Medical Nutrition Therapy Diet

Phenylketonuria (PKU)

1. Purpose
   a. Nutrition Indicators
      High levels of the amino acid phenylalanine are an indicator for PKU. Nurses collect blood from the baby's heel or crook of its arm using a needle to diagnosis babies with PKU.
   b. Criteria to Assign the Diet
      A diet with low levels of phenylalanine is followed when blood levels are above 20 mg/dL. For normal newborns the upper limit is 2 mg/dL, with most children unaffected by PKU having levels below 1 mg/dl. Individuals with high blood levels of Phe are tested further to distinguish further.
   c. Rationale for Diet
      With PKU the body does not contain the enzyme to breakdown the amino acid phenylalanine. Too much phenylalanine in the body can cause many health problems such as mental retardation, behavioral or social problems, seizures, tremors, hyperactivity, stunted growth, skin rashes, small head size, fair skin and blue eyes, and a musty odor in breath, skin or urine.

2. Population
   a. Overview
      PKU is a genetic disorder that's characterized by the inability of the body to breakdown and turn the amino acid phenylalanine (Phe) into tyrosine. When this enzyme does not exit it results in Phe building up in the body. Too much of Phe is toxic for the brain. It can cause problems such as lower intelligence, bad moods, feeling foggy, thinking and responding slower, being depressed, feeling anxious, and not being able to focus or pay attention.
   b. Disease Process
      It is an inherited recessive gene that affects one out of every 20,000 births.
   c. Biochemical and Nutrient Needs
      Avoid foods that are high in Phe.

3. General Guidelines
   a. Nutrition Rx
      Keep diet low in Phe, which increases the intake of CHO and fat.
   b. Adequacy of Nutrition Rx
      The Phe free formulas help the patients with PKU obtain all of the vitamins and minerals they are not able to get through oral intake. Increased weight gain can be seen because of the high intake of CHO and fat.
   c. Goals
      Keep the blood Phe levels low and on a case to case basic determine the amount of Phe in the diet. This can be done by avoiding food
have a high Phe value; contain the sweetener aspartame (products such as Equal, NutraSweet, and Caderel), and high protein foods.

d. Does it Meet DRI

Low blood levels of iron, vitamin A, zinc, and essential fatty acids are seen in those with PKU. Low bone mass can also be seen even though they obtain enough calcium and other minerals through their diet.

4. Education Material

a. Nutrition Therapy

Talk with the patient about the importance of keeping Phe levels low. Provide them with a PKU food guide that they can carry with them as a quick and easy tool. Also encourage them to bring Phe free foods with them wherever they go.

b. Ideas for Compliance

- Have them keep a food journal or use a computer program to help keep track of the amount of Phe intake.
- Try and not plan events focused around food. Encourage various hobbies, music, and sporting events.
- Pick 3-4 foods you like that are easy for non-PKU friends to offer you, or that you can find at most restaurants.
- Be prepared for potlucks, picnics and car trips.
- Talk with friends, family, and teachers about PKU and the PKU diet.
- Low protein products that resemble other foods help PKU individuals fit in with their peers.
- Look for a local support group for individuals with PKU.
- Talk about financial aid resources to pay for the price of the formula.
- Maintain a positive food attitude.
- Make your grocery list and your meals with the whole family in mind.

5. Sample Menu

a. Foods Recommended

Many Phe free formulas are recommended. For example comida-PKU A formula is used for infants during their first year of life and comida-PKU B is for toddlers and children between one and 14 years old. Some of these formulas can be found as a pre made mix or some you mix with water. Also low protein bread pastas, and cereal can be included in the diet.

b. Foods to Avoid

Individuals with PKU should stay away from high protein foods containing a large amount of phenylalanine. Examples of food to avoid with PKU include milk, eggs, cheese, nuts and soybeans. Beans, chicken, steak and other beef products, fish, chocolate, peas, pasta, rice, bread, cookies, and certain fruits and vegetables. Check with pharmacist for any products containing phe, avoid low calorie foods that contain aspartame (equal and NutraSweet)
c. Example of a meal plan

![Meal Plan Image]

6. Websites
   a. Organizations with Websites
      MayoClinic – Phenylketonuria
   b. Government Websites
      [http://www.nichd.nih.gov/health/topics/phenylketonuria.cfm](http://www.nichd.nih.gov/health/topics/phenylketonuria.cfm)
      www.FDA.gov
      www.CDC.gov
7. References
   a. Journal articles references
