Lesson 4: Understanding Inborn Errors of Metabolism

### Lesson-at-a-Glance

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<td><strong>Objective 1:</strong> Define inborn errors of metabolism, identify the more common errors, and explain dietary treatment for children with these disorders.</td>
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<td>Objective 1</td>
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<td><strong>Objective 2:</strong> Describe cafeteria accommodations for these children and understand the need for a professional consultant in difficult cases.</td>
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Lesson Plan

Overview
This lesson is intended to provide background information on inborn errors of metabolism. It can be used alone or in a series with the other lessons in this manual. This lesson complements material on pages 27-31 in the *Handbook for Children with Special Food and Nutrition Needs*.

You may decide to invite your school nurse to attend this lesson since inborn errors of metabolism are serious disorders with very strict dietary implications. Inclusion of the school nurse will reinforce the need to collaborate when serving meals to students with special dietary needs.

Pre-Lesson Activities
As participants check in for the class, distribute a copy of the Pre-Training Assessment to each participant. Collect completed Pre-Training Assessments prior to beginning the lesson. Copies of the Pre- and Post-Training Assessments and answer keys are found at the end of the lesson. Allow approximately 5-10 minutes for the Pre-Training Assessment. Stress that the purpose of the Pre-Training Assessment is to help the trainer plan for the presentation of future lessons. It is to measure how much class participants have learned by the end of the lesson.

Introduction  5 minutes

Slide 1

**Meeting Children’s Special Food and Nutrition Needs in Child Nutrition Programs**  
**Lesson 4: Understanding Inborn Errors of Metabolism**

Learning Objectives
- Define inborn errors of metabolism, identify the more common errors, and explain dietary treatment for children with inborn errors of metabolism.
- Describe cafeteria accommodations for these children and understand the need for a professional consultant in difficult cases.

**Tell:** Welcome to *Meeting Children’s Special Food and Nutrition Needs in Child Nutrition Programs*. This Breakfast Lunch Training Module, or BLT, provides information on how you can meet the needs of children in your school who have inborn errors of metabolism.
Inborn errors of metabolism are rare genetic disorders that require dietary restrictions. Failure to follow these restrictions can result in serious complications, including mental retardation, organ failure, and death. If a student in your school has an inborn error of metabolism, it is important for you to understand the condition and the foods that are allowed.

**Note to Instructor:** Conduct the Introductory Activity by following the directions listed on the Introductory Activity Instruction Sheet found at the end of the lesson. Give directions, complete the activity, and summarize. After the activity is completed, continue with the lesson.

Inborn errors of metabolism are considered disabilities under Section 504 of the Rehabilitation Act of 1973 and the Americans with Disabilities Act of 1990 (U.S. Department of Agriculture [USDA], 2001). These Acts require schools to make accommodations for students with inborn errors of metabolism.

**Read:** The learning objectives for this lesson are

- to define inborn errors of metabolism, to identify the more common errors, and to explain dietary treatment for children with inborn errors of metabolism, and
- to describe cafeteria accommodations for these children and to understand the need for a professional consultant in difficult cases.

**Objective 1: Define inborn errors of metabolism, identify the more common errors, and explain dietary treatment for children with inborn errors of metabolism.** 12 minutes

**Slide 2**

<table>
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<tr>
<td>• Rare genetic disorders in which the body cannot metabolize food normally</td>
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<tr>
<td>• By-products of metabolism, amino acids, sugars, fatty acids build up in the body, causing serious complications</td>
</tr>
<tr>
<td>• Dietary treatment: strict diet management to avoid toxic buildup of dietary by-products</td>
</tr>
<tr>
<td>• Special foods or formulas may be needed</td>
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</table>

**Tell:** Inborn errors of metabolism are rare genetic disorders in which the body cannot normally turn food into energy (A.D.A.M., 2004). The process of turning food into energy is called metabolism. During normal metabolism, enzymes, which are proteins that help in chemical reactions, break down the food components of carbohydrate, fat, and protein into fuel, and all waste from this process is eliminated from the body. Carbohydrates are broken down into
starches and sugars, fats into fatty acids, and proteins into amino acids—all of which are further broken down to be burned or stored as energy.

Inborn errors of metabolism are caused by defects in one or more metabolic enzymes. It is crucial that people with inborn errors of metabolism avoid eating anything that their bodies cannot completely break down. If they eat these foods, unmetabolized nutrients like sugars, fatty acids, or amino acids can build up in their bodies and cause serious complications such as mental retardation or death (A.D.A.M., 2004).

In many states, infants are screened at birth for several inborn errors of metabolism. If an error is detected an infant will immediately be placed on a special diet, which must be followed for the rest of his or her life. Dietary treatment is different for each disorder and can even vary among individuals with the same inborn error of metabolism.

Typically, treatment involves strict dietary management to prevent that person from eating anything that will cause unmetabolized nutrients to build up. Special products like formulas or modified foods are often needed to provide complete nutrition to an individual with such a restricted diet. The type and extent of the changes depend on the specific metabolic error. Registered dietitians and physicians can help with the diet modifications needed for each disease (National Food Service Management Institute [NFSMI], 2006; A.D.A.M., 2004).

Today we will discuss four specific inborn errors of metabolism so that you can become familiar with the required dietary modifications. There are many other disorders that may affect students. Always follow the strict guidelines given for any child with an inborn error of metabolism.

**Slide 3**

**Phenylketonuria (PKU)**

- Cannot process the amino acid phenylalanine
- Dietary treatment:
  - low-protein diet (to prevent increase in phenylalanine)
  - special formula to provide protein

**Tell:** Phenylketonuria (pronounced fen-il-key-toe-noo-ree-a), or PKU, is a deficiency of the enzyme that processes the amino acid phenylalanine (pronounced fen-il-al-a-nee). The resulting elevated levels of phenylalanine in the blood and other tissues can cause mental retardation, delayed speech, seizures, eczema, behavior abnormalities, and other symptoms (National Institutes of Health, 2000).
Lesson 4: Understanding Inborn Errors of Metabolism

Dietary treatment includes adherence to a strict low-protein diet including special low- or no-phenylalanine products and modified foods. Protein sources like milk, cheese, eggs, meat, and fish are not allowed. Special high-protein formulas that do not contain phenylalanine are required daily to provide essential nutrients. Some vegetables, fruits, and grain products may be allowed depending on the individual’s phenylalanine tolerance (March of Dimes, 2006).

**Slide 4**

**Galactosemia**
- Cannot process the sugar galactose
- Dietary treatment: no milk or dairy products

**Tell:** Galactosemia (pronounced ga-lak-toe-see-me-a) is an enzyme deficiency leading to elevated levels of galactose in the blood. Galactose is a sugar found in milk. Untreated galactosemia can cause liver, brain, eye, and kidney damage. Individuals with galactosemia cannot have any milk or dairy products in their diet (American Liver Foundation, 2006).

It is extremely important to read labels when feeding an individual with galactosemia since dairy ingredients include many items other than milk and can be present in unexpected foods. Dairy ingredients include butter, nonfat milk, milk, cheese, nonfat dry milk solids, lactose, casein, whey and whey solids, sodium caseinate, calcium caseinate, and lactalbumin.

**Slide 5**

**Hereditary Fructose Intolerance**
- Cannot process the sugar fructose
- Dietary treatment:
  - no fructose (high-fructose corn syrup, honey, fruit)
  - no sucrose (table sugar)
Tell: Individuals with hereditary fructose intolerance do not produce an enzyme that metabolizes the sugar fructose, which causes extremely low blood sugar and liver damage or failure. The treatment is complete elimination of fructose and sucrose from the diet (A.D.A.M., 2005a). Fructose is typically found in fruit, honey, and commercial sweeteners such as high-fructose corn syrup. Sucrose, the scientific name for common table sugar, is broken down into fructose. It is important to read labels since sugar and high-fructose corn syrup are common ingredients in many foods and beverages.

Slide 6

Maple Syrup Urine Disease
- Cannot process the branched chain amino acids leucine, isoleucine, valine
- Dietary treatment:
  - low-protein diet
  - special formula to provide protein

Tell: Individuals with maple syrup urine disease are unable to metabolize the branched-chain amino acids leucine, isoleucine, and valine. Buildup of these amino acids can lead to neurological damage and death. The name of this disorder comes from the distinct odor of an affected individual’s urine. Individuals with maple syrup urine disease require a special diet similar to the one for PKU, which calls for a severe limitation of protein sources and supplementation with a special formula to meet protein needs (A.D.A.M., 2005b).

Note to Instructor: Conduct the Matching Activity by following the directions listed on the Matching Activity Instruction Sheet found after the lesson. Give directions, complete the activity, and summarize. After the activity is completed, continue with the lesson.

Objective 2: Describe cafeteria accommodations for these children and understand the need for a professional consultant in difficult cases.

Note to Instructor: Using the Video Clip
At this point, the class participants will view the video clip that corresponds with this lesson. The two purposes of this video clip are to summarize what the class participants have learned about inborn errors of metabolism in the course and to explain what food service assistants can do to help students with inborn errors of metabolism.
Lesson 4: Understanding Inborn Errors of Metabolism

Summary of the Video Clip
The video clip begins by introducing a child with PKU who explains what it is like to have such a restricted diet. She will go through the lunch line and show what accommodations are being made for her diet.

Tell: We will now view a video clip about a student who has PKU. Pay particular attention to what you learn about the role of the food service assistant in helping a student with an inborn error of metabolism.

Show Video Clip

Ask: How did the food service assistants in the video clip help this student?

Slide 7

Tell:
1. Communication among the parents, teacher, school nurse, food service, and consultant is essential to promote understanding of the precise dietary needs of the student with an inborn error of metabolism. Because the diet for students with inborn errors of metabolism can be complex, a nutrition or genetic specialist may be brought in as a consultant to review menus and determine what additional foods, products, or formulas, if any, are needed to follow the physician’s orders (NFSMI, 2006). This consultant may also provide special training to the food service staff.

2. A plan that details these dietary restrictions will be on file with the school nurse. This plan will be updated as dietary restrictions are adjusted over time due to growth, illness, or changes in nutrient tolerance. The school nurse will notify food service of changes and will explain how to make proper accommodations. A list of specific accommodations for students with inborn errors of metabolism should be kept in the cafeteria for food service assistants to reference.
3. Special formulas or foods may be needed to meet the student’s nutritional needs, and these may be provided by the parent or obtained by food service. Students should not be charged extra for these special formulas or foods. A consultant may need to train food service staff in preparation of special meals or dietary products. If food service assistants are unsure of any instructions regarding a student with an inborn error of metabolism, they should discuss the issue with the food service supervisor.

4. It is critical to follow prescribed portion sizes for all foods when serving a student with an inborn error of metabolism. Even though certain foods are allowed, excessive portions of these foods may cause dangerous levels of unmetabolized nutrients in the body. The medical prescription must be followed exactly as written, using only allowed substitutions (NFSMI, 2006).

5. Mistakes occasionally happen, and it is important to immediately communicate any mistakes to the school nurse or other school personnel in charge of the student’s health care. The rest of the day’s diet may need to be altered to correct an accidentally large portion of an allowed food item at lunch. Medical care may be needed if the student eats something that is not allowed (NFSMI, 2006).

6. Remember that all information obtained about a child who needs special meal accommodations should be kept confidential. The documentation should be kept in a secure location and should not be discussed with other students or anyone else that does not have a need to know (USDA, 2001).

Note to Instructor: Conduct the What Should You Do? Activity by following the directions listed on the What Should You Do? Instruction Sheet found after the lesson. Give directions, complete the activity, and summarize. After the activity is completed, continue with the lesson.

Summary and Conclusion 1 minute

Note to Instructor: Summarize and close the lesson as follows.

Tell: Students with inborn errors of metabolism need to follow highly specialized diets to avoid complications that are often life threatening. Careful food selection and preparation by food service assistants ensure that these students can safely eat school meals in the cafeteria.

Post-Lesson Activity: Distribute the Post-Training Assessment. Allow the class participants 5 minutes to complete it. Have the participants score their own assessments as the class reviews the correct answers found in the Post-Training Assessment Answer Key. Collect the assessments to evaluate the effectiveness of the class.
Lesson 4: Understanding Inborn Errors of Metabolism

References


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Preparation Checklist

<table>
<thead>
<tr>
<th>Task</th>
<th>Completed</th>
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</thead>
<tbody>
<tr>
<td>Announce meeting time and purpose in advance.</td>
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<tr>
<td>Make arrangements for meeting room.</td>
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<tr>
<td><strong>Reserve equipment and gather supplies.</strong></td>
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<tr>
<td><strong>Equipment needed:</strong></td>
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<tr>
<td>Computer and projector for the DVD video and slides</td>
<td></td>
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<tr>
<td>Screen or other surface for projection</td>
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<tr>
<td>DVD player and television to play video clip, if not using computer and projector</td>
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<tr>
<td><strong>Supplies needed:</strong></td>
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<td>BLT 2006 video</td>
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<tr>
<td>BLT 2006 slide presentation</td>
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<tr>
<td>For Introductory Activity: white board and marker OR chalkboard and chalk OR large paper, marker and tape</td>
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<tr>
<td>Name tags (optional), pencils or pens, folders to hold handouts (one for each class member)</td>
<td></td>
</tr>
<tr>
<td><strong>Make copies of all class handouts and display materials.</strong></td>
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<tr>
<td><strong>Handout and display materials needed:</strong></td>
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<tr>
<td>Introductory Activity Handout and the list of foods written for display on a white board, chalkboard, or large piece of paper</td>
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<tr>
<td>Matching Activity Handout</td>
<td></td>
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<tr>
<td>What Should You Do? Activity Handout</td>
<td></td>
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<tr>
<td>Fact Sheet for Lesson 4</td>
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<tr>
<td>Resource Sheet for Lesson 4</td>
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<tr>
<td>Pre-Training Assessment</td>
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<tr>
<td>Post-Training Assessment</td>
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<tr>
<td><strong>Preview video clip for Lesson 4</strong></td>
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</tbody>
</table>
**Preparation**
Before you begin the class, make copies of the *Introductory Activity Handout* on the following page. Make one copy for each participant. Write the following words on a white board, chalkboard, or large sheet of paper taped to the wall: low-fat vanilla yogurt, baked beans, multi-grain crackers.

**Activity Instructions**
Show participants the list of the three foods.

**Tell:** Imagine that you have a disorder that does not allow you to eat any sugar or fructose (which is a type of sugar). Which of these foods would you be able to eat?

**Note to Instructor:** Ask for a volunteer to give an answer. Ask the rest of the participants if they agree with it. Distribute the *Introductory Activity Handout* containing ingredient labels for the three foods. Have participants examine the labels for a few seconds, and then ask if they would change their answers.

**Tell:** As you can see, all of these foods contain sugar. The yogurt contains high fructose corn syrup, the beans contain sugar, and the crackers contain sugar and high fructose corn syrup. None of these foods would be allowed on a diet that eliminates sugar or fructose. Imagine how difficult it is to follow this diet. It is important to check all labels when serving a student with a serious dietary restriction.
Introductory Activity Handout

Low-fat Vanilla Yogurt
INGREDIENTS: CULTURED PASTEURIZED GRADE A NONFAT MILK, HIGH FRUCTOSE CORN SYRUP, MODIFIED CORN STARCH, WHEY PROTEIN CONCENTRATE, KOSHER GELATIN, NATURAL FLAVOR, ASPARTAME, POTASSIUM SORBATE ADDED TO MAINTAIN FRESHNESS, VITAMIN A ACETATE, COLORED WITH TURMERIC AND ANNATTO EXTRACT, VITAMIN D3.

Baked Beans
INGREDIENTS: WATER, PREPARED WHITE BEANS, SUGAR, MUSTARD, SALT, CORN STARCH, ONION POWDER, CARAMEL COLOR, TAPIOCA MALTODEXTRIN, AUTOLYZED YEAST EXTRACT, NATURAL FLAVORS.

Multi-grain Crackers
INGREDIENTS: WHOLE GRAIN WHEAT FLOUR, ENRICHED FLOUR, BARLEY FLAKES, SOYBEAN OIL, SUGAR, HIGH FRUCTOSE CORN SYRUP, RYE, TRITICALE (A GRAIN), MILLET, MOLASSES, SALT, WHOLE WHEAT, LEAVENING, EMULSIFIERS, ROLLED OATS, ONION POWDER, CORNSTARCH.
Matching Activity Instruction Sheet

Preparation
Before you begin the class, make copies of the Matching Activity Handout on the following page. Make one copy for each participant.

Activity Instructions
Divide participants into teams of three or four people. Distribute the Matching Activity Handout to participants.

Tell: In this activity you will determine which school meal is appropriate for students with each of the inborn errors of metabolism we just discussed.

Read: Below are two lists. The first one lists the inborn errors of metabolism that we have already discussed. The second one lists school meals that have been specially modified. Work together with your teammates to match the meal to the appropriate inborn error of metabolism, and write the letter of that meal on the line.

Note to instructor: Give participants 3 minutes to complete the activity. The correct answers can be found on the Matching Activity Handout Answer Key. Review the correct answers with the participants.

Tell: Although labels were not provided for this activity, it is important to check the ingredient list on a label when serving a food to a student who cannot have common ingredients like sugar or milk.
### Instructions:

Below are two lists. The first one lists the inborn errors of metabolism that we have already discussed. The second one lists school meals that have been specially modified. Work together with your teammates to match the meal to the appropriate inborn error of metabolism, and write the letter of that meal on the line.

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<td>2. Galactosemia – inability to metabolize the sugar galactose</td>
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<td>4. Maple syrup urine disease – inability to metabolize the branched-chain amino acids leucine, isoleucine, and valine</td>
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</tr>
</tbody>
</table>

**Meals**

A. Cheeseburger on sugar-free bun, low-fat white milk, and carrot sticks

B. Specially modified, low-protein pizza, green beans, fruit salad, and low-phenylalanine formula

C. Specially modified, low-protein mushroom burger, salad with oil and vinegar, apple, and low-branched-chain amino acid formula

D. Baked chicken with brown rice, broccoli, and apple juice
Matching Activity Handout Answer Key

**Instructions:** Below are two lists. The first one lists the inborn errors of metabolism that we have already discussed. The second one lists school meals that have been specially modified. Work together with your teammates to match the meal to the appropriate inborn error of metabolism, and write the letter of that meal on the line.

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**Meals**

A. Cheeseburger on sugar-free bun, low-fat white milk, and carrot sticks

B. Specially modified, low-protein pizza, green beans, fruit salad, and low-phenylalanine formula

C. Specially modified, low-protein mushroom burger, salad with oil and vinegar, apple, and low-branched-chain amino acid formula

D. Baked chicken with brown rice, broccoli, and apple juice
**Preparation**
Before you begin the class, make copies of the *What Should You Do? Activity Handout* on the following page. Make one copy for each participant.

**Activity Instructions**
Distribute the *What Should You Do? Activity Handout* to participants.

**Tell:** In this activity you will read three scenarios and decide what your next step should be. We will discuss the scenarios briefly after you have had a couple of minutes to think about them.

**Read:** Use the same teams you were in for the last activity. As a team, read through each scenario below and decide what the food service assistant should do. Write down your suggestion after each scenario.

**Note to instructor:** Give participants 5 minutes to complete the activity. The correct answers can be found on the *What Should You Do? Activity Handout Answer Key*. Review the correct answers with the participants. You should customize the responses so they reflect your department’s policies.
What Should You Do? Activity Handout

Instructions: Use the same teams you were in for the Matching Activity. As a team, read through each scenario below and decide what the food service assistant should do. Write down your suggestion after each scenario.

1. Caroline, a student with phenylketonuria (PKU), tells you that she would like one of the salads you are serving to her friends, but this item is not on her list of approved foods. What should you do?

2. As the cafeteria cashier you notice that the point-of-sale computer system is alerting you that George, the student checking out, has galactosemia and cannot have milk or any item containing dairy. What should you do?

3. The lunch period was hectic, and as the students are throwing away their trash you realize that Travis, a student with maple syrup urine disease, was mistakenly given regular pizza instead of his specially modified low-protein pizza. What should you do?
What Should You Do? Activity Handout Answer Key

Instructions: Use the same teams you were in for the last activity. As a team, read through each scenario below and decide what the food service assistant should do. Write down your suggestion after each scenario.

1. Caroline, a student with phenylketonuria (PKU), tells you that she would like one of the salads you are serving to her friends, but this item is not on her list of approved foods. What should you do?

   *It is essential that you never serve non-approved foods to a student with PKU, regardless of the student’s request. Serve Caroline her approved meal, and notify the food service manager or food service director of the situation so it can be discussed with the school nurse or Caroline’s parents. Try not to make a scene that will draw attention to the student and her health condition.*

2. As the cafeteria cashier you notice that the point-of-sale computer system is alerting you that George, the student checking out, has galactosemia and cannot have milk or any item containing dairy. What should you do?

   *This is a good opportunity to do a final check of George’s tray to make sure it only contains approved items. Be discreet so information about his health condition remains confidential.*

3. The lunch period was hectic, and as the students are throwing away their trash you realize that Travis, a student with maple syrup urine disease, was mistakenly given regular pizza instead of his specially modified low-protein pizza. What should you do?

   *Notify the school nurse and Travis’s teacher IMMEDIATELY so they can proceed according to his medical plan. Evaluate the serving line and cafeteria to determine how Travis got regular pizza so the situation can be avoided in the future.*
Fact Sheet

Inborn errors of metabolism are rare genetic disorders in which the body cannot normally turn food into energy (A.D.A.M. Medical Encyclopedia, 2004).

The process of turning food into energy is called metabolism. During normal metabolism, enzymes break down the food components of carbohydrate, fat, and protein into fuel, and all waste is eliminated from the body. Carbohydrates are broken down into starches and sugars, fats into fatty acids, and proteins into amino acids—all of which will be used or stored as energy.

Inborn errors of metabolism are caused by defects in one or more metabolic enzymes. People with inborn errors of metabolism need to avoid eating anything that their bodies cannot completely break down (A.D.A.M. Medical Encyclopedia, 2004).

Phenylketonuria, or PKU, is a deficiency of the enzyme that processes the amino acid phenylalanine (National Institutes of Health, 2000). Dietary treatment requires a low-protein diet including special low- or no-phenylalanine products and modified foods. Special high-protein formulas are required daily (March of Dimes, 2006).

Galactosemia is an enzyme deficiency leading to elevated levels of galactose, a sugar in milk, in the blood. Dietary treatment eliminates all dairy products and ingredients derived from milk from the diet (American Liver Foundation, 2006).

Hereditary fructose intolerance is a deficiency of the enzyme that metabolizes the sugar fructose. Dietary treatment eliminates fructose and sucrose from the diet (A.D.A.M. Medical Encyclopedia, 2005).

Maple syrup urine disease is a deficiency of the enzyme that breaks down the branched-chain amino acids leucine, isoleucine, and valine. Dietary treatment requires a low-protein diet including special low- or no-phenylalanine products and modified foods and special high-protein formulas (A.D.A.M. Medical Encyclopedia, 2005).

Important role of food service assistants
- Communicate with food service manager, parents, teacher, and school nurse
- Keep a plan on file detailing dietary restrictions for each child with an inborn error of metabolism as well as the foods that may be substituted
- Obtain and serve special formulas or foods to meet the student’s nutritional needs
- Follow prescribed portion sizes for all foods when serving a student with an inborn error of metabolism (National School Food Service Institute [NFSMI], 2006)
- Report mistakes immediately to the school nurse or other school personnel in charge of the student’s health care (NFSMI, 2006)
- Maintain confidentiality (U.S. Department of Agriculture, 2001)
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Resource Sheet

**National Newborn Screening and Genetics Resource Center (NNSGRC)**
The National Newborn Screening and Genetics Resource Center (NNSGRC) maintains an updated report that provides tables identifying which diagnoses are screened for in the United States.
http://genes-r-us.uthscsa.edu/nbsdisorders.pdf

**Registered Dietitians of the American Dietetic Association (ADA)**
The ADA’s toll free Consumer Nutrition Information/Hotline provides referrals for qualified registered dietitians in your area as well as daily nutrition messages. Call for more information: 800-366-1655.
http://www.eatright.org

**National Organization for Rare Diseases (NORD)**
The National Organization for Rare Disorders (NORD), a 501(c)3 organization, is a unique federation of voluntary health organizations dedicated to helping people with rare “orphan” diseases and assisting the organizations that serve them. The NORD Web site maintains reports on more than 1,150 diseases.
http://www.rarediseases.org
Lesson 4: Understanding Inborn Errors of Metabolism

Pre-Training Assessment

1. Inborn errors of metabolism are
   a. illnesses that make you burn more calories.
   b. rare genetic disorders.
   c. food allergies.
   d. conditions that children grow out of.

2. A student with phenylketonuria (PKU) cannot have
   a. sugar.
   b. protein.
   c. fat.
   d. water.

3. A student with galactosemia cannot have
   a. sugar.
   b. meat.
   c. dairy products.
   d. water.

4. When serving a student with an inborn error of metabolism
   a. substitute any salad dressing for an allowed brand.
   b. serve him/her the same meal as another student with the same condition.
   c. allow him/her a second portion of an acceptable food once the first has been eaten.
   d. keep information about his/her condition confidential.

5. During metabolism, enzymes break down proteins into
   a. amino acids.
   b. sugars.
   c. starches.
   d. calcium.
Correct answers are underlined.

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Slide Presentation

Slide 1
Meeting Children’s Special Food and Nutrition Needs in Child Nutrition Programs
Lesson 4: Understanding Inborn Errors of Metabolism

Learning Objectives
• Define inborn errors of metabolism, identify the more common errors, and explain dietary treatment for children with inborn errors of metabolism.
• Describe cafeteria accommodations for these children and understand the need for a professional consultant in difficult cases.

Slide 2
Inborn Errors of Metabolism
• Rare genetic disorders in which the body cannot metabolize food normally
• By-products of metabolism, amino acids, sugars, fatty acids build up in the body, causing serious complications
• Dietary treatment: strict diet management to avoid toxic buildup of dietary by-products
• Special foods or formulas may be needed

Slide 3
Phenylketonuria (PKU)
• Cannot process the amino acid phenylalanine
• Dietary treatment:
  – low-protein diet (to prevent increase in phenylalanine)
  – special formula to provide protein
Lesson 4: Understanding Inborn Errors of Metabolism

Slide 4

Galactosemia

- Cannot process the sugar galactose
- Dietary treatment: no milk or dairy products

Slide 5

Hereditary Fructose Intolerance

- Cannot process the sugar fructose
- Dietary treatment:
  - no fructose (high-fructose corn syrup, honey, fruit)
  - no sucrose (table sugar)

Slide 6

Maple Syrup Urine Disease

- Cannot process the branched chain amino acids leucine, isoleucine, valine
- Dietary treatment:
  - low-protein diet
  - special formula to provide protein

Meeting Children’s Special Food and Nutrition Needs in Child Nutrition Programs
Food Service Assistants

- Maintain communication among parents, teacher, school nurse, food service, and consultant if needed
- Understand the dietary restrictions prescribed in the plan kept on file with the school nurse
- Obtain and serve special formula or foods
- Follow prescribed portion sizes
- Report mistakes immediately
- Keep information confidential
Meeting Children’s Special Food and Nutrition Needs In Child Nutrition Programs

Certificate of Completion

is recognized for successful completion of the NFSMI professional development seminar,
Meeting Children’s Special Nutrition Needs In Child Nutrition Programs

Instructor(s)

Date Issued

Continuing Education Credit Hours

National Food Service Management Institute
The University of Mississippi