challenge may be needed. Children with AD could have a food allergy that can be diagnosed using a skin prick test and double-blind food challenge. Assist as needed in obtaining information; teach how to keep a food diary to track reactions to food.
- The therapy is avoidance of incriminating foods, plus education to avoid inadvertent exposures. Exclude or avoid the offending allergen. If it is not known, use an elimination diet to discover the cause. Note that "rotation diets" are not effective and are potentially dangerous.
- Monitor speed of onset of reactions, delayed versus immediate. The onset of delayed reaction may take from several hours to as long as 5 days. An immediate response is more common with raw foods; patient history may include diarrhea, urticaria, dermatitis, rhinitis, and asthma (see Asthma, Section 5). Allergic diarrhea is almost entirely IgE and mast-cell dependent, mediated by platelet-activating factor (PAF) and serotonin (Finkelman, 2007).
- Treatment of GI allergic disorders includes strict dietary elimination of offending food (Garcia-Careaga and Kerner, 2005). Use of protein hydrolysates has not been proven to be necessary (Osborn and Sinn, 2006).
- Breastfeeding should be promoted for primary prevention of allergic infants who are not lactose-intolerant (Garcia-Careaga and Kerner, 2005).
- Treat nutritional deficiencies or ensure adequate supplementation. Children who have multiple food allergies tend to have growth problems. The nutritional consequences of food allergy by various allergens are listed in Table 2-9.

Follow the new guidelines at http://www.niaid.nih.gov/topics/foodAllergy/clinical/Documents/guidelines.pdf.

FOOD AND NUTRITION

- The most common allergens in infants are eggs, wheat, milk, and fish. For children, cow's milk, eggs, soy, peanuts, wheat, TNs, and fish are often a problem. For adults, common allergens include shellfish, peanuts, and TNs. Peanuts are implicated in approximately one third of all cases of anaphylaxis.
- After skin testing, a double-blind, placebo-controlled food challenge (DBPCFC) can be useful (Perry et al, 2004). This should only be used under the guidance

of a physician in case there are immediate or severe reactions.
- For an elimination diet, use an unflavored elemental diet as a hypoallergenic base to which other foods are added as test challenges. Foods that seldom cause an allergic reaction include apples, apricots, artichokes, carrots, gelatin, lamb, lettuce, peaches, pears, rice, squash, and turkey; they may be used in this protocol.
- Read labels of foods prepared for the patient. Check all menu items served to patients. See Table 2-10.
- Monitor food preparation methods to exclude possible cross contact with the allergen.
- Monitor nutrient needs specific for the patient's age; evaluate for possible "hidden" ingredients.
- For infants, exclusive breastfeeding is best as it is nonallergenic.
- Lactating mothers may want to omit cow's milk, eggs, fish, and nuts from their own diets. See Table 2-11. Do not give cow's milk to infants until after they are 1-year old.
- Children should not eat peanuts, nuts, or fish until they are 3-year old. Toddlers should not eat eggs until after 2 years of age.
- Persons with rhinitis may be sensitive to monosodium benzoate in fruit juice, pie filling, pickles, olives, salad dressings, and fruit drinks.

Common Drugs Used and Potential Side Effects

- Aspirin may trigger skin reactions, which are associated with the inhibition of cyclooxygenase-1 (COX-1) and characterized by overproduction of cysteinyl leukotrienes; these reactions are due to the interference of aspirin-like drugs with arachidonic acid metabolism (Mastalerz, 2005).

TABLE 2-10 Major Food Allergens and Nutritional Consequences

Most Common	Nutrients of concern
Milk	Check for deficiencies in protein, riboflavin, calcium, and vitamins A and D.
Eggs	Check for iron from other sources.
Fish and shellfish	Other protein sources will be needed. Niacin, vitamin B_6, vitamin B_{12}, omega-3 fatty acids, phosphorus, and selenium should be available from other foods.
Nuts, tree	Protein, fatty acids, and other nutrients will be needed from other sources in the diet. Often, children outgrow a tree nut allergy.
Peanuts	Protein, fatty acids, and other nutrients will be needed from other sources in the diet.
Soy	Protein and other nutrients may be needed from other sources.
Wheat	Check for sufficiency of B vitamins and iron from other sources.
Less Common	Most frequently tied to adverse reactions that can be confused with food allergy are yellow dye number 5, monosodium glutamate (MSG), and sulfites.
Food additives: Tartrazine (not a true food allergen)	Yellow dye number 5 can cause hives, although rarely. FD&C Yellow No. 5, or tartrazine, is used to color beverages, dessert powders, candy, ice cream, custards, and other foods. The color additive may cause hives in fewer than one out of every 10,000 people. By law, whenever the color is added to foods or taken internally, it must be listed on the label so those who may be sensitive to FD&C Yellow No. 5 can avoid it (http://www.cfsan.fda.gov/~dms/qa-top.html).
MSG (not a true food allergen)	Dietary glutamate is a major energy source for the intestines and placenta. The brain is well protected against a flux of glutamate, and it is not toxic. Glutamate is found naturally in foods such as tomatoes and cheeses and is released in protein hydrolysis during stock or soup preparation. It is added to foods in crystalline form as MSG. MSG, which is 14% sodium, is used as a flavor enhancer, known as "umami." Glutamate helps to stimulate the vagus nerve and helps to facilitate digestion and nutrient absorption (Fernstrom and Garattini, 2000). MSG enhances flavor, but when consumed in large amounts, it can cause flushing, sensations of warmth, lightheadedness, headache, facial pressure, and chest tightness; these effects are temporary. These adverse reactions, "Chinese restaurant syndrome," have not been confirmed in double-blind studies (Geha et al, 2000).
Mustard	Mustard allergy is not as uncommon as previously believed (Figueroa et al, 2005). There is a relationship with mugwort pollinosis and plant-derived food allergies. A relationship between this syndrome and food-dependent exercise-induced anaphylaxis has also been reported (Figueroa et al, 2005).
Rice	Certain ethnic groups may have sensitivities to foods that may not be as allergenic for other populations. An example is an Asian person who develops an allergy to rice. Some of this may be dose-related exposure.
Spices	Spices may cause delayed-typed contact allergic or immediate allergic reaction. Sesame seed is a fairly common allergen. Carmine/cochineal is another minor allergen.
Sulfites (not a true food allergen)	Although not an IgE-mediated allergic response, sulfites can produce life-threatening reactions similar to the major food allergens. To help sulfite-sensitive people avoid problems, FDA requires the presence of sulfites in processed foods to be declared on the label and prohibits the use of sulfites on fresh produce intended to be sold or served raw to consumers (FDA Consumer: http://vm.cfsan.fda.gov/~dms/wh-alrg1.html). Foods such as wine, beer, dried fruits and vegetables, maraschino cherries, and dried or frozen potatoes may contain sulfites. No specific nutrient deficits are likely if omitted from the diet.

TABLE 2-11 Food Processing Concerns

Manufacturing processes	The food industry has taken steps to address the needs of consumers with food allergies, including changes to manufacturing processes to reduce the potential for cross-contact with major food allergens. Under existing good manufacturing practice (GMP) regulations, reasonable precautions must be taken to prevent cross-contact with major allergenic proteins. In instances when cross-contact cannot be avoided, even when complying, food and ingredient manufacturers use labeling that informs the food allergic consumer of the possible presence of allergens in the food. Food manufacturers label the ingredients in their products in accordance with existing regulatory requirements. The rule is, "no protein, no problem."
Oils in processing	Most oils used in food processing and for sale to the public contain no protein and are extracted from the oilseed or nut using solvents and then are degummed, refined, bleached, and deodorized. Some oils are mechanically extracted (cold pressed) and left unrefined to purposely maintain the flavor; these oils may contain protein and be allergenic.
Product recalls	Undeclared food allergens have been responsible for many food product recalls during the past decade, and the food industry has made significant investment, effort, and improvements in allergen control during this time (Hefle and Taylor, 2004). More research will be important.

- Cyclosporine, when used, can be steroid sparing; monitor BUN, Creat, and BP (Kaplan, 2004).
- Epinephrine is the synthetic version of naturally occurring adrenaline. It is the first line of defense for anaphylaxis and often requires an emergency room visit. Injectable epinephrine should be carried by those who are prone to allergic reactions to food and other allergens. An Epi-Pen provides a single dose; the Ana-Kit provides two doses.
- Oral antihistamines, such as Benadryl or Atarax or Vistaril, should be taken with food. Dry mouth, constipation, and GI distress are potential side effects.
- H_1-antihistamines (such as ranitidine, cimetidine) are adjunctive treatment therapy for acute anaphylactoid reactions, but they have a slow onset of action when compared with epinephrine, the medication of choice (Winbery and Lieberman, 2002). They are a mainstay of therapy for urticaria; nonsedating products include fexofenadine, loratadine, and desloratadine (Monroe, 2005).
- Treatment with topical or systemic steroids is used if all dietary measures are unsuccessful (Garcia-Careaga and Kerner, 2005).
- Consumption of omega-3 fatty acids can reduce the severity of asthma symptoms. With fish allergy, use nonfish oil sources.
- Probiotics have possible use for treatment of AD (Vanderhoof, 2008). Other new treatments for AD, urticaria and angioedema include biologics, vitamin D, and skin creams (Sicherer and Leung, 2009).
- Scientists at NIH fed mice a mixture of whole peanut extract (WPE) and a toxin called staphylococcal enterotoxin B (SEB) to simulate the human anaphylactic reaction to peanuts in mice. They are trying to develop a method for desensitizing people who are allergic to peanuts over time.

Herbs, Botanicals, and Supplements

- The pathway that activates transcription factors can be interrupted by phytochemicals derived from turmeric (curcumin); red pepper (capsaicin); cloves (eugenol); ginger (gingerol); cumin, anise, and fennel (anethol); basil and rosemary (ursolic acid); garlic (diallyl sulfide, S-allylmercaptocysteine, ajoene); and pomegranate (ellagic acid) (Aggarwal and Shishodia, 2004).
  - Bee pollen does not prevent allergies. It may, in fact, cause asthma, urticaria, rhinitis, or anaphylaxis after eating plants that cross-react with ragweed, such as sunflowers or dandelion greens.
  - Food/plant sensitivities are common (e.g., melon/ragweed, apple/birch, wheat/grasses). Be wary of herbal teas.
  - Jewelweed, parsley, stinging nettle, amaranth, gingko, chamomile, and feverfew have been proposed for use in allergies or with hives; no long-term studies are needed.
  - Sweeteners are not usually allergenic. After reviewing scientific studies, the FDA determined in 1981 that aspartame is safe for use in foods. Persons who have phenylketonuria (PKU) should not use it because it is made from phenylalanine.
  - For conditions such as allergic asthma, clinical trials are underway to determine effectiveness of Traditional Chinese Medicines (TCM).

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Education on reading ingredient labeling is essential. Ensure extensive nutrition counseling and health education for those who have food allergies to avoid nutrient deficiencies, to limit unnecessary restrictions, and to prevent reactions. Nutrient deficiencies will depend on the food groups involved and omitted.
- Signs of anaphylaxis (hoarseness, throat tightening, difficulty breathing, tingling in hands or feet or scalp, wheezing) should be taken seriously; call 911 immediately.
- Teach about possible cross-reactivity, such as cow's milk with goat's milk, or various types of fin fish.
- Tips to share with individuals who have allergies are found in Table 2-12.

Patient Education—Food Safety

- Intake of food allergens is actually "intake of unsafe food" in susceptible individuals. With GI disturbances and reactive symptoms, individuals with food allergies may be more sensitive to foodborne illness. Teach good hand washing, food preparation and storage tips.
- Children who have allergies should wear a Medic Alert tag or bracelet.

TABLE 2-12 Specifics of Food Allergies

Egg	Reactions are usually mild. Flu shots may contain egg albumin. Yolks are often tolerated. ALWAYS CONTAINS IT: Eggs, egg whites, dried eggs, egg solids, egg nog, albumin, cake, some candies or creamed foods, cookies, custard, doughnuts, egg rolls, some frostings, hollandaise sauce, some ice creams, lecithin, mayonnaise, meringue, some puddings, pretzels, Simplesse sweetener, souffle, waffles. MAY CONTAIN IT: Egg may appear as "albumin" in marshmallows, frozen dinners, and dry food mixes. Egg washes are often used on bakery goods to make them look shiny. Eggs are often used in glazes and icings.
Fish and shellfish	Avoid seafood restaurants. Abalone, clams, crab, crawfish, lobster, oysters, scallops, shrimp, cockle (sea urchin), and mussels are the shellfish that should be avoided. ALWAYS CONTAINS IT: Fish, shellfish, agar, alginic acid, ammonium alginate, anchovies, calcium alginate, caviar, disodium ionsinate, potassium alginate, propylene glycol alginate sodium alginate, imitation crab or "surimi," roe. MAY CONTAIN IT: Asian sauces, Caesar salad dressing, omega-3 fatty acid capsules or oils; Chinese, Vietnamese, Japanese, Indian, Indonesian, and Thai foods; fried foods, i.e., French fries, chicken nuggets (often cooked in the same oil as fish/shellfish); steak or Worcestershire sauces.
Latex	Natural rubber latex contains more than 35 proteins that may be related to type IgE–mediated allergy (Perkin, 2000). Latex-specific IgF may be responsible. Cross-reactivity has been documented with banana, avocado, kiwi, and European chestnuts; less commonly with potatoes; tomatoes; and peaches, plums, cherries, and other pitted fruits. Individuals with latex allergy also tend to report food allergies, including fish and shellfish (Kim and Hussain, 1999). Children with spina bifida and atopic dermatitis are a high-risk group for latex sensitization. Increasing age, additional sensitization to ubiquitous inhaled allergens, and enhanced total serum IgE values seem to be important variables for latex sensitization and further sensitization to the latex-associated foods (Tucke et al, 1999).
Milk	ALWAYS CONTAINS IT: Casein, caseinates, lactalbumins, lactoglobulins, lactose, nougat, rennet, milk, milk solids, nonfat or powdered milks, buttermilk, evaporated milk, condensed milk, yogurt, cream, cream cheese, sour cream, cheese, cheese sauces, cottage cheese, butter, butter fat, curds, whey, white sauces. MAY CONTAIN IT: Artificial butter flavor. Caramel color or flavoring, flavorings or seasonings, puddings, custards, sauces, sherbet. It may be necessary to acquire sufficient calcium from greens and broccoli or clams, oysters, shrimp, and salmon if not allergic to fish Calcium supplementation may also be warranted. Persons with a milk allergy can add vanilla or other flavorings to soy milk. Goat's milk has less lactalbumin, vitamin D, and folacin than cow's milk and supplements may be required. Some people may also be allergic to goat's milk, so caution must be used. Avoid early introduction of cow's milk in infancy.
Nuts, tree	Tree nuts include almonds, Brazil nuts, cashews, chestnuts, filberts, hazelnuts, hickory nuts, macadamia nuts, pecans, pine nuts, pistachios, and walnuts. Monitor food labels for nut paste, nut oil, and nut extracts. Avoid nut butters also. Read labels for ground or mixed nuts.
Peanut	Peanuts are a type of legume, but a person is more likely to be allergic to tree nuts than to beans, peas, and lentils. Avoid nut butters also; aflatoxins can cause an allergic-like reaction. For the food industry, new inexpensive kits are available to test for presence of peanut proteins in cookies, cereal, ice cream, and milk chocolate. Despite severity and reaction frequency to peanut and tree nut allergy, only 74% of children and 44% of adults in a large study sought evaluation for the allergy, and fewer than half who did were prescribed self-injectable epinephrine (Sicherer et al, 2003). It may be recommended that those children who outgrow their peanut allergy be encouraged to eat peanut frequently and carry epinephrine until they demonstrate true peanut tolerance (Fleischer et al, 2004). ALWAYS CONTAINS IT: Peanuts, mixed nuts, peanut butter, peanut oil, peanut flour, ground or mixed nuts, artificial nuts, nougat, many types of candy or cookies, ethnic dishes made with peanut oil, some egg rolls, marzipan. MAY CONTAIN IT: Peanut butter may be used to keep egg rolls from falling apart or in chili as a thickener.
Soybean	Some people are also allergic to legumes such as chickpeas, navy beans, kidney beans, black beans, pinto beans, lentils, and peanuts. ALWAYS CONTAINS IT: Soybeans, soybean oil, margarines made from soybean oil, soy sauce, soy nuts, soy milk. Reading food labels will be very important. COMMON SOURCES: Soy protein, textured vegetable protein, hydrolyzed plant protein, lecithin, miso, soy sauce, Worcestershire sauce, tofu, tempeh, some vegetable broths.
Wheat	Wheat-dependent, exercise-induced anaphylaxis (WDEIA) and baker's asthma are different clinical forms of wheat allergy (Mittag et al, 2004). ALWAYS CONTAINS IT: Whole-wheat or enriched flour, high-gluten flour, high-protein flour, bran, farina, bulgur, durum, wheat malt, wheat starch, modified starch, wheat germ, graham flour, wheat gluten, matzoh/matzoh meal, semolina, bread crumbs, cereal extract, dextrin, malt flavoring, modified starch. COMMON SOURCES: Baby food, baked goods, baking mixes, breaded foods, processed meats, pastas, snack foods, soups, breads, cookies, cakes and other baked goods made with wheat flour; crackers, many cereals, some couscous, cracker meal, pasta, gelatinized starch, hydrolyzed vegetable protein, wheat gluten, vegetable gum, vegetable starch.

- Always work with an RD to identify foods and ingredients to avoid, and develop an eating plan to ensure that each child gets all the nutrients needed to grow and develop properly.

For More Information

- American Academy of Allergy, Asthma, and Immunology
  http://www.aaaai.org/
- American College of Allergy, Asthma, and Immunology
  http://www.acaai.org/
- Asthma and Allergy Foundation of America
  http://www.aafa.org/
- Cherrybrook Kitchens: Allergy free
  https://www.cherrybrookkitchen.com/index.html
- Food Allergy and Anaphylaxis Network
  http://www.foodallergy.org/

- Food Allergies Database
  http://allergyadvisor.com/
- Food Allergy Initiative
  http://www.foodallergyinitiative.org/
- Food and Nutrition Information Center
  http://www.nal.usda.gov/fnic/pubs/bibs/gen/allergy.htm
- Food Labeling
  http://www.fda.gov/fdac/special/foodlabel/food_toc.html
- Grocery Manufacturers Association
  http://www.gmaonline.org/
- Hidden Allergens
  http://allergyadvisor.com/hidden.htm
- International Food Information Council Foundation
  http://ific.org
- Kids with Allergies
  http://www.kidswithfoodallergies.org
- Mayo Clinic
  http://www.mayoclinic.com/health/food-allergy/DS00082
- Medline: Food Allergy
  http://www.nlm.nih.gov/medlineplus/foodallergy.html
- National Institute on Allergy and Infectious Diseases
  http://www3.niaid.nih.gov/
- Nutrition MD—allergies
  http://www.nutritionmd.org/nutrition_tips/nutrition_tips_managing_diseases/allergies.html
- Nutrition MD—Diet Makeover
  http://www.nutritionmd.org/makeover/basics/not.html
- RAST Testing
  http://www.labtestsonline.org/understanding/analytes/allergy/test.html
- Teen Allergies
  http://kidshealth.org/teen/food_fitness/nutrition/food_allergies.html

FOOD ALLERGY AND INTOLERANCES—CITED REFERENCES

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MÉNIÈRE'S SYNDROME (AUTOIMMUNE INNER-EAR DISEASE)

NUTRITIONAL ACUITY RANKING: LEVEL 1

DEFINITIONS AND BACKGROUND

Ménière's syndrome is also known as autoimmune inner-ear disease (AIED;) it affects the inner ear, causing disturbed fluid flow. Patient may have a history of otitis media, smoking, or allergies. Attacks last from a few hours to several days, but the vertigo causes disability in many patients. Ménière's disease affects about 1% of the population and presents with episodic vertigo, fluctuating hearing loss, tinnitus and aural fullness (Hamid, 2009).

Sodium restriction and diuretic treatment are early management measures (Devaiah and Ator, 2000; Minor et al, 2004).

Aggressive medical therapy can prevent disease progression and hearing loss. Treatment options are limited and usually targeted toward reducing endolymphatic hydrops (Hamid, 2009).

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Clinical/History

Height
Weight
BMI
Diet history
Fluctuating hearing loss
Tinnitus with roaring sensation
Vertigo, blurred vision
Nausea and vomiting
Known allergies?
BP

Lab Work

IgE levels
H & H, serum Fe
Alb
Electrocochleography (ECOG)
Electronystagmography, or balance test (ENG)
Auditory brainstem response
CT scan or MRI
Serum Na^{++}, K^{+}
I & O

INTERVENTION

OBJECTIVES

- Correct nausea and vomiting; replace any electrolyte losses.
- Avoid or decrease edema and fluid retention, which can aggravate an attack.
- Omit any known food allergens from the diet.

FOOD AND NUTRITION

- Low-sodium (1000–2000 mg) diet may be useful.
- Use a multivitamin–mineral supplement and foods that are nutrient dense. Calcium and vitamin D strengthen the bones of the inner ear. Folate, vitamins B_6 and B_{12} reduce high levels of tHcy, which can reduce blood flow to the cochlea. Vitamin B_{12} also protects the nerves of the ear.
- Provide a diet that is free of known allergens, specific for the individual.
- Some people report feeling better after eliminating caffeine, aspartame, or alcohol.

SAMPLE NUTRITION CARE PROCESS STEPS

Knowledge Deficit

Assessment: Food diaries and frequency of intake of high sodium foods, history of previous anaphylaxis or food allergens.

Nutrition Diagnosis: Knowledge deficit related to high sodium foods as evidenced by chronic fluid retention in the ear.

Intervention: Education and counseling about high sodium foods, food labeling, recipes, and ingredients. Teach how to keep a food diary, keeping records of foods eaten and any adverse symptoms related to Ménière's.

Monitoring and Evaluation: Review of food diaries; reports of fewer problems related to Ménière's syndrome.

Common Drugs Used and Potential Side Effects

- Diuretics are used to reduce edema in the ear. If thiazides are used (such as furosemide, Lasix), monitor for the need for a potassium replacement. Other treatments to lower the pressure within the inner ear include antihistamines, anticholinergics, or steroids.
- Diazepam (Valium) may cause nausea, drowsiness, fatigue, and other effects. Limit caffeine.

Herbs, Botanicals, and Supplements

- Herbs and botanicals may be used; identify and monitor side effects. Counsel about use of herbal teas, especially regarding toxic substances.
- For *earache*: ephedra, goldenseal, forsythia, gentian, garlic, honeysuckle, and Echinacea are sometimes recommended but have not been proven as effective.
- For *tinnitus*: black cohosh, sesame, goldenseal, and spinach have been suggested; no long-term studies are on record that prove effectiveness. Gingko biloba has been approved for tinnitus in Europe but research has proven little effect (Karkos et al, 2007).

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Discuss how a balanced diet can affect general health status.
- Discuss sources of sodium and hidden ingredients that could aggravate the condition.
- Relaxation and biofeedback techniques may be useful for enhancing pain tolerance.

For More Information

- Ear Surgery Center
  http://www.earsurgery.org/meniere.html
- Ménière's Disease Information Center
  http://www.menieresinfo.com/
- National Institute on Deafness
  http://www.nidcd.nih.gov/
- NIH—Ménière's
  http://www.nidcd.nih.gov/health/balance/meniere.asp

MÉNIÈRE'S SYNDROME—CITED REFERENCES

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FOODBORNE ILLNESS

FOODBORNE ILLNESS

NUTRITIONAL ACUITY RANKING: LEVEL 1 (PREVENTIVE COUNSELING); LEVEL 2 (CORRECTIVE THERAPY)

DEFINITIONS AND BACKGROUND

True foodborne illness involves GIT insults, infections, or intoxications resulting from contaminated beverages or food. Millions of cases occur annually, but only a few hundred are reported. The Centers for Disease Control and Prevention report that there are millions of cases each year in the United States alone. The most vulnerable are elderly people, pregnant women, immune-compromised people, and children. Bacterial pathogens cause the largest percentage of outbreaks; chemical agents, viruses, and parasites are often implicated. In addition, multistate outbreaks caused by contaminated produce and outbreaks caused by *Escherichia coli* O157:H7 remain a concern. See Table 2-13.

Pathogens often transmitted via food contaminated by infected food handlers are *Salmonella typhi* and other species, *Shigella*, *Staphylococcus aureus*, *Streptococcus pyogenes*, hepatitis A virus, norovirus, *Listeria*, and *E. coli* O157:H7. Personal hygiene is one of the most important steps in food safety. The Centers for Disease Control and Prevention and most health departments require that food handlers and preparers with gastroenteritis ***not*** work until 2 or 3 days after they feel better. Strict hand washing after using the bathroom and before handling food items is important in preventing contamination. Food handlers who were recently sick can be given different duties in the foodservice operation so that they do not have to handle food.

An outbreak occurs when two or more individuals develop the same symptoms over the same time period. Infants and children younger than age 6, people with chronic illnesses (such as HIV infection or cancer), pregnant women, and elderly individuals are most at risk. For most infections, reported incidence is highest among children aged <4 years (CDC, 2010). Table 2-14 lists the most common foodborne illnesses, their onset and duration.

Nausea, vomiting, diarrhea, abdominal cramping, vision problems, fever, chills, dizziness, and headaches may occur. Some people attribute their symptoms mistakenly to "stomach flu." The Foodborne Diseases Active Surveillance Network (FoodNet) of CDC's Emerging Infections Program conducts active, population-based surveillance in the United States. The incidence of Vibrio infection continues to increase while there have been declines in reported incidence of infections caused by Campylobacter, Listeria, Salmonella, Shiga toxin-producing Escherichia coli (STEC) O157, Shigella, and Yersinia have been observed (CDC, 2010).

A recent legislative provision requires school food authorities participating in the National School Lunch Program (NSLP) or the School Breakfast Program (SBP) to develop a school food safety program for the preparation and service of school meals served to children, based on the hazard analysis and critical control point (HACCP) system (Food and Nutrition Services, 2009). Figure 2-11 shows the Food Safety Pyramid of how issues are handled by health departments and the Centers for Disease Control and Prevention (CDC).

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Clinical/History

- Height
- Weight
- BMI
- Usual weight
- Weight loss/changes during illness
- Diet history
- Vomiting
- Diarrhea
- Nausea
- Abdominal cramps
- Blood or parasites in stools?
- Fever?
- Timing of symptoms after suspected meal
- Signs of dehydration; I & O

Lab Work

- Na^+, K^+
- Chloride (Cl^-)
- H & H, serum Fe

SAMPLE NUTRITION CARE PROCESS STEPS

Foodborne Illness

Assessment: Food diary reveals intake of unsafe food or beverage, altered GI function (diarrhea, nausea, vomiting).

Nutrition Diagnosis: Intake of unsafe food related to salmonella as evidenced by onset of multiple episodes of vomiting and foodborne gastroenteritis, diarrhea, and lethargy in past several days.

Intervention: Education about rehydration with foods containing sodium, potassium, fluids (such as Gatorade). Counseling about food safety measures, including hand washing, avoidance of cross-contamination, safe food storage.

Monitoring and Evaluation: Review of food diaries; fewer problems related to foodborne illness from intake of unsafe food.

TABLE 2-13 Tips for Educating Individuals about Food Allergies

Shopping in Food Stores
DELI: Ask to have the deli slicer cleaned before preparing the order. Avoid prepared foods because they often share bins and serving utensils. Request that clean gloves to be worn.
ICE CREAM SHOPS: Make sure they do not share scoops for different flavors.
PACKAGED FOODS: Read labels to detect hidden allergens. Choose foods made in facilities that do not make other problematic products. Re-read labels often as ingredients may change; if unsure, call the manufacturer.
SALAD BARS: Be careful with severe allergies because food can drop from one container into another.
Dining Out
AVOID FRIED FOODS, which often share oil with other problem foods.
INQUIRE AHEAD if possible and consult the chef on best menu picks for safe dining.
USE A PLEASANT BUT ASSERTIVE MANNER in explaining the situation to wait staff. Let them know that eating even a small amount of a certain food(s) will make you severely ill.
BE CAREFUL of sauces and soups. Make sure you know exactly what is in them before eating.
REGULAR PATRONAGE. Choose a favorite eatery that accommodates well and visit often.
At School
EDUCATE. Schools need to educate their entire staff, improve prevention and avoidance measures, make sure epinephrine is readily available and that the staff knows how to administer it, and use consumer agency resources (Munoz-Furlong, 2004). The Food Allergy Network has educational kits targeted at schools to assist in the training of the staff on food allergies.
MEDIC ALERT. Students should be encouraged to wear a Medic-Alert bracelet.
CAFETERIA MEALS. Food allergy continues to rise in childhood, and careful meal planning is needed.
At Home
KEEP A FOOD DIARY. Identify all symptoms, timing, and foods eaten.
READ FOOD LABELS every time a food is purchased and used.
FIND RECIPE BOOKS THAT PROVIDE ALTERNATIVES. Recipe books are available from formula companies, food manufacturers, the Food Allergy and Anaphylaxis Network, and registered dietitians.
PATIENT OR PARENT EDUCATION. Patients and parents must stay informed about how to handle allergic reactions (Stone, 2004).
At the Doctor's Office
TESTING. Cytotoxic testing, sublingual provocative tests, pulse tests, kinesiologic testing, yeast hypersensitivity, and brain allergy theories should be dismissed entirely.
After Anaphylaxis
To work with anaphylaxis, remember the "3 Rs": RECOGNIZE symptoms; REACT quickly; REVIEW what happened to prevent it from happening again.

INTERVENTION

OBJECTIVES

- Allow the GIT to rest after rehydration; progress diet as tolerated.
- Prepare and store all foods using safe food-handling practices and good personal hygiene. Temperatures should be maintained below 40°F or above 140°F for safe food handling, storage, and holding.
- Teach the importance of hand washing, care of food contact surfaces, and insect or rodent extermination. This is especially important in foodservice operations where members of the public are fed.
- Any person operating a foodservice operation should know and use Hazard Analysis and Critical Control Point (HACCP) procedures to evaluate critical control points where foodborne illness risk is high and use precautions and safeguards (McCluskey, 2004). Careful monitoring is recommended. For the aging population in particular, barriers against the use of HACCP should be minimized (Strohbehn et al, 2004).
- Sanitize all surfaces before food preparation; sanitize after each food item is prepared when using the same surface (e.g., cutting boards and slicers). See Table 2-15 for more safe food practices.

TABLE 2-14 Symptoms, Sources, and Pathogens That Cause Foodborne Illness

General Source of Illness	Symptoms	Bacteria
Raw and undercooked meat and poultry	Abdominal pain and cramping, diarrhea, nausea, and vomiting	*Campylobacter jejuni, Escherichia coli* O157:H7, *Listeria monocytogenes, Salmonella*
Raw (unpasteurized) milk and dairy products, such as soft cheeses	Nausea and vomiting, fever, abdominal cramps, and diarrhea	*L. monocytogenes, Salmonella, Shigella, Staphylococcus aureus, C. jejuni*
Fresh or minimally processed produce	Diarrhea, nausea, and vomiting	*E. coli* O157:H7, *L. monocytogenes, Salmonella, Shigella, Yersinia enterocolitica*, viruses, and parasites

Specific Illness	Symptoms	Bacteria	Onset and Duration
Meats, milk, vegetables, and fish (diarrhea). Rice products; potato, pasta, and cheese products (vomiting) Consider also: sauces, puddings, soups, salads, casseroles, pastries	Watery diarrhea, abdominal cramping, vomiting	*Bacillus cereus* Gram-positive, aerobic spore former	6–15 hours after consumption. Duration = 24 hours
Raw and undercooked meat and poultry; raw milk and soft cheeses	Abdominal pain and cramping, diarrhea (often bloody), nausea, and vomiting. Note: 40% of Guillain–Barré syndrome (GBS) cases in the United States are caused by campylobacteriosis	*Campylobacter jejuni*	2–5 days after exposure. Duration = 2–10 days
Improperly canned goods especially with low acid content—asparagus, green beans, beets, and corn; chopped garlic in oil; chile peppers; improperly handled baked potatoes wrapped in aluminum foil; home-canned or fermented fish. Honey contains spores	Muscle paralysis caused by the bacterial toxin; double vision, inability to swallow, slurred speech and difficulty speaking, and inability to breathe Infants appear lethargic with poor muscle tone, feed poorly, are constipated, and have a weak cry	*Clostridium botulinum*	18–36 hours after eating contaminated food; can occur as early as 6 hours or as late as 10 days. Duration may be weeks or months
Canned meats, contaminated dried mixes, gravy, stews, refried beans, meat products, and unwashed vegetables	Nausea with vomiting, diarrhea, acute gastroenteritis	*Clostridium perfringens*	Within 6–24 hours from ingestion; lasting 1 day
Contaminated food from poor handling	Watery stools, diarrhea, nausea, vomiting, slight fever, and stomach cramps; especially in immunocompromised patients	*Cryptosporidium parvum* (protozoa)	2–10 days after infection
Contaminated water with human sewage may lead to contamination of foods; infected food handlers. More common with travel to other countries	Watery diarrhea, abdominal cramps, low-grade fever, nausea, and malaise. In infants or debilitated elderly persons, electrolyte replacement therapy may be necessary	*Escherichia coli*; Enterotoxigenic *E. coli* (ETEC)	With high infective dose, diarrhea can be induced within 24 hours
Undercooked ground beef and meats; unpasteurized fruit juices such as apple cider; unwashed fruits and vegetables (lettuce, alfalfa sprouts); dry-cured salami, game meat; cheese curds. *E. coli* O157:H7 can survive in refrigerated acid foods for weeks	Hemorrhagic colitis (painful, bloody diarrhea) The condition may progress to hemolytic anemia, thrombocytopenia, and acute renal failure requiring dialysis and transfusions	*E. coli* O157:H7; Enterohemorrhagic *E. coli* (EHEC)	Onset is slow, 3–8 days after ingestion Antibiotics are not used as they can spread the infection Hemolytic uremic syndrome can be fatal, especially in young children

(continued)

TABLE 2-14 Symptoms, Sources, and Pathogens That Cause Foodborne Illness *(continued)*

Specific Illness	Symptoms	Bacteria	Onset and Duration
Processed, ready-to-eat products (undercooked hot dogs, deli or lunchmeat, unpasteurized dairy products). Postpasteurization contamination of soft cheeses, milk, or commercial coleslaw. Cross-contamination between food surfaces	Mild fever, headache, vomiting, and severe illness in pregnancy; sepsis in immunocompromised patients; febrile gastroenteritis in adults; meningoencephalitis in infants. May lead to meningitis or septicemia if untreated	*Listeria monocytogenes* (LM)	Onset is 2–30 days. Can be fatal
Direct contact or droplets from contaminated hands or work surfaces (stool or vomit). Most common on cruise ships	Gastroenteritis with nausea, vomiting, diarrhea; fever with chills; abdominal cramps; headache; muscle aches. Vomiting may be frequent and quite violent, but subsides within a few days. Drink liquids to prevent dehydration	*Norovirus* Virus cannot multiply outside human body. Once on food, it can be transmitted easily to humans	24–48 hours after ingestion of the virus but may appear as early as 12 hours. Lasts only 1 or 2 days
Raw or undercooked meat, poultry, fish, unpasteurized dairy products; unwashed fruits and raw vegetables (melons and sprouts). Need thorough cooking, hygiene, and sanitation	Diarrhea, fever, and abdominal cramps. Most people recover without treatment. However, elderly, infants, and those with impaired immune systems are more likely to have a severe illness requiring hospitalization and antibiotics	*Salmonella typhimurium*	12–72 hours after infection. Duration = 4–7 days
Raw or undercooked eggs; eggs in foods such as homemade hollandaise sauce, caesar and other salad dressings, tiramisu, homemade ice cream, homemade mayonnaise, cookie dough, frostings	Nausea and vomiting, fever, abdominal cramps, and diarrhea	*Salmonella enteriditis*	12–72 hours after infection. Duration = 4–7 days
Milk and dairy products; cold mixed egg, tuna, chicken, potato, and meat salads	Bloody diarrhea, fever, and stomach cramps	*Shigella* (causes Shigellosis)	24–48 hours after exposure
Meat, pork, eggs, poultry, tuna salad, prepared salads, gravy, stuffing, cream-filled pastries	Nausea, vomiting, retching, abdominal cramping, and prostration	*Staphylococcus aureus* Cooking does not destroy the toxin. Refrigerate foods immediately after preparation and meal service	Within 1–6 hours; rarely fatal. Duration = 1–2 days
Milk, ice cream, eggs, steamed lobster, ground ham, potato salad, egg salad, custard, rice pudding, shrimp salad Foodstuffs at room temperature for several hours between preparation and consumption	Sore and red throat, pain on swallowing, tonsillitis, high fever, headache, nausea, vomiting, malaise, rash, rhinorrhea Complications are rare and are treated with antibiotics	*Streptococcus pyogenes* Entrance into the food is the result of poor hygiene, ill food handlers, or the use of unpasteurized milk	Onset = 1–3 days
Raw or undercooked shellfish, especially raw clams and oysters, contaminated with human pathogen	Vomiting, diarrhea; chills, fever, and collapse Can be fatal in immunocompromised individuals	*Vibrio vulnificus, V. parahaemolyticus* This bacterium is in the same family as cholera. It yields a norovirus	16 hours after eating contaminated food. Duration = 48 hours
Raw or undercooked pork products. Postpasteurization contamination of chocolate milk, reconstituted dry milk, pasteurized milk, and tofu	Fever, abdominal pain, and diarrhea (often bloody) in children In older children and adults, right-sided abdominal pain and fever may be predominant, mimicking appendicitis. Rarely, skin rash, joint pains, and sepsis may occur	*Yersinia enterocolitica* Occurs most often in young children. Cold storage does not kill the bacteria	1–2 days after exposure. Duration = 1–3 weeks or longer

Adapted from the following Web sites (accessed April 7, 2009):
CDC: http://www.cdc.gov/az.do.
FDA: http://www.cfsan.fda.gov/~MOW/intro.html.
NIDDK: http://digestive.niddk.nih.gov/ddiseases/pubs/bacteria/#10.

TABLE 2-15 Safe Food Handling and Food Safety Guidelines

Food Preparation

- Clean hands, food contact surfaces, and fruits and vegetables.
- Meat and poultry should not be washed.
- Because bacteria are commonly found on foods such as cantaloupe, cilantro, and imported produce, wash all fresh fruits and vegetables. Scrub the outside of produce such as melons and cucumbers before cutting. Scallions have been linked to hepatitis A outbreak; cook them thoroughly.
- Discard cracked eggs; avoid using products from dented cans.
- Avoid food preparation when sick with viral or bacterial infections; use gloves if needed.
- Sanitize work surfaces and sponges daily with a mild bleach solution (2 teaspoons per quart of water is sufficient). However, if a work surface comes into contact with raw food, it should be sanitized after contact with each food.
- Separate raw, cooked, and ready-to-eat foods while shopping, preparing, and storing foods.
- Sanitize work surfaces after each food. Ideally, keep one board for poultry, another for meats, and another for produce to prevent cross-contamination. Discard cutting boards that are badly damaged.
- Chill (refrigerate) perishable food promptly and defrost foods properly. Thaw meats and poultry in the refrigerator, not at room temperature. If necessary, thaw in a sink with cold running water that allows continuous drainage or thaw quickly in the microwave and use immediately.
- Do not partially cook meat or poultry in advance of final preparation. Bacteria may still grow rampantly.
- Cook foods to a safe temperature to kill micro-organisms. Cook beef to proper internal temperature of 160°F, pork to 165°F, and poultry to 175°F. Cook hamburger to the proper temperature of 165°F; "pink in the middle, cooked too little." Monitor internal temperatures with an accurate food thermometer placed correctly into the meat or poultry.
- Boil water used for drinking when necessary; hold at boiling temperature for 1 minute.
- Avoid raw or partially cooked eggs, raw or undercooked fish or shellfish, and raw or undercooked meats because of potential foodborne illnesses.
- Avoid raw (unpasteurized) milk or products made from it.
- Avoid serving unpasteurized juices and raw sprouts.

Holding and Serving Foods

- Hold and serve foods at 140–165°F during meal service.
- Reheat foods to at least 165°F. Discard leftovers after the first reheating process.
- Keep hot foods above 140°F and cold foods below 40°F.
- Discard cooked foods that are left at room temperature for more than 2 hours.
- Reheat home-canned foods appropriately. In institutional settings, do not allow home-cooked foods at all.
- Only serve certain deli meats and frankfurters that have been heated to steaming hot temperature.
- Keep pet foods and utensils separate from those for human use.
- Use clean plates and separate utensils between raw and cooked foods.
- Cool foods quickly in shallow pans (2–4 inches deep). Temperature should reach 70°F within 2 hours. If food has not cooled to that level, place in the freezer for a short time. Then, wrap lightly and return to refrigerator.

Other Tips

- When traveling, avoid tap water and ice made from tap water, uncooked produce such as lettuce, and raw or undercooked seafood.
- Airline water may not be free from contamination. Use of bottled water is recommended. Coffee and tea may not be hot enough to kill all bacteria.
- See Fight BAC guidelines at http://www.fightbac.org.

FOOD AND NUTRITION

- For patients with extreme diarrhea or vomiting, feed with intravenous glucose (NPO) until progress has been made.
- Oral rehydration therapy may be a useful adjunct treatment in the recovery process.
- Start with bland or soft foods and then progress to a normal diet.
- Prolonged inability to eat orally may require tube feeding.

Common Drugs Used and Potential Side Effects

- Hydrochloric acid in the stomach protects against pathogens ingested with food or water. A gastric fluid pH of 1–2 is deleterious to many microbial pathogens. Neutralization of gastric acid by antacids or the inhibition of acid secretion by various drugs can alter stomach pH and may increase the risk of acquiring food- or waterborne illnesses (Smith, 2003).
- Octreotide (Sandostatin) may be used parenterally only. It may alter fat absorption and fat-soluble vitamin absorption.

- Antibiotics such as Puromycin, erythromycin, or a fluoroquinolone may be prescribed.
- For *Salmonella*, ampicillin, gentamicin, trimethoprim/sulfamethoxazole, or ciprofloxacin may be used. New strains of this bacteria have evolved so that they are more resistant to antimicrobial treatment (Foley and Lynne, 2008).
- *V. vulnificus* infection is treated with doxycycline or ceftazidime.

Herbs, Botanicals, and Supplements

- Note that herbs and botanicals themselves could be a source of foodborne bacteria and thus exacerbate an existing foodborne infection. If herbs and botanicals are used, identify and monitor for potential contamination and side effects.
- Counsel about use of herbal teas, especially regarding toxic substances.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Encourage safe methods of food handling. More males, African Americans, and adults aged between 30 and 54 years consume raw/undercooked ground beef than any other group; males, Caucasians, Hispanics, and young adults aged between 18 and 29 years often engage in poor hygienic practices (Patil et al, 2004).
- Monitor water supply for unexpected odors or color changes; report to authorities.
- Discuss ways to prevent further episodes of foodborne illness. See Table 2-15.
- Hand washing is important. Wash hands with soap before handling raw foods of animal origin, after handling raw foods of animal origin, and before touching anything else. Prevent cross-contamination in the kitchen. Proper refrigeration and sanitation are also essential.
- Avoid raw milk and cook all meats and poultry thoroughly. Drink only pasteurized milk.
- Bacteria may be found in raw vegetables and fruits. Wash before eating.
- Throw out bulging, leaking, or dented cans and jars that are leaking. Safe home canning tips can be obtained from county extension services or from the U.S. Department of Agriculture.
- Commercial mayonnaise, salad dressings, and sauces appear to be safe due to their content of acetic acid and lesser amounts of citric or lactic acids (Smittle, 2000). Thus, if items are prepared using cold dressings, they stay at proper temperatures longer.
- To prevent parasite infestation (such as *Giardia*), sewage treatment, proper hand washing, and use of bottled water is recommended (Kucik et al, 2004).
- Food handlers need more education to understand their role in food safety, especially non-English speaking staff members (DeBess et al, 2009). Correct hand washing procedures, involvement of both managers and food handlers, and support from health departments are all important (Pragle et al, 2007). Errors in methods of washing hands, utensils, and preparation surfaces between food preparation tasks are common (Kendall et al, 2004).
- Three key practices are needed for safe food handling: careful hand washing, using thermometers, and proper handling of food surfaces (Pilling et al, 2008). Barriers in the workplace often include time constraints, inconvenience, inadequate training, and insufficient resources (Howells et al, 2008).
- Cancer and imunocompromised patients are especially vulnerable to foodborne illness. Risk-reducing behaviors, better food handling of routine foods, and hand washing should be encouraged (Medeiros et al, 2008).
- Young adults also need education about food safety for themselves and for their future families (Byrd-Bredbenner et al, 2007).
- Low socioeconomic status has been shown to be linked with poorer quality produce and an increased reliance on small retail stores (Koro et al, 2010).
- Biotechnology has developed food crops that are more resistant to pests and have better nutritional value as well as having longer shelf-life for food safety. Some consumers are concerned about the safety of food irradiation, genetically modified foods (GMOs), and potential allergens. Nutrition professionals should reassure the public that GM items are safe to eat.

TABLE 2-16 Recommended Refrigerator Food Storage

Food	Time Period
Butter	1–3 months
Cheese, hard	6 months unopened; 3–4 weeks opened
Cheese, soft	1 week
Chicken, turkey	1–2 days
Eggs in shell	3–5 weeks
Eggs, raw	2–4 days
Fish, cooked	3–4 days
Fish, raw	1–2 days
Gravy	1–2 days
Hamburger, raw	1–2 days
Juices, chilled	3 weeks unopened, 7–10 days opened
Luncheon meat	3–5 days opened, 2 weeks in sealed package
Margarine	4–5 months
Milk	1 week
Pizza	3–4 days
Roast or steak	3–5 days (uncooked)
Sausage, raw	1–2 days
Sausage, smoked	1 week
Shellfish, fresh	1–2 days
Soups, stews	3–4 days
Yogurt	1–2 weeks

For More Information

- American Dietetic Association Home Food Safety Program
  http://www.homefoodsafety.org/index.jsp
- Biotechnology and Food Safety
  http://www.foodsafety.gov/~lrd/biotechm.html
- Bioterrorism and Food Safety
  http://www.fda.gov/oc/opacom/hottopics/bioterrorism.html
  http://www.foodsafety.gov/~fsg/bioterr.html
- Botulism
  http://www.cdc.gov/nczved/dfbmd/disease_listing/botulism_gi.html
- CDC Foodborne, Bacterial and Mycotic Diseases
  http://www.cdc.gov/nczved/dfbmd/disease_listing.html
- CDC Wonder—single access site
  http://wonder.cdc.gov/
- Codex Alimentarius—International Food Regulations
  http://www.fsis.usda.gov/Codex_Alimentarius/index.asp
  http://www.fsis.usda.gov/codex_alimentarius/Codex_Publications/index.asp
- Division of Emerging Infections and Surveillance Services (DEISS)
  http://www.cdc.gov/ncpdcid/deiss/index.html
- Drinking Water Safety
  http://www.epa.gov/safewater/dwh/index.html
- Frequently Asked Questions—Drinking Water
  http://www.epa.gov/safewater/faq/faq.html
- Federal Consumer Information Center
  http://www.pueblo.gsa.gov/food.htm
- Fight BAC
  http://www.fightbac.org/
- Food and Drug Administration Center for Food Safety and Applied Nutrition (CFSAN)
  http://www.cfsan.fda.gov/
- Food Defense and Emergency Response
  http://www.fsis.usda.gov/food_defense_&_emergency_response/index.asp
- FoodNet Incidence Figures
  http://www.cdc.gov/foodnet/factsandfigures.htm
- Food Safety Education
  http://www.fsis.usda.gov/Food_Safety_Education/index.asp
- Food Safety and Inspection Service (FSIS)
  http://www.fsis.usda.gov/
- Food Safety and Inspection—Risk Assessment
  http://www.fsis.usda.gov/Science/Risk_Assessments/index.asp
- Food Safety for Kids, Teenagers and Educators
  http://www.foodsafety.gov/~fsg/Fsgkids.html
- Food Safety for Seniors
  http://www.pueblo.gsa.gov/cic_text/food/foodsafetyfs/seniors.htm
- Government Food Safety Website
  http://www.foodsafety.gov/
- HACCP
  http://www.fsis.usda.gov/Science/Hazard_Analysis_&_Pathogen_Reduction/index.asp
  http://www.who.int/foodsafety/fs_management/haccp/en/index.html
  http://foodsafety.nal.usda.gov/nal_display/index.php?info_center=16&tax_level=1&tax_subject=178
- HACCP—International Alliance
  http://www.haccpalliance.org/sub/index.html
- Image Library
  http://www.fsis.usda.gov/News_&_Events/FSIS_Images/index.asp?src_location=IWT&src_page=FSE
- International Food Safety Sites
  http://www.foodsafety.gov/?7Efsg/fsgintl.html
- North Carolina State University
  http://www.ces.ncsu.edu/depts/foodsci/agentinfo/
- USDA Home Canning Guide
  http://www.uga.edu/nchfp/publications/publications_usda.html
- USDA Food Safety Index
  http://www.fsis.usda.gov/help/site_map/index.asp
- Water Quality Association
  http://www.wqa.org/
- World Health Organization—Biotechnology and GM Foods
  http://www.who.int/foodsafety/biotech/en/
- World Health Organization—Foodborne Illnesses
  http://www.who.int/topics/foodborne_diseases/en/index.html
- World Health Organization—Food Safety
  http://www.who.int/topics/food_safety/en/
- World Health Organization—International Travel and Health
  http://www.who.int/ith/en/index.html
  http://www.who.int/foodsafety/publications/consumer/travellers/en/index.html
- World Health Organization—Water Sanitation
  http://www.who.int/water_sanitation_health/mdg1/en/index.html

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SECTION 3

Pediatrics: Birth Defects and Genetic and Acquired Disorders

BACKGROUND AND CONSIDERATIONS

Because good nutrition is essential for achieving growth and development, screening and assessment should be an integral part of health care. Efforts should be made to enhance appetite and intake in children who are not with their families; familiarity is important. Simple nutritional screening tools can help identify children at risk for malnutrition, affecting one fourth to one third of children admitted to a hospital. When there are problems with growth, proper interventions and referrals are important. A checklist of "ABCDE" factors can be used to assure completion of all assessments. Poor health habits, limited access to services, and long-term use of multiple medications are common health risk factors (American Dietetic Association, 2010). See Table 3-1 for assessments and calculations and see Table 3-2 for common pediatric problems. Nationally credentialed dietetics professionals are best able to provide appropriate nutrition information as it pertains to wellness, good health, and quality of life (American Dietetic Association, 2010). In particular, children with developmental disabilities and special health care needs frequently have growth alterations (failure to thrive (FTT), obesity, or growth retardation), metabolic disorders, poor feeding skills, medication–nutrient interactions, or dependence on enteral or parenteral nutrition (PN; American Dietetic Association, 2010). Nutrition services should be provided throughout life in a manner that is interdisciplinary, family-centered, community-based, and culturally competent (American Dietetic Association, 2010).

TABLE 3-1 Useful Assessments in Pediatrics

Anthropometric

Use age-, gender-, and disease-specific growth charts from the Centers for Disease Control and Prevention (CDC) with trained personnel and appropriate equipment. *Figure 1 shows how to measure the length of an infant properly.*

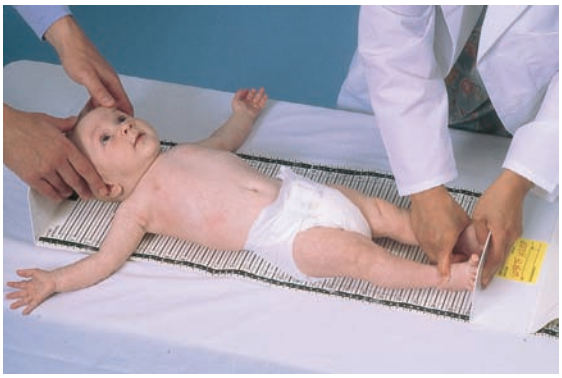

Figure 1. Measuring an infant. (From Bickley LS, Szilagyi P. *Bates' guide to physical examination and history taking,* 8th ed. Philadelphia: Lippincott Williams & Wilkins, 2003).

- Birth data (Weight, length, head circumference, size, gestational age)
  - Low birthweight ≤2500 g or 5.5 lb
  - Very low birth weight ≤1500 g or 3.5 lb
- Growth parameters: *Figure 2 provides the pediatric BMI tables*
  - Current height (ht) and weight (wt)
  - Wt/age <10th percentile or >85th percentile (overweight) or >95% (obese)
  - Ht/age <5th percentile
  - Wt/ht <5th percentile (underweight or FTT) or >85th percentile (overweight)
  - Head circumference <5th percentile (under 3 years of age)
- Pubertal staging (Tanner Stages), skeletal maturity staging
- Small for gestational age—need catch up growth to normalize length and weight; may later have CKD or metabolic syndrome as adults
- Unintentional weight loss

Behavioral—Psychosocial

- Developmental disorders: mental retardation, learning disorders, motor skills disorder, communication disorders, or PDDs
- Growth and development milestones

CALCULATED BODY MASS INDEX (BMI) FOR CHILDREN AND ADOLESCENTS

HEIGHT (in inches) \ WEIGHT (in pounds)	50	55	60	65	70	75	80	85	90	95	100	105	110	115	120	125	130	135	140	145	150	155	160	165
3'9"	17.4	19.1	20.8	22.6	24.3	26	27.8	29.5	31.2	33	34.716	36.452	38.2	39.9	41.66	43.395	45.131	46.87	48.6	50.34	52.07	53.81	55.546	57.28
	17	18.7	19	20	23.8	25.5	27.2	28.9	30.6	32.3	33.957	35.655	37.4	39.1	40.75	42.447	44.144	45.84	47.54	49.24	50.94	52.63	54.332	56.03
3'10"	16.6	18.3	18	20	23.3	24.9	26.6	28.2	29.9	31.6	33.223	34.884	36.5	38.2	39.87	41.529	43.19	44.85	46.51	48.17	49.83	51.5	53.157	54.82
	16.3	17.9	18	19.5	22.8	24.4	26	27.6	29.3	30.9	32.512	34.138	35.8	37.4	39.01	40.641	42.266	43.89	45.52	47.14	48.77	50.39	52.02	53.65
3'11"	15.9	17.5	19.1	20.7	22.3	23.9	25.5	27.1	28.6	30.2	31.824	33.416	35	36.6	38.19	39.78	41.372	42.96	44.55	46.15	47.74	49.33	50.919	52.51
	15.6	17.1	18.7	20.3	21.8	23.4	24.9	26.5	28	29.6	31.158	32.716	34.3	35.8	37.39	38.947	40.505	42.06	43.62	45.18	46.74	48.29	49.853	51.41
4'0"	15.3	16.8	18.3	19.8	21.4	22.9	24.4	25.9	27.5	29	30.512	32.038	33.6	35.1	36.61	38.14	39.666	41.19	42.72	44.24	45.77	47.29	48.819	50.35
4'1"	14.6	16.1	17.6	19	20.5	22	23.4	24.9	26.4	27.8	29.279	30.743	32.2	33.7	35.14	36.599	38.063	39.53	40.99	42.46	43.92	45.38	46.847	48.31
4'2"	14.1	15.5	16.9	18.3	19.7	21.1	22.5	23.9	25.3	26.7	28.12	29.526	30.9	32.3	33.74	35.15	36.556	37.96	39.37	40.77	42.18	43.59	44.992	46.4
4'3"	13.5	14.9	16.2	17.6	18.9	20.3	21.6	23	24.3	25.7	27.028	28.379	29.7	31.1	32.43	33.785	35.136	36.49	37.84	39.19	40.54	41.89	43.245	44.6
4'4"	13	14.3	15.6	16.9	18.2	19.5	20.8	22.1	23.4	24.7	25.999	27.298	28.6	29.9	31.2	32.498	33.798	35.1	36.4	37.7	39	40.3	41.598	42.9
4'5"	12.5	13.8	15	16.3	17.5	18.8	20	21.3	22.5	23.8	25.027	26.278	27.5	28.8	30.03	31.283	32.535	33.79	35.04	36.29	37.54	38.79	40.043	41.29
4'6"	12.1	13.3	14.5	15.7	16.9	18.1	19.3	20.5	21.7	22.9	24.108	25.314	26.5	27.7	28.93	30.135	31.341	32.55	33.75	34.96	36.16	37.37	38.573	39.78
4'7"	11.6	12.8	13.9	15.1	16.3	17.4	18.6	19.8	20.9	22.1	23.24	24.402	25.6	26.7	27.89	29.05	30.212	31.37	32.54	33.7	34.86	36.02	37.183	38.35
4'8"	11.2	12.3	13.5	14.6	15.7	16.8	17.9	19.1	20.2	21.3	22.417	23.538	24.7	25.8	26.9	28.021	29.142	30.26	31.38	32.5	33.63	34.75	35.867	36.99
4'9"	10.8	11.9	13	14.1	15.1	16.2	17.3	18.4	19.5	20.6	21.637	22.719	23.8	24.9	25.96	27.047	28.129	29.21	30.29	31.37	32.46	33.54	34.62	35.7
4'10"	10.4	11.5	12.5	13.6	14.6	15.7	16.7	17.8	18.8	19.9	20.898	21.943	23	24	25.08	26.122	27.167	28.21	29.26	30.3	31.35	32.39	33.436	34.48
4'11"	10.1	11.1	12.1	13.1	14.1	15.1	16.2	17.2	18.2	19.2	20.195	21.205	22.2	23.2	24.23	25.244	26.254	27.26	28.27	29.28	30.29	31.3	32.313	33.32
5'0"	9.76	10.7	11.7	12.7	13.7	14.6	15.6	16.6	17.6	18.6	19.528	20.504	21.5	22.5	23.43	24.41	25.386	26.36	27.34	28.32	29.29	30.27	31.244	32.22
5'1"	9.45	10.4	11.3	12.3	13.2	14.2	15.1	16.1	17	17.9	18.893	19.837	20.8	21.7	22.67	23.616	24.561	25.51	26.45	27.39	28.34	29.28	30.228	31.17
5'2"	9.14	10.1	11	11.9	12.8	13.7	14.6	15.5	16.5	17.4	18.288	19.203	20.1	21	21.95	22.86	23.775	24.69	25.6	26.52	27.43	28.35	29.261	30.18
5'3"	8.86	9.74	10.6	11.5	12.4	13.3	14.2	15.1	15.9	16.8	17.712	18.598	19.5	20.4	21.25	22.14	23.026	23.91	24.8	25.68	26.57	27.45	28.34	29.23
5'4"	8.58	9.44	10.3	11.2	12	12.9	13.7	14.6	15.4	16.3	17.163	18.021	18.9	19.7	20.6	21.454	22.312	23.17	24.03	24.89	25.74	26.6	27.461	28.32
5'5"	8.32	9.15	9.98	10.8	11.6	12.5	13.3	14.1	15	15.8	16.639	17.471	18.3	19.1	19.97	20.799	21.631	22.46	23.29	24.13	24.96	25.79	26.622	27.45
5'6"	8.07	8.88	9.68	10.5	11.3	12.1	12.9	13.7	14.5	15.3	16.139	16.946	17.8	18.6	19.37	20.173	20.98	21.79	22.59	23.4	24.21	25.01	25.822	26.63
5'7"	7.83	8.61	9.4	10.2	11	11.7	12.5	13.3	14.1	14.9	15.661	16.444	17.2	18	18.79	19.576	20.359	21.14	21.92	22.71	23.49	24.27	25.057	25.84
5'8"	7.6	8.36	9.12	9.88	10.6	11.4	12.2	12.9	13.7	14.4	15.203	15.963	16.7	17.5	18.24	19.004	19.764	20.52	21.28	22.04	22.8	23.57	24.325	25.09
5'9"	7.38	8.12	8.86	9.6	10.3	11.1	11.8	12.6	13.3	14	14.766	15.504	16.2	17	17.72	18.457	19.196	19.93	20.67	21.41	22.15	22.89	23.625	24.36
5'10"	7.17	7.89	8.61	9.33	10	10.8	11.5	12.2	12.9	13.6	14.347	15.064	15.8	16.5	17.22	17.934	18.651	19.37	20.09	20.8	21.52	22.24	22.955	23.67
5'11"	6.97	7.67	8.37	9.06	9.76	10.5	11.2	11.9	12.6	13.2	13.946	14.643	15.3	16	16.73	17.432	18.129	18.83	19.52	20.22	20.92	21.62	22.313	23.01
6'0"	6.78	7.46	8.14	8.81	9.49	10.2	10.8	11.5	12.2	12.9	13.561	14.239	14.9	15.6	16.27	16.951	17.629	18.31	18.99	19.66	20.34	21.02	21.698	22.38
6'1"	6.6	7.26	7.92	8.57	9.23	9.89	10.6	11.2	11.9	12.5	13.192	13.852	14.5	15.2	15.83	16.49	17.15	17.81	18.47	19.13	19.79	20.45	21.107	21.77
6'2"	6.42	7.06	7.7	8.34	8.99	9.63	10.3	10.9	11.6	12.2	12.838	13.48	14.1	14.8	15.41	16.047	16.689	17.33	17.97	18.61	19.26	19.9	20.541	21.18
6'3"	6.25	6.87	7.5	8.12	8.75	9.37	10	10.6	11.2	11.9	12.498	13.123	13.7	14.4	15	15.622	16.247	16.87	17.5	18.12	18.75	19.37	19.996	20.62
6'4"	6.09	6.69	7.3	7.91	8.52	9.13	9.74	10.3	11	11.6	12.171	12.78	13.4	14	14.61	15.214	15.822	16.43	17.04	17.65	18.26	18.87	19.474	20.08
6'5"	5.93	6.52	7.11	7.71	8.3	8.89	9.49	10.1	10.7	11.3	11.857	12.45	13	13.6	14.23	14.821	15.414	16.01	16.6	17.19	17.79	18.38	18.971	19.56
6'6"	5.78	6.36	6.93	7.51	8.09	8.67	9.24	9.82	10.4	11	11.555	12.133	12.7	13.3	13.87	14.444	15.021	15.6	16.18	16.75	17.33	17.91	18.488	19.07
6'7"	5.63	6.2	6.76	7.32	7.88	8.45	9.01	9.57	10.1	10.7	11.264	11.827	12.4	13	13.52	14.08	14.643	15.21	15.77	16.33	16.9	17.46	18.023	18.59

Figure 2. Pediatric BMI tables (*continued*)

(continued)

TABLE 3-1 Useful Assessments in Pediatrics *(continued)*

- Hunger and satiety; use of food for reward or as pacifier
- Home environment and family economics (access to food)
- Access to interdisciplinary, family-centered, community-based and culturally relevant services (American Dietetic Association, 2004)

Clinical

- Altered gastrointestinal function: nausea, vomiting, acute diarrhea, constipation, GERD
- Altered nutrition-related biochemical values—such as serum cholesterol. Total serum cholesterol should be <170 mg/dL in children and teens. If >170–199 mg/dL, take a second total serum cholesterol, and average the two together. If >200 mg/dL, a fasting lipid profile is needed
- Birth defects: some can be diagnosed before birth, using prenatal ultrasound, amniocentesis, and chorionic villus sampling (CVS). Ultrasound can help diagnose structural birth defects, such as spina bifida and heart or urinary tract defects. Amniocentesis and CVS are used to diagnose chromosomal abnormalities, such as DS
- Chewing and swallowing difficulties (such as from cleft lip or palate, oral lesions)
- Chronic illnesses (cancer, cardiac disease or heart failure, diabetes, elevated lipids, FTT, hypertension, kidney disease, malabsorption, HIV/AIDS, trauma)
- Congenital or chromosomal abnormalities, inborn metabolic disorders
- Digestive and malabsorptive problems from celiac disease, lactose deficiency, or inflammatory bowel disease; sugar intolerance; foul-smelling, bulky stools indicate fat malabsorption
- Food allergies
- Inadequate intake because of depression, pain or dyspnea, poor appetite >3 days
- Increased nutrient demands, as from protein–energy malnutrition, pressure ulcers

WEIGHT (in pounds)																			
170	175	180	185	190	195	200	205	210	215	220	225	230	235	240	245	250	255	260	265
59.017	60.753	62.489	64.225	65.96	67.7	69.432	71.1679	72.9037	74.64	76.375	78.111	79.84691	81.583	83.31852	85.054321	86.7901	88.5259	90.2617	91.998
57.727	59.425	61.123	62.821	64.52	66.22	67.915	69.6124	71.3102	73.01	74.706	76.404	78.10168	79.8	81.4974	83.195266	84.8931	86.591	88.2889	89.987
56.479	58.14	59.802	61.463	63.12	64.78	66.446	68.1073	69.7684	71.43	73.091	74.752	76.41304	78.074	79.73535	81.396503	83.0577	84.7188	86.38	88.041
55.271	56.897	58.522	60.148	61.77	63.4	65.025	66.6505	68.2761	69.9	71.527	73.153	74.77859	76.404	78.02983	79.655451	81.2811	82.9067	84.5323	86.158
54.101	55.693	57.284	58.875	60.47	62.06	63.649	65.2399	66.8311	68.42	70.014	71.605	73.19602	74.787	76.37845	77.96967	79.5609	81.1521	82.7433	84.335
52.968	54.526	56.084	57.642	59.2	60.76	62.316	63.8737	65.4316	66.99	68.547	70.105	71.66316	73.221	74.77895	76.336842	77.8947	79.4526	81.0105	82.568
51.871	53.396	54.922	56.447	57.97	59.5	61.024	62.5499	64.0755	65.6	67.127	68.652	70.17795	71.704	73.22917	74.754774	76.2804	77.806	79.3316	80.857
49.775	51.239	52.703	54.167	55.63	57.09	58.559	60.0229	61.4869	62.95	64.415	65.879	67.34277	68.807	70.27072	71.734694	73.1987	74.6626	76.1266	77.591
47.804	49.21	50.616	52.022	53.43	54.83	56.24	57.646	59.052	60.46	61.864	63.27	64.676	66.082	67.488	68.894	70.3	71.706	73.112	74.518
45.948	47.299	48.651	50.002	51.35	52.7	54.056	55.4075	56.7589	58.11	59.462	60.813	62.16455	63.516	64.86736	66.218762	67.5702	68.9216	70.273	71.624
44.197	45.497	46.797	48.097	49.4	50.7	51.997	53.297	54.5969	55.9	57.197	58.497	59.7966	61.097	62.39645	63.696376	64.9963	66.2962	67.5962	68.896
42.545	43.797	45.048	46.299	47.55	48.8	50.053	51.3047	52.5561	53.81	55.059	56.31	57.56141	58.813	60.06408	61.315415	62.5667	63.8181	65.0694	66.321
40.984	42.19	43.395	44.6	45.81	47.01	48.217	49.4222	50.6276	51.83	53.038	54.244	55.44925	56.655	57.86008	59.065501	60.2709	61.4763	62.6818	63.887
39.507	40.669	41.831	42.993	44.16	45.32	46.479	47.6413	48.8033	49.97	51.127	52.289	53.45124	54.613	55.77521	56.93719	58.0992	59.2612	60.4231	61.585
38.109	39.23	40.351	41.472	42.59	43.71	44.834	45.955	47.0759	48.2	49.318	50.438	51.55931	52.68	53.80102	54.921875	56.0427	57.1636	58.2844	59.405
36.784	37.865	38.947	40.029	41.11	42.19	43.275	44.3567	45.4386	46.52	47.602	48.684	49.76608	50.848	51.92982	53.011696	54.0936	55.1754	56.2573	57.339
35.526	36.571	37.616	38.661	39.71	40.75	41.795	42.8404	43.8853	44.93	45.975	47.02	48.0648	49.11	50.15458	51.199465	52.2444	53.2892	54.3341	55.379
34.332	35.342	36.352	37.361	38.37	39.38	40.391	41.4005	42.4102	43.42	44.43	45.44	46.4493	47.459	48.46883	49.478598	50.4884	51.4981	52.5079	53.518
33.197	34.174	35.15	36.126	37.1	38.08	39.056	40.0319	41.0083	41.98	42.961	43.938	44.91389	45.89	46.86667	47.843056	48.8194	49.7958	50.7722	51.749
32.118	33.062	34.007	34.952	35.9	36.84	37.786	38.7302	39.6748	40.62	41.564	42.509	43.45337	44.398	45.34265	46.287288	47.2319	48.1766	49.1212	50.066
31.09	32.004	32.919	33.833	34.75	35.66	36.576	37.4909	38.4053	39.32	40.234	41.149	42.06296	42.977	43.89178	44.806191	45.7206	46.635	47.5494	48.464
30.111	30.996	31.882	32.768	33.65	34.54	35.425	36.3102	37.1958	38.08	38.967	39.853	40.73822	41.624	42.50945	43.395062	44.2807	45.1663	46.0519	46.938
29.177	30.035	30.894	31.752	32.61	33.47	34.326	35.1843	36.0425	36.9	37.759	38.617	39.4751	40.333	41.19141	42.049561	42.9077	43.7659	44.624	45.482
28.286	29.118	29.95	30.782	31.61	32.45	33.278	34.1101	34.942	35.77	36.606	37.438	38.26982	39.102	39.93373	40.76568	41.5976	42.4296	43.2615	44.093
27.436	28.243	29.05	29.857	30.66	31.47	32.277	33.0843	33.8912	34.7	35.505	36.312	37.11892	37.926	38.73278	39.539715	40.3466	41.1536	41.9605	42.767
26.623	27.406	28.189	28.972	29.75	30.54	31.321	32.104	32.8871	33.67	34.453	35.236	36.01916	36.802	37.58521	38.368233	39.1513	39.9343	40.7173	41.5
25.846	26.606	27.366	28.126	28.89	29.65	30.407	31.1667	31.9269	32.69	33.447	34.207	34.96756	35.728	36.48789	37.248054	38.0082	38.7684	39.5285	40.289
25.102	25.84	26.578	27.317	28.06	28.79	29.532	30.2699	31.0082	31.75	32.485	33.223	33.96135	34.7	35.43793	36.176223	36.9145	37.6528	38.3911	39.129
24.39	25.107	25.824	26.542	27.26	27.98	28.694	29.4112	30.1286	30.85	31.563	32.281	32.99796	33.715	34.43265	35.15	35.8673	36.5847	37.302	38.019
23.708	24.405	25.102	25.799	26.5	27.19	27.891	28.5886	29.2859	29.98	30.68	31.378	32.07499	32.772	33.46955	34.166832	34.8641	35.5614	36.2587	36.956
23.054	23.732	24.41	25.088	25.77	26.44	27.122	27.8	28.478	29.16	29.834	30.512	31.1902	31.868	32.5463	33.224344	33.9024	34.5804	35.2585	35.937
22.426	23.086	23.746	24.405	25.06	25.72	26.384	27.0435	27.7031	28.36	29.022	29.682	30.34153	31.001	31.66072	32.320323	32.9799	33.6395	34.2991	34.959
21.824	22.466	23.108	23.75	24.39	25.03	25.676	26.3176	26.9595	27.6	28.243	28.885	29.52703	30.169	30.81081	31.452703	32.0946	32.7365	33.3784	34.02
21.246	21.871	22.496	23.121	23.75	24.37	24.996	25.6204	26.2453	26.87	27.495	28.12	28.74489	29.37	29.99467	30.619556	31.2444	31.8693	32.4942	33.119
20.691	21.299	21.908	22.516	23.13	23.73	24.342	24.9507	25.5592	26.17	26.776	27.385	27.99342	28.602	29.21053	29.819079	30.4276	31.0362	31.6447	32.253
20.157	20.75	21.343	21.935	22.53	23.12	23.714	24.3068	24.8996	25.49	26.085	26.678	27.27104	27.864	28.45674	29.049587	29.6424	30.2353	30.8281	31.421
19.643	20.221	20.799	21.377	21.95	22.53	23.11	23.6875	24.2653	24.84	25.421	25.999	26.57627	27.154	27.73176	28.3095	28.8872	29.465	30.0427	30.62
19.149	19.712	20.276	20.839	21.4	21.97	22.528	23.0917	23.6549	24.22	24.781	25.344	25.90771	26.471	27.03413	27.59734	28.1606	28.7238	29.287	29.85

(continued)

TABLE 3-1 Useful Assessments in Pediatrics *(continued)*

- Marked weight loss (malabsorption, IBD, hyperthyroidism, or malignancy)
- Medications with nutritional side effects:

  Antibiotics (energy, protein, minerals; GI problems)

  Anticonvulsants (vitamins C, K, D, and B-complex, and calcium)

  Corticosteroids (calcium, phosphorus, glucose levels; weight gain or stunting)

  Diuretics (potassium, magnesium, calcium, energy; GI problems)

  Stimulants such as ritalin (energy and protein intake, growth, appetite)

  Sulfonamides (vitamin C, protein, folate, and iron)

  Tranquilizers (energy intake; weight gain)
- Inability to consume oral diet:

  Pediatric tube feeding: prematurity, developmental delays, orofacial defects, CP, anorexia nervosa, cystic fibrosis, metabolic disorders, renal failure, HIV infection, or inflammatory bowel disorders

  Pediatric TPN: biliary atresia, Hirschsprung's disease with enterocolitis, Crohn's disease, ulcerative colitis, congenital short-bowel, GI ischemia or fistulas, severe burns or trauma, and bowel transplantation. It may be possible to wean from TPN to tube or oral feeding in some conditions; for others, PN may be permanent. Children cannot tolerate fasting as long as adults
- Increased nutrient needs for trauma, surgery, recent hospitalizations, acute illnesses, chemotherapy or radiation

Developmental Disabilities

- Altered nutritional status, feeding skills, feeding behaviors including positioning
- Use specific screening tools for each physical, motor, sensory, or developmental delays. Use arm span where height is difficult to measure
- In children whose weight is hard to maintain, catch-up growth is important with a focus on protein and kilocals
- Individualize care: design the desired outcomes, determine necessary resources, and seek regular feedback on progress or obstacles. Personal control, independence, and choice must be considered

Eating and Feeding Skills

- Avoidance of easily aspirated foods
- Biting, chewing or swallowing difficulty requiring texture modifications
- Coordination for safe and proper chewing, sucking, swallowing
- Feeding: length of time, feeding method, skill level, persons involved
- Food allergies, multiple or severe
- Food Intake: ability to eat and retain food
- Food preferences, dislikes, and intolerances
- Special formula or supplements, tube feeding or PN

Genetic and Metabolic Disorders

- Growth failure, skin rashes, developmental delays, vomiting or diarrhea and other concerns affect nutrition and health status in:

  Amino acid metabolism: phenylketonuria (PKU); maple syrup urine disease (MSUD); glutaric acidemia type 1(GA1); argininosuccinic academia (ASA); tyrosinemia (TYR); propionic academia (PA); isovaleric academia (IVA); citrullinemia type 1 (CIT)

  Carbohydrate metabolism: galactosemia; glycogen storage disease (GSD); galactose-phosphate uridyltransferase (GALT)

  Fatty acid metabolism: medium chain acyl dehydrogenase deficiency (MCAD); carnitine uptake deficiency (CUD); very long-chain acyl-CoA dehydrogenase (VLCAD); abetalipoproteinemia (ABL)
- Presently, screening for 29 disorders is recommended for newborns in the United States; many states are doubling or tripling the number of tests offered (Isaacs and Zand, 2007). Newborn screening consists of testing; follow-up of abnormal screening; diagnostic testing; disease management; continuous evaluation and improvement of the system. Clues that suggest a genetic condition or an inherited susceptibility to a common disease include the following:

  Two or more seemingly unrelated medical conditions (e.g., hearing loss and renal disease, diabetes and muscle disease)

  A medical condition and dysmorphic features

  Developmental delay with dysmorphic features and/or physical birth anomalies

  Developmental delay associated with other medical conditions

  Progressive mental retardation, loss of developmental milestones

  Progressive behavioral problems

  Unexplained hypotonia

  A movement disorder

  Unexplained seizures

  Unexplained ataxia

  Two or more major birth anomalies

  Three or more minor birth anomalies

  One major birth defect with two minor anomalies

  A cleft palate, or cleft lip with or without cleft palate

  Unusual birthmarks (particularly if associated with seizures, learning disabilities, or dysmorphic features)

  Hair anomalies (hirsute, brittle, coarse, kinky, sparse or absent)

  Congenital or juvenile deafness

  Congenital or juvenile blindness

  Cataracts at a young age

  Primary amenorrhea

  Ambiguous genitalia

(continued)

TABLE 3-1 Useful Assessments in Pediatrics *(continued)*

	A Fetus With
Proportionate short stature with dysmorphic features and/or delayed or arrested puberty	A major structural anomaly
Disproportionate short stature	Significant growth retardation
Premature ovarian failure	Multiple minor anomalies
Proportionate short stature and primary amenorrhea	(Gilchrist, 2002; March of Dimes, 2009)
Males with hypogonadism and/or significant gynecomastia	
Congenital absence of the vas deferens	
Oligozoospermia/azoospermia	

TABLE 1A: Examples of Single Gene Disorders in Adults[a]

Neurology

Muscular dystrophy
Spinocerebellar ataxia
Hereditary neuropathy
Dystonia
Early onset Alzheimer's disease
Familial multiple sclerosis
Familial amyotrophic lateral sclerosis
Neurofibromatosis

Nephrology

Autosomal dominant polycystic kidney disease
Hereditary nephritis
Disorders of renal physiology

Hematology

Hemoglobinopathies
Hereditary disorders of hemostasis
Hereditary hypercoagulability

Pulmonary disorders

Adult-onset cystic fibrosis
Alpha-1-antitrypsin deficiency

Cardiac disorders

Conduction abnormalities
Cardiomyopathy

Infectious disease

Immune deficiencies

Metabolic disorders

Hemochromatosis
Lipid disorders
Homocysteine

Gastroenterology

Osler-Weber-Rendu disease
Polyposis

Oncology

BRCA1/2
Familial adenomatous polyposis and hereditary nonpolyposis colon cancer
Familial prostate cancer
Multiple endocrine neoplasia
Hippel-Lindau disease
Li-Fraumeni syndrome

Musculoskeletal disorders

Inherited disorders of connective tissue—
Marfan's, Ehlers-Danlos, osteogenesis imperfecta

Dermatology

Icthyosis
Bullous disorders

Notes: Inborn metabolic disorders are usually due to defects of single genes that code for enzymes, intended to convert substrates into products.
[a]This is a far from complete list.

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Lucas B, ed. *Children with special care needs: nutrition care handbook*. Chicago: The American Dietetic Association, 2004.
March of Dimes. Accessed May 9, 2009, at http://www.marchofdimes.com/.

(continued)

TABLE 3-1 Useful Assessments in Pediatrics *(continued)*

Estimating Daily Energy Requirements (EER) and Total Energy Expenditure (TEE) for Infants and Children (Derived from Institute of Medicine, 2002; Lucas, 2004)

Age (months)	Equation
0–3	(89 × Wt − 100) + 175
4–6	(89 × Wt − 100) + 56
7–12	(89 × Wt − 100) + 22
13–35	(89 × Wt − 100) + 20

Boys: Age (years)	Equation
3–8	EER = 88.5 − 61.9 × age (y) + PA × (26.7 × Wt + 903 × Ht) + 20
9–19	EER = 88.5 − 61.9 × age (y) + PA × (26.7 × Wt + 903 × Ht) + 25
3–19, overweight	TEE = 114 −50.9 × age (y) + PA × (19.5 × Wt + 116.4 × Ht)

Girls: Age (years)	Equation
3–8	EER = 135.3 −30.8 × age (y) + PA × (10.0 × Wt + 934 × Ht) + 20
9–19	EER = 135.3 − 30.8 × age (y) + PA × (10.0 × Wt + 934 × Ht) + 25
3–19, overweight	TEE = 389 − 41.2 × age (y) + PA × (15.0 × Wt + 701 × Ht)

Physical Activity (PA) Coefficients for Children Aged 3–19 Years

	Coefficient for Boys Aged 3–19 years	Boys	Coefficient for Girls Aged 3–19 years	Girls
Activity Level	Normal Wt	Overweight	Normal Wt	Overweight
Sedentary	1.0	1.0	1.0	1.00
Low active	1.13	1.12	1.16	1.18
Active	1.26	1.24	1.31	1.35
Very Active	1.42	1.45	1.56	1.60

Acceptable Macronutrient Ranges

	Range (% of energy)		
Age	CHO	Fat	Protein
Full-term infant	35–65	30–55	7–16
1–3 years	45–65	30–40	5–20
4–18 years	45–65	25–35	10–30

TABLE 3-2 Nutritional Risks Associated with Selected Pediatric Disorders

	Low weight	Over-weight	Short Stature	Low Energy Needs	High Energy Needs	Feeding Problems	Constipation	Chronic Meds
Autism spectrum disorders	X				X	X		X
Bronchopulmonary dysplasia	X	X			X	X		X
Cerebral palsy	X	X	X	X	X	X	X	X
Cystic fibrosis	X		X		X	X		
Down syndrome		X		X		X		
Fetal alcohol syndrome	X		X					
Heart disease, congenital	X				X	X		X
HIV infection, AIDS	X				X			X
Phenylketonuria	X					X		
Prader-Willi syndrome		X	X	X				
Prematurity or low birth weight	X		X		X	X		
Seizure disorder								X
Spina bifida; neural tube defects	X	X	X	X			X	X

Adapted from: Baer M, Harris A. Pediatric nutrition assessment: identifying children at risk. *J Am Diet Assoc.* 97:107A, 1997.

For More Information About Birth Defects and Genetic Disorders

- Centers for Disease Control and Prevention (CDC) Birth Defects Research
  http://www.cdc.gov/ncbddd/bd/research.htm
- Coalition of State Genetics Coordinators
  http://www.stategeneticscoordinators.org
- Family Voices
  http://www.familyvoices.org/
- Genetic Alliance Disease InfoSearch
  http://www.geneticalliance.org
- Human Genome Project
  http://www.ornl.gov/sci/techresources/Human_Genome/home.shtml
- March of Dimes
  http://www.modimes.org/
- National Center for Education in Maternal and Child Health
  http://www.ncemch.org/
- National Dissemination Center for Children with Disabilities
  http://www.nichcy.org/
- National Folic Acid Campaign
  http://www.cdc.gov/ncbddd/folicacid/council.htm
- National Coalition for Health Professional Education in Genetics
  http://www.nchpeg.org
- National Institutes of Health Office of Rare Diseases
  http://rarediseases.info.nih.gov
- National Newborn Screening and Genetics Resource Center
  http://genes-r-us.uthscsa.edu
- National Urea Cycles Disorders Foundation
  http://www.nucdf.org/
- Online Mendelian Inheritance in Man (OMIM)
  http://www.ncbi.nlm.nih.gov/entrez/dispomim
- Organic Acidemia Association
  http://www.oaanews.org

For More Information About Feeding Problems and Assistance

- American Occupational Therapy Association, Inc.
  http://www.aota.org/
- The Oley Foundation for Home Enteral/Parenteral Therapy
  http://www.oley.org/

For More Information About Specialty Foods and Formulas

- Abbott Pediatric Products
  http:// www.ross.com/productHandbook/default.asp
- Applied Nutrition
  http://www.medicalfood.com
- Cambrooke Foods
  http://www.cambrookefoods.com/
- Dietary Specialties
  http://www.dietspec.com/
- Ener-G Foods
  http://www.ener-g.com/
- Glutino
  http://www.glutino.com/
- Kingsmill Foods
  http://www.kingsmillfoods.com/
- Loprofin (SHS)
  http://www.shsna.com/pages/loprofin.htm
- MedDiet
  http://www.med-diet.com/
- Mead Johnson
  http://www.meadjohnson.com/professional/prodinfo.html
- Milupa North American
  http://www.milupana.com
- Novartis
  http://www.novartis.com
- Scientific Hospital Supply
  http://www.shsna.com

For More Information About Rare Disorders and Health Laws

- Alliance of Genetic Support Groups
  http://geneticalliance.org/
- Genetics Home Reference
  http://ghr.nlm.nih.gov/
- Metabolic Disorders
  http://themedicalbiochemistrypage.org/inborn.html
- National Health Law Program
  http://www.healthlaw.org

- National Organization for Rare Disorders
  http://www.rarediseases.org/
- Newborn Screening
  http://www.savebabies.org/
- Office of Rare Diseases—NIH
  http://rarediseases.info.nih.gov/

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ABETALIPOPROTEINEMIA

NUTRITIONAL ACUITY RANKING: LEVEL 2–3

DEFINITIONS AND BACKGROUND

Abetalipoproteinemia (ABL) is a rare, inherited disease characterized by the inability to make low-density lipoproteins (LDL), very low–density lipoproteins (VLDL), and chylomicrons or to fully absorb dietary fats through the gut. Other names for this condition are Bassen-Kornzweig syndrome, acanthocytosis, or apolipoprotein B deficiency.

Acanthocytosis refers to the altered shape of the normal erythrocyte into one with a few irregularly shaped external projections that are thorny in appearance (Rampoldi et al., 2002; Wong, 2004). Infants present with FTT and fatty and pale stools that are frothy and foul smelling. They may also have a protruding abdomen, developmental delays, slurred speech, and problems with balance and muscle coordination after age 10. Mental deterioration and scoliosis also occur.

Prognosis is related to the progression of neurological and visual problems. Severe forms lead to irreversible neurological disease before age 30 and often to blindness. Progressive ataxic neuropathy and retinopathy are related to oxidative damage resulting from deficiencies of vitamins E and A (Granot and Kohen, 2004). Therefore, vigorous nutritional supplementation is essential (Chardon et al, 2008). Other treatments such as stem-cell therapy and gene product replacement are under evaluation.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: Microsomal triglyceride transfer protein (MTTP or MTP) deficiency

Clinical/History

- Height
- Weight
- Growth chart
- Diet/intake history
- Scoliosis
- Retinal degeneration, retinitis pigmentosa
- Low vision or blindness
- Developmental delay?
- Neurological changes

Lab Work

- CBC with abnormal, thorny-shaped cells
- Serum apolipoprotein B levels (low or absent)
- Albumin (Alb)
- Total Cholesterol (TC)
- LDL Cholesterol (may be low)
- VLDL (may be low)
- Fatty acid profile
- Triglycerides (Trig)
- Fecal fat study: high levels with steatorrhea
- Serum retinol and vitamin E
- Serum vitamin K; Pro-time (PT) or international normalized ratio (INR)
- Alkaline phosphorus (for vitamin D status)
- Ca^{++}, Mg^{++}
- EMG or nerve conduction velocity testing (demyelination of peripheral nerves)
- Hemoglobin and hematocrit (H & H)

INTERVENTION

OBJECTIVES

- Decrease rapid progression of disorder by giving large doses of fat-soluble vitamin supplements. This may help prevent deterioration of vision and degeneration of the retina (retinitis pigmentosa).

SAMPLE NUTRITION CARE PROCESS STEPS

Steatorrhea

Assessment Data: Food records indicating poor intake; changes in weight; cholesterol levels.

Nutrition Diagnoses (PES): Inadequate fat soluble vitamin intake (especially E) related to fat malaborption in ABL as evidenced by frothy stools four to five times daily, low serum cholesterol level, low serum levels of vitamin E, and abdominal distention.

Intervention: Education of parents about the need for fat-soluble vitamin supplementation and for linoleic acid supplementation for condition.

Monitoring and Evaluation: Improved lab reports for vitamin E and cholesterol; weight improvement; fewer frothy stools and less abdominal distention.

- Avoid use of long-chain triglycerides; use medium-chain triglycerides (MCT) instead.
- Prevent nutrient deficiency symptoms and conditions, such as FTT; impaired balance; difficulty walking, and other complications. Provide linoleic acid supplementation for essential fatty acids (EFAs).

FOOD AND NUTRITION

- The diet should contain no more than 5 oz of lean meat, fish, or poultry per day.
- Use skim milk instead of whole milk; reduce fats from other types of dairy products.
- Use MCT oil in food preparation and with gravies, sauces, and other cooked foods. Avoid excesses to prevent liver problems.
- The diet should be supplemented with fat-soluble vitamins A, D, E, and K plus linoleic acid to prevent deficiency. Water-miscible forms will be needed.

Common Drugs Used and Potential Side Effects

- Large doses of supplemental vitamins A and E may be needed and prescribed by the physician (Chowers et al, 2001).

Herbs, Botanicals, and Supplements

- Herbs and botanicals are not recommended for this condition because there are no clinical trials proving efficacy.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Advise that MCT products should be consumed slowly to avoid side effects such as diarrhea.
- Discuss the need for intake of linoleic acid.
- A multivitamin–mineral supplement will be recommended. Identify food sources of the fat-soluble vitamins and discuss how the disorder prevents use of these vitamins accordingly.
- For persons with low vision, teaching with food models or large pictures may be more beneficial than use of text. Audiotapes may also be developed.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants to decrease potential risk of botulism.

For More Information

- Prevent Blindness
  http://www.preventblindness.org/
- Prevent Blindness Foundation
  http://www.pbf.org.au/

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ATTENTION-DEFICIT DISORDERS

NUTRITIONAL ACUITY RANKING: LEVEL 1

DEFINITIONS AND BACKGROUND

Attention-deficit disorder (ADD) and attention-deficit hyperactivity disorder (ADHD) were formerly called "hyperkinesis." Adults can have symptoms as well and may find relief from certain medications and therapies. ADD is a neurobiological condition characterized by developmentally inappropriate level of attention, concentration, activity, distractibility, and impulsivity; it is more common in males.

ADHD is the most commonly diagnosed behavioral disorder of childhood, affecting an estimated 7% of school-aged children (Schonwald, 2005). Three types are designated: predominantly hyperactive-impulsive, predominantly inattentive, and combined hyperactive-impulsive and inattentive; most children have the combined form. The Preschool ADHD Treatment Study (PATS) provides guidance for diagnostic considerations and intervention strategies for children between the ages of 3 and 5. Most ADHD is identified by age 6. Children with ADHD with a particular version of a certain gene have thinner brain tissue in the areas of the brain associated with attention, which is usually outgrown.

Glucose is the brain's energy source. In ADD, brain regions that inhibit impulses and control attention actually use less glucose; this decreased activity in the brain leads to inattention. PET scan comparisons between the brain of a normal child and the brain of an ADHD child show a significant difference. Children should be assessed for brain injury or seizure disorders, which may cause inattention and sleep disturbances (Schubert, 2005).

Exposure to lead is neurotoxic. Lead is found in trace amounts in everything from children's costume jewelry to imported candies to soil and drinking water (Nigg et al, 2010). It attaches to sites in the brain's striatum and frontal cortex, causing genes there to turn on or remain inactive, disrupting brain activity, decreasing cognitive control and contributing to hyperactivity and lack of vigilance (Nigg et al, 2010).

Children with ADHD may have low levels of other nutrients. Iron deficiency causes abnormal dopaminergic neurotransmission and may also contribute to ADD (Konofal et al, 2004). Zinc and EFAs (EPA and DHA) may be needed along with pharmacotherapy (DiGirolamo and Ramirez-Zea, 2009).

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: ADHD often runs in families. Several genes make people more susceptible: SLC6A1, SLC9A9, HES1, ADRB2, HTR1E, DDC, ADRA1 A, DBH, DRD2, BDNF, TPH2, HTR2 A, SLC6A2, PER1, CHRNA4, SNAP25, and COMT but SLC9A9 has the strongest relationship (Lasky-Su et al, 2008). While genes account for 70% of hyperactivity and inattention in children, 30% is environmental (Nigg et al, 2010).

Clinical/History

- Height
- Weight
- Growth chart
- Diet/intake history
- Mental retardation, other developmental delay?
- Head injury?
- Seizures or hx of epilepsy?
- EEG
- Sleep disturbances
- Middle ear infections with hearing loss
- Learning disabilities
- Anxiety or depression
- Nocturnal enuresis

Lab Work

- Glucose (Gluc)
- H & H, serum Fe
- Ferritin levels
- Serum lead (elevated?)
- Serum zinc (low?)
- Alb
- Chol
- Liver function tests (LFTs)

SAMPLE NUTRITION CARE PROCESS STEPS

Poor Food Choices

Assessment Data: Food records indicating poor intake; weight loss and food jags. Low H & H levels.

Nutrition Diagnoses (PES):

NI 1.4. Inadequate energy intake related to insufficient energy intake from a diet compared to estimated needs as evidenced by weight loss and dietary intake records.

NI 55.1. Inadequate iron intake related to a disordered eating pattern as evidenced by a low Hgb of 10 g/L and low iron food choices.

Interventions: Food-nutrient delivery to include iron-rich foods. Educate parents about introducing new iron-rich food items, reducing distractions at mealtime, scheduling of activities to give structure, and the use of small snacks throughout the day to improve overall intake of nutrients and energy. Counseling about the importance of nutrient density in children's diets. Coordinate care with medical team to test and identify other risks, such as high serum lead or low zinc levels.

Monitoring and Evaluation: Improved nutrient density in food intake records. Better attention in school, fewer complaints about hyperactivity.

INTERVENTION

OBJECTIVES

- Prevent nutrient deficiencies if diet is inadequate or with extensive documented food allergies.
- Address poor intake and appetite, where present. Offer foods that are liked along with one to two new tastes to encourage expanding preferences.
- Correct zinc deficiency and iron deficiency anemia, where indicated.
- Rule out lead poisoning.
- Provide sufficient intake of omega-3 fatty acids and other micronutrients.

FOOD AND NUTRITION

- The diet should be balanced and sufficient in energy and protein for age and sex. The MyPyramid food guidance system should be the basis for planning; see Section 2.
- Omit any food allergens that have been medically diagnosed and verified (McCann et al, 2007).
- Elimination of sugar is not required; moderation is reasonable. Small, frequent healthy snacks may be beneficial.
- EFA supplementation may be useful (Murphy et al, 2005; Schnoll et al, 2003). Include DHA and EPA in the diet from tuna, mackerel, herring, sardines, and salmon (Ramakrishnan et al, 2009). The use of fortified foods requires some long-term studies (Riediger et al, 2009).
- Offer plenty of whole grains, low-fat dairy, fruits, and vegetables in greater proportion than sugary foods to provide more micronutrients and phytochemicals.
- Discuss good food sources of zinc and iron, especially for children with a limited diet or food jags. They may not eat meats, poultry, fish, eggs, and fortified cereals in sufficient amounts.

Common Drugs Used and Potential Side Effects

- Stimulants such as Ritalin (methylphenidate) have been used for many years. They may cause weight loss, appetite change, sleep problems or irritability. Newer, long-acting medications will alleviate some of the burden of ADD (Connor and Steingard, 2004). See Table 3-3.
- Nonstimulant Strattera (atomoxetine) is used to increase attention and the ability to focus; long-term use may cause liver damage or suicidal thoughts, so monitor carefully.
- Antiepileptic drugs phenobarbital, gabapentin, and topiramate may alter attention whereas lamotrigine and carbamazepine may have beneficial effects on attention span.
- Mood stabilizers, beta-blockers, or serotonin reuptake inhibitors be used in managing aggression or self-injury. There are many side effects, such as weight gain or loss, elevation of glucose with possible diabetes, gastrointestinal distress; monitor closely.

Herbs, Botanicals, and Supplements

- Many parents try complementary medicine (CAM) as an alternative to stimulant medicines as the marketing for herbal remedies, elimination diets, and food supplements for ADHD has increased. There are key questions regarding safety and efficacy of these treatments in children (Sawni, 2008). Herbs and botanicals are not recommended for this condition because there are no clinical trials proving efficacy.

TABLE 3-3 ADHD Medications Approved by U.S. Food and Drug Administration (FDA)[a]

Trade Name	Generic Name	Approved Age
Adderall	Amphetamine	3 and older
Adderall XR	Amphetamine (extended release)	6 and older
Concerta	Methylphenidate (long acting)	6 and older
Daytrana	Methylphenidate patch	6 and older
Desoxyn	Methamphetamine hydrochloride	6 and older
Dexedrine	Dextroamphetamine	3 and older
Dextrostat	Dextroamphetamine	3 and older
Focalin	Dexmethylphenidate	6 and older
Focalin XR	Dexmethylphenidate (extended release)	6 and older
Metadate ER	Methylphenidate (extended release)	6 and older
Metadate CD	Methylphenidate (extended release)	6 and older
Methylin	Methylphenidate (oral solution and chewable tablets)	6 and older
Ritalin	Methylphenidate	6 and older
Ritalin SR	Methylphenidate (extended release)	6 and older
Ritalin LA	Methylphenidate (long acting)	6 and older
Strattera	Atomoxetine	6 and older
Vyvanse	Lisdexamfetamine dimesylate	6 and older

[a]Not all ADHD medications are approved for use in adults.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Different causes of the symptoms of ADD may include allergies, food intolerances, anxiety, depression, family problems, poor discipline, or acute illnesses. A healthy, balanced diet is important (Marcason, 2005).
- Since glucose is the brain's source of energy, a sufficient intake of carbohydrate is needed. Assure that healthy choices are made from dairy, fruit and vegetable, and bread and cereal items. Reduce intake of sugary sweets, beverages, and snacks as a commonsense approach.
- Identify and remove sources of lead in the environment, especially if serum levels are found to be high.
- EFAs are important; include adequate amounts of fats in the daily diet (Ross et al, 2007).
- Zinc intake also plays an important role (Arnold and DiSilvestro, 2005; DiGirolamo and Ramirez-Zea, 2009).
- Identify and treat nocturnal enuresis, which may be present.
- Children need clear, consistent rules and praise when rules are followed.
- Individual psychotherapy may be quite beneficial. Encourage full participation.
- Children need help to stay organized and follow directions. Use of a schedule is important; organize everyday items; use notebook organizers.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing meals and snacks. Use clean utensils and containers.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants to decrease potential risk of botulism.

For More Information

- Attention Deficit Disorder Association
  http://www.add.org/
- National Institute for Mental Health
  http://www.nimh.nih.gov/publicat/adhd.cfm

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AUTISM SPECTRUM DISORDER

NUTRITIONAL ACUITY RANKING: LEVEL 1–2

DEFINITIONS AND BACKGROUND

Autism spectrum disorder (ASD) begins in childhood as developmental disabilities caused by abnormalities in the brain. Generalized enlargement of gray and white matter cerebral volumes is present at 2 years of age. Increased rate of brain growth occurs in the latter part of the first year of life (Hazlett et al, 2005). The ASDs are part of a broader category of pervasive developmental disorders (PDDs), affecting over 500,000 individuals in the United States.

Autism is a neurodevelopmental condition affecting 1 in 160 children in the United States (West et al, 2009). Children with ASDs have unusual ways of learning, paying attention, or reacting to different sensations. They like to repeat certain behaviors and do not want change in their daily activities. They are hypersensitive to sensory stimuli (tastes, smells, sounds, sights) and withdraw from what is perceived as distressing or painful. People with ASDs have problems with social and communication skills. Up to 40% of persons with an ASD do not speak. Rather than conversing in a dialogue fashion, they may repeat back what has been said (echolalia).

Autism is considered to be genetic. Environmental factors with exposure to toxins, infections such as measles, mumps, or rubella, and diet play a role (White, 2003). Mental retardation and seizures of mild-to-moderate intensity can be present, especially in **fragile X syndrome** (found mostly in males). **Rett syndrome** occurs primarily in girls and is evident by repetitive hand movements. In **Asperger's disorder**, speech occurs at the usual time, intelligence is normal or above average, but social skills are stunted and interests are limited or obsessive.

Pica with anemia is common, such as eating paper, string, or dirt. Ritualistic eating behaviors, food limitations, messy eating habits and food jags are common; variety in texture or colors may not be accepted. Foods that could cause choking should be avoided. A quiet environment for eating is best tolerated.

High use of oral antibiotics or higher mercury exposure can aggravate oxidative stress and decrease detoxification capacity, leading to decreased plasma methionine, glutathione, cysteine, SAM, and sulfate (Adams et al, 2007; James et al, 2004). A methylation problem is likely (Deth et al, 2007). Allergies or sensitivities are also relevant. Autistic children often have IgA deficiency, decreased natural killer (NK) cell numbers, antibodies against serotonin receptors, and a tumor necrosis factor (TNF) response to casein, gluten, and soy (Schneider, 2007; Vojdani et al, 2008). Mega doses of nutrients are not helpful.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: At least 10 genes and enzymes have been implicated (Campbell, 2006; Muhle et al, 2004; Schneider, 2007): Adenosine Deaminase; Cystathionine-B-Synthase; Catecholamine-O-Methyltransferase (COMT 472G > A); FOXP2, RAY1/ST7, IMMP2 L; GABA(A) receptor; Glutathione-S-transferase (GST M1); MET Receptor Tyrosine Kinase (MET); Methionine Synthase (MS); Methionine Synthase Reductase (MSR); MTHFR C677 T and A1298 C; Paraoxonase (PON1); Reduced folate carrier (RFC 80G > A); Reelin and RELN genes at 7q22-q33; Serotonin transporter gene (5-HTT); Transcobalamin II (TCN2 776G > C); UBE3 A genes.

Clinical/History

- Height
- Weight
- Head circumference
- Growth chart
- Diet/intake history
- Childhood Asperger Syndrome Test (CAST), a parental questionnaire
- MRI or CT scan of the brain
- Pica
- Endoscopy
- Sleep patterns

Lab Work

- CBC with differential
- Gluc
- H & H, Serum Fe, Ferritin
- Celiac screening panel
- Lactose breath test
- Thyroid studies
- LFTs Lipid profile
- RAST or allergy testing
- Alb
- Chol, Trig
- Serum lead (with pica)
- C-reactive protein (CRP)

SAMPLE NUTRITION CARE PROCESS STEPS

Intake of Unsafe Foods

Assessment Data: Food records indicating intake of gluten; loss of weight; chronic rashes, infections and diarrhea; small bowel biopsy indicating celiac disease.

Nutrition Diagnoses (PES):

NI 5.3. Inadequate protein-energy intake related to highly restricted eating behaviors and pickiness as evidenced by parental report of insufficient intake, below 50% for age for weight, growth failure, and frequent lack of interest in food.

NB 3.1. Intake of unsafe foods related to sensitivity to gluten in autistic child as evidenced by biopsy positive for celiac disease, rashes, chronic diarrhea.

Intervention: Educate parents about gluten-free diet, food labeling, simple meal preparation. Counseling about use of food diaries and routines; how to include frequent nutrient-dense snacks of desired food items.

Monitoring and Evaluation: Improved weight records; fewer loose stools, infections and rashes.

INTERVENTION

OBJECTIVES

- Prevent or lessen complications of the disorder, such as feeding problems. Offer consistency in food textures and tastes to prevent sensory overload.
- Evaluate carefully and analyze which nutrients should be replaced in the diet. If the diet has been severely limited, nutritional status may be at risk.
- Correct constipation if fiber intake is low and if symptoms are present.
- Work with other therapies, such as speech therapy or occupational therapy, to determine how to best offer foods of greater texture and variety that can be consumed by the child or offered by the caregiver.
- Monitor food jags, pica, history of choking on foods, and intolerances for varied textures, and adapt meals and menu items accordingly. Goal is to eat foods from all parts of the food pyramid regardless of texture.
- Autism seems to be related to altered immunity (Ashwood et al, 2004). Frequent infections, gastro-intestinal (GI) concerns (including chronic constipation or diarrhea) thyroid problems, and allergies are common and require nutritional management. A high prevalence of non-IgE-mediated food allergies exist in young children with ASDs, with GI symptoms (Jyonouchi, 2008).
- Monitor use of the popular gluten casein–free diet, as more clinical trials are needed to confirm efficacy (Millward et al, 2008).

FOOD AND NUTRITION

- Offer foods of texture and variety that are desired by the child. Follow a usual pattern and enhance with nutrient-dense additives in food preparation that will not alter flavor and texture.
- If a multivitamin–mineral supplement is needed, use one that has acceptable taste to the individual.
- Gluten-casein-soy elimination diets have some success, but can also lead to nutrient deficiencies. Use under supervision of a dietitian.
- Offer extra energy if weight is low, a common finding. Assess needs according to activity levels, weight and nutritional status, and medications that are prescribed.
- Since the brain requires omega-3 fatty acids for membrane integrity and to reduce inflammation, it is beneficial to include them in the diet or to use them in supplemental form.
- Some autistic children have disaccharide deficiencies; alter diet accordingly.

Common Drugs Used and Potential Side Effects

- Only risperidone has Food and Drug Administration approval for the pharmacologic management of autism in children (West et al, 2009).
- Atypical antipsychotic agents, selective-serotonin reuptake inhibitors, stimulants, and *N*-methyl-D-aspirate receptor antagonists may be used off-label to decrease core behaviors and associated symptoms (West et al, 2009). If clonidine, clomipramine, haloperidol, naltrexone, or desipramine is prescribed for behavioral or learning problems, monitor for GI side effects, nausea, and diarrhea. Olanzapine (Zyprexa) may cause weight gain, dry mouth, and constipation. Fluoxetine (Prozac) may cause anorexia or weight loss, GI distress, and diarrhea.
- Medications to control seizures may be needed (Tuchman, 2004).
- D-cycloserine treatment may improve social withdrawal without side effects (Posey et al, 2004).
- Folinic acid, betaine, and methylcobalamin may be needed normalize metabolic imbalances or to treat cerebral folate deficiency (James et al, 2004; Moretti et al, 2005).

Herbs, Botanicals, and Supplements

- Over half of children with autism are using CAM (Golnick and Ireland, 2009). Melatonin, sulfation glutathione, amino acids, chelation, probiotics, thyroid supplements, antifungals are common. In one report (Golnick and Ireland, 2009) physicians encouraged multivitamins (49%), EFAs (25%), melatonin (25%), and probiotics (19%) and discouraged withholding immunizations (76%), chelation (61%), anti-infectives (57%), delaying immunizations (55%), and secretin (43%). Evidence-based studies are needed.
- Herbs and botanicals are not recommended for these conditions because clinical trials have not proven efficacy.
- "Improved diet" techniques may include organic foods, which adds expense to the cost of meals.
- Vitamin–mineral mega doses such as vitamin B_6 and magnesium may have been used; discuss the implications and potential risks.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Evaluate for behaviors such as pica; discuss how this may lead to anemia.
- Assist with tips on how to handle picky eating, rigid food behaviors, and nutrient insufficiency. Discuss various ways to include nutrient-dense foods in the diet.
- Discuss the importance of maintaining a quiet environment with few interruptions or distractions.
- Keep language simple and concrete; do not offer abstract text. Pictures and simple words are more effective when working with an older child or teen.
- Artificial colors and preservatives may have a detrimental effect on behavior, especially red and yellow food dyes. Limit their use in the diet.
- Avoid allergens, where documented.
- Support various therapies, such as speech therapy, occupational therapy, use assistive technology and biomedical applications that are evidence-based.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers. Sanitize work surfaces before food preparation.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer. To avoid wasting foods, serve small portions more frequently.

For More Information

- Asperger's Syndrome
  http://www.aspergers.com/
- Asperger Syndrome Coalition of the United States
  http://www.irsc.org/
- Autism Research Institute
  http://www.autism.com/
- Autism Society of America
  http://www.autism-society.org/
- CDC—About Autism
  http://www.cdc.gov/ncbddd/autism/
- Center for Collaborative Genetic Studies on Mental Disorders
  http://www.nimhgenetics.org
- Defeat Autism Now (DAN)
  http://www.defeatautismnow.com/
- National Fragile X Foundation
  http://www.nfxf.org/
- Report to Congress on Autism
  http://www.nimh.nih.gov/autismiacc/autismreport2004.pdf
- Rett Syndrome
  http://www.rettsyndrome.org/
- Vaccines
  http://www.cdc.gov/ncbddd/autism/documents/vaccine_studies.pdf

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BILIARY ATRESIA

NUTRITIONAL ACUITY RANKING: LEVEL 2–3

DEFINITIONS AND BACKGROUND

Biliary atresia (neonatal hepatitis) is a serious condition, affecting one in 15,000 births. Incidence is highest in the Asian population. Unconjugated hyperbilirubinemia occurs in approximately 60% of normal-term infants and in 80% of preterm infants; persistence beyond 2 weeks of age demands evaluation (Gubernick et al, 2000).

Complete degeneration or incomplete development of one or more of the bile duct components occurs due to arrested fetal development. CD4(1) lymphocytes and CD56(1) NK cells predominate in the liver of infants with extrahepatic biliary

atresia (Davenport et al, 2001). Lymphocyte-mediated inflammatory damage of the bile ducts plays a role (Shinkai et al, 2006), as does altered *HLA-DR* gene expression in bile ductules (Feng et al, 2004).

Biliary atresia results in persistent jaundice, enlarged spleen, liver damage, portal hypertension, clay-colored stools, dark urine, irritability, and swollen abdomen. The condition becomes evident between 2 and 6 weeks after birth. Treatment involves having a surgical procedure done, the Kasai procedure, which bypasses the ducts to connect the liver to the small intestine. It is more successful if performed early. Complications of the surgery can include liver failure, infections, and sepsis.

If a donor is available, the patient may be a candidate for a liver transplantation. This is the most common disease in childhood that requires liver transplantation. Malnutrition is a critical predictor of mortality and morbidity in children with biliary atresia. Immunosuppressive drugs are then necessary to overcome organ rejection.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: *HLA-DR* gene in bile ductules has been noted, but otherwise not likely inherited.

Clinical/History

- Birth weight
- Height
- Growth (%)
- Diet/intake history
- Dark urine
- Steatorrhea
- Swollen abdomen; edema
- Jaundice >1 month
- Clay-colored stools
- Nuclear hepato imino diacetic acid (HIDA) test
- Liver scan or biopsy

Lab Work

- Alb
- Transthyretin
- H & H
- Alk phos
- Chol
- Trig
- Transferrin
- Blood urea nitrogen (BUN)
- Aspartate aminotransferase (AST)
- Alanine aminotransferase (ALT)
- Alpha-1 antitrypsin deficiency
- PT or INR
- Serum zinc
- Serum copper

INTERVENTION

OBJECTIVES

Preoperatively

- Correct malabsorption and alleviate steatorrhea from decreased bile.
- Correct malnutrition of fat-soluble vitamins and zinc. Prevent rickets, visual disturbances, peripheral neuropathy, and coagulopathies.
- Prevent hemorrhage from high blood pressure (BP) if there is portal hypertension.
- Prepare for surgery or transplantation.

SAMPLE NUTRITION CARE PROCESS STEPS

Altered GI Function

Assessment Data: Stool and urine tests; abdominal girth; weight changes; poor intake.

Nutrition Diagnoses (PES): Altered GI function related to degenerating bile ducts and biliary atresia as evidenced by clay-colored stools, abdominal distention, jaundice, and dark urine.

Intervention: Educate parents about the needed surgery and need for protein, fat-soluble vitamins.

Monitoring and Evaluation: Improved weight and intake after surgery; reduced abdominal distention and normalized stools and urine color.

Postoperatively

- Support proper wound healing by providing all necessary nutrients (e.g., vitamin C, zinc) using appropriate and tolerated feeding method.
- Promote normal growth and development.
- Provide regular nutritional assessments to evaluate progress and improvement or decline.
- Reduce inflammation, which may continue even after surgery (Asakawa et al, 2009).

FOOD AND NUTRITION

Preoperative

- Infants need 1.5–3.0 g protein/kg dry weight to avoid protein catabolism, dependent on enteral versus parenteral source. This translates to 2–2.5 g/kg for PN and 2.5–3 g/kg for enteral nutrition, 1–1.5 g/kg if encephalopathic.
- Identify products enriched with branched-chain amino acids (BCAAs).
- Small, frequent feedings may be useful.
- Use low total fat from the diet. Supplement with oil high in MCTs; add EFAs for age and body size. Pregestimil or Alimentum or other elemental formulas may be needed to decrease fiber and prevent hemorrhage anywhere along the GI tract.
- With edema, limit intake of sodium to 1–2 g/d.
- Supplement with vitamins A, D, E, and K. Intravenous or water-miscible forms can be used.
- Provide antioxidants as serum levels of minerals, such as selenium, zinc, and iron, tend to be low. Avoid use of copper in total parenteral nutrition (TPN) or supplements to prevent toxic build-up.
- Tube feed especially if recurrent or prolonged bleeding from the GI tract occurs. If nasogastric (NG) feeding is not tolerated, a percutaneous gastrostomy (PEG) tube may be used. Failing nutrition should prompt aggressive support (Utterson et al, 2005).

Postoperative

- Control sodium, protein, and other nutrients only if necessary based on symptoms such as edema and renal failure. Carefully monitor vitamin and mineral requirements.
- Use of antioxidants will be needed to reduce inflammatory processes (Asakawa et al, 2009).

- For needed catch-up growth, tube feeding may be beneficial. Assure that all key nutrients are included over a long-term basis.

Common Drugs Used and Potential Side Effects

- Ursodiol (Actigall, Urso) promotes bile flow and may be used after surgery. Side effects are minimal.
- Phenobarbital and cholestyramine are often used to control hyperlipidemia and pruritus. Increase vitamin D, calcium, vitamin B_{12} and folate intake. Constipation can result.
- Corticosteroids may be needed to stimulate independent bile flow. Long-term use can deplete stores of calcium and phosphorus; may elevate glucose, cause stunting, or cause weight gain.
- Diuretics may be used; monitor for depletion of potassium, magnesium, calcium, and folate. Anorexia can occur.
- Antibiotics such as Bactrim or Septra may be needed to manage cholangitis, common following the Kasai procedure. Anorexia, nausea, or vomiting may result. Use of acidophilus and probiotic products may alleviate loss of intestinal bacteria.
- Growth hormone (GH) may be useful to promote catch-up growth.

Herbs, Botanicals, and Supplements

- Herbs and botanicals are not recommended with this condition because the liver is not able to perform its usual role of detoxification.
- Probiotics may be helpful; more studies are needed.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Teach parents about proper feedings or supplements. Indicate which foods provide antixodants, including vitamins C and E, selenium.
- If bile flow improves after surgery or transplantation, a regular diet may be used, although continuing use of MCT oil may be better tolerated for awhile.
- Teach that the fat-soluble vitamins A, D, E, and K can be used only when they are bound to fat. It may be important to take supplemental forms.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants to decrease potential risk of botulism.

For More Information

- American Liver Foundation
  http://www.liverfoundation.org/
- Canadian Liver Foundation
  http://www.liver.ca/Home.aspx
- Children's Liver Association for Support Services
  http://www.classkids.org/library/biliaryatresia.htm

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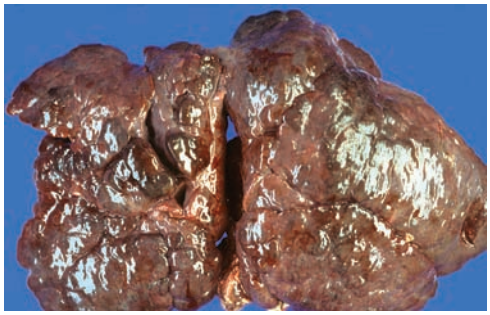
BRONCHOPULMONARY DYSPLASIA

NUTRITIONAL ACUITY RANKING: LEVEL 3–4



Adapted from: Cagle PT, MD. *Color Atlas and Text of Pulmonary Pathology.* Philadelphia: Lippincott Williams & Wilkins, 2005.

DEFINITIONS AND BACKGROUND

Bronchopulmonary dysplasia (BPD) is a chronic lung disease with abnormal growth of the lungs, often following the respiratory distress syndrome of prematurity. One third to one half of all infants born before the 28th week of gestation develop BPD and inflammatory regulators appear to be involved in the pathogenesis in the fetus (Cohen et al, 2007). Chronic aspiration of GI contents is common; measurement of pepsin is one marker (Farhath et al, 2008). The use of supplemental oxygen, while necessary, may also aggravate the condition (Bancalari and Claure, 2006).

Very low-birth-weight (VLBW) infants with severe respiratory disease need extra nutrients for epithelial cell repair and to support catch-up growth. VLBW infants should be given adequate nutritional attention (e.g., parenteral or enteral nutrition, fluid restriction) from the first day of life to enhance growth and minimize respiratory morbidity (Biniwale and Ehrenkranz, 2006). Respiratory failure, supplemental oxygen use, mechanical ventilation, endotracheal intubation, and congenital heart disease all affect nutritional status (D'Angio and Maniscalco, 2004). Slow growth occurs and feeding problems are common. Long-term chronic care is required.

Infants with BPD benefit from comprehensive nutrition and feeding therapy with adequate energy, parental support and education, and feeding evaluation (Biniwale and Ehrenkranz, 2006). Between 10% and 25% of preterm infants with BPD have malnutrition after 2 years of age, and 30–60% of them will continue to suffer from persistent airway obstruction or asthma (Bott et al, 2004).

Glutamine is the main source for lung energy; inositol is necessary for surfactant synthesis; vitamin E and selenium have antioxidant effects (Bott et al, 2004). Vitamin A provides benefit in these patients (Biniwale and Ehrenkranz, 2006; Spears et al, 2004). Long-chain polyunsaturated fatty acids and surfactant replacement therapy are sometimes used to prevent BPD in susceptible infants (D'Angio and Maniscalco, 2004), but controlled trials are needed to verify efficacy (Biniwale and Ehrenkranz, 2006).

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: Gene expression of the newborn umbilical cord implicates chromatin remodeling pathways in premature infants with BPD (Cohen et al, 2007).

Clinical/History

- Gestational age
- Length
- Body mass index (BMI)
- LBW or VLBW growth charts for height and weight
- Size for gestational age (use intrauterine growth chart if available)
- Head circumference
- Diet/intake history
- Emesis
- Stool pattern
- Urinary output
- BP
- Pulmonary hypertension

Lab Work

- H & H
- Pepsin levels
- pH
- Chol, Trig
- K^+ and Cl^- (tend to be low)
- Na^+
- Alb
- Alkaline phosphatase (alk phos)
- White blood cell (WBC) count
- Gluc
- Oxygen saturation levels
- Partial pressure of carbon dioxide (pCO_2)
- Partial pressure of oxygen (pO_2)
- Ca^{++}, Mg^{++}
- Urine-specific gravity

SAMPLE NUTRITION CARE PROCESS STEPS

Overfeeding

Assessment Data: Weight and growth charts, presence of Gastroesophageal reflux disease (GERD) or vomiting, low serum potassium and chloride.

Nutrition Diagnoses (PES): Excessive oral intake of formula in VLBW infant related to respiratory distress and BPD as evidenced by vomiting after most feedings and GERD.

Intervention: Educate parents about need for appropriate amounts of formula, feeding tips for discharge to home, appropriate rate of growth and weight change.

Monitoring and Evaluation: Weight records, decreased vomiting and episodes of GERD, normalized labs for K^+ and Cl^- and other labs.

INTERVENTION

OBJECTIVES

- Increase energy intake to improve growth and respiratory functioning by correcting nutritional deficits (Lai et al, 2006).
- Correct gastroesophageal reflux. Position an infant carefully if formula fed.
- Avoid pareneral overfeeding, which may lead to PN-associated cholestasis (Robinson and Ehrenkranz, 2008).
- Achieve desirable growth. Infants with BPD tend to have delayed development (Bott et al, 2004). Energy needs are approximately 25–50% above normal. Correct malnutrition and anorexia from respiratory distress and ventilator support.
- Provide optimal protein for linear growth, development, and resistance to infection. Improve lean body mass if depleted.
- Spare protein by providing extra energy from fat and carbohydrate. However, excesses of carbohydrate can increase CO_2 production and prevent extubation; calculate needs carefully.
- Replace lost electrolytes, especially chloride, which may lead to death if uncorrected.
- Prevent EFA deficiency.
- Prevent complications, such as aspiration pneumonia or choking during feeding.
- Fluid restriction may be needed if fluid retention is noted; monitor closely.
- Prevent metabolic bone disease by including sufficient calcium and vitamin D intake.

FOOD AND NUTRITION

- Energy requirements will be 25% above normal; provide 120–160 kcal/kg to achieve optimal weight.
- Within the first few days of life, TPN or tube feeding may be required. Initially: 70 (PN) or 95 (enteral) kcal/kg, increasing gradually to 130–180 kcal/kg after acuity subsides.

- Protein requirements may be slightly higher than usual. Careful formula management is needed; Trophamine is beneficial (Blau et al, 2007). Initially, use 2.0 g protein/kg, increasing to 2.5–3.5 g/kg.
- Decrease total carbohydrate (CHO) intake if glucose intolerance develops; monitor blood glucose levels.
- Provide at least the normal recommended allowances for antioxidant and other important nutrients. Include vitamins A, D, and E (use water-miscible sources if necessary); provide adequate calcium, phosphorus, and iron if needed. Nutrient or energy-enriched infant formulas may be needed for catch-up growth.
- Fluid intake (may be restricted to <150 cc/kg/d) and sodium levels may need to be restricted if there is pulmonary edema or hypertension.
- With decreased suck and swallowing ability, tube feeding may be better tolerated. Infants can tolerate most formulas. Nocturnal tube feeding may be useful, especially with growth failures. With gastroesophageal reflux (GERD) a gastrostomy feeding tube may be appropriate.
- Increase fat:CHO ratio with respiratory distress. To meet EFA needs, start with 0.5–1 g/kg and progress to 3 g/kg.
- Omega-3 fatty acids, selenium, inositol, vitamins A and E have been suggested for use with infants who have chronic pulmonary insufficiency (Biniwale and Ehrenkranz, 2006; Bott et al, 2004).
- When ready to progress to an oral diet, use of solids may be better tolerated than liquids. If necessary, thicken liquids or formula (e.g., with baby cereal or other thickeners). Use a supine position to avoid aspiration.

Common Drugs Used and Potential Side Effects

- Exogenous steroid therapy (dexamethasone or methylprednisone) is only used for pulmonary compliance in ventilated premature infants (Grier and Halliday, 2005). This may compromise vitamin A status and restrict bone growth. Sodium retention, anorexia, edema, hypertension, and potassium losses are side effects. Take with food to decrease GI effects. Use more protein and less sodium; enhance potassium if needed.
- Antibiotics are needed during infections; acidophilus and probiotic products may alleviate losses of intestinal bacteria.
- Bronchodilators or caffeine may be used for apnea of prematurity. Anorexia can occur.
- Diuretics may be needed to lessen pulmonary edema. Monitor those that deplete serum potassium, such as furosemide (Lasix). Magnesium, calcium, and folate may be also depleted; appetite may decline.
- Cysteine, N-aceyl cystiene, or cystine may be used in combination with chloride.

Herbs, Botanicals, and Supplements

- Herbs and botanicals should not be used for BPD because the lungs are not able to perform their role in oxygenation of cells.
- Use of acidophilus and probiotic products may be useful with chronic antibiotic therapy.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Diet must be re-evaluated periodically to reflect growth and disease process. Assure adequacy of vitamins and related nutrients for lung health (for example, vitamin A).
- Ensure that all foods and beverages are nutrient dense.
- New foods may be introduced gradually; thicken as needed to avoid aspiration.
- Fluid intake should be adequate to meet needs but not excessive.
- Discuss signs of overhydration and dehydration with the parent/caregiver.
- Oral–motor skills may be delayed from long-term ventilator use; discuss how to make adjustments with caregiver.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants to decrease potential risk of botulism.

For More Information

- American Lung Association
  http://www.lungusa.org/
- National Blood, Heart, and Lung Institute
  http://www.nhlbi.nih.gov/health/dci/Diseases/Bpd/Bpd_WhatIs.html

BRONCHOPULMONARY DYSPLASIA—CITED REFERENCES

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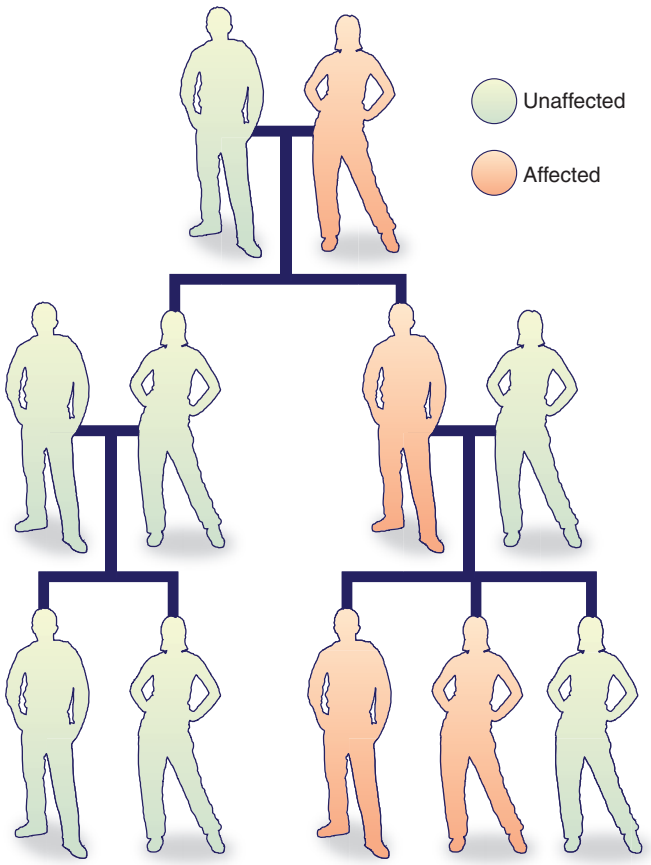
CARBOHYDRATE METABOLIC DISORDERS

NUTRITIONAL ACUITY RANKING: LEVEL 4

Condition affecting members of a family



DEFINITIONS AND BACKGROUND

Abnormalities of glucose metabolism are the most common errors of carbohydrate metabolism; causes include both environmental and genetic factors and generally lead to one of the forms of diabetes (see chapter 9). Glucose is transported across cell membranes by active sodium-facilitated transport in the intestinal or renal cells; in all other cells, the GLUT family of glucose transporters are needed (Brown, 2000). Diagnosis of "carbohydrate malabsorption" occurs during infancy or childhood, with hypoglycemia, hepatomegaly, poor physical growth, and deranged biochemical profiles.

Congenital glucose–galactose malabsorption (congenital renal glycosuria) is an extremely rare, autosomal recessive trait. Watery, profuse diarrhea occurs from deficiency in the sodium-coupled cotransport of glucose and galactose in the intestinal mucosa. There is no cure, but removal of lactose, sucrose, and glucose lessons symptoms. **Glucose transporter type 1 (Glut1) deficiency syndrome** produces a seizure disorder with low glucose in the cerebrospinal fluid, developmental delay, and acquired microcephaly (Wang et al, 2004). **Glut2 deficiency** produces Fanconi–Bickel syndrome, which resembles type I glycogen storage disease (GSD; Brown, 2000).

Fructose intolerance results from a defect in the enzyme converting fructose to glucose (1-phosphofructaldolase). It is an autosomal recessive disease, as common as 1 in 20,000 persons in some European countries. Fructose intolerance causes GI discomfort, nausea, malaise, and growth failure. Ingesting fructose causes profound hypoglycemia; if left untreated, progressive liver disease results.

Galactosemia (galactose-1-phosphate uridylyltransferase deficiency or GALT deficiency) causes cataracts, hepatomegaly, and mental retardation. It occurs in one of 60,000 births. People with galactosemia are unable to fully metabolize the simple sugar galactose. The enzyme galactokinase may play a role (Holden et al, 2004). High levels of the sugar alcohol, galactitol, may be present. FTT, vomiting or diarrhea, jaundice, liver disease occur after milk ingestion. Bone density may decline over time (Panis et al, 2004). Cataracts, encephalopathy, or death from *E. coli* sepsis may occur (Berry, 2008). In females, serum FSH levels may be elevated and primary ovarian insufficiency (POI) may prevent successful pregnancy (Berry, 2008).

GSDs are rare genetic disorders in which glycogen cannot be metabolized to glucose in the liver because of enzyme deficits. Glycogen is the storage form of glucose and is found in the liver and muscles; a small amount is also found in the kidney and intestine. In severe cases of GSD, liver transplantation may be needed. Table 3-4 lists the various types of GSD.

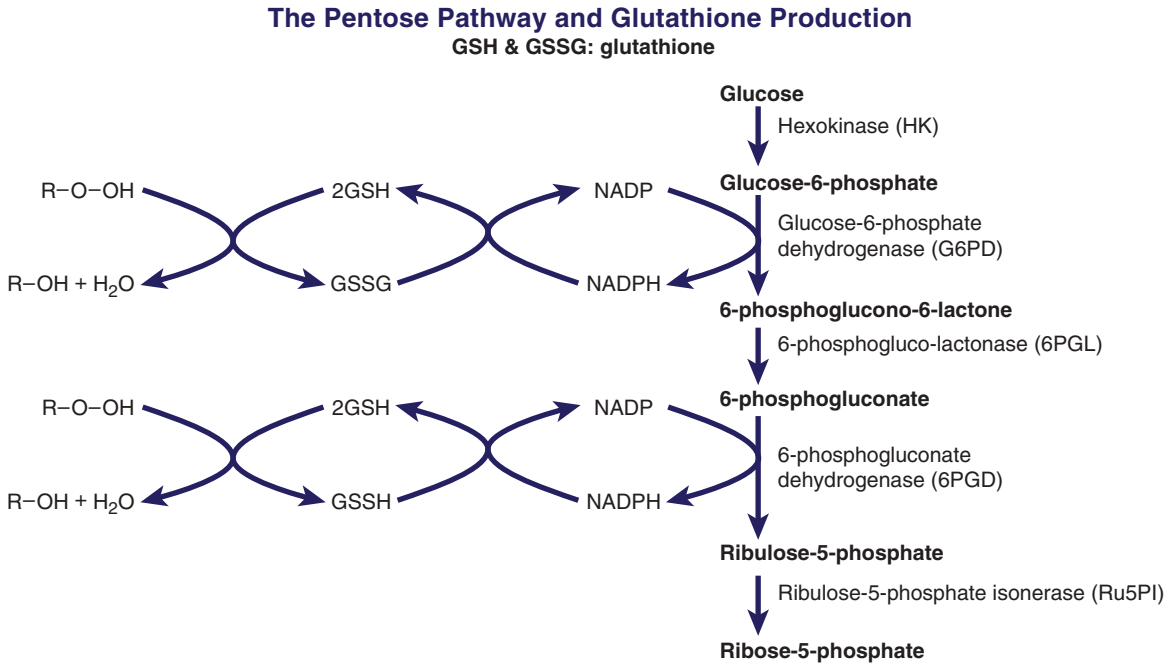
Sucrose intolerance occurs rarely as a genetic defect or temporarily after GI flu or irritable bowel distress. Sucrase and maltase deficiency may occur simultaneously, with an osmotic diarrhea.

Lactose intolerance is discussed in chapter 7; the autosomal recessive trait deficit is encoded by the lactase gene on chromosome 2 with a frequency of 5% to 90% in humans.

TABLE 3-4 Signs of Cerebral Palsy

Struggles with fine motor skills: handling scissors, using crayons, buttoning a shirt, and any other movement that uses fingers and hands.
Struggles with gross motor skills: walking, riding a tricycle, kicking a ball, and other movements using legs and arms.
Trouble sitting upright; it takes a lot of muscle tone to sit up without toppling over.
Shakes a lot or has uncontrollable jerking of her legs, arms, or torso.
Muscles are weak.
Body tremors, drooling, weakened muscles in his face; may lose control of his tongue.
Trouble moving from one position to another.
Trouble sucking.

Source: http://www.cerebralpalsy.org/what-is-cerebral-palsy/symptoms/.



ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: The congenital defects relate to aberrant transporter genes: glucose–galactose malabsorption syndrome, *SGLT1*; glucose transporter 1 deficiency syndrome, *GLUT1*; Fanconi–Bickel syndrome, *GLUT2;* fructose intolerance, *fructose-1-phosphate aldolase.* The GSDs have deficiency in various types of microsomal *glucose-6-phosphatase (G6Pase)* activity.

Clinical/History

- Height or length
- Weight
- BMI
- Growth (%)
- Diet/intake history
- Infections
- Nausea and vomiting
- Jaundice
- Infantile seizures
- Acquired microcephaly
- Development delay or FTT?
- Edema
- Demyelinating neuropathy?

Lab Work

- Gluc (decreased in fructosemia)
- HgbA1 c (decreased erythrocyte glucose uptake)
- Hypoglycorrhachia (CSF glucose <40 mg/dL)
- Trig, Chol (elevated in Von Gierke's disease)
- LFTs: ALT, AST, CK
- Urinary and serum galactose or fructose
- Acetone
- Serum phosphate
- Serum lactate
- Serum ammonia
- Serum bilirubin
- Uric acid
- Alb

INTERVENTION

OBJECTIVES

- Eliminate the offending macronutrient that cannot be digested; adjust the other macronutrients to promote growth and health maintenance. Prevent hypoglycemia, where indicated.
- Read labels carefully.
- Fructose intolerance requires omission of fructose from the diet.
- For galactosemia, correct the diet to prevent physical and mental retardation, cataracts, portal hypertension, and cirrhosis. Vitamin E seems to have positive, protective effects. Read labels carefully; galactose is not always reported. Consider infant formulas containing glucose, without lactose and maltose; lactose-restricted diet products;

SAMPLE NUTRITION CARE PROCESS STEPS

Abnormal GI Function

Assessment Data: Weight and growth charts, nausea and vomiting, elevated LFTs, and frequent episodes of hypoglycemia.

Nutrition Diagnoses (PES): Abnormal GI function related to metabolic disorder and GSD as evidenced by nausea and vomiting.

Intervention: Educate parents about frequency and timing for meals and snacks, enhancing energy intake through six to eight small meals daily plus nightly gastrostomy feeding of a complete nutritional supplement with cornstarch, DHA, and special oil.

Monitoring and Evaluation: Weight records, growth, tolerance for various food consistencies, less nausea and vomiting.

rice-based milk substitutes; lactose-free products that contain glucose.
- For (Glut1) deficiency syndrome, a higher fat intake is useful. The ketogenic or a modified Atkins diet should be introduced early and continue into adolescence (Ito et al, 2008; Klepper, 2008).
- For the GSDs, maintain glucose homeostasis, prevent hypoglycemia, promote positive nitrogen balance and growth, and correct or prevent fatty liver. Prevent EFA deficiency (Abdel-Ghaffar et al, 2003). Consider carbohydrate-modified products such as cornstarch (as a low-release glucose source).
- Sucrose/maltose intolerance requires omission of sucrose and maltose from the diet.

FOOD AND NUTRITION

Congenital Glucose–Galactose Malabsorption

- Use a diet free from sucrose, lactose, and glucose. Add fructose to a CHO-free formula incrementally as tolerated.
- Fructose may be used for older children; the other CHO sources should be avoided.

Fructosemia

- Diet must exclude fructose, sucrose, sorbitol, invert sugar, maple syrup, honey, and molasses.
- Read labels carefully. Tube feedings or intravenous solutions may contain sources of fructose.

Galactosemia

- Use a lactose and galactose-free diet—no milk, milk products, soybeans, peaches, lentils, liver, brains, or breads or cereals containing milk or cream cheese. Omit fresh blueberries and honeydew melon; fresh cherries, citrus, mango, red plums, and strawberries are allowed (Stepnick-Gropper et al, 2000).
- For infants, try Isomil or ProSobee, Elecare, Nutramigen, or formulas containing casein hydrolysate.
- Supplement with calcium, vitamin D, vitamin E, and riboflavin. In some disorders, galactose can often be reintroduced later in life.
- Read labels carefully; galactose is not reported on labels. Formulas labeled "low lactose" are *not* good substitutes; they contain lactose in amounts that can seriously harm patients with galactosemia.
- Be careful when using tube feedings or intravenous solutions; they may contain lactose.

Glycogen Storage Disorders

- Increase protein intake to improve muscle strength (Bembi et al, 2003).
- Use small, frequent feedings and, if steroids are used in treatment, a low-sodium diet. Long-term use of steroids can deplete stores of calcium and phosphorus and may elevate glucose, cause stunting, or cause weight gain.
- Avoid lactose and sucrose. Read all product labels.
- Glucose may be used. Concentrated sweets may be restricted unless made with pure glucose syrup.
- Cornstarch is used to prevent hypoglycemia.
- Sometimes, night feedings with additional daytime meals work effectively. Giving 4 tbsp cornstarch in 5 oz of fluid and 3 oz of juice, carnitine, DHA via gastrostomy (24 mL/h) at night may help the liver to maintain a normal blood glucose level (Isaacs and Zand, 2007).
- A multivitamin–mineral supplement with vitamin C, iron, and calcium may be needed because fruits and milk are limited. As necessary, replete nutrients such as vitamin B_{12}, folate, calcium, and iron.

Sucrose/Maltose Intolerance

- Omit sucrose and maltose from the diet.
- For the nongenetic form, gradually add these sugars back into the diet.
- Tube feedings or intravenous solutions may contain sources of sucrose or maltose; read labels.

Common Drugs Used and Potential Side Effects

- For persons with galactosemia, eliminate drugs containing lactose; supplement with calcium and riboflavin.
- Sucrose and maltose are added to many drugs; check carefully.
- All vitamin–mineral supplements must be free of the nontolerated carbohydrates.
- If liver transplantation is needed, support the immunosuppression with appropriate nutrition interventions. Changes in fluid or sodium or other nutrients may be required.
- In Pompe disease, Myozyme (alglucosidase alfa) may be prescribed.

Herbs, Botanicals, and Supplements

- Herbs and botanicals should not be used for these conditions because there are no controlled trials to prove efficacy.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Explain which sources of carbohydrate are allowed specific to the disorder.
- Read labels carefully. Many foods contain milk solids, galactose (e.g., luncheon meats, hot dogs), and other sugars; omit according to the disorder. Contact formula companies regarding product updates.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants to decrease potential risk of botulism.

For More Information

- Association for Glycogen Storage Disease—United States
  http://www.agsdus.org/
- International Pompe Association
  http://www.worldpompe.org/
- March of Dimes
  http://www.marchofdimes.com

INBORN CARBOHYDRATE METABOLIC DISORDERS—CITED REFERENCES

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CEREBRAL PALSY

NUTRITIONAL ACUITY RANKING: LEVEL 3

Adapted from: Weber J RN, EdD and Kelley J RN, PhD. *Health Assessment in Nursing,* 2nd ed. Philadelphia: Lippincott Williams & Wilkins, 2003.

DEFINITIONS AND BACKGROUND

Cerebral palsy (CP) results from brain damage to motor centers before, during, or after birth. Human epidemiological data suggest a relationship between CP and cytokines or inflammation (Gaudet and Smith, 2001). One of 500 live births may be affected. Each year, 1200 to 1500 preschool-age children in the United States are identified as having CP, which causes physical and mental disabilities that are nonprogressive.

Infants may present with early abnormal rolling, stiffness, irritability, and developmental delays (see Table 3-5). Seizures, mental retardation, hyperactive gag reflex, tongue thrust, poor lip closure, inability to chew properly, behavioral problems, visual or auditory problems may occur. Symptoms may be mild or severe, and vary from one person to the next. Skeletal maturation is frequently delayed.

Spastic (uncontrolled shaking or difficult, stiff movement) affects about 75%, **athetoid** (involuntary worm-like movement) affects 15%, **ataxic** (impaired coordination and balance) affects about 10%, and many individuals have a **mixed** form of CP. In many individuals, wasting of voluntary muscles contributes to reduced resting energy needs (Hogan, 2004). The potential for malnutrition exists, wherein indirect calorimetry is useful to assure adequacy of intake (Hogan, 2004).

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: The condition is not genetic in origin.

Clinical/History

- Low birth weight (LBW)
- Low 5-minute Apgar score (below 7)
- Height or length
- Weight and growth chart (%)

Shaking, worm-like, or stiff movements
Impaired balance and coordination
Skull x-ray
Chewing problems
Diet/intake history
Self-feeding problems
Seizures
GERD
Constipation

Lab Work

Gluc
Alb
Serum Ca^{++}, Mg^{++}
Transferrin
Alk phos
H & H, serum Fe, ferritin

INTERVENTION

OBJECTIVES

- Alleviate malnutrition resulting from the patient's inability to close lips, suck, bite, chew, or swallow.
- Promote independence through use of adaptive feeding devices. Eye–hand coordination is often lacking, and grasp may not be strong.
- Assess appropriate energy and nutrient needs. When adequately nourished, children and adolescents with CP appear more tranquil and require decreased feeding time (Hogan, 2004). Promote mealtimes in a quiet, unhurried environment.

TABLE 3-5 Medications for Congenital Heart Disease

Generic Name	Brand Name	Concerns
Acebutol	Sectral	GI distress or nausea
Atenolol	Tenormin	GI distress or nausea
Azathioprine	Imuran	
Baby aspirin	Bayer	
Captopril	Capoten	GI distress or nausea
Cisapride	Propulsid	
Digoxin	Lanoxin	
Enalapril	Vasotec	
Furosemide	Lasix	GI distress or nausea; potassium, magnesium, calcium, and folate may be depleted
Hydrochlorothiazide	Hydrodiuril	GI distress or nausea; potassium, magnesium, calcium, and folate may be depleted
Lisinopril	Zestril	
Metoprolol	Lopressor	
Prednisone	Deltasone	Depletes calcium and phosphorus; may elevate glucose, cause stunting, or cause weight gain.
Propranolol	Inderal	
Spironolactone	Aldactone	
Warfarin	Coumadin	Need steady intake of vitamin K; no big fluctuations

SAMPLE NUTRITION CARE PROCESS STEPS

Difficulty with Feeding Self and Soy Allergy

Assessment Data: Weight and growth charts, medical history of aspiration, difficulty consuming adequate intake orally or by tube, soy allergy.

Nutrition Diagnosis (PES): NB 2.6. Self-feeding difficulty related to inability to bite properly and use utensils in CP as evidenced by weight loss of 4 lb in 6 months, current BMI of 13, ht and wt percentiles both <5%, history of aspiration when tube fed, allergy to soy.

Interventions:

Food and Nutrient Delivery

ND 1.2. Increase caloric intake through bolus feeding.

ND 1.3. Provide high-calorie, high protein formula free of soy.

Nutrition Education.

E 1.1. Discuss importance of nutrition and foods child is able to tolerate.

Counseling

C 2. RD to counsel mother and home nurse on importance of enhancing high calorie foods as necessary to prevent weight and muscle loss.

Care Management

RC 1.3. RD to collaborate with MD and home nurse for highest quality of care for patient, RD to collaborate with formula company to find high calorie high protein formula that is soy-free.

Monitoring and Evaluation: Weight records, improved intake of sufficient energy and protein to rebuild muscle mass and improve in growth percentiles; improved BMI for age.

- Correct nutritional deficits, altered growth rate, developmental delays, or retardation.
- Prevent or correct constipation, aspiration pneumonia, gastroesophageal reflux, pressure ulcers.

FOOD AND NUTRITION

- Energy requirements of children and adolescents will vary depending on functional capacity, degree of mobility, severity of disease, and level of altered metabolism (Hogan, 2004). Reduce energy intake for spastic patients or those with severely limited activity, 11 kcal/cm for ages 5–11. For moderately active patients, use 14 kcal/cm for ages 5–11. Increase energy intake (up to 45 kcal/kg) to accommodate for added movements of the athetoid patient over age 18.
- Breast milk is recommended for infants with CP (Vohr et al, 2006).
- Feeding gastrostomy tubes are a reasonable alternative for severe feeding and swallowing problems and poor weight gain (Rogers, 2004; Sullivan et al, 2004). Night feedings may allow more normal daytime routines. Daytime bolus feedings of high-calorie, high-protein formulas at scheduled times may also be needed to provide adequate nutrition in some cases.

- For chewing problems, eliminate coarse, stringy foods. Puree foods as needed.
- With frequent vomiting, assess actual intake; anti-emetic medications may be needed.
- For constant dribbling, add cereal or yogurt to fluids. Replace fluids, thickened if needed.
- For constipation, use laxative or high-fiber foods such as bran in the diet. Provide extra fluids. In younger children, too much fiber can displace intake of adequate nutrition.
- Supplement with a general multivitamin–mineral supplement, especially for B-complex vitamins, calcium, and vitamin D (Henderson et al, 2005).
- For pressure ulcers or skin breakdown from minimal positioning of the body, ensure adequate protein, vitamins C and A, and zinc. Work with caregivers to turn and reposition every 2 hours.

Common Drugs Used and Potential Side Effects

- Dantrolene (Dantrium) inhibits the release of calcium in muscle and skeletal tissue, preventing muscle cramping and spasms. Diarrhea, changes in BP, weight loss, and constipation may all occur.
- Klonopin (clonazepam) is a benzodiazepine used to slow down the central nervous system (CNS) for treatment of spasticity. Side effects may include constipation or diarrhea, dizziness, drowsiness, clumsiness, unsteadiness, a "hangover" effect, headache, nausea, and vomiting.
- Antibiotics such as baclofen may cause or aggravate diarrhea. Use of acidophilus and probiotic products may alleviate loss of intestinal bacteria.
- Laxatives may often be needed; monitor fiber and fluid needs. Milk of magnesia can be used safely in a pediatric dosage. Avoid using laxatives containing mineral oil.
- Anticonvulsants may increase risk of osteomalacia. Nutrient deficiencies are common: vitamins D, B_6, B_{12}, and K, folate, calcium, and biotin are often insufficient and should be replaced.

Herbs, Botanicals, and Supplements

- Herbs and botanicals should not be used; there are no controlled trials to prove efficacy.
- Probiotics may be used to alleviate loss of intestinal bacteria. Encourage natural sources such as yogurt or acidophilus milk, if tolerated.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Remind older patients to keep lips closed to avoid losing food from their mouths as they try to chew.
- Fortify the diet with dry or evaporated milk, wheat germ, and other nutrient enhancers when intake is inadequate.
- Allow extra time for feedings. Use of adaptive feeding equipment may be beneficial. Provide special training as needed for a specific feeding procedure (e.g., a preemie nipple for poor suck).
- Help parent or caretaker with problems related to dental caries, drugs, constipation, pica, or weight.
- Exercise can be beneficial, such as recreational sports, yoga, and hippotherapy.
- Tube feeding may be needed. Ensure proper positioning to avoid aspiration or GERD.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants to decrease potential risk of botulism.

For More Information

- American Academy of Developmental Medicine and Dentistry
  http://www.aadmd.org
- American Association on Health and Disabilities
  http://www.aahd.us/
- American Cerebral Palsy Information Center
  http://www.cerebralpalsy.org
- CP Connection
  http://www.cpconnection.com/
- CP Resource Center
  http://twinenterprises.com/cp/
- Developmental Disabilities Nurses Association
  http://www.ddna.org/
- Disability Resource Network
  http://www.d-r-d.com/
- Easter Seals
  http://www.easter-seals.org
- Hemiplegic Cerebral Palsy
  http://www.hemikids.org/
- United Cerebral Palsy Association, Inc.
  http://www.ucpa.org/

CEREBRAL PALSY—CITED REFERENCES

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Henderson RC. Longitudinal changes in bone density in children and adolescents with moderate to severe cerebral palsy. *J Pediatr.* 146:769, 2005.

Hogan SE. Energy requirements of children with cerebral palsy. *Can J Diet Pract Res.* 65:124, 2004.

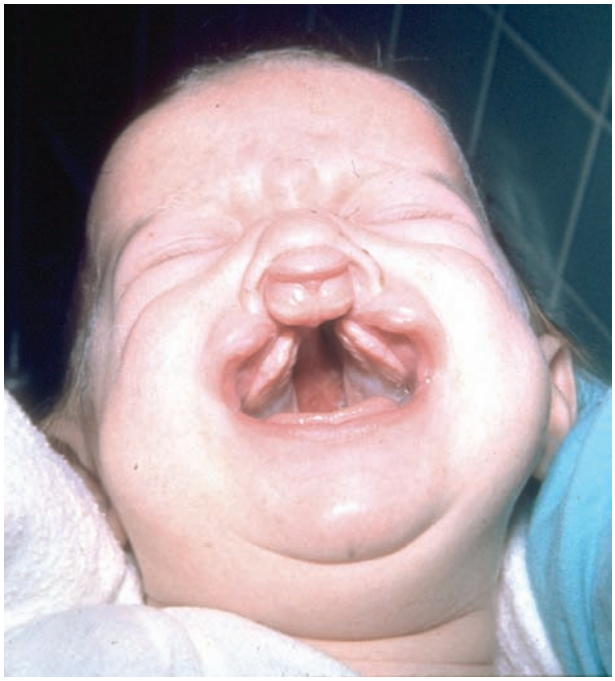
Rogers B. Feeding method and health outcomes of children with cerebral palsy. *J Pediatr.* 145:S28, 2004.

Sullivan PB, et al. Impact of gastrostomy tube feeding on the quality of life of carers of children with cerebral palsy. *Dev Med Child Neurol.* 46:796, 2004.

Vohr BR, et al. Beneficial effects of breast milk in the neonatal intensive care unit on the developmental outcome of extremely low birth weight infants at 18 months of age. *Pediatrics.* 118:115, 2006.

CLEFT LIP AND PALATE (OROFACIAL CLEFTS)

NUTRITIONAL ACUITY RANKING: LEVEL 3



Adapted from: Rubin E MD and Farber JL MD. *Pathology,* 3rd ed. Philadelphia: Lippincott Williams & Wilkins, 1999.

DEFINITIONS AND BACKGROUND

Cleft lip and palate are congenital malformations occurring during the embryonic period of development. They result in a fissure in the lip and roof of the mouth, which may be unilateral or bilateral. Incidence is approximately one in 700 births in Caucasians, or about 5000 births annually in the United States. Infants with cleft palate are often smaller in size and weight than other infants.

Periconceptional folate and folic acid intake prevents orofacial clefts (OFC; Krapels et al, 2004). Other nutrients also play a role, and many mothers who eat poorly risk having a baby with OFC. Sufficient preconceptual intake of macronutrients and key micronutrients may decrease OFC risk (Krapels et al, 2006; Mitchell et al, 2003). There is also a risk from inadequate zinc intake (Tamura et al, 2005).

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: Interferon regulatory factor 6 (IRF6) gene may be related.

Clinical/History

- Length (height)
- Growth (%)
- Weight
- Weight changes
- Diet/intake history
- Head circumference
- Cleft type (unilateral or bilateral; complete or incomplete)
- Otitis media (OM) or other infections
- Chewing difficulty

Lab Work

- Gluc
- Alb
- H & H
- Serum Ca^{++}, Mg^{++}

INTERVENTION

OBJECTIVES

- Cleft palate is more of a problem than cleft lip. Compensate for the patient's inability to suck because of the air space between the mouth and nose.
- Prevent choking, air swallowing, coughing, and fatigue as much as possible.
- Encourage breastfeeding where possible to protect against OM (Aniansson et al, 2002).
- Supply the child with energy to heal and to grow; offer tips for meal planning and resources because feeding will be a challenge (Redford-Badwal et al, 2003).
- For surgery, allow extra energy and protein for healing; use a multivitamin supplement. Before surgery, a custom retainer device may be placed in the mouth and is intended to gradually pull the edges of the cleft closer to achieve better lip repair. The device also aids in the feeding process.

FOOD AND NUTRITION

- Provide a normal diet in accordance with the patient's age and dietary recommendations. Monitor diet carefully

SAMPLE NUTRITION CARE PROCESS STEPS

Inability to Bite or Chew

Assessment Data: Weight and growth charts, difficulty chewing and biting into foods.

Nutrition Diagnoses (PES): NC 1.2. Biting/chewing difficulty related to craniofacial malformations as evidenced by prolonged feeding time and decreased intake.

Intervention: Educate parents about texture changes, timing for meals, enhancing energy intake through high calorie foods and supplements, oral health and hygiene.

Monitoring and Evaluation: Weight records, growth, tolerance for various food consistencies.

because mother may have had a poor diet during preconceptual period and pregnancy.
- For infant feeding, use a medicine dropper or plastic bottle with a soft nipple and enlarged hole. The use of a squeezable, collapsible bottle with a longer nipple and a large crosscut opening, which allows parents to control the flow of milk, can help. Release formula or milk a little at a time, in coordination with the infant's chewing movements. Burp infant frequently to release swallowed air. Feed the infant in an upright position to prevent aspiration.
- When the infant is 4–6 months of age, begin to add solids in the diet. Pureed baby foods can be used, or the infant can be spoon fed with milk used to dilute the baby foods. Feed solids from a spoon and avoid use of a bottle or commercial syringe feeder, unless prescribed for unique circumstances.
- Avoid fruit peelings, nuts, peanut butter, leafy vegetables, heavy cream dishes, popcorn, grapes, biscuits, cookies, and chewing gum as they may get lodged in the palate. Avoid spicy, acidic foods if they cause irritation.

Common Drugs Used and Potential Side Effects

- No specific medicines are used for cleft lip and palate; surgery is the primary treatment. After surgery, there may be a need for antibiotics if infection sets in.
- Women who are taking valproate, lithium, carbamezine and other bipolar disorder medicines should discontinue use during pregnancy to reduce risk for cleft lip or palate.
- If genetic testing indicates an MTHFR allele, L-methylfolate (such as in Deplin) may be prescribed.

Herbs, Botanicals, and Supplements

- Herbs and botanicals are not required for cleft lip or palate.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Explain how to feed the infant with a special nipple as needed.
- Solids may be started at 4–6 months. Using thickened baby food or pureed items as tolerated.
- Supplement the diet with vitamin C if citrus juices are not taken well.
- Have the parents use only small amounts of liquid when they are feeding an infant. To prevent choking, slow swallowing should be encouraged and proper positioning should be taught.
- Discuss the impact of surgery and how to promote effective healing by using a nutrient-dense diet with adequate amounts of protein, energy, vitamins A and C, and zinc.
- Because of the types of problems that may occur (teeth in the area of the cleft may be missing or improperly positioned, affecting biting and chewing ability; speech difficulties; frequent colds, sore throats, OM, tonsillitis), assistance from a variety of therapists and professionals is needed. The dietitian can assist with nutrition and feeding-related issues. Nutrient density and texture assessments should be ongoing.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants; this will decrease the risk of botulism.

For More Information

- AboutFace USA
  http://www.aboutfaceusa.org/
- About Smiles
  http://www.aboutsmiles.org/
- American Cleft Palate-Craniofacial Association
  http://www.cleftline.org/
- Center for Craniofacial Development and Disorders
  http://www.hopkinsmedicine.org/craniofacial/Home/Index.cfm
- Cleft Lip and Palate Resources
  http://www.widesmiles.org/
- Cleft Palate Foundation
  http://www.cleftline.org/aboutclp/
- FACES: The National Craniofacial Organization
  http://www.faces-cranio.org/
- Federation for Children with Special Needs
  http://www.faces-cranio.org/
- Forward Face: The Charity for Children with Craniofacial Conditions
  http://www.nffr.org/ForwardFace.htm
- Smile Train
  http://www.smiletrain.org/library/PublicLibrary.html

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Aniansson G, et al. Otitis media and feeding with breast milk of children with cleft palate. *Scand J Plast Reconstr Surg Hand Surg.* 36:9, 2002.

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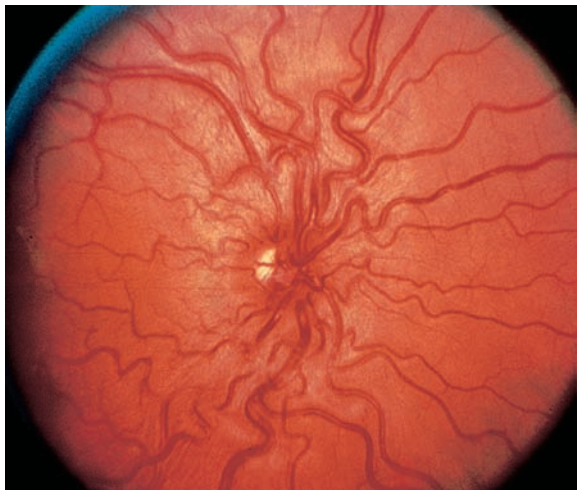
Tamura T, et al. Plasma zinc concentrations of mothers and the risk of nonsyndromic oral clefts in their children: a case-control study in the Philippines. *Birth Defects Res A Clin Mol Teratol.* 73:612, 2005.

CONGENITAL HEART DISEASE

NUTRITIONAL ACUITY RANKING: LEVEL 2



Adapted from: Gold DH, MD, and Weingeist TA, MD, PhD. *Color Atlas of the Eye in Systemic Disease.* Baltimore: Lippincott Williams & Wilkins, 2001.

DEFINITIONS AND BACKGROUND

Congenital heart disease is the most common birth defect in the United States, with an estimated incidence of approximately 12–14 of 1000 live births per year (Owens and Musa, 2009). Usually, some developmental defect occurred between weeks 5 and 8 of pregnancy (e.g., from rubella). An increased risk for malnutrition, growth failure, or pulmonary hypertension occurs.

Energy expenditure is significantly elevated and feeding difficulties are common. Increased levels of grehlin and other factors may lead to growth retardation and FTT. Supplementary oxygen is often needed, especially during feeding; the child will not grow if oxygen is inadequate.

Surgical repair may be delayed to allow weight gain. Surgery is performed when a patient reaches an ideal weight and age, or if FTT precludes further waiting. The neonate undergoing cardiopulmonary bypass surgery experiences profound metabolic response to stress and has less metabolic reserves for wound healing and growth (Owens and Musa, 2009).

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: Some congenital heart defects are related to an abnormality of an infant's chromosomes (5–6%), single-gene defects (3–5%), or environmental factors (2%) while the majority are multifactorial. Down syndrome (DS), Williams syndrome, trisomy 13 or trisomy 18 are examples of conditions that are often linked with congenital heart disease (Children's Hospital Boston, 2009).

Clinical/History

- Height
- Weight
- Head circumference
- Growth pattern
- FTT?
- Diet/intake history
- BP
- Weight changes
- Edema
- Intake and output (I & O)
- Ultrasound
- Echocardiography
- Heart murmur?
- Cyanosis?
- Ventilator dependency?

Lab Work

- Gluc
- Urinary Osm
- Na^+, K^+
- BUN, creatinine (Creat)
- Chol
- Trig
- Serum folate, B_{12}
- H & H, serum Fe or ferritin
- O_2, CO_2
- Serum zinc

INTERVENTION

OBJECTIVES

- Support normal growth and weight gains; growth failure is common, especially with associated heart failure. An 8–16 oz gain in one month might be acceptable.
- Improve oral intake. Poor sucking may occur in infants, but it is possible to breastfeed with education and support of the mother (Barbas and Kelleher, 2004).

SAMPLE NUTRITION CARE PROCESS STEPS

High Energy Requirements

Assessment Data: Weight and growth charts, need for ventilator support, fluid requirements, estimated needs to increase rate of growth.

Nutrition Diagnoses (PES):

NI 1.2. Increased energy expenditure related to breathing and inadequate weight gain as evidenced by low oxygen saturation levels, inadequate rate of growth, <5% weight for height and age.

NC 1.3. Breastfeeding difficulty related to poor sucking ability as evidenced by observation of mother during attempts to breastfeed, with infant unable to latch on and sustain intake longer than a few seconds.

Intervention: Assist mother with breastfeeding tips and explain how to use supplemental formulas if needed to support growth of infant. Educate mother/parents about increasing nutrient density, frequency of meals or snacks, types of formula needed when required, allowing extra feeding time due to dyspnea.

Monitoring and Evaluation: Weight records and improved growth rate allowing child to have heart surgery.

- Lessen fatigue associated with mealtimes. Assure adequate oxygen replacement, especially during feeding.
- Meet energy needs from increased metabolic rate and the need for catch-up growth, without creating excessive cardiac burden or excessive renal solute overload.
- Improve appetite, which can be decreased from the medications.
- Promote good oral hygiene to prevent infections.

FOOD AND NUTRITION

- Determine and provide calories as needed for age (e.g., 100 kcal/kg in second year of life). See Table 3-1. Most formulas contain 67 kcal/dL or 20 kcal/oz. Severe FTT cases may need an extra 30–60 kcal/kg/d over usual; follow standard mixing recommendations for formula concentration and add modular products to reach a desired level. For infants, a formula up to 90–100 kcal/dL can be used while carefully monitoring adequacy of fluid ingestion.
- Energy should contain approximately 10% protein (avoid overloading), 35–50% fat as vegetable oils (known to be readily absorbed), and 40–55% CHO.
- Sodium intake should be approximately 6–8 mEq daily, dependent on diuretic use and cardiopulmonary status.
- Continuous 24-hour tube feeding may be useful. PEG tube feeding can be a useful adjunctive therapy, especially using formulas with a lower mineral to protein ratio (e.g., partially demineralized whey).

Common Drugs Used and Potential Side Effects

- Diuretics are often prescribed. Other drugs are specific to the individual patient's requirements; see Table 3-6. Give medicines before feedings to be sure they have been taken.
- With Nesiritide, used to cause diuresis; hypotension may result (Feingold and Law, 2004).
- Infective endocarditis (IE) among children with *Staphylococcus aureus* bacteremia may require treatment with antibiotics; use of probiotic products may alleviate loss of intestinal bacteria.

Herbs, Botanicals, and Supplements

- Herbs and botanicals should not be used because there are no controlled trials to prove efficacy.
- With prolonged use of antibiotic therapy, probiotic products may be useful. Encourage intake of yogurt, acidophilus milk, and related products.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Discuss the role of nutrition in achieving adequate growth and controlling heart disease.
- Discuss growth patterns and goals.

TABLE 3-6 Normal Growth Rates for Height and Weight in Children

Age	Growth in Length or Height (mm/d)[a]	Growth in Length or Height (inches/yr)
0–6 months	1.06 declining to 0.77	7–10
6–12 months	0.47	6–7
1–2 years	0.35 declining to 0.30	4–5
2–3 years	—	3–4
3–4 years	—	2–3
4–10 years	—	2

Age	Daily Growth in Weight (oz/d)[a]	Growth in Weight
0–4 months	1.0 declining to 0.61	1½ lb/mo
4–10 months	0.61 declining to 0.47	1 lb/mo
10–24 months	0.47 declining to 0.25	½ lb/mo
2–8 years	—	3–4 lb/y

Formula for Measurement Conversion

One pound = 0.455 kg OR 2.2 lb = 1 kg

One inch = 2.54 cm

Multiply each inch by 2.54 to come up with length in centimeters

Multiply each pound by .455 to come up with weight in kilograms or take the pounds and divide by 2.2

[a]See Russell-Silver Syndrome Growth Charts, based on the Centers for Disease Control and Prevention's federally authorized growth charts, http://www.magicfoundation.org.

- Provide support for breastfeeding mothers who wish to continue as long as possible.
- Discuss the role of nutrition in oral health and overall immunity.
- Women who wish to become pregnant should be sure they are immunized against rubella, which can cause congenital heart disease.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants to decrease potential risk of botulism.

For More Information

- Children's Cardiomyopathy Foundation
  http://www.childrenscardiomyopathy.org/site/overview.php
- Children's Heart Institute
  http://www.childrenheartinstitute.org/educate/heartwrk/elechhse.htm
- Children's Organ Transplant Network
  http://www.cota.org/customsites/cota/index.asp

- Congenital Heart Defects
  http://www.congenitalheartdefects.com/
- Congenital Heart Information Network
  http://www.tchin.org/
- Heart Center Encyclopedia
  http://www.cincinnatichildrens.org/health/heart-encyclopedia/default.htm
- Heart Institute for Children
  http://www.thic.com/Default.htm
- Kids with Heart, National Association for Children's Heart Disorders, Inc
  http://kidswithheart.org/
- United Hearts
  http://kidswithheart.org/

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Barbas KH, Kelleher DK. Breastfeeding success among infants with congenital heart disease. *Pediatr Nurs.* 30:285, 2004.

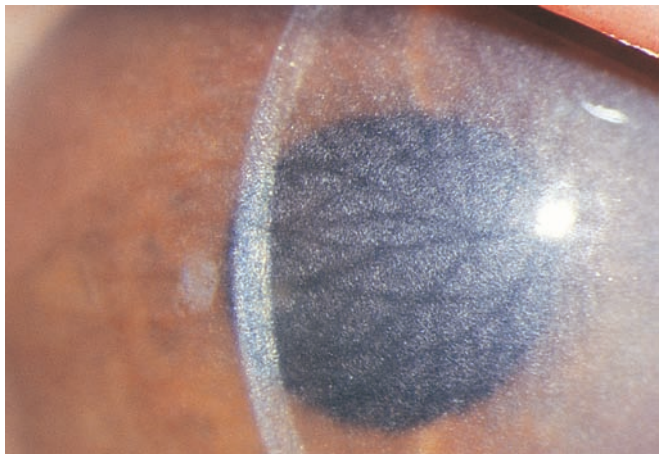
Children's Hospital Boston. Accessed April 27, 2009, at http://www.childrenshospital.org/az/Site2116/mainpageS2116P0.html.

Feingold B, Law YM. Nesiritide use in pediatric patients with congestive heart failure. *J Heart Lung Transplant.* 23:1455, 2004.

Owens JL, Musa N. Nutrition support after neonatal cardiac surgery. *Nutr Clin Pract.* 24:242, 2009.

CYSTINOSIS AND FANCONI'S SYNDROME

NUTRITIONAL ACUITY RANKING: LEVEL 3



Adapted from: Tasman W, Jaeger E. *The Wills Eye Hospital Atlas of Clinical Ophthalmology,* 2nd ed. Lippincott Williams & Wilkins, 2001.

DEFINITIONS AND BACKGROUND

In cystinosis, crystals of cystine are deposited throughout the body. If left untreated, the disease may lead to kidney failure. Toxic accumulations of copper in the brain and kidney account for neurological symptoms. Cystinosis may be inherited or acquired, such as by lead poisoning. Manifestations are also seen in hereditary fructose intolerance. Myopathy leads to restrictive lung disease in adults who have not received long-term cystine depletion. Whether or not oral cystamine therapy can prevent this complication remains to be determined (Anikster et al, 2001).

Infantile nephropathic cystinosis, the most severe form, is a lysosomal membrane transport defect. FTT, rickets, metabolic acidosis, unexplained glucosuria of renal tubular origin, loss of color in the retina of the eyes, and severe photophobia can appear as early as 3–18 months of age. In **intermediate cystinosis**, kidney and eye symptoms become apparent during the teenage years or early adulthood. Polyuria, growth retardation, rickets, acidosis, and vomiting are present. In **benign or adult cystinosis**, crystalline cystine accumulates primarily in the cornea of the eyes and adults may present with acidosis, hypokalemia, polyuria, or osteomalacia.

Fanconi's syndrome, a generalized tubular dysfunction, can be either acquired or inherited. The hereditary form may accompany Wilson's disease, galactosemia or glycogen storage diseases. Nephrotoxic drugs, such as use of some chemotherapy agents, streptozocin, antiretrovirals, valproate, or outdated tetracycline, may cause the acquired form (Knorr et al, 2004). Vitamin D deficiency, myeloma, amlyoidosis, and heavy metal intoxication may also be triggers. Regardless of origin, Fanconi's syndrome results in multiple organ damage, with profound renal damage. Excessive urination (polyuria), excessive thirst (polydipsia), and severe hypokalemia occur. Renal transplantation may be needed.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: The affected gene in the inherited form is *CTNS*, located on chromosome 17, which codes for cystinosin.

Clinical/History

- Birth weight (infant or child)
- Present weight
- Length or Height
- Growth (%), head circumference
- Abnormal sensitivity to light (photophobia)
- Loss of color in the retina
- Rickets
- Dehydration
- Dysphagia
- Patchy brown skin
- Diet/intake history
- Polydipsia, polyuria

Lab Work

- Gluc
- Ca^{++}, Mg^{++}
- Aminoaciduria

Serum phosphorous (decreased)	K^+ (decreased)	I & O
	CO_2	Uric acid (decreased)
Phosphaturia	Alb	BUN, Creat
Aminoaciduria	H & H	Ceruloplasmin
Na^+	Serum Fe	WBCs
	Serum vitamin D	

INTERVENTION

OBJECTIVES

- Remove the offending nephrotoxin in the acquired forms.
- Prevent bone demineralization and kidney failure. Correct hypokalemia, hypophosphatemia, and vitamin D insufficiency.
- Manage swallowing dysfunction.
- Support growth, which tends to be stunted in children.
- Prevent or delay corneal damage.
- Provide sufficient volumes of fluid and supplemental nutrients.
- Prepare for renal transplantation if needed. Postoperatively, promote wound healing and prevent graft rejection.

FOOD AND NUTRITION

- Use a diet low in cystine, with protein-free diet, PFD1 or PFD2 from Mead Johnson.
- Provide sufficient fluid intake. Input and output should be checked by standards for age.
- Supplement with vitamin D_3 (cannot convert 25-dihydroxycholecalciferol); give phosphate and calcium as appropriate. Bicarbonate is also needed.
- Provide sufficient sodium and potassium replacements.
- Alter consistency (liquids, solids) as needed.
- Prepare for wound healing with sufficient vitamins A, C, zinc, protein, and energy.

Common Drugs Used and Potential Side Effects

- Sodium bicarbonate or citrate should be used to correct acidosis. Take separately from iron supplements. Edema can occur.

SAMPLE NUTRITION CARE PROCESS STEPS

Inadequate Vitamin and Mineral Intakes

Assessment Data: Weight and growth charts, lab reports showing losses of K^+ and phosphorus in the urine, evidence of rickets (bowed legs).

Nutrition Diagnoses (PES): Inadequate mineral intake (potassium and phosphorus) related to excessive urinary losses from cystinosis as evidenced by urine tests, insufficient vitamin D metabolism and rickets.

Intervention: Educate parents about sources of vitamin D, potassium and phosphorus from medications and diet.

Monitoring and Evaluation: Weight records, growth, labs for potassium, phosphorus, and vitamin D.

- K depletion may require replacement therapy with a K-containing salt.
- Cysteamine (Cystagon), administered orally, halts glomerular destruction and decreases cystine content in cells. It mitigates morbidity and death (Gahl et al, 2007).
- Long-term GH treatment can be safe and effective; it should be started early in the course of the disease (Wuhl et al, 2001).

Herbs, Botanicals, and Supplements

- Herbs and botanicals should not be used for this condition because there are no controlled trials to prove efficacy.
- Use of Chinese herbs may be problematic, causing some forms of cystinosis in susceptible individuals. Discourage use.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Emphasize the importance of correcting fluid and electrolyte imbalances.
- Discuss any necessary changes in consistency to assist with dysphagia.
- Discuss diet for managing renal failure if necessary.
- If transplantation is needed, discuss guidelines for managing side effects such as graft–host resistance.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants to decrease potential risk of botulism.

For More Information

- American Foundation for Urologic Disease
  http://www.afud.org
- Cystinosis Central
  http://medicine.ucsd.edu/cystinosis/Index.htm\
- Cystinosis Foundation
  http://www.cystinosisfoundation.org/
- Cystinosis Research Foundation
  http://www.cystinosis.org/

CYSTINOSIS AND FANCONI'S SYNDROME—CITED REFERENCES

Anikster Y, et al. Pulmonary dysfunction in adults with nephropathic cystinosis. *Chest.* 119:394, 2001.

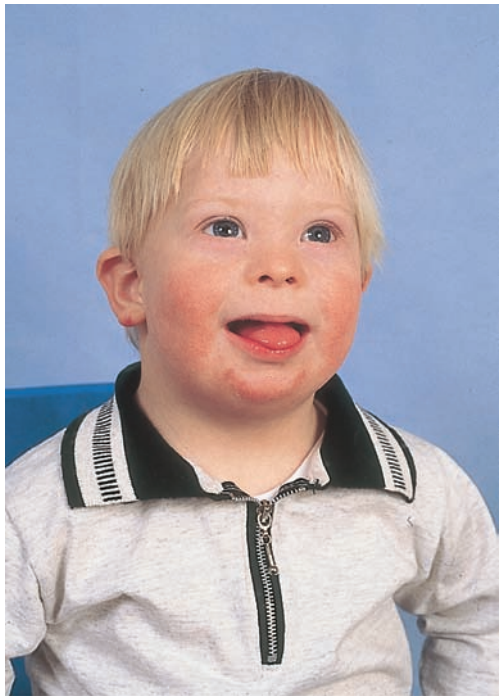
Gahl WA, et al. Nephropathic cystinosis in adults: natural history and effects of oral cysteamine therapy. *Ann Int Med.* 144:242, 2007.

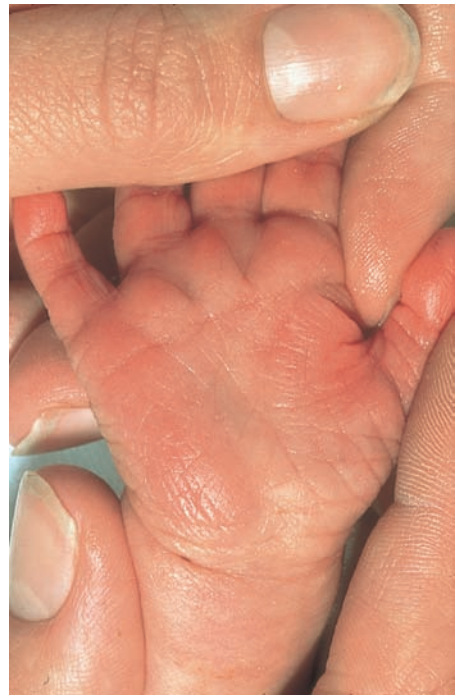
Knorr M, et al. Fanconi syndrome caused by antiepileptic therapy with valproic acid. *Epilepsia.* 45:868, 2004.

Wuhl E, et al. Long-term treatment with growth hormone in short children with nephropathic cystinosis. *J Pediatr.* 138:880, 2001.

DOWN SYNDROME

NUTRITIONAL ACUITY RANKING: LEVEL 2





DEFINITIONS AND BACKGROUND

DS is a congenital defect in which patients carry an altered chromosome; trisomy patients have an extra chromosome 21. Children with DS have short stature, decreased muscle tone, constipation, intestinal defects, weight changes, and mental retardation. There is a higher risk for congenital heart disease, gum disease, celiac disease, Hirschsprung's disease, hypothyroidism, leukemia, respiratory problems, and gastroesophageal reflux. Incidence of the syndrome is often related to maternal age.

Chronic oxidative stress is a consideration; antioxidants, such as selenium, vitamins C and E, are important to include. Research also suggests that folate has a relationship. Women of childbearing age should consume 400 mg folic acid daily though food sources and/or supplementation. (Czeizel and Puho, 2005).

Compared with other individuals, those with DS may have lower levels of vitamin A, thiamin, folate, vitamin B_{12}, vitamin C, magnesium, manganese, selenium, zinc, carnitine, carnosine, and choline; excesses of copper, cysteine, phenylalanine (Phe), and superoxide dismutase are sometimes encountered (Thiel and Fowkes, 2004). Disorders of metabolism involving vitamin B_6, vitamin D, calcium, and tryptophan may play a role (Thiel and Fowkes, 2004).

The use of chromosome engineering to generate trisomic mouse models and large-scale studies of genotype–phenotype relationships in patients will contribute to the future understanding of DS (Wiseman et al, 2009). First-trimester screening is generally recommended.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: DS is caused by trisomy of chromosome 21 (Hsa21). There are other alleles that may have an impact.

Clinical/History

- Length or height
- Birth weight
- Present weight
- BMI
- Diet/intake history
- Head circumference
- DS growth chart
- Growth (%)
- Hyperextensibility of joints
- History of prematurity?
- Large tongue, eye slant
- Endocardial defects
- Developmental delay
- Small nose with flat bridge
- Pica
- Skin prick test

Lab Work

- Gluc
- Uric acid (increased)
- Plasma zinc
- Chol, Trig
- Na^+, K^+
- Ca^{++}, Mg^{++}
- I & O
- Serum folate

SAMPLE NUTRITION CARE PROCESS STEPS

Overweight

Assessment Data: Weight and growth charts, BMI >normal range.

Nutrition Diagnoses (PES): NC 3.3. Overweight related to inadequate energy expenditure in DS as evidenced by BMI >28, limited activity levels, and frequent consumption of high fat foods and snacks.

Intervention: Discuss differing growth patterns from usual which may lead to excessive weight gain. Discuss optimal nutrition goals and physical activity, encouraging plenty of daily activity. Review foods to avoid because of risks for choking.

Monitoring and Evaluation: Weight records, growth and improved BMI levels, tolerance for various foods and consistencies.

INTERVENTION

OBJECTIVES

- Provide adequate energy and nutrients for growth. Short stature is not caused by nutritional deficiencies; use appropriate DS growth charts.
- Monitor introduction of solid food, which may be delayed. Fruits and vegetables may not be consumed in adequate amounts.
- To avoid lowered intake of vitamins and minerals, treat obesity in children with DS with a balanced diet plus vitamin and mineral supplements; no energy restriction; and an increase in physical activity.
- Assist with feeding problems; tongue thrust and poor suck are common.
- Reduce emotional problems that lead to overeating. Overfeeding should be avoided. Use proper positioning.
- Manage constipation, diarrhea, gluten enteropathy, urinary tract infections (UTIs), gum and periodontal diseases, which are common. Prevent osteoporosis and bone disease.

FOOD AND NUTRITION

- Supply adequate amounts of energy for age; for children aged 5–11 years, use 14.3 kcal/cm for girls and 16.1 kcal/cm for boys (Lucas, 2004, p. 41).
- Use protein according to age-dependent dietary reference intakes.
- Use a gluten-free diet if celiac disease is present (Hill et al, 2005).
- Monitor pica, overeating, and idiosyncrasies.
- Provide supplemental sources of folate, vitamin A, vitamin E, zinc, iron, and calcium if intake of fruits, vegetables, meats, dairy products, or whole grains is limited.
- Provide feeding assistance if needed. Tube feed if the patient is unable to eat orally; gradually wean to solids when possible.
- Provide extra fluid for drooling, diarrhea, or spillage.
- Encourage complex carbohydrates, prune juice, etc., if constipation is a problem.

Common Drugs Used and Potential Side Effects

- Aricept may have some benefit in individuals with DS. Nausea or diarrhea are sometimes side effects.
- For MTHFR alleles, products such as L-methylfolate (Deplin) may be given.

Herbs, Botanicals, and Supplements

- Herbs and botanicals should not be used; because there are no controlled trials to prove efficacy.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Explain feeding techniques that may be beneficial. Discuss use of self-feeding utensils if needed.
- Help control energy intake and physical activity for appropriate levels.
- Never rush mealtime. Encourage socialization.
- Discuss how growth patterns differ from usual; FTT (Krugman and Dubowitz, 2003) or excessive weight gain may result as the child grows older.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature >2 hours.
- Do not use honey in the diets of infants to decrease potential risk of botulism.

For More Information

- Down Syndrome
  http://www.nas.com/downsyn/
- Down Syndrome Quarterly
  http://www.denison.edu/dsq
- Drexel University—Down Syndrome Growth Charts
  http://www.growthcharts.com/
  http://www.growthcharts.com/charts/DS/charts.htm
- National Association for Down Syndrome
  http://www.nads.org/
- National Down Syndrome Society
  http://www.ndss.org/content.cfm
- Special Olympics
  http://www.specialolympics.org/Special+Olympics+Public+Website/default.htm

DOWN SYNDROME—CITED REFERENCES

Czeizel AE, Puho E. Maternal use of nutritional supplements during the first month of pregnancy and decreased risk of Down's syndrome: case-control study. *Nutrition.* 21:698, 2005.

Hill ID, et al. Guideline for the diagnosis and treatment of celiac disease in children: recommendations of the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition. *J Pediatr Gastroenterol Nutr.* 40:1, 2005.
Krugman SD, Dubowitz H. Failure to thrive. *Am Fam Physician.* 68:879, 2003.
Lucas B, ed. *Children with special care needs: nutrition care handbook.* Chicago: The American Dietetic Association, 2004.
Thiel RJ, Fowkes SW. Down syndrome and epilepsy: a nutritional connection? *Med Hypotheses.* 62:35, 2004.
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FAILURE TO THRIVE

NUTRITIONAL ACUITY RANKING: LEVEL 4

DEFINITIONS AND BACKGROUND

FTT is a diagnostic term used to describe infants and children who fail to grow and develop at a normal rate; it indicates protein, energy, vitamin, and mineral insufficiency. In many pediatric centers, one third of the referred children are malnourished. FTT is a complex problem that can be caused by many medical or social factors; without treatment, chronic illnesses or death may ensue.

Prompt diagnosis and intervention are important for preventing malnutrition and developmental delays. Careful attention must be paid to growth charts and medical histories (Krugman and Dubowitz, 2003). Food refusal, poor feeding, vomiting, gagging, irritability, and FTT are commonly found in both infantile feeding disorders (IFD) and common treatable medical conditions (Levy et al, 2009).

Weight is the most reliable marker for FTT. According to the American Academy of Pediatrics (2004), FTT is established when weight (or weight/height) is less than 2 standard deviations below the mean for sex and age and/or the weight curve has dropped more than 2 percentile lines on the National Center for Health Statistics growth charts after a previously stable pattern. Other indices include a small head circumference, muscular wasting, apathy, weight loss, or poor weight gain. Learning failure (e.g., slow to talk, behavior problems) can occur. Infants with DS, intrauterine growth retardation (IUGR), or premature birth follow different growth patterns than usual; monitor carefully to evaluate for FTT (Krugman and Dubowitz, 2003). About 25% of normal infants will shift to a lower growth percentile in the first 2 years of life and then remain at that percentile; this is not FTT (Krugman and Dubowitz, 2003).

Primary FTT originates from social/environmental deficits, inadequate feeding procedures, or caretaker behaviors. Adolescent mothers may need a lot of support and education. Proximity and touch are especially disturbed in feeding disorders (i.e., mothers provide less touch that supports growth), and children demonstrate signs of touch aversion (Feldman et al, 2004). Early interventions by trained home visitors may promote a more nurturing environment and reduce developmental delays.

Secondary FTT originates from some disease states (e.g., cancer, allergies, chronic infections, cystic fibrosis, cleft lip or palate, DS, or other physical or mental disability). Growth failure plus fever of unknown origin and anemia in older children or teens may suggest the onset of Crohn's disease; evaluation is recommended. About half of the causes of FTT are organic; the other 50% are from inorganic causes. The American Dietetic Association suggests at least five medical nutrition therapy visits for infants and children with FTT.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: Genes causing FTT would be related to the specific condition, such as s congenital heart defect, spina bifida, DS, cystic fibrosis.

Clinical/History

- Height
- Very low birth weight?
- Apgar scores
- Premature or small for gestational age (SGA)?
- Current or goal weights
- Growth grid
- Percent height for age (actual height/expected height)
- Head circumference; microcephaly?
- Skinfold thickness
- Diet and intake history
- Feeding schedule and timing
- Food allergy, especially milk allergy
- GERD
- Medical history
- Breastfed or bottle fed?
- Solid food introduction pattern
- Diarrhea or vomiting?
- Constipation
- Dehydration
- Inadequate access to food?
- Infections, parasites?
- Frequent UTIs
- Maternal depression?

Lab Work

- H & H
- Serum Fe, ferritin
- Anemia (iron, sickle cell, other)
- Alb
- Gluc
- Chol, Trig
- BUN
- Thyroid function tests
- I & O
- Sweat test

INTERVENTION

OBJECTIVES

- All children with FTT need additional calories for catch-up growth at about 150% of the energy requirement for

SAMPLE NUTRITION CARE PROCESS STEPS

FTT

Assessment Data: Weight and growth charts, medical conditions causing excessive energy expenditure or requirements, feeding methods used for the child, available financial resources to buy food or formula, access to safe and sufficient food supply.

Nutrition Diagnoses (PES): NI 2.1. Inadequate oral food/beverage intake related to minimal intake of formula and age-appropriate foods as evidenced by drop in more than 2 percentile lines on the National Center for Health Statistics growth chart after having achieved a previously stable pattern and medical Dx of FTT.

Interventions:

Food and Nutrition Delivery:

ND 5.7. Feeding environment to support growth in 12-month-olds.

ND 1.2. To follow increased caloric intake and frequent snacks.

Nutrition Education:

E 2.2. RD to provide nutrition counseling to support weight gain in patient and teach mother and caretaker how to properly feed according to infant's nutritional needs. Discuss adequate timing for feeding child. Educate and teach mother about appropriate feeding behaviors and practices for 12-month-olds.

Counseling:

C 2. Counsel mother/caretaker on how to provide nutritional needs for patient and environment to support those need. Goal is to achieve daily gains of weight, 30 g/d.

Coordination of Care:

RC 1.3. RD to collaborate with MD; RD will refer patient for in-home assessment and follow-through. Correct environmental causes of FTT. Refer to WIC or SNAP (food stamps) programs to help with financial challenges and food insecurity.

Monitoring and Evaluation: Weight records, growth, tolerance for various foods or formulas, financial access to food.

their expected, not actual, weight (Krugman and Dubowitz, 2003). Use calculations for determining needs from the most current *Pediatric Manual of Clinical Dietetics.*

- Identify and correct etiologies such as decreased energy intake, increased nutrient losses, increased metabolic demands. Determine if malnutrition is primary (from faulty feeding patterns or dietary inadequacy) or secondary (from disease process interfering with intake).
- Teach the parent or caretaker how to properly feed and how to determine needs. Advise parents to support nurturing during feeding.
- Provide the most optimal nutrition compatible with a normal growth pattern. Achieve daily gains of 30 g for young infants; extra may be desirable for catch-up. Nutrient-enriched formulas are probably not necessary (Henderson et al, 2007).
- Provide a schedule of feeding for infant's age to support catch-up growth and improved brain development (Powers et al, 2008). See Table 3-7 for more details.

FOOD AND NUTRITION

- Conduct a thorough nutrition assessment and acquire actual intake records when possible. Evaluate the child's nutritional history and growth in comparison with the percentiles of other same-age children. If special growth charts are needed, use those instead (as for DS). Discuss findings with parent or caretaker.
- Calculate energy and protein needs carefully. While not easy to do, indirect calorimetry may be needed.
- Check recommended intakes for all nutrients. Provide adequate zinc and vitamin B_6, as determined by the infant's age; 120–130% is a common practice.
- Monitor growth (weight) weekly; feeding behaviors.
- If the infant is dehydrated, provide adequate amounts of fluids. However, FTT can be aggravated by excessive consumption of fruit juice and sweetened beverages (often 12–30 oz daily) which may replace other nutrient-dense foods. Limit to 4–6 oz daily until overall diet quality and growth rate have improved.
- Provide meals and snacks at scheduled times; support a comfortable social and emotional environment. Family meals and allowing children to be a part of meal preparation are also important.
- If FTT children are strictly vegan, monitor for vitamins B_{12}, D, B_6, iron, zinc, and calcium deficiencies.
- Tube feeding may be useful as a supplemental or alternative feeding method; nightly feeding is an effective recommendation if it can be managed by the caregiver.

Common Drugs Used and Potential Side Effects

- Evaluate medications given for any reason to determine if some or all affect nutritional intake. Adjust diet as needed.
- Endogenous cannabinoids or other appetite enhancers are being studied for their safety and effectiveness in FTT.

Herbs, Botanicals, and Supplements

- Herbs and botanicals should not be used for FTT; there are no controlled trials to prove efficacy.
- Probiotics are useful for their live micro-organisms with a health benefit for GI disorders, cancer, infant allergies, FTT, and infections (Brown and Valiere, 2004).
- Zinc supplementation may be needed during catch-up growth in malnourished children (Castillo-Duran and Weisstaub, 2003). Avoid prolonged or excessive doses.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Describe appropriate nutritional intake according to age and any predisposing medical conditions.
- Encourage the use of appropriate growth charts at home to monitor success. Develop a progress chart for developmental milestones. Growth spurts follow sustained weight gains; monitor growth frequently.

TABLE 3-7 Grading and Treatment for Hirschsprung's-Associated Enterocolitis

Grade	Clinical Symptoms
I	Mild explosive diarrhea, mild or moderate abdominal distention; no systemic manifestations
II	Moderate explosive diarrhea, moderate-to-severe abdominal distention; mild systemic symptoms
III	Severe explosive diarrhea, marked abdominal distention, and shock or impending shock
Fiber enhancement	
Breakfast	Whole wheat waffles with fresh fruit. Cereal choices: oatmeal, Frosted Mini Wheats, Kashi Mighty Bites, Raisin Bran, Wheat Chex. Whole wheat bagel, or whole wheat English muffin with chunky peanut butter. Bran muffins. Dried fruit to increase fiber content. Add fresh fruit
Lunch or dinner	Vegetable soup with whole wheat crackers. Whole wheat sandwich with leaf lettuce, tomato, meat of choice. Whole wheat macaroni and cheese with peas. Whole wheat spaghetti with sautéed zucchini and tomatoes in sauce. Fresh fruit. Whole wheat pizza with sauce, cheese, and vegetable toppings (green, red, yellow or orange peppers, mushrooms, tomatoes, olives). For tacos, use whole wheat tortillas, add vegetables (tomatoes, lettuce, olives, avocado). Brown rice, whole wheat pasta, legumes, beans; add a vegetable as a side. Green, red, yellow or orange peppers and cucumber slices with vegetable dip. Sliced pears, peaches, strawberries, or cantaloupe with fruit dip. Celery, 2 Tbsp peanut butter, raisins. Whole grain crackers

Fiber supplements:

Name	Active Ingredient	Serving Size	Amount of Fiber
Metamucil wafers	Psyllium 50% soluble	2 wafers	6 g
Metamucil powder	Psyllium 65% soluble	1 tbsp	3 g
Ground flax seed	45% insoluble, 55% soluble	1 tbsp	3 g
Benefiber	Wheat dextrin, 100% soluble	2 tbsp	3 g
Citrucil	Methocellulose, 100% soluble	1 scoop or 4 caplets	2 g
Pectin	100% soluble	1.75 oz package	4.3 g

Sources:

1. American Dietetic Association. *Fiber facts: soluble fiber and heart disease*. Chicago: American Dietetic Association, 2007.
2. Children's Hospital of Boston. Accessed April 27, 2009, at http://www.childrenshospital.org/az/Site2116/mainpageS2116P0.html.
3. Li BW, Andrews KW, Pehrsson PR. Individual sugars, soluble, and insoluble dietary fiber contents of 70 high consumption foods. *J Food Comp Anal.* 15:715–723, 2002.

- Offer simple, specific instructions when needed, such as mechanics of breastfeeding and typical intakes for children of same age. If formula is used, improper mixing of formula is common; help correct any misunderstandings.
- Discuss nutrient density (e.g., milk vs. sweetened carbonated beverages; whole fruit vs. juice).
- Explain proper use of over-the-counter vitamin–mineral supplements, age-appropriate for the child.
- Address any harmful or unusual dietary beliefs (Feld and Hyams, 2004).
- Practical suggestions should be offered regarding nurturing and emotional support for the child. Parenting classes may be beneficial.
- Coordinate referral to child welfare services where neglect is suspected (Block et al, 2005). Refer to WIC programs, La Leche League, SNAP (food stamps) whenever appropriate.
- Follow-up should be provided at outpatient clinics or by home visits.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants; this will decrease the potential risk of botulism.

For More Information

- Kids Health
  http://kidshealth.org/parent/nutrition_fit/nutrition/failure_thrive.html
- Medline—FTT
  http://www.nlm.nih.gov/MEDLINEPLUS/ency/article/000991.htm

FAILURE TO THRIVE—CITED REFERENCES

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Castillo-Duran C, Weisstaub G. Zinc supplementation and growth of the fetus and low birth weight infant. *J Nutr.* 133:1494S, 2003.

Feld LG, Hyams JS, eds. *Growth assessment and growth failure. Consensus in pediatrics.* Evansville, IN: Mead Johnson & Company, 2004.

Feldman R, et al. Mother-child touch patterns in infant feeding disorders: relation to maternal, child, and environmental factors. *J Am Acad Child Adolesc Psychiatry.* 43:1089, 2004.
Henderson G, et al. Nutrient-enriched formula versus standard term formula for preterm infants following hospital discharge. *Cochrane Database Syst Res.* 17(4):CD004696, 2007.
Krugman SD, Dubowitz H. Failure to thrive. *Am Fam Physician.* 68:879, 2003.
Levy Y, et al. Diagnostic clues for identification of nonorganic vs organic causes of food refusal and poor feeding. *J Pediatr Gastroenterol Nutr.* 48:355, 2009.
Powers GC, et al. Postdischarge growth and development in a predominantly Hispanic, very low birth weight population. *Pediatrics.* 122:1258, 2008.

FATTY ACID OXIDATION DISORDERS

NUTRITIONAL ACUITY RANKING: LEVEL 4

DEFINITIONS AND BACKGROUND

Fatty acid oxidation disorders disrupt mitochondrial energy generation and ketone production (Isaacs and Zand, 2007). Muscle protein breaks down and may lead to death if the heart muscle is involved. The fatty acid disorders with dietary implications include medium-chain acyl-CoA dehydrogenase deficiency (MCAD), long-chain 3-hydroxyacyl-CoA dehydrogenase (LCHAD) deficiency, and very long chain acyl-CoA dehydrogenase deficiency (Isaacs and Zand, 2007).

MCAD is caused by the lack of an enzyme required to convert fat to energy. Children with MCAD cannot use MCTs to make energy, so the body begins to malfunction when they fast (i.e., they have no more long-chain dietary fats available from the diet). MCAD occurs in approximately one in every 10,000 live births. MCAD occurs mostly among Caucasians of Northern European background. Symptoms typically begin in infancy or early childhood, often with simple lethargy. While some affected individuals have no symptoms at birth, disorders such as hypoglycemia, seizures, coma, brain damage, or cardiac arrest can occur very quickly with illness. If not detected and treated appropriately, MCAD can result in death. About 1 in 100 sudden infant death syndrome (SIDS) deaths are probably a result of undiagnosed MCAD (Nennstiel-Ratzel et al, 2005).

Early detection allows treatment and a normal life expectancy. Medical nutrition therapy to lower dietary fats does not decrease toxic metabolites because the body can make triglycerides from carbohydrates, proteins, or fats (Isaacs and Zand, 2007). The appropriate fatty acids must be omitted.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: MCAD involves medium-chain acyl-coenzyme A (CoA) dehydrogenase deficiency; adenosine replaces guanosine at position 985 of the MCAD gene.

Clinical/History	Seizures?	Trig
Length (height)	Retardation?	Lipid Panel
Birth weight		H & H
Present weight	**Lab Work**	Serum Fe
Growth (%)	Gluc	
Diet/intake history	Alb	
	Chol	

INTERVENTION

OBJECTIVES

- Avoid periods of fasting, day and night (Roe and Ding, 2001). Use IV glucose when food cannot be tolerated, such as with colds or flu.
- Customize protocol for the individual. LCHAD requires a severe dietary restriction of long-chain fats, to the lowest level that can deliver the EFAs and fat-soluble vitamins (Isaacs and Zand, 2007). MCT can be used in LCHAD but not in MCAD.
- Provide EFAs.

FOOD AND NUTRITION

- Restrict periods of fasting by offering small, frequent feedings (Oey et al, 2005).

SAMPLE NUTRITION CARE PROCESS STEPS

Excessive Intake of Types of Fats

Assessment Data: Weight and growth charts.

Nutrition Diagnoses (PES): Excessive intake of medium chain fatty acids related to MCAD deficiency as evidenced by signs of lethargy and elevated levels of triglycerides.

Intervention: Educate parents about avoiding sources of MCT; document in medical record about formulas to avoid.

Monitoring and Evaluation: Weight records, growth, improvement in lipid levels, reduced lethargy, normal mental development for age.

- A diet with avoidance of the specific, problematic fatty acids will be needed. For example, do not use enteral formulas that contain MCTs in MCAD.
- The diets will be higher in carbohydrates and fat-free protein foods.
- Supplement linoleic and a-linolenic acids; monitor by laboratory measurements of fatty acids.
- Supplemental carnitine has been recommended.
- Waking the child at least once during the night, or feeding by gastrostomy or NG tube overnight, is required for most of the fatty acid oxidation disorders (Isaacs and Zand, 2007).
- Monitor weight and growth closely to prevent obesity, but do not skip meals or feedings.

Common Drugs Used and Potential Side Effects

No specific drugs are used. Dietary alterations are the management.

Herbs, Botanicals, and Supplements

- Herbs and botanicals should not be used for MCADD because there are no controlled trials to prove efficacy for any related problems.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Educate about the dangers of fasting, including periods during illness.
- Share information about frequent feedings and how to avoid the designated fatty acids from supplemental products, formulas, etc.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants to decrease potential risk of botulism.

For More Information

- Fatty Oxidation Disorder
  http://www.fodsupport.org/
- MCAD
  http://www.mcadangel.com/mcad-links.html
- National Newborn Screening
  http://genes-r-us.uthscsa.edu/

FATTY ACID OXIDATION DISORDERS—CITED REFERENCES

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FETAL ALCOHOL SYNDROME

NUTRITIONAL ACUITY RANKING: LEVEL 1–2

Adapted from: Sadler T, PhD. *Langman's Medical Embryology,* 9th ed. Image Bank. Baltimore: Lippincott Williams & Wilkins, 2003.

DEFINITIONS AND BACKGROUND

Generally noted shortly after birth, fetal alcohol syndrome (FAS) is a syndrome in infants with developmental delay, ocular anomalies, LBW, tremors, short stature, retarded intellect, seizures, and microcephaly. There is a continuum of FAS recognized as fetal alcohol spectrum disorders (FASD).

FAS is the third leading cause of mental retardation in the United States; it is the most preventable (Centers for Disease Control and Prevention, 2004).

No level of alcohol consumption during pregnancy is safe (Centers for Disease Control and Prevention, 2004). Exposure to alcohol during brain development can permanently alter the physiology of the hippocampal formation, thus promoting epileptic activity and depression (Bonthius et al, 2001). Disrupted cholesterol homeostasis may contribute

to neurotoxicity; the developing brain requires cholesterol for proper cell proliferation (Guizzetti and Costa, 2005). Approximately 10% of pregnant women use alcohol and 2% engage in binge drinking or frequent use of alcohol (Centers for Disease Control and Prevention, 2004).

Acetaldehyde damages RNA (Eriksson, 2001; Wang et al, 2009). The steady concurrent use of tobacco and alcohol by young women emphasizes the need for enhanced efforts to reduce initial tobacco and alcohol use by young people. Women who report abuse of tobacco or alcohol should be evaluated for abuse of both substances, and interventions should address abuse of both substances, especially to prevent FAS (Ebrahim et al, 2000).

Early risk assessment is needed, though it may be difficult to find and treat children who have FASD. Using the combination of weight and head circumference below the 10th percentile at birth is useful for identifying children at substantial risk for growth and developmental delays (Weiss et al, 2004). Children with FAS may have more social and medical needs. They often have more facial dysmorphology, growth deficiency, central nervous system dysfunction, muscular problems, hospitalizations for OM, pneumonia, dehydration, and anemia (Kvigne et al, 2004).

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: FAS is considered to be environmental.

Clinical/History

- Birth weight
- Current weight (often below 10th percentile)
- Length
- Growth (%)
- Diet/intake history
- Head circumference (<10th percentile)
- Seizures
- Physical growth delay
- Functional deficits (motor, social, memory, etc.).

Lab Work

- Alb
- Na^+, K^+
- Gluc
- H & H
- Serum Fe
- Ca^{++}, Mg^{++}
- Serum folate

INTERVENTION

OBJECTIVES

- Promote effective family coping skills and effective parental bonding.
- Prevent additional retardation or developmental delays, blindness, other complications.
- Improve intake and nutritional status.
- Prevent or correct vomiting, cardiac symptoms, other problems.
- Encourage normal growth patterns; prevent FTT.

SAMPLE NUTRITION CARE PROCESS STEPS

Inadequate Energy Intake

Assessment Data: Weight and growth charts, head circumference below 10% tile.

Nutrition Diagnoses (PES): Inadequate energy intake related to increased energy demands from FAS as evidenced by microcephaly (head circumference below 10% tile) and slow growth rate for age.

Intervention: Educate parents about enhancing energy intake through nutrient-dense foods and supplements.

Monitoring and Evaluation: Weight records, growth, head circumference more closely normal for age.

FOOD AND NUTRITION

- Provide a diet appropriate for age and status. Ensure adequate protein and energy for catch-up growth.
- If necessary, provide tube feeding or TPN while hospitalized. Some infants may require additional nutrition support in the home setting to promote better growth and development.

Common Drugs Used and Potential Side Effects

- Anticonvulsants may be needed to correct seizures. Monitor for depletion of vitamins C, D, B_6, B_{12}, and K, folic acid, and calcium.

Herbs, Botanicals, and Supplements

- Herbs and botanicals should not be used for FAS because there are no controlled trials to prove efficacy.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Discuss appropriate feeding techniques for age of infant.
- Discuss importance of diet in aiding normal growth and development.
- Encourage mother's participation in alcohol rehabilitation if needed. Discuss her future plans for additional pregnancies and encourage counseling to avoid continued alcohol intake during that time.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.

- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants; this will decrease potential risk of botulism.

For More Information

- CDC–Division of Birth Defects and Developmental Disabilities, FAS Site http://www.cdc.gov/ncbddd/fas/
- CDC Diagnosis and Referral Guide http://www.cdc.gov/ncbddd/fas/documents/FAS_guidelines_accessible.pdf
- Fetal Alcohol and Drug Unit http://depts.washington.edu/fadu
- Fetal Alcohol Syndrome Handbook http://www.usd.edu/cd/publications/fashandbook.cfm
- Fetal Alcohol Syndrome websites http://www.come-over.to/FAS/faslinks.htm
- FAS Community Resource Center http://www.come-over.to/FASCRC/
- National Center for Family Support http://www.familysupport-hsri.org/
- National Clearinghouse for Alcohol and Drug Information (NCADI) http://www.health.org/
- National Council on Alcoholism and Drug Dependence (NCADD) http://www.ncadd.org/
- National Organization of Fetal Alcohol Syndrome http://www.nofas.org/

FETAL ALCOHOL SYNDROME—CITED REFERENCES

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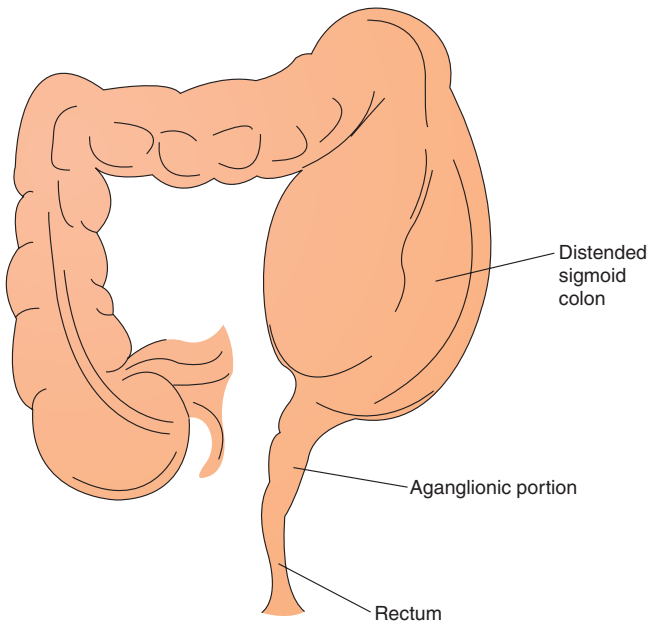
Weiss M, et al. The Wisconsin Fetal Alcohol Syndrome Screening Project. *WMJ.* 103:53, 2004.

HIRSCHSPRUNG'S DISEASE (CONGENITAL MEGACOLON)

NUTRITIONAL ACUITY RANKING: LEVEL 4



DEFINITIONS AND BACKGROUND

Hirschsprung's disease (HD) is characterized by the absence of ganglion cells and the presence of hypertrophic nerve trunks in the distal bowel. HD is also known as jejunal gangliosus or congenital megacolon. Its incidence is 1 in 5000 live births. HD may reoccur in other babies born to the same family with a child with HD (Stewart and von Allmen, 2003).

As a congenital malformation, HD interferes with normal mass peristalsis and functional obstruction. Normally, ganglia stimulate the gut and allow peristalsis to occur. In HD, the ganglia are missing, and segments of bowel become obstructed. This creates abdominal distention, failure to pass meconium stool, vomiting, and constipation. If diagnosed when older, growth failure may be a presenting sign.

Surgical removal may be required to alleviate bowel obstruction, followed by a temporary colostomy. Complications after a definitive pull-through procedure for HD include stricture formation, enterocolitis, and occasionally, wound infection (Finck et al, 2001). Often, removal of the affected area and reconnection of the colon occurs at age 6 months or older.

Over the long term, one in five patients will have continued constipation, occasional soiling, and incontinence, and one in 10 patients may have severe problems. A special type of enterocolitis, HD-associated enterocolitis (HAE), may also be a concern (Nofech-Mozes et al, 2004). The condition can be life threatening, and signs include hypoalbuminemia, diarrhea and vomiting, and anorexia and weight loss. See Table 3-8.

TABLE 3-8 Glycogen Storage Diseases (GSDs): Deficiency of a Glycogen Synthase That Normally Converts Glycogen to Glucose

Disease	Description
GSD1: glucose-6 phosphatase deficiency (G6PD), von Gierke disease	Slow or stunted growth, enlarged liver, delayed or absent pubertal development, gout, kidney failure, and a poor ability to withstand fasting due to low blood sugar occur. Patients with this condition are prone to frequent infections, hemolytic anemia, and inflammatory bowel disease. Brain damage can result from low glucose availability (Melis et al, 2004). Early death was common. Portacaval shunt may be considered in patients with height for age <3rd percentile (Corbeel et al, 2000).
GSD 2: alpha-glucosidase deficiency, Pompe disease	Onset in infancy is the most severe; most patients present with hypotonia and cardiomyopathy. Recombinant human GAA (rhGAA) can be tested for enzyme replacement (Raben et al, 2005).
GSD 3: debrancher enzyme deficiency, Cori disease or Forbes disease	There may be low bone density and a high risk for osteoporosis (Cabrera-Abreu et al, 2004).
GSD4: brancher enzyme deficiency, Andersen disease	Glycogen branching enzyme (GBE) deficiency results in the accumulation of an amylopectin-like polysaccharide and presents with liver disease, progressing to cirrhosis (Bruno et al, 2004).
GSD5: muscle glucagon phosphorylase deficiency, McArdle disease	X-linked liver glycogenosis (XLG) is one of the most common forms; onset is often in adults. Low levels of phosphorylase result in abnormal storage of glycogen in muscle tissue, muscle pain, cramping, stiffness, and poor exercise tolerance. Avoid strenuous exercise.
GSD6: liver phosphorylase deficiency, Hers disease	Gross hepatomegaly and hypoglycemia occur with reduced liver phosphorylase activity.
GSD7: muscle phosphofructokinase deficiency, Tarui disease	This syndrome presents often with exertional myopathy and hemolytic syndrome.
GSD9 a: liver glycogen phosphorylase kinase deficiency	Growth retardation, abdominal distention, and hepatomegaly may be present (Schippers et al, 2003). Liver transplantation results in normal fasting glucose production and normal glucose and insulin concentrations.
GSD9b: β-subunit phosphorylase kinase Fanconi-Bickel syndrome	Hepatorenal glycogenosis is abnormal.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: HD develops before a child is born but is not thought to be genetic.

Clinical/History

Birth weight
Length
Present weight
FTT?
Growth (slow)
Diet/intake history
Failure to pass meconium after birth (newborn)
Watery diarrhea (newborn)
Constipation
Temperature (fever?)
Vomiting
Abdominal distention
Rectal bleeding?
Dehydration; I & O
Abdominal x-ray
Barium enema
Malabsorption
Enterocolitis?

Lab Work

H & H
Serum Fe, ferritin
Alb
Na^+, K^+
Ca^{++}, Mg^{++}
Gluc
LFTs

INTERVENTION

OBJECTIVES

- Provide adequate nutrition for the patient's age and development. Growth may be inhibited.
- Replace electrolytes and fluids, especially with diarrhea and enterocolitis.

SAMPLE NUTRITION CARE PROCESS STEPS

Altered GI Function

Assessment Data: Weight and growth charts, constipation and stool records.

Nutrition Diagnoses (PES):

NC 1.4. Altered GI function related to megacolon as evidenced by current complaints of constipation.

NB 1.7. Undesirable food choices related to lack of fruit and vegetable intake and increase in cookies and crackers in diet.

Intervention: Educate parents about high fiber foods and increased use of fruits, vegetables, whole grains and fluids.

Monitoring and Evaluation: Weight records, decreased symptoms of constipation, improved stooling pattern.

- Compensate for poor absorption of nutrients; water-miscible forms of fat-soluble vitamins may be needed.
- Prevent complications after surgery, especially constipation, incontinence, or enterocolitis.

FOOD AND NUTRITION

- Use a high-energy/high-protein diet. Enteral products, oral supplements, or TPN can be used.
- Monitor serum electrolytes, especially potassium, if laxatives are used. Encourage a diet high in fiber and fluid to wean off medication if possible.
- Provide fluids adequate for the patient's age, hydration status, and extra fluid requirements.
- Use a laxative diet (Cincinnati Children's, 2009); see Table 3-8.
- Provide TPN if large sections of the bowel are removed. Advance infant feedings as tolerated using human milk or preterm or standard infant formulas, and then gradually progress to soft/bland foods.
- Monitor calcium, magnesium, and other nutrients if long-term TPN is needed.

Common Drugs Used and Potential Side Effects

- Antibiotics may be needed if perforation has occurred or when there is enterocolitis. Monitor for side effects.
- In constipation, laxatives can deplete numerous nutrient reserves; monitor carefully. Encourage a diet high in fiber and fluid to wean off medication if possible.

Herbs, Botanicals, and Supplements

- Herbs and botanicals should not be used for megacolon because there are no controlled trials to prove efficacy.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Teach patient about sources of protein, energy, potassium, and other key nutrients from diet.
- Discuss wound healing or colostomy procedures after surgery.
- For constipation and bowel incontinence, a high-fiber diet may be useful; discuss signs and symptoms of obstruction to report immediately to a doctor. Initial suggestion: age plus 10, example 4 years + 10 = 14 g/d.
- Extra fluids will be needed with high-fiber intake.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants to decrease potential risk of botulism.

For More Information

- Hirschsprung's & Motility Disorders Support Network
  http://www.hirschsprungs.info/index.html
- International Foundation for Functional Gastrointestinal Disorders
  http://www.iffgd.org/
- National Digestive Diseases Clearinghouse
  http://digestive.niddk.nih.gov/ddiseases/pubs/hirschsprungs_ez/index.htm
- United Ostomy Association
  http://uoa.org/

HIRSCHSPRUNG'S DISEASE—CITED REFERENCES

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HIV INFECTION, PEDIATRIC

NUTRITIONAL ACUITY RANKING: LEVEL 4

DEFINITIONS AND BACKGROUND

There are unique considerations related to HIV infection in infants, children, and adolescents. With the use of highly active antiretroviral therapy (HAART), there is minimal mother-to-child transmission of HIV infection in developed countries (i.e., 1–2% only) (King et al, 2004; Newell and Thorne, 2004). In developed nations, HIV infection is more of a chronic disease, with extensive medications, costs, and side effects to consider. A high proportion of HIV-infected individuals are African or African American.

Developing nations still have a battle to address. Infants who are breastfed by HIV-infected mothers have the risk of acquiring the infection. In addition, HIV-infected mothers may transmit opportunistic pathogens to their infants. FTT and protein–calorie malnutrition are common. Every child with HIV infection should be assessed at baseline and every 4–6 months thereafter to determine risk of nutritional compromise. Severity or degree of nutritional risk is measured with anthropometric, biochemical, dietary intake, and medical data. Salivary gland disease is a common finding related to HIV infection; gland enlargement or xerostomia may present, and the reduction in saliva must be addressed (Pinto and DeRossi, 2004).

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: HIV infection is not genetic but is transmitted prenatally by the infected mother, or by contaminated needles or blood transfusions.

Clinical/History

- Height
- Weight
- Weight for height, BMI
- Growth percentile and pattern
- Diet/intake history; energy intake
- Head circumference (infants)
- Stunting
- FTT
- Mid-arm muscle circumference (MAMC)
- Opportunistic infections

Lab Work

- H & H
- Serum Fe
- Alb
- Na^+, K^+
- Ca^{++}, Mg^{++}
- Gluc
- Immunological status
- CD4+ T-cell counts
- Vitamin A level

SAMPLE NUTRITION CARE PROCESS STEPS

Inadequate Energy Intake

Assessment Data: Weight and growth charts, frequent infections.

Nutrition Diagnoses (PES): Inadequate energy intake related insufficient intake, diarrrheal losses, and high metabolic demand of HIV as evidenced by weight loss and frequent opportunistic infections.

Intervention: Educate parent/caregiver about use of tolerated high calorie foods and supplements.

Monitoring and Evaluation: Weight records, growth, tolerance of formulas or supplemental products.

INTERVENTION

OBJECTIVES

- Maternal factors, including Vitamin A level and CD4+ T-cell counts during pregnancy, as well as infant viral load and CD4+ T-cell counts in the first several months of life, can help identify those infants at risk for rapid disease progression who may benefit from early aggressive therapy.
- Achieve a normal growth pattern; allow for catch-up growth and monitor growth patterns closely.
- Prevent opportunistic infections by improving or maintaining immune status with good nutrition.
- Alleviate wasting syndrome, diarrhea, malabsorption, enteric infections, malnutrition, and immune deficiency. Preserve lean body mass.
- Follow the CDC evidence-based guidelines (CDC, 2009):
  1. Emphasize the important role of effective antiretroviral therapy in augmenting immune function
  2. Support diagnosis and management of immune reconstitution inflammatory syndrome, a condition in which the immune system begins to recover, but then responds aggressively to a previously acquired opportunistic infection
  3. Prevent Hepatitis B and C, *Mycobacterium tuberculosis* infection, malaria
  4. Manage drug–drug interactions and drug–nutrient interactions

FOOD AND NUTRITION

- Use a high-protein diet. Enteral products, oral supplements, and frequent snacks should be used if required. Protein needs may be 1.5–2 times the usual for age and gender.
- Energy needs may vary from 50% to 200% of the usual requirements. Children with severe encephalopathy may be bed bound and require fewer total calories.
- Assure adequacy of fluid intake, especially with many medications to be taken each day.
- A multivitamin supplement is needed to provide at least 100% of the daily needs. Poor absorption may be a problem for vitamins A, C, B_6, and B_{12}, folate, iron, selenium, and zinc. Calcium is needed to prevent loss of bone mass (O'Brien et al, 2001).
- Naturally occurring antioxidants are safe when consumed in normal amounts. For example, include nuts for vitamin E and selenium and citrus fruits for vitamin C. Be aware of excesses from pills and other supplemental forms because excesses can deplete immunity. Mega doses are up to 10 times the RDA (American Academy of Pediatrics, 2004) and have not proven to be of benefit.
- Aggressive nutritional support is critical. Nocturnal, continuous feedings may be useful.

Common Drugs Used and Potential Side Effects

- Few HIV medicines are produced in pediatric formulations. Those drugs available as syrups have limitations,

such as short shelf-life, objectionable taste, difficult measuring of correct doses, and expense. For a list of FDA-approved medications used in HIV infection, see Section 15.

- HIV-infected mothers may transmit opportunistic pathogens to their infants. There may be antibiotics or antiviral agents prescribed that should be closely monitored for nutritional and GI side effects. Adherence to complex antiretroviral (HAART) therapy requires addressing developmental, psychosocial, and family factors (Mellins et al, 2004). Early treatment saves lives.

Herbs, Botanicals, and Supplements

- Herbs and botanicals should not be used for HIV; there are no clinical trials proving efficacy.
- HIV-infected individuals may be attracted to the many possible supplements on the market. Carefully review all items and discuss their viability or potential for harm.
- Use of acidophilus and probiotic products may alleviate loss of intestinal bacteria.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- There will be a need for medication management, a nutrient-dense diet, doctor visits, and other intervention and therapies. Provide support to the child and family or caregivers.
- Encourage formula feeding for mothers who has HIV infection.
- Discuss HIV infection prevention strategies, especially with noninfected teens. Researchers are working on a vaccine for HIV prevention.
- Children should receive all of their usual vaccinations to prevent other illnesses or complications.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard beverages and foods that left at room temperature for 2+ hours.
- Avoid honey in the diets of infants to decrease the risk of botulism.

For More Information

- AIDS Pediatric guidelines
  http://www.aidsinfo.nih.gov/Guidelines/GuidelineDetail.aspx?GuidelineID=8
- AIDS Vaccine Advocacy Coalition (AVAC)
  www.avac.org
- American Foundation for AIDS Research (amFAR)
  www.amfar.org
- Baylor AIDS Curriculum
  http://bayloraids.org/curriculum/
- Baylor International Pediatric AIDS Initiative
  http://bayloraids.org/
- Johns Hopkins University School of Medicine AIDS Site
  www.hopkins-aids.edu/
- National Institute of Allergy and Infectious Diseases (NIAID)
  www.niaid.nih.gov/daids/
- National Institutes of Health. Pediatric guidelines
  http://aidsinfo.nih.gov/contentfiles/Pediatric_OI.pdf
- National Pediatric AIDS Network
  http://www.npan.org/
- Pediatric AIDS Foundation
  http://www.pedaids.org/
- U.S. Coalition for Child Survival
  www.child-survival.org

HIV INFECTION, PEDIATRIC—CITED REFERENCES

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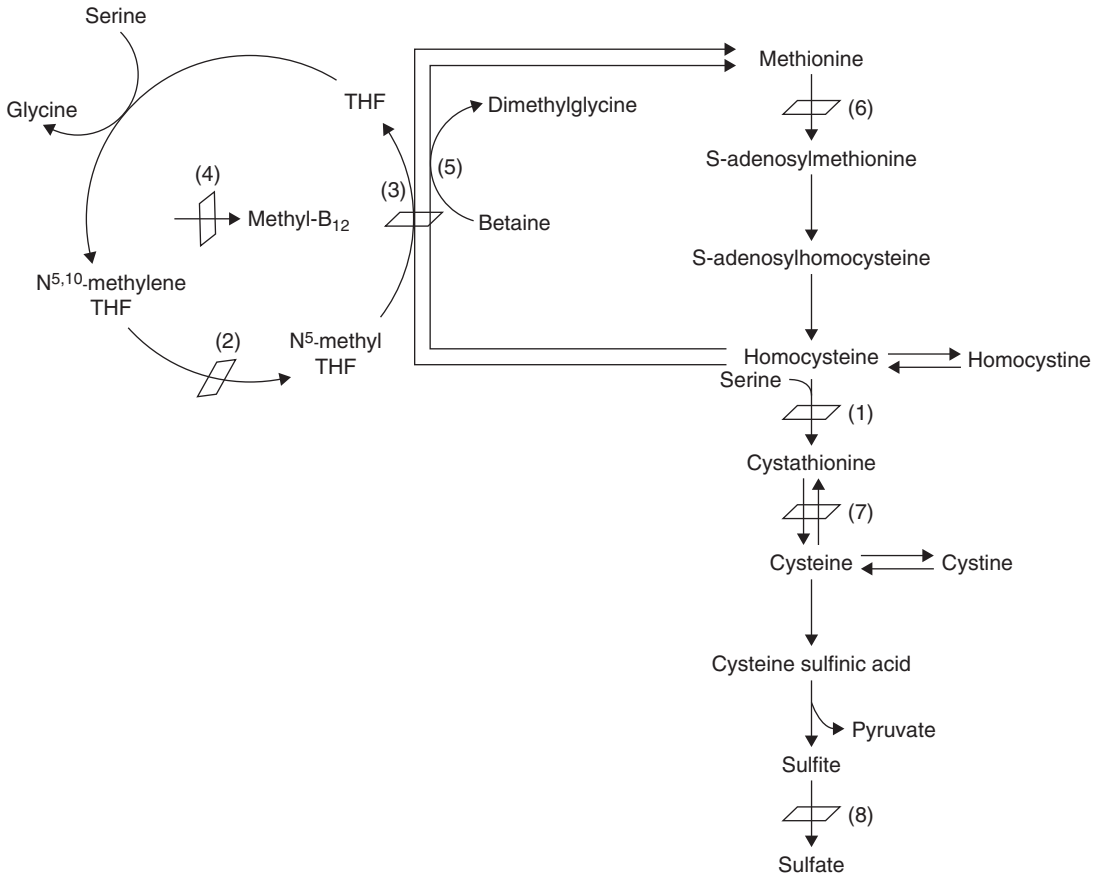
HOMOCYSTINURIA

NUTRITIONAL ACUITY RANKING: LEVEL 3–4



DEFINITIONS AND BACKGROUND

The significance of homocysteine (Hcy) in human disease was unknown until 1962, when cases of homocystinuria were correlated with vascular disease (McCully, 2007). Hcy is usually converted to cysteine and partly remethylated to methionine with the help of vitamin B_{12} and folate.

Homocystinuria (HCU) is a rare, autosomal recessive metabolic disorder of amino acid metabolism. In HCU type I from deficiency of cystathionine-beta-synthase (which requires vitamin B6 for activation). Hcy accumulates in the blood, methionine builds up, cysteine decreases, mental retardation and eye changes can occur from a lack of glutathione production (Ramakrishnan et al, 2006). In types II, III, and IV, methionine is decreased and no mental retardation occurs; here, treatment involves folate, vitamin B_{12} and avoiding excess of methionine (Ramakrishnan et al, 2006). Newborn screening is recommended (Refsum, Fredriksen et al, 2004).

HCU due to deficiency of CBS is inherited as an autosomal recessive trait. Human CBS is an *S*-adenosylmethionine–regulated enzyme that plays a key role in the metabolism of Hcy. HCU type 1 occurs in 1 in 200,000 births worldwide, with stronger prevalence in Ireland, Norway, and Qatar. Untreated, it leads to mental retardation, seizures, altered growth, hepatic disease, osteoporosis, thromboses, glaucoma, cataracts, and strokes. Individuals with HCU may be unusually tall in stature, with long arms and legs; this growth is directly mediated by Hcy.

Deranged vitamin B_6 metabolism or low levels of reductase enzyme may also cause HCU. A single biochemical test is not available; abnormal urinary tHcy response after methionine loading is the most sensitive test (Guttormsen et al, 2001). Urinary excretion of Hcy occurs but is unusual.

5,10-Methylene-tetrahydrofolate reductase deficiency (MTHFR) affects many enzyme systems. It can present with mental retardation, microcephaly, gait disturbance, psychiatric disturbances, seizures, abnormal EEG, and limb weakness. Therapy usually involves administration of folinic acid to enhance enzyme activity; 5-methyl-tetrahydrofolate to replace the missing end product; extra betaine, hydroxycobalamin, carnitine, and riboflavin to assist with related enzymatic actions.

For some patients, medications can reduce the excretion of Hcy in the urine, increase body weight, and improve mental function. Methionine may be given to correct low serum levels, and pyridoxine may lower serum Hcy levels if needed. If individuals do not respond to combinations of these drugs, supportive care is offered to reduce symptoms.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: Mutations in the cystathionine beta-synthase (CBS) on chromosome 21 are most common. Methylene-tetrahydrofolate reductase (MTHFR), 5-methyltetrahydrofolate-homocysteine methyltransferase (MTR), and 5-methyltetrahydrofolate-homocysteine methyltransferase reductase (MTRR) genes cause HCU less frequently. Patients with a cobalamin C (CblC) defect have combined methylmalonic aciduria and HCU (Heil et al, 2007).

Clinical/History

- Birth weight
- Present weight
- Length
- Growth (%); FTT?
- Scoliosis
- Diet/intake history
- Nearsightedness
- Lens dislocation
- Blood clots in veins
- Mental retardation
- Cognitive changes or psychiatric problems
- Osteopenia or osterporosis (DEXA)
- Marflan syndrome (long limbs, tall stature)

Lab Work

- ALT, AST
- Gluc
- Plasma methionine (fluctuates)
- Plasma cysteine
- Serum Hcy (elevated)
- Urinary methylmalonic acid
- Urinary tHcy after methionine load
- Serum folate
- Macrocytic anemia?
- MTHFR activity
- Serum B_{12}
- Serum B_6
- Serum Ca^{++}, Mg^{++}

SAMPLE NUTRITION CARE PROCESS STEPS

Inadequate Intake of Bioactive Substances

Assessment Data: Weight and growth chart showing long limbs, tall stature for age, lab tests showing HCU and low serum levels of vitamins B_6, B_{12}, and folate, myopia and hx of thrombotic clots in legs.

Nutrition Diagnoses (PES): Inadequate intake of bioactive substances related to genetic defect as evidenced by HCU and low serum levels of B_6, B_{12}, and folate.

Intervention: Educate parents about dietary enhancements for foods rich in B_6, B_{12}, and folate. Counseling about appropriate drug therapy and desirable nutritional outcomes.

Monitoring and Evaluation: Weight and growth records showing slower increments in added height; improved serum levels of vitamins; decreased or minimal Hcy in the urine.

INTERVENTION

OBJECTIVES

- Prevent mental retardation, growth delays, fractures, lens changes. Fractures occur because of defective collagen formation. A lens may become dislocated in CBS deficiency.
- Prevent cardiovascular complications (arterial and venous thrombosis, stroke, hypertension). Note: Dramatic decline in cardiovascular mortality in the United States since 1950 may be attributable in part to voluntary fortification of the food supply with vitamin B_6 and folic acid (McCully, 2007). Supplement with essential nutrients. Low folic acid intake aggravate the symptoms.
- In **Type I HCU,** reduce methionine in the diet to prevent accumulation of Hcy.
- **Metabolic disorders of cobalamin** will require intramuscular vitamin B_{12}.
- **Alterations of the MTHFR alleles** may require use of L-methylfolate.

FOOD AND NUTRITION

- Increase fluid intake.
- **HCU type I** is treated with supplementation of vitamin B_6 and cystine (to supply sulfur). If nonresponsive to B_6, use a low-methionine diet with a supplement of cystine. Reduce intake of methionine from meat, poultry, fish, and eggs. Soy products (e.g., Isomil, ProSobee, Soyalac) can be used. XMET Maxamaid (SHS North America), Hominex 1 for infants or Hominex 2 for children (Ross Laboratories), or Product HOM 1 or HOM 2 (Mead Johnson) is also useful.
- **For cobalamin metabolic disorders:** Intramuscular vitamin B_{12} is needed; dietary or supplemental vitamin B_{12} is not effective.
- **For MTHFR:** Methylated folic acid, Vitamins B_6 and B_{12}, riboflavin, choline, and betaine are useful supplements.

Common Drugs Used and Potential Side Effects

- Dipyridamole may be used to decrease thrombosis.
- Pyridoxine therapy (vitamin B_6) for longer than 1 month is useful for some forms. The doctor may prescribe 100–500 mg or higher.
- Folic acid and vitamin B_{12} should be supplied in a methylated form if needed. For example, Cerefolin (Pan Am Labs) contains methylated folate and B_{12} plus *N*-acetylcysteine; Deplin contains 7.5 mg L-methylfolate.
- Choline and betaine may be useful (Alfthan et al, 2004; Busby et al, 2005).

Herbs, Botanicals, and Supplements

- Herbs and botanicals should not be used for HCU because there are no controlled trials to prove efficacy.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Emphasize the importance of controlling diet, snacks, using proper forms of supplemental nutrients.
- Discuss good food sources of folic acid and other B-complex vitamins. Therapy with folic acid, vitamin B_6, l-carnitine, and intramuscular vitamin B_{12} often results in improvement of symptoms (Heil et al, 2007).
- Newborn screening of tHcy is useful to detect vitamin B_{12} deficiency or HCU (Refsum et al, 2004).
- Because of increased incidence of osteoporosis, high serum Hcy levels interfere with collagen cross-linking (McLean et al, 2004). Controlling serum Hcy is important for bone health.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants to decrease potential risk of botulism.

For More Information

- Children Living with Inherited Metabolic Diseases http://www.climb.org.uk/
- NIH Genetics Home Reference http://ghr.nlm.nih.gov/condition=homocystinuria
- Save Babies http://www.savebabies.org/diseasedescriptions/homocystinuria.php

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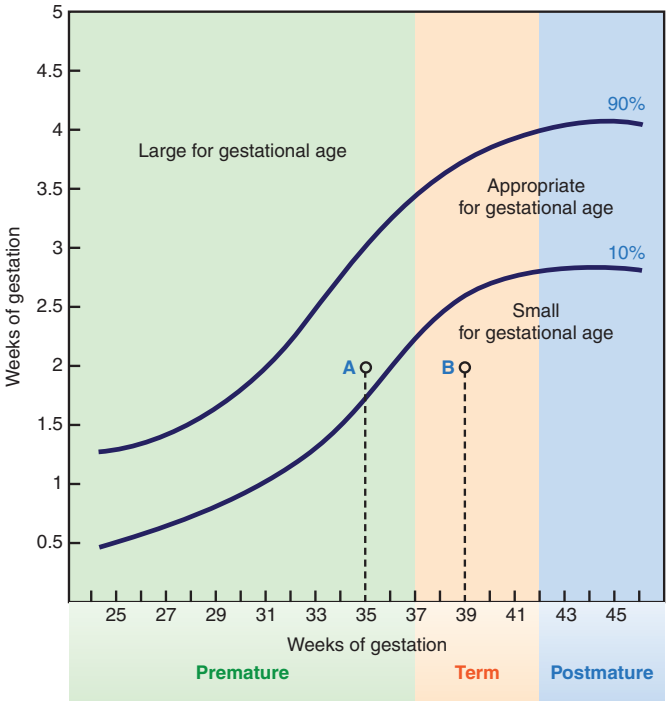
LARGE FOR GESTATIONAL AGE INFANT (INFANT MACROSOMIA)

NUTRITIONAL ACUITY RANKING: LEVEL 1–3



DEFINITIONS AND BACKGROUND

Infants whose weight is more than the 90th percentile for gestational age are classified as large for gestational age (LGA). Birth weight is high (3300–4000 g) at 40 weeks. Obesity and gestational diabetes increase the risk of an LGA delivery (Ehrenberg et al, 2004). LGA infants may also be born to mothers who are multiparous.

Macrosomia in newborns raises the risk for birth-related problems (Samaras et al, 2004). Problems may include hypoglycemia, respiratory distress, aspiration pneumonia, bronchial paralysis, or facial paralysis. LGA neonates usually have higher body fat and lower lean body mass than appropriate for gestational age (AGA) infants. Cord plasma adiponectin and leptin levels are very high in LGA infants; adiponectin is involved in regulating fetal growth (Tsai et al, 2004).

After birth, rapid adaptation is necessary for infants to be able to maintain independent glucose homeostasis; this process is compromised in LGA infants (Beardsall et al, 2008). Controlling maternal weight gain remains an important goal for successful pregnancy outcome. High birth weight may eventually promote impaired glucose tolerance, diabetes, obesity, or cancer (Samaras et al, 2004).

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: Mitrochondrial RNA deletions may be involved, but no specific gene has been identified.

Clinical/History		Lab Work
Head circumference	Neonatal Behavior Assessment Scale (motor maturity, autonomic stability, and withdrawal)	Serum Gluc; hypoglycemia?
Length	Respirations	Elevated serum insulin
Birth weight more than the 90th percentile for gestational age	pCO_2, pO_2 levels	Chol, Trig
Neonatal Growth Assessment Scores	BP	Alb
	Diet/intake history	Hemoglobin
	I & O	Hct (elevated)?
		Hyperbilirubinemia?
		Urinary acetone

INTERVENTION

OBJECTIVES

- Allow adequate growth rate and development.
- Prevent rapid phase of hypoglycemia.
- Maintain energy intake at a desired level while allowing adequate growth in the infant. Prevent obesity and its consequences for the infant as much as possible.
- Monitor serum lipids or bilirubin as deemed necessary.

SAMPLE NUTRITION CARE PROCESS STEPS

Abnormal Nutritional Labs

Assessment Data: Abnormal labs for blood glucose, insulin, bilirubin, hematocrit in LGA infant.

Nutrition Diagnoses (PES): Abnormal nutritional lab values related to macrosomia as evidenced by neonatal hyperinsulinism after termination of maternal glucose at birth.

Interventions:

Prophylactic IV infusion of 10% dextrose in water until early frequent feedings can be established.

Educate mother about monitoring for signs of hypoglycemia, hyperbilirubinemia.

Monitoring and Evaluation:

Blood glucose levels should be closely monitored by bedside testing.

Evaluate over first few weeks for blood glucose control and normalization of serum insulin, bilirubin, hematocrit.

FOOD AND NUTRITION

- Feed the infant often, as indicated by infant's appetite and goal weight pattern. Control total glucose intake if infant shows signs of hyperglycemia.
- Alter intake of fat as determined by lipid profile.
- Maintain a sufficient level of protein if energy needs to be restricted from CHO or fat.

Common Drugs Used and Potential Side Effects

- Insulin may be necessary to control hyperglycemia. Beware of any excesses of insulin, which could aggravate hypoglycemia.

Herbs, Botanicals, and Supplements

- Herbs and botanicals should not be used for LGA infants because there are no controlled trials to prove efficacy for any related problems.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Discuss normal growth patterns as appropriate for the infant, reviewed in concert with the pediatrician. Signs of hyperglycemia and hypoglycemia should be discussed.
- Review risks inherent in another pregnancy, especially if the mother has diabetes. Counseling may be beneficial.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants to decrease potential risk of botulism.

For More Information

- American College of Obstetricians and Gynecologists
  http://www.acog.org
- Large for Gestational Age
  http://www.chp.edu/CHP/P02383

LARGE FOR GESTATIONAL AGE INFANT—CITED REFERENCES

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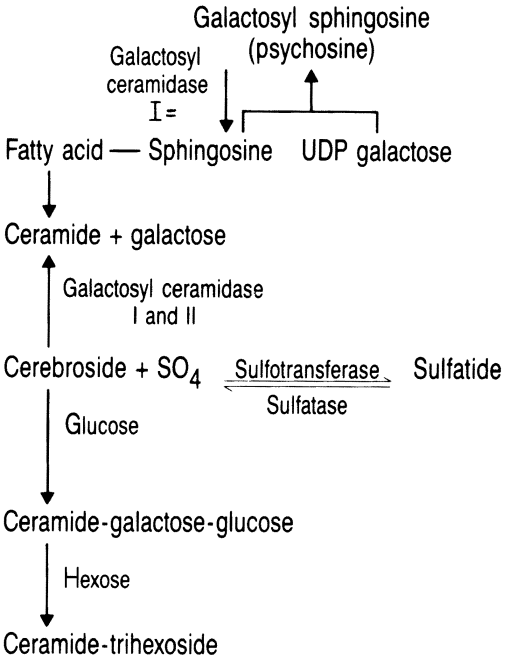
LEUKODYSTROPHIES

NUTRITIONAL ACUITY RANKING: LEVEL 1



DEFINITIONS AND BACKGROUND

Leukodystrophies (peroxisome biogenesis disorders) are genetic disorders that affect the myelin sheath. While the genetic defect and biochemical abnormalities have been defined, there is a wide range of phenotypic expression (Moser et al, 2005). Neonatal adrenoleukodystrophy and infantile **Refsum's disease** are mild phenotypes. The most severe form is **Zellweger's syndrome**, which may be fatal and is characterized by an enlarged liver, high serum levels of iron and copper, and visual changes. Noninvasive and presymptomatic diagnosis and prenatal diagnosis are available; family screening and genetic counseling are important (Moser et al, 2005).

X-linked adrenoleukodystrophy (X-ALD) is one of the autosomal recessive disorders with an enzymatic defect in very long–chain fatty acid (VLCFA) oxidation, which is usually abundant in sphingomyelin. Ultimately, the myelin sheath surrounding the nerves is destroyed, causing demyelination and neurological problems. Adrenal gland malfunction causes Addison's disease (adrenal insufficiency). Accumulation of saturated VLCFA, especially hexacosanoate (C26:0), occurs because there is a missing or defective protein (ALD protein) to process that fatty acid. The incidence of X-ALD is estimated to be one in 17,000 in all ethnic groups (Moser et al, 2005).

Onset of X-ALD is usually in childhood, with a rapid, progressive demyelination, hypotonia, and psychomotor retardation. However, at least half of patients with X-ALD are adults with somewhat milder manifestations, and women who are carriers may become symptomatic (Moser et al, 2005). X-ALD is often misdiagnosed as ADHD in boys and as multiple sclerosis in men and women (Moser et al, 2005). Prognosis is poor and death may occur up to a decade after onset.

The observation that dietary fatty acids affect membrane composition has led to the use of modified diets in these conditions. Lorenzo's oil is a mixture of oleic and erucic (rapeseed, or canola) oils, which reduces the production of VLCFA. Early oral administration helps infants and children with the neonatal form (Suzuki, 2001). In addition, the omega-3 fatty acid, DHA, is present in large amounts in infant brains but is often absent in infant formulas. Therefore, intake of omega-3 fatty acids is now recommended. Gene therapy of endogenous hematopoietic stem cells, pharmacological upregulation of other genes encoding proteins involved in peroxisomal beta-oxidation, reduction of oxidative stress, and possibly lovastatin are under study (Semmler et al, 2008).

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: Most of the leukodystrophies are genetic. X-linked adrenoleukodystrophy (X-ALD) is caused by defects of the ABCD1 gene on chromosome Xq28. See website http://www.ulf.org/types/types.html for details about the 34 known types.

Clinical/History

- Height
- Weight
- Growth chart
- Diet/intake history
- Bronzing of skin (Addison's disease)
- Cataracts or glaucoma?
- Poor sucking; feeding problems

Lab Work

- Plasma phosphatidylcholine
- Fatty acid profile; VLCFA (elevated?)
- Alb
- Chol
- Trig
- Plasma sphingomyelin
- H & H
- Pipecolic acid testing

INTERVENTION

OBJECTIVES

- Decrease rapid progression of demyelination of CNS by offering sufficient fatty acids (DHA). Overall, maintain total VLCFA levels while altering fatty acid sources.
- Prevent or lessen complications of the disorder, including adrenal dysfunction.
- Support the physical therapy by maintaining strength with an adequate diet.

SAMPLE NUTRITION CARE PROCESS STEPS

Self-Feeding Difficulty

Assessment Data: Abnormal weight and growth, difficulty with self-feeding.

Nutrition Diagnoses (PES): Self-feeding difficulty related to low vision and limited mobility.

Intervention: Educate parents about DHA and appropriate fat ratios. Counsel about tips for self-feeding, including special adaptive equipment.

Monitoring and Evaluation: Weight records, growth, slower disease progression.

FOOD AND NUTRITION

- Increase endogenous VLCFA synthesis of monounsaturated fatty acids by restricting exogenous (dietary) VLCFA (C26:0) to less than 3 mg and by increasing oleic acid (C18:1). The typical American diet yields 35–40% total energy from fat with 12–40 mg C26:0 daily.
- Offer a low- to very low–fat diet, with supplementation of oleic and erucic acids (Lorenzo's oil), plus DHA (Deon et al, 2008). Include sources of omega-3 fatty acids, such as salmon, tuna, or mackerel, for older children and adults. Use good food sources of vitamins C, E, selenium, zinc for antioxidant properties.
- Lorenzo's oil is similar to olive oil (87% C18:1, 4.8% linoleic acid) but lacks measurable fatty acids with a chain length greater than C20. It can be used in cooking, as a supplement in juice, as an oil for salad dressings, or in food preparation instead of margarine, butter, mayonnaise, or shortening.
- If the patient requires tube feeding, a formula can be developed that contains nonfat milk, specialty oils, corn syrup or sugar, and a vitamin–mineral supplement.

Common Drugs Used and Potential Side Effects

- Hormone-replacement therapy is necessary in all patients with adrenal insufficiency (Semmler et al, 2008). Long-term prednisone and spironolactone may cause hyperglycemia and osteoporosis.
- Use of lovastatin is being tested.

Herbs, Botanicals, and Supplements

- Herbs and botanicals should not be used for this condition because there are no clinical trials proving efficacy.
- Studies suggest that vitamin E, selenium, and carnitine (antioxidants) should be considered.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- The whole family can support the diet; it can be adapted for everyone. Discuss cooking methods using Lorenzo's oil.
- Restaurant dining can be a problem and special meals may have to be developed for travel.
- If nausea occurs, the oil can be taken in an emulsion.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants to decrease potential risk of botulism.

For More Information

- Genetic Fairness
  http://www.geneticfairness.org/
- Myelin Project
  http://www.myelin.org/
- National Institute of Neurological Disorders and Stroke
  http://www.ninds.nih.gov/disorders/adrenoleukodystrophy/adrenoleukodystrophy.htm
- Rare Diseases
  http://www.rarediseases.org/
- United Leukodystrophy Foundation
  http://www.ulf.org/

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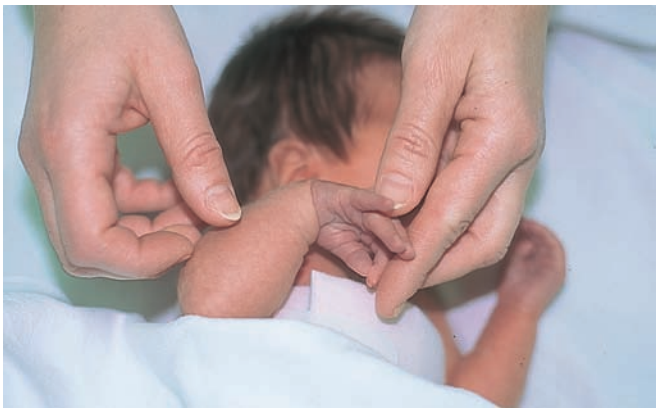
Suzuki Y. The clinical course of childhood and adolescent adrenoleukodystrophy before and after Lorenzo's oil. *Brain Dev.* 23:30, 2001.

LOW BIRTH WEIGHT OR PREMATURITY

NUTRITIONAL ACUITY RANKING: LEVEL 3–4



DEFINITIONS AND BACKGROUND

Every newborn is classified as one of the following: premature (<37 weeks of gestation), full-term (37–42 weeks of gestation), or postterm (>42 weeks of gestation). **Prematurity** is generally correlated with LBW. **LBW** infants may be small for date, have IUGR, or have dysmaturity. LBW infants weigh less than 2500 g or 5.5 lb (<10th percentile for gestational age) at birth. VLBW infants (<1300–1500 g) are especially prone to nutritional deficits. Infants who weigh 1000 g are called "micropreemies."

Low weight or BMI at conception or delivery, as well as poor weight gain during pregnancy, are associated with LBW, prematurity, and maternal delivery complications (Ehrenberg et al, 2003).

In the United States, infants born to mothers younger than 20 or older than 35 years are more likely to be preterm than infants born to mothers 20–35 years old (March of Dimes, 2009). Teens should be counseled to delay pregnancy until they are adults.

LBW infants have higher risk of mortality, morbidity, and poor growth. Typical problems of the LBW or premature infant include hypoglycemia, hypothermia, jaundice, dry skin, decreased subcutaneous fat, and anemia. Admission to neonatal intensive care units (NICUs) is common, especially for respiratory distress.

Adequate nutrition support almost immediately after birth is important to prevent growth restriction.

Undernutrition at critical stages of development (especially protein) produces long-term short stature, organ growth failure, neuronal deficits of number and dendritic connections, later behavioral and cognitive outcomes (Hay, 2008). Without nutrition starting immediately after birth, the infant enters a catabolic condition, which limits normal development and growth (Hay, 2008). During the first months after discharge, VLBW babies need to have nutrition support to help promote early catch-up growth and mineralization. Careful and frequent monitoring of growth patterns is needed to prevent developmental delays.

Premature breast milk has higher electrolyte, protein, and MCT levels than mature breast milk. Breastfeeding in the NICU is very beneficial and has long-term benefits for the child (Vohr et al, 2006). Feeding "on demand" is best (Crosson and Pickler, 2004). Preterm infants have lower energy expenditure when they are fed breast milk than when they are fed formula (Lubeztsky et al, 2003). Early feeding increases intestinal lactase activity, which is a marker of intestinal maturity and may influence clinical outcomes. Nearly all studies have shown that minimal enteral feeding approaches (e.g., "trophic feeds") promote the capacity to feed enterally (Hay, 2008).

Omega-3 fatty acids are important for healthy infants (Fewtrell et al, 2004). Maternal dietary intake of DHA is important. The brain, retina, and neural tissues are rich in DHA and arachidonic acid (ARA). Table 3-9 lists the nutritional deficits found in premature or LBW infants.

Milk has distinct advantages over formulas in avoiding necrotizing enterocolitis (NEC); minimal enteral feeding regimens produce less NEC than more aggressive enteral feeding (Hay, 2008). Caution must also be used in feeding as overfeeding has the potential to produce adipose tissue, or obesity, which then leads to insulin resistance, glucose intolerance, and diabetes (Hay, 2008).

Supplementation for premature infants remains controversial. Glutamine does not decrease sepsis (Poindexter et al, 2004), whereas selenium supplementation does (Darlow and Austin, 2003). Overall, the American Dietetic Association recommends at least five medical nutrition therapy visits for high-risk, premature infants.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: There are no specific markers that predict LBW.

Clinical/History

- Birth weight
- Gestational age
- Birth length
- % weight/length (Olsen et al, 2009)
- Diet/intake history
- Swallowing reflex
- Temperature (often decreased)
- Sucking reflex
- Apgar scores
- I & O
- Hypotension?
- Infection?
- Respiratory distress?
- Retinopathy of prematurity (ROP)?

Lab Work

- Gluc
- H & H; No. of RBCs
- Anemia?
- Alb
- Ca^{++}, Mg^{++}
- Na^{+}, K^{+}
- Transthyretin
- ALT, AST
- Serum folic acid and vitamin B_{12}
- Serum phosphorus
- Lecithin to sphingomyelin ratio (L:S ratio)
- Bilirubin

TABLE 3-9 Nutritional Deficits in the Premature or Low Birth Weight Infant

Problem	Implication
Immaturity at the cellular level	Altered biochemical needs
Underdeveloped digestive/absorptive abilities	Malabsorption
Essential fatty acid deficiency	Slowed growth, renal and lung changes, fatty liver, impaired water balance, RBC fragility, and dermatitis
Delayed oral neuromuscular development and small gastric capacity	Limited ability to consume adequate amounts of nutrients
Marginal nutrient stores at birth	Fat, glycogen, and minerals such as calcium and phosphorus
Slow growth	Higher metabolic demands
Poor nutritional intake of the mother	Deficiencies such as folate

INTERVENTION

OBJECTIVES

- Begin feedings of distilled water or colostrum as soon as possible for infants without respiratory distress. Early feeding (3–5 days after birth) tends to allow babies to mature faster; they have fewer days of intolerance, a shorter hospital length of stay, and earlier tolerance of full feedings.
- Encourage the mother to breastfeed as long as possible. Taurine is an especially important nutrient in breastmilk (Verner et al, 2007). If tube feeding is needed, mother can express milk to be given to the infant; it can also be supplemented to meet special needs. Supplement with EFAs and DHA. Assure adequate intake of folate and vitamin B_{12} to prevent anemias.
- Provide glucose as soon after birth as possible; adjust according to frequent measurements of plasma glucose to achieve and maintain concentrations >45 mg/dL but <120 mg/dL to avoid hypoglycemia, and hyperglycemia respectively (Hay, 2008).

SAMPLE NUTRITION CARE PROCESS STEPS

Inadequate Protein-Energy Intake

Assessment Data: Low birth weight, inadequate growth, poor feeding skills.

Nutrition Diagnoses (PES): Inadequate intake of protein and energy related to prolonged feeding time and difficulty latching-on, as evidenced by premature birth, underweight (<3 percentile) and irritability after feeding.

Intervention: Educate staff and parents about nutritional requirements and appropriate formula choice if mother has discontinued breastfeeding.

Monitoring and Evaluation: Weight records, catch-up growth, tolerance of formula feedings, less irritability after feeding.

- When possible, use enteral feedings instead of parenteral to reduce onset of cholestasis and osteopenia.
- Gradually increase energy and protein to meet the needs of rapid growth. For protein, ensure a proper whey to casein ratio.
- With parenteral feeding, include amino acids in proper amounts, especially cysteine, taurine (Wharton et al, 2004), tyrosine, glycine, and arginine (Wu et al, 2004). Intravenous TrophAmine can be used. Be sure selenium is provided (Darlow and Austin, 2003).
- Promote catch-up growth and development. While long-term effects such as metabolic syndrome occur among LBW or premature infants later in life, effective nutrition support is needed (Greer, 2007).
- Prevent illness, rickets, respiratory distress, hypoglycemia or hyperglycemia, NEC, infections, obstructive jaundice, and tyrosinemia.

FOOD AND NUTRITION

- While in the radiant warmer, provide water at 60–80 mL/kg body weight/d; gradually increase to 150 mL/kg. Add electrolytes (sodium, potassium, and chloride) on at least the second day.
- Day 1: Breastfeed or give glucose at 6–8 mg/kg/min. Advance by no more than 20 mL/kg daily.
- Progress to special formulas such as Similac Special Care 24 or Enfamil Premature Formula (24 kcal/oz) to yield 120–150 mL/kg up to 180–200 mL/kg/d. NeoSure or Enfacare are helpful for transition to home (22 kcal/oz with added Ca^{++} and phosphorus).
- Within 7 days, the diet should provide 120–150 kcal/kg BW daily; carbohydrate should be 40–45% total kcal (10–30 g/kg). Protein should be age specific.
- Tube feeding initiation: Start at 10–15 mL/hr at one quarter strength. Progress as tolerated to desired rate. Specialty products have been developed for VLBW infants, such as Mead Johnson's Enfamil Human Milk Fortifier (Berseth et al, 2004). See Table 3-10 for estimated energy and macronutrient calculations.
- Use TPN if not feeding by day 3; glucose infusion rate is 15 mg/kg/min. TPN needs are similar to enteral needs. Crystalline amino acid infusions promote positive nitrogen balance by use of 1 g/kg/d as soon as possible. Use up to 3 g/kg/d of lipid infused continuously or early enteral feeding to prevent cholestatic liver disease. Monitor for carnitine deficiency. See Table 3-11 for recommended vitamin and mineral intakes.
- There may be subtle and delayed hunger cues from the infant. If poor sucking or swallowing instincts exist, the infant may need gavage feeding. Feed every 2 hours or use continuous drip feeding and change to bolus feedings when full strength is tolerated. If infant weighs 1000–1750 g, feed more vigorously; if infant weighs 1750 g or more, feed as a normal-term infant.
- The micronutrient needs of a stable, preterm LBW infant may be as follows: high levels of calcium 120–230 mg/kg; vitamin E (water soluble), 6.0–12.0 mg/kg; 2.5 mg iron/100 kcal in formula (necessary only if stores are depleted); vitamin D, 400 IU/d; folic acid, 25–50 μg/kg; sodium to avoid hyponatremia, 2–3 mEq/kg; vitamin C,

TABLE 3-10 Nutrient Needs of Preterm Infants

	Under 2.5 kg body weight	Over 2.5 kg body weight
Daily basal needs	60–80 kcal/kg	40–70 kcal/kg
+ Fecal losses	10–20 kcal/kg	10–20 kcal/kg
+ Growth (tissue synthesis, energy stores)	10 kcal/kg	10 kcal/kg
Total energy needs[a]	110–130 kcal/kg enteral	100–120 kcal/kg enteral
Parenteral needs (no fecal losses, no thermogenic effect of food)	90–110 kcal/kg	80–100 kcal/kg
Protein	3.5–4.0 g/kg enteral	3.6–4.0 g/kg enteral
Amino acids	2–3 g/kg parenteral	3.2–3.5 g/kg parenteral
Lipid	0.5 g/kg for essential fatty acids	0.5 g/kg for essential fatty acids
Fluid	80 mL/kg; increase 10–20 mL/kg daily to 120–160 mL/kg	60 mL/kg; increase 10–20 mL/kg daily to 120–150 mL/kg

[a]Add extra kilocals for fever (7% per 1-degree elevation); cardiac failure; sepsis; failure to thrive; major surgery; BPD.

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Pierro A. Metabolism and nutritional support in the surgical neonate. *J Ped Surg*. 37:811, 2002
Mirtollo J, et al. Safe practices for parenteral nutrition. *J Parenter Enteral Nutr.* 28:39S, 2004.

TABLE 3-11 Parenteral Vitamin and Mineral Needs in Preterm Infant

Nutrient	Recommended Intake for Infants <2.5 kg	Recommended Intake for Infants >2.5 kg
Vitamin A	280 μg	700 μg
Vitamin D	160 IU	400 IU
Vitamin E	2.8 mg	7 mg
Vitamin K	80 μg	200 μg
Vitamin C	32 mg	80 mg
Thiamin	0.48 mg	1.2 mg
Riboflavin	0.56 mg	1.4 mg
Niacin	6.8 mg	17 mg
Pyridoxine (B_6)	0.40 mg	1.0 mg
Folic acid	56 μg	140 μg
Vitamin B_{12}	0.40 μg	1 μg
Biotin	8 μg	20 μg
Pantothenate	2 mg	5 mg
Calcium	80–100 mg/kg	80–100 mg/kg
Phosphorus	43–62 mg/kg	43–62 mg/kg
Magnesium	6–10 mg/kg	6–10 mg/kg
Chromium	0.05–0.2 μg/kg	0.2 μg/kg
Copper	20 μg/kg	20 μg/kg
Iodide	1 μg/kg	1 μg/kg
Manganese	1 μg/kg	1 μg/kg
Molybdenum	0.25 μg/kg	0.25 μg/kg
Selenium	1.5–2 μg/kg	2 μg/kg
Zinc	400 μg/kg	50–250 μg/kg

Needs are estimated for use with a solution of 2.5 g/dL of amino acids infused at 120–150 mL/kg/dL.
Derived from: Hay WW Jr. Strategies for feeding the preterm infant. *Neonatology*. 94:245, 2008, and Mirtollo, et al. Safe practices for parenteral nutrition. *J Parenter Enteral Nutr.* 28:39S, 2004.

18–24 mg/kg; and phosphorus, 60–140 mg/kg. Monitor need for vitamins A and B_{12}, magnesium, zinc, selenium, and copper. Other nutrients should be provided according to DRI tables for the newborn.

- Total fat should be 5–7 g/kg to meet half of energy needs without excess carbohydrate. Soybean oil provides EFAs (1–2% kcals as EFA) in the form of linoleic acid. Exogenous carnitine may be needed to take EFAs into the mitochondria. Inositol may be needed in respiratory distress.

Common Drugs Used and Potential Side Effects

- With hyperkalemia and hyperglycemia, continuous intravenous insulin is needed.
- Sometimes, supplemental thyroid hormone or iodide may be prescribed.
- Other medications may be used for underlying disease states. Monitor for side effects.
- Use caution with vitamin supplements. Early vitamin supplementation may promote increased asthma, especially in black children (Milner et al, 2004).

Herbs, Botanicals, and Supplements

- Herbs and botanicals should not be used for LBW or premature infants because there may be allergic or asthmatic reactions.
- Glutamine supplementation does not seem to alleviate NEC.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Teach the caretaker or parent about increased nutrient needs of LBW or premature infant. Special formulas have

80 kcal/dL (usual is 67 kcal/dL) and have MCT, extra protein, calcium, phosphorus, and sodium.
- Emphasize progression of infant feeding for an adequate growth pattern and weight. Catch-up is common by 2–3 years of age in this population. VLBW infants experience catch-up growth and attain predicted genetic height during adolescence (Anderson, 2008).
- Emphasize the importance of nutrient density for growth (e.g., zinc, vitamin B_6, and vitamin E).
- Do not overfeed; excess nonprotein energy is stored as fat regardless of its source (fat or carbohydrate). High-energy or MCT intake in otherwise healthy, growing preterm infants should be avoided (Romero et al, 2004).
- Monitor for the tendency to aspirate, for lactose intolerance, and for other problems. Feed slowly to avoid NEC.
- The child may benefit from the WIC program if available.
- Follow-up clinic or home visits are recommended. Offer tips such as using small, frequent feedings; using a quiet, calm environment for feeding; supporting the jaw; and trying special feeding equipment if needed (angle-neck bottle).

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any formula, beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey because of the potential risk of botulism.

For More Information

- March of Dimes–Prematurity
  http://www.modimes.org/prematurity/
- Prematurity
  http://www.prematurity.org/
- UNICEF
  http://www.unicef.org

LOW BIRTH WEIGHT OR PREMATURITY—CITED REFERENCES

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MAPLE SYRUP URINE DISEASE

NUTRITIONAL ACUITY RANKING: LEVEL 3–4

DEFINITIONS AND BACKGROUND

Maple syrup urine disease (MSUD) results from an autosomal recessive trait, causing an inborn error of metabolism of the BCAAs leucine, isoleucine, valine. Byproduct ketoacids become elevated and may cause life-threatening cerebral edema and dysmyelination (Riazi et al, 2004). The elevated BCAAs (leucinosis) cause an infant or child with MSUD to become symptomatic. Neurotransmitter deficiencies related to BCAA accumulation, and energy deprivation through Krebs cycle disruption from ketoacid accumulation, are mechanisms (Zinnanti et al, 2009).

In the United States, MSUD occurs in one in 225,000 births. The Mennonite population from eastern Pennsylvania has a high percentage of births with this disorder. MSUD also occurs in other populations throughout the world. Onset occurs in children between the ages of 1 and 8 years.

Symptoms in an infant include poor sucking reflex, anorexia, FTT, listlessness, irritability, and a characteristic odor (sweet, burnt maple syrup odor of the urine and sweat).

TABLE 3-12 Types and Nutrition Interventions for Maple Syrup Urine Disease (MSUD)

Type	Nutrition Intervention
Classic MSUD	Most common. Little or no enzyme activity (usually <2% of normal). Protein from branched-chain amino acids (BCAAs) must be severely restricted.
Intermediate MSUD	Higher level of enzyme activity (approximately 3–8% of normal). Tolerance for leucine is slightly better. Management is the same as for the classic form.
Intermittent MSUD	Milder form; greater enzyme activity (8–15% of normal). Few symptoms until 12–24 months of age, often in response to an illness or larger protein intake. During episodes of illness or fasting, the BCAA levels elevate, the characteristic odor becomes evident, and the child can go into a metabolic crisis.
Thiamin-responsive MSUD	Rare form. Giving large doses of thiamin to the thiamin-responsive child will increase the enzyme activity. Moderate protein restriction is needed.

Derived from: data available at http://www.msud-support.org/overv.htm, accessed January 2, 2005.

Afflicted infants have a high-pitched cry and may alternate between being limp or rigid. Without treatment, symptoms progress rapidly to seizures, coma, and death (Schonberger et al, 2004). With earlier diagnosis and treatment, there is a lower risk of peripheral neuropathy.

Nutrition therapy is lifelong. Supplemental isoleucine and valine are usually required (Chuang and Shih, 2001). The amino acid mixture contributes 30–40% of total energy, leaving 60–70% from food (Isaacs and Zand, 2007). Note that thiamin is the coenzyme for BCAAs and should be made available. There are four classifications for the types of MSUD: classic, intermediate, intermittent, and thiamin-responsive; these refer to the amount and type of enzyme activity present. See Table 3-12.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: The branched-chain alpha-keto acid dehydrogenase (BCKD) is missing.

Clinical/History

- Length (height)
- Birth weight
- Present weight
- Growth (%)
- Diet/intake history
- Perspiration with maple odor
- Grand mal seizures?
- Hypertonicity
- Cerebral edema?

Lab Work

- Plasma leucine, isoleucine, valine (therapeutic range of 100–300 μmol/L)
- Plasma L-alloisoleucine 0.5 μmol/L (most specific and most sensitive for MSUD)
- Urinary excretion of ketoacids
- Urine odor of burnt maple syrup
- Alb
- Globulin
- Uric acid (increased?)
- H & H
- Serum Fe
- Serum osmolality

INTERVENTION

OBJECTIVES

- Prevent endogenous protein catabolism (Morton et al, 2002).
- Prevent toxic concentrations of BCAAs (Riazi et al, 2004) by using an appropriate medical formula, special intravenous feeding, or low-BCAA diet. Monitor serum levels of leucine frequently to determine current status.
- Support normal growth and development with adequate protein synthesis and prevention of essential amino acid deficiencies. Overcome any difficulty with feeding related to poor sucking reflex.
- Control intake of BCAAs for life. As the child grows, add BCAAs individually in a controlled manner.
- Maintain normal serum osmolality (Morton et al, 2002).
- In emergencies, hemodialysis is sometimes necessary (Hmiel et al, 2004).

FOOD AND NUTRITION

- Restrict intake of BCAAs in the diet to 45–62 mg/d (Riazi et al, 2004). Use Mead Johnson's MSUD powder or Ross Laboratories' Maxamaid MSUD. Use the latter with PFD1 or PFD2 (Mead Johnson) because it contains no cholesterol or fat.
- When BCAA levels are high (during illness or fasting), it may be necessary to use a specific IV solution that allows the excess leucine, valine, and isoleucine to be used for protein synthesis in the body, thereby rapidly decreasing the elevated levels.

SAMPLE NUTRITION CARE PROCESS STEPS

Excessive Protein Intake

Assessment Data: Weight and growth charts, elevated serum leucine.

Nutrition Diagnoses (PES): Excessive protein intake related to MSUD and genetic inability to metabolize large amounts of dietary BCAAs as evidenced by high serum leucine and elevated urinary keto-acids.

Intervention: Educate parents about appropriate formula to manage MSUD. Counsel about appropriate formula and food sources of leucine and needed supplements, including thiamin.

Monitoring and Evaluation: Weight records, growth, improved appetite, improved serum and urinary lab test results.

- Provide adequate energy intake from CHO and fat to spare amino acids for building tissue, etc.
- Use small amounts of milk in the diet to support growth. Cow's milk contains 350 mg of leucine, 228 mg of isoleucine, and 245 mg of valine per 100 mL.
- Avoid eggs, meat, nuts, and other dairy products. Gelatin, a form of protein low in BCAAs, may be used in the diet.
- If hemodialysis is needed, monitor fluid, protein, and electrolytes carefully.

Common Drugs Used and Potential Side Effects

- Sometimes insulin or a similar agent is given to speed up the utilization of excess BCAAs.
- The doctor may prescribe large doses of thiamin for children who are thiamin-responsive.
- Avoid use of aspirin with MSUD; individuals with this condition are more prone to Reye's syndrome.
- Norleucine is being tested as a possible treatment for emergency management of MSUD crises.

Herbs, Botanicals, and Supplements

- Herbs and botanicals should not be used for MSUD because there are no controlled trials to prove efficacy.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Educate caregiver and patient that the diet must be maintained for life.
- Discuss the diet's total energy and protein intake that are appropriate for the patient's age and stage of development.
- Illness or infection can cause elevations of BCAAs. This can lead to vomiting, diarrhea, irritability, sleepiness, unusual breathing, staggering, hallucinations, and slurred speech. This is an emergency and must be treated immediately.
- With knowledge of the pathophysiology of MSUD and understanding of what to do for cerebral edema, fluid and electrolyte management, nutrition, and psychosocial issues, a full life is possible (Robinson and Drumm, 2001).

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants to decrease potential risk of botulism.

For More Information

- Cambrooke Foods
  http://www.cambrookefoods.com/
- MSUD Family Support Group
  http://www.msud-support.org/
- National Newborn Screening
  http://genes-r-us.uthscsa.edu/
- Nutrient Data
  http://www.nal.usda.gov/fnic/foodcomp/search/
- Save Babies
  http://www.savebabies.org/
- Screening
  http://www.msud-support.org/testing.htm

MAPLE SYRUP URINE DISEASE—CITED REFERENCES

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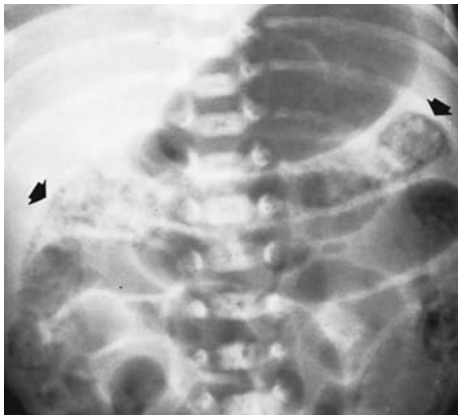
NECROTIZING ENTEROCOLITIS

NUTRITIONAL ACUITY RANKING: LEVEL 4



Adapted from: Ronald L. *Eisenberg, an Atlas of Differential Diagnosis,* 4th ed. Philadelphia: Lippincott Williams & Wilkins, 2003.

DEFINITIONS AND BACKGROUND

NEC involves ischemia of the intestinal tract and invasion of the mucosa with enteric pathogens. This is a common GI problem in preterm and SGA infants with tissue injury and inflammation, congenital heart disease, or with Hirschsprung's disease. Symptoms and signs include a distended abdomen, lethargy, respiratory distress, pallor, hyperbilirubinemia, vomiting, diarrhea, grossly bloody stools, and sepsis.

NEC is the leading cause of short bowel syndrome in infancy; it is a medical emergency. NEC affects about 1–8% of all admissions to NICUs. Thrombocytopenia within the first 3 days after a diagnosis of NEC suggests a higher likelihood of bowel gangrene, morbidity, and mortality (Kenton et al, 2005). Neonatal endotoxemia and release of proinflammatory cytokines are important contributors to NEC; gut barrier failure plays an important role in adverse outcomes (Sharma et al, 2007).

If feeding intolerance is significant, it is beneficial to use breast milk rather than formula. Milk has distinct advantages over formulas; minimal enteral feeding regimens produce less NEC than more aggressive enteral feeding (Hay, 2008). There is no conclusive evidence about the use of special formulas that include glutamine or arginine. Preventive strategies include amino acid or polyunsaturated fatty acid administration (Reber and Nankervis, 2004).

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: NEC is not genetic in origin.

Clinical/History

- Height/length
- Head circumference
- Vomiting
- Diarrhea?
- Distended abdomen
- Lethargy
- Respiratory distress
- Pallor
- Hyperbilirubinemia
- Grossly bloody stools
- Sepsis

Lab Work

- Weight/birth weight
- CBC; elevated WBCs
- Thrombocytopenia (low platelet count)
- Neutropenia
- Hemoglobin (decreased)
- Hct (altered?)
- Abdominal x-rays

Diet/intake history

- Pro time (prolonged), INR
- Guaiac test for blood in stools
- Na^+ (decreased)
- K^+ (increased)
- Platelets (decreased)
- Gluc
- Lactic acidosis
- pO_2, pCO_2

INTERVENTION

OBJECTIVES

- TPN is needed for 14–21 days while the intestine heals.
- Prevent or correct dehydration, hypoglycemia, and electrolyte imbalances.
- Correct diarrhea and further malnutrition.
- Prepare patient for bowel surgery, for wound healing, and for the possibility of ostomy feeding if surgery becomes necessary, as for perforation or after peritonitis.
- Because breastfeeding is more protective than formula feeding, promote maternal breastfeeding or use of donor milk after TPN (Updegrove, 2004).

FOOD AND NUTRITION

- Acute: No oral feedings; use IVs to support circulation. TPN as appropriate with periods of extensive intestinal inflammation and peritonitis.
- Recovery: Use two times RDA of protein; 25% more kilocals than normal for age; frequent feedings. Where possible, offer donor milk if mother cannot breastfeed.

SAMPLE NUTRITION CARE PROCESS STEPS

Increased Energy Expenditure

Assessment Data: Weight loss, sepsis, diarrhea and vomiting, fever, stooling pattern.

Nutrition Diagnoses (PES): Increased energy expenditure related to NEC with losses from diarrhea and vomiting as evidenced by fever, bloody stools, distended abdomen, and altered lab values.

Intervention: Provide support for use of breastmilk, where possible.

Monitoring and Evaluation: Weight stabilization or gains in growth, cessation of bloody stools and diarrhea or vomiting, resolution of fever and sepsis, lab values returning to normal.

- Some partially elemental formulas are available, such as Pregestimil or Nutramigen, or more elemental nutrients may be required if the digestive tract has not recovered fully. Among infants between 1000 and 2000 g at birth, giving feedings at 30 mL/kg/d seems to be a safe practice and is faster than using 20 mL/kg/d (Caple et al, 2004).
- Ensure adequate iron, copper, and zinc. Iron-fortified products may reduce the need for blood transfusions in VLBW infants (Berseth et al, 2004). Copper seems to protect against TPN-related liver damage from intrauterine growth deficits (Zambrano et al, 2004).
- Occasionally, a colostomy or ileostomy must be performed, and tube feeding may be needed.

Common Drugs Used and Potential Side Effects

- Aggressive treatment of hyperglycemia may be needed, as with insulin (Hall et al, 2004). Monitor for side effects.
- Systemic antibiotics are started, usually a β-lactam antibiotic and an aminoglycoside. Some outbreaks may be infectious.

Herbs, Botanicals, and Supplements

- Studies suggest use of probiotics (Henry and Moss, 2004). A systematic Cochrane review promotes enteral feeding of probiotics in preterm infants >1000 g at birth but not for smaller infants (Alfaleh and Bassler, 2008). *Lactobacillus acidophilus* and *Bifudus infantis* may be beneficial.
- Herbs and botanicals should not be used for NEC.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Promote breastfeeding or use of donor milk.
- When formula is used, assure that the parent/caretaker understands the differences between ready-to-feed and concentrated formula related to, hypertonicity of the solution.
- If surgery and short-gut is the resolution, long-term TPN may be needed.
- Careful monitoring of growth is important. Besides bowel sequelae, VLBW infants who survive NEC are at risk for impairment of growth and neurodevelopment (Yeh et al, 2004).
- Monitor weight and stool changes; advise physician when necessary.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer. Powdered infant formulas are not sterile and may contain pathogenic bacteria; milk products are also media for bacterial proliferation (Agostoni et al, 2004).
- Do not use honey in the diets of infants to decrease potential risk of botulism.

For More Information

- Merck Manual
  http://www.merck.com/mrkshared/mmanual/section19/chapter260/260n.jsp
- Necrotizing Enterocolitis
  http://www.pediatrie.be/NECROT_%20ENTEROCOL.htm

NECROTIZING ENTEROCOLITIS—CITED REFERENCES

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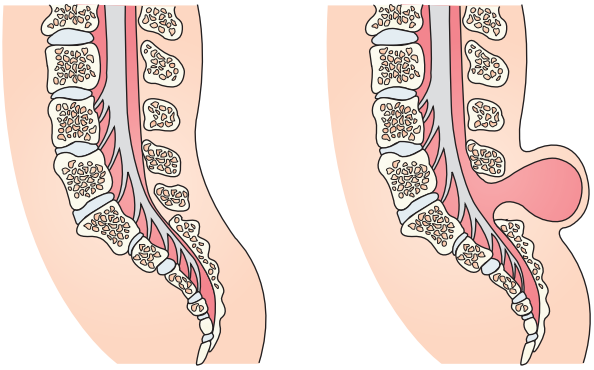
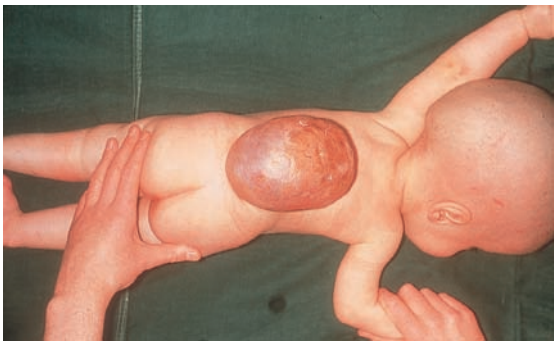
NEURAL TUBE DEFECTS: SPINA BIFIDA AND MELOMENINGOCELE

NUTRITIONAL ACUITY RANKING: LEVEL 3





Reprinted with permission from: Pillitteri A. *Maternal and Child Health Nursing,* 4th ed. Philadelphia: Lippincott Williams & Wilkins, 2003.

The neural tube folds and closes during the third and fourth weeks of pregnancy to form the brain and spinal cord (Li et al, 2006). Neural tube defects (NTDs) are serious birth defects of the spine (spina bifida) and brain (anencephaly), affecting approximately 3000 pregnancies each year in the United States. Women at risk for having a baby with NTDs are of English/Irish ancestry or Hispanic ancestry, may have diabetes mellitus, are obese, have poor dietary habits, or take medications that are folate antagonists.

The natural folates are chemically unstable and poorly bioavailable in contrast to the chemical form, folic acid (Scott, 2009). Periconceptional consumption of folic acid (400 mg daily) reduces the occurrence of NTDs by 50–70% (Centers for Disease Control and Prevention, 2004; Hamner et al, 2009). No more than one fifth of women take supplements effectively, largely due to the fact that over half of pregnancies are unplanned (Scott, 2009).

A remarkable successes of epidemiology was the demonstration in the late twentieth century that spina bifida and anencephaly are caused primarily by folate deficiency (Oakley, 2009). The genetic MTHFR polymorphisms may cause congenital folate malabsorption, severe MTHFR deficiency, and formiminotransferase deficiency (Whitehead, 2006). In these cases, diets deficient in folate *do not* influence the incidence or severity of NTDs; folic acid and methionine cycle genes for SNP genotyping alter the effects of diet (Boyles et al, 2006).

Low levels of betaine-tHcy methyltransferase (BHMT) and choline may be associated with NTDs (Boyles et al, 2006; Innis et al, 2007; Shaw et al, 2004). Betaine (trimethylglycine) is formed from choline, or from the diet (seafood, wheat germ, bran, and spinach). Risks may be greater when vitamin B_{12} status is low (van der Linden et al, 2006). Because results vary from one population to another, studies are needed to clarify how different populations respond to betaine and choline from diet or supplements.

Spina bifida includes any congenital defect involving insufficient closure of the spine (usually laminae of the vertebrae). Most defects occur in the lower lumbar or sacral areas of the back (the lowest areas of the back) because this area is the last part of the spine to close. **Spina bifida occulta** involves the bones of the spine not closing completely while the spinal cord and meninges remain in place; skin covers the defect. **Spina bifida cystica** is more severe. **Anencephaly** forms when the brain does not close; the baby lacks parts of the brain, skull, or scalp, may be deaf, blind, and usually will not thrive.

In **meningoceles**, the meninges (the membranes covering the spinal cord) protrude through the vertebral defect but the spinal cord remains in place. Clubfoot, dislocated hip, scoliosis, and other musculoskeletal deformities may also be present. **Myelomeningocele (MMC)** is one of the most severe forms of birth defects of the brain and spinal cord. MMC accounts for about 75% of all cases of spina bifida, affecting 1 of every 800 infants. Pregnant women may show a positive alpha-fetoprotein level during prenatal testing in a triple screen. The spinal canal is incomplete. This causes decreased or lack of function of body areas controlled at or below the defect. Partial or complete paralysis of the legs, partial or complete lack of sensation, loss of bowel or bladder control, meningitis, hydrocephalus, and hip dislocation may also be present. Sometimes, surgery for the tethered cord or to repair the hydrocephaly improves quality of life. MMC patients are usually wheelchair bound, wear braces, or use crutches. Obesity can increase likelihood of pressure ulcers or make ambulation and surgery more difficult.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: Folate status is a significant determinant of NTD risk; genetic variation, folate nutriture, and specific metabolic, and/or genomic pathways are involved (Beaudlin and Stover, 2007). Women who have a methylenetetrahydrofolate reductase (MTHFR) allele tend to have a higher risk for giving birth to an infant with an NTD. Hispanic and Latina women,

women from the United Kingdom and Italy may have this allele and should be tested before pregnancy.

Clinical/History

Height, weight
Birth weight/length
Growth percentile
Weight changes
Diet/intake history (folate, choline, B_{12}, betaine)
Temperature
I & O
Vertebral defect
Spinal x-rays or computed tomography (CT) scan
Diarrhea or Constipation
Incontinence
Skin integrity
Hydrocephaly

Lab Work

Gluc
Alb, transthyretin
H & H
Serum Fe
Chol
Na^+, K^+
Ca^{++}, Mg^{++}
Serum folic acid
MTHFR allele?
Hcy
Serum B_{12}, Methylmalonic acid

INTERVENTION

OBJECTIVES

- Manage feeding problems, which are common. Assure proper positioning for all feedings.
- Achieve and maintain ideal BMI for age. There are decreased energy needs because of short stature and limited mobility. Obesity is common.
- Reduce impact of the defect. Promote any and all possible ambulation or activity.

SAMPLE NUTRITION CARE PROCESS STEPS

Inadequate Folic Acid Intake

Assessment Data: Prenatal weight gain, dietary records, and nutrient analysis, MTHFR allele.

Nutrition Diagnoses (PES): NI 54.2. Inadequate vitamin (folic acid) intake related to diet history and increased needs during pregnancy as evidenced by diet recall with folate intake much lower than needs in pregnancy (600 mcg) and prior birth of an infant with NTD.

Interventions:

Food and Nutrient Delivery:

ND 1.3. Add natural sources of folate to diet—orange juice, carrots, green vegetables.

ND 1.3. Eat fortified cereal at breakfast and whole grains such as oatmeal, brown rice.

ND 3.2. Recommend folic acid supplement of 400 μg/d.

Education: Rich folate sources from food; importance of taking a daily multivitamin supplement.

Counseling: Investigate need for L-methylfolate during pregnancy related to genetic allele.

Monitoring and Evaluation: Weight and health records. Prenatal care and intake of supplemental folic acid. Note of willingness to comply with recommendations.

- Correct infections; prevent or correct sepsis and pressure ulcers.
- Correct nutrient deficiencies.
- Alter diet to prevent or correct constipation, obesity, and UTIs.
- Increase independence and self-care potentials.
- Preserve brain function as far as possible.
- Correct swallowing problems (from Arnold–Chiari malformation of the brain).
- Initiate treatment or surgical intervention as appropriate.

FOOD AND NUTRITION

- Individualize diet for proper nutrition to achieve a desirable weight and monitor carefully.
- Decrease energy to control weight. Use special growth charts. The standard CDC charts will be irrelevant. For children under age 8 who are minimally active, use 9–11 kcal/cm or 50% fewer calories than recommended for child of a similar age (Lucas, 2004, p. 41). For older teens or adults, 7 kcal/cm may be needed for weight loss; generally, needs are about 50% of normal.
- Provide adequate protein, folic acid, B-complex vitamins, choline, betaine, zinc, and other nutrients for age.
- Low-calorie snacks may be the only between-meal snacks allowed.
- Provide adequate nutrients for wound healing if surgery has been performed. For healing of any pressure ulcers, adequate zinc, vitamins A and C, and protein are required.
- For females of childbearing age, pay attention to folic acid, choline, vitamin B_{12}, betaine intakes. A multivitamin with minerals should be recommended for those who lack variety in their diets.
- Ensure adequate fiber intake and fluid to prevent or correct problems with diarrhea or constipation.
- Individuals with spina bifida are often allergic to latex; they may require a diet that limits use of apples, avocado, banana, bell pepper, cherries, chestnut, kiwi, nectarines, peach, plums, potato, and tomato.

Common Drugs Used and Potential Side Effects

- Antibiotics may be required if the patient develops sepsis. Use of acidophilus and probiotic products may alleviate loss of intestinal bacteria.
- Avoid zinc and iron with parenteral administration in sepsis; these are bacterial nutrients.
- Botulinum-A toxin injections have been used in cases of neurogenic detrusor overactivity to manage some bladder incontinence (Leippold et al, 2003). Otherwise, medications for managing urinary incontinence may be used; monitor for side effects.
- Medications prescribed may affect the utilization of folate: anticonvulsant medications (e.g., phenytoin), metformin, sulfasalazine, triamterene, and methotrexate. Other medications deplete important nutrients prenatally, such as antibiotics, antihypertensives, cathartics, corticosteroids, stimulants, sulfonamides, and tranquilizers. Vitamin and mineral supplements are needed to compensate for the specific nutrient alteration.

- Sulfonamides may crystallize vitamin C in the bladder; extra vitamin C, protein, iron, and folate may be needed.

Herbs, Botanicals, and Supplements

- Herbs and botanicals should not be used because there are no controlled trials to prove efficacy for any related problems.
- Use of low-calorie cranberry juice may help reduce UTIs.

NUTRITION EDUCATION, COUNSELING, CARE MANAGEMENT

- Behavior modification, low-calorie food and snack preparation, rewards, and activity/exercise factors should be reviewed with the parent/caretaker. Food lists with green "go" foods, red "stop" foods, and yellow "caution" foods have been used with success in weight management.
- Discuss potential medical conditions, such as fractures, seizures, lazy eye, early puberty, and latex allergy. Bone health and allergies can be managed with some nutritional interventions.
- Family counseling may be needed in preparation for future pregnancies. Referral to a local chapter of the March of Dimes may be beneficial.
- Even with food fortification, women of childbearing age should be advised to take a folic acid–containing supplement on a daily basis (Schuaibi et al, 2008). Choline, betaine, and B_{12} should be included as well.

Patient Education—Food Safety

- Hand washing with soap and hot water is recommended before preparing formula or meals. Use clean utensils and containers for mixing formula.
- Before using tap water for formula preparation or to give as a beverage, let cold tap water run for 2 minutes to remove any lead that may be in the pipes.
- Follow the 2-hour rule: discard any beverage or food that has been left at room temperature for 2 hours or longer.
- Do not use honey in the diets of infants to decrease potential risk of botulism.

For More Information

- Association for Spina Bifida and Hydrocephalus
  www.asbah.org
- CDC—Spina Bifida
  http://www.cdc.gov/ncbddd/birthdefects/SpinaBifida.htm
- CDC Folic Acid National Campaign
  http://www.cdc.gov/ncbddd/Folicacid
- Management of Myelomenigocele Study
  http://www.spinabifidamoms.com/english/index.html
- NIH—Spina Bifida
  http://www.ninds.nih.gov/disorders/spina_bifida/spina_bifida.htm
- Pregnancy Planning
  http://www.cdc.gov/ncbddd/pregnancy/
- Spina Bifida Association
  http://www.sbaa.org

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Schuaibi AM, et al. Folate status of young Canadian women after folic acid fortification of grain products. *J Am Diet Assoc.* 108:2090, 2008.

Shaw GM, et al. Periconceptional dietary intake of choline and betaine and neural tube defects in offspring. *Am J Epid.* 160(2):102, 2004.

Van Der Linden IJ, et al. The methionine synthase reductase 66 A>G polymorphism is a maternal risk factor for spina bifida. *J Mol Med.* 84(12): 1047, 2006.

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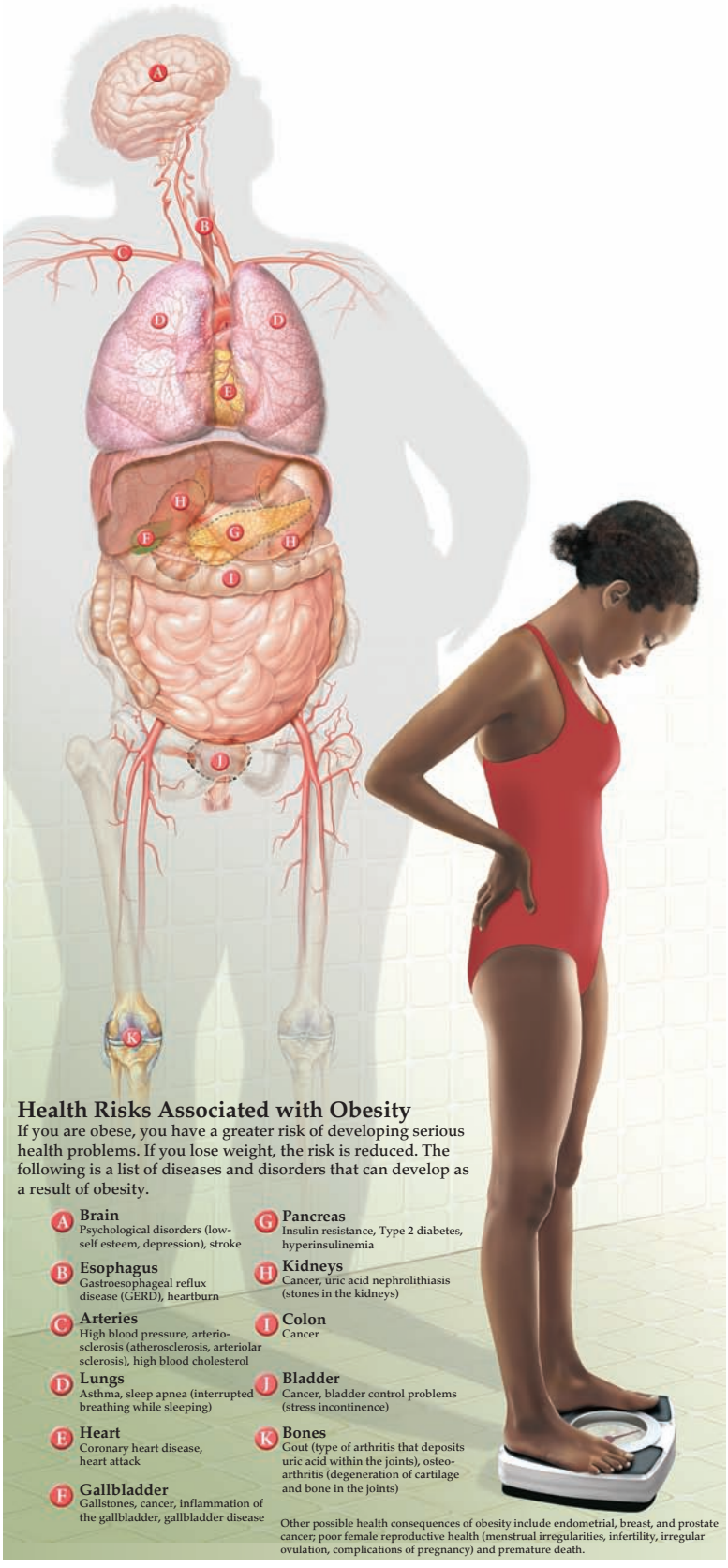
OBESITY, CHILDHOOD

NUTRITIONAL ACUITY RANKING: LEVEL 3–4 (COUNSELING)



Asset provided by Anatomical Chart Co.

DEFINITIONS AND BACKGROUND

The prevalence of overweight is increasing for children and adolescents in the United States. "At risk for overweight" is defined by the sex- and age-specific ≥85th percentile cutoff points of the CDC growth charts or of BMI for age; overweight or obese is defined as ≥95th percentile of growth charts or BMI for age. BMI increases during the first year of life and then decreases; it begins to rise again at 6–6.5 years of age. BMI tables are not useful before age 2; they are a screening tool and do not reflect body composition well. While BMI tables have limitations, they are considered a reasonable place to begin; an increase in BMI of three to four units is a reason to investigate further.

The preferred weight gain pattern in childhood is as follows: infant doubles birth weight by 6 months and triples birth weight at 12 months. Tripling birth weight before 1 year is associated with increased risk of obesity. In year 2, gain is 8–10 lb (3.5–4.5 kg); in year 3, gain is 4.5–6.5 lb (2–3 kg); annually thereafter, the gain is about 4.5–6.5 lb (2–3 kg). Until 6 years of age, the number of fat cells increases (hyperplasia). After 6 years of age, the size of fat cells increases (hypertrophy). Hormones play a role. Leptin, insulin, and adiponectin regulate lipid metabolism.

TABLE 3-13 Complications of Childhood Obesity

Psychosocial	Poor self-esteem Anxiety Depression Eating disorders Social isolation Lower educational attainment
Neurologic	Pseudotumor cerebri
Endocrine	Insulin resistance Type 2 diabetes Precocious puberty Polycystic ovaries (girls) Hypogonadism (boys)
Cardiovascular	Dyslipidemia Hypertension Coagulopathy Chronic inflammation Endothelial dysfunction
Pulmonary	Sleep apnea Asthma Exercise intolerance
Gastrointestinal	Gastroesophageal reflux Steatohepatitis Gallstones Constipation
Renal	Glomerulosclerosis
Musculoskeletal	Slipped capital femoral epiphysis Blount's disease[a] Forearm fracture Back pain Flat feet

[a]Blount's disease is a growth disorder of the tibia that causes the lower leg to angle inward (tibia vara).

Obesity is an epidemic. Rates of unhealthy body weight among children and adolescents have tripled since the 1980s. Three critical periods for prevention of adult obesity are: ages 5–7 years, adolescence, and pregnancy. Interaction between genes in the fetus and maternal overnutrition or undernutrition are relevant; obese women should attempt healthy weight loss before they become pregnant (American Dietetic Association, 2009). Women should then be encouraged to breastfeed.

After birth, overfeeding for catch-up growth in a premature or underweight child can contribute to obesity; weight gain proceeds at a rate that is too fast for linear growth. Overnutrition, resulting from high birth weight or gestational diabetes, is associated with subsequent fatness in the child. Many adult-onset disorders are showing up in obese children; see Table 3-13.

Research suggests that phthalates from soft plastics disrupt endocrine glands and affect hormones that regulate bodily functions; long-term exposure may contribute to obesity. In addition to genetics and environment, social factors in childhood also strongly influence adult obesity. Parents should provide children with access to nutrient-dense foods and beverages and high-fiber foods; reduce children's access to high-calorie, nutrient-poor beverages and foods both at home and at restaurants; avoid excessive food restriction; do not use food as a reward; and encourage children to eat breakfast on a daily basis (Ritchie et al, 2005). A Lifestyle Behavior Checklist may be used to identify problems that parents have in managing the behavior of their children and their confidence in doing so (West and Sanders, 2009). Management of preadolescent obesity seems to be most successful when it is started during preschool years.

Counseling must distinguish between "simple obesity" and severe or "morbid" obesity in the child, as well as the comorbidities. Major attitudinal changes are often needed in parents or caretakers when a child has reached the severe/morbid phase. Table 3-14 offers tips for weight loss plans.

ASSESSMENT, MONITORING, AND EVALUATION

CLINICAL INDICATORS

Genetic Markers: Twin studies have shown that 40–70% of the variation in BMI is inherited. The genetics of obesity is a work in progress; see Web site http://obesitygene.pbrc.edu/. Research suggests an association between the FTO, MC3R, seven other genes, and overweight.

Clinical/History

- Maternal gestational diabetes
- Gestational age at birth
- Birth length, birth weight
- Height
- Present weight
- Weight hx
- Diet/intake history
- BMI 85–94% = at risk of continuing overweight into adulthood
- BMI ≥95% = overweight with need for in-depth assessment