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 phenylalaine 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 phenylainaine. Too much phenylainaine 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 phenylainanine (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 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 state 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

What a typical low Phe diet might look like:

		mg Phe
Breakfast	1 cup corn flakes	94
	6 oz diluted orange juice	15
	8 oz phenylalanine-free formula	0
Lunch	2 slices low-protein bread	30
	1 slice low-protein cheese	29
	5 potato chips	29
	1 medium peach	19
	12 oz sweetened beverage (aspartame free)	0
Snacks	8 oz phenylalanine-free formula	0
	1 medium apple	7
	1 fruit juice bar	0
Dinner	8 oz phenylalanine-free formula	0
	1 cup low-protein pasta	15
	1/4 cup marinara	17
	1/2 cup green beans	43
	1/2 cup iceberg lettuce	14
	1/2 cup raw tomato	21
	2 tbsp Italian dressing	8
Snacks	8 oz phenylalanine-free formula	0
	2 chocolate chip cookies	54
Total		**401**

6. Websites
   a. Organizations with Websites

      MayoClinic – Phenylketonuria
      http://www.mayoclinic.com/health/phenylketonuria/DS00514/DSECTION=symptoms

      BioMarin Pharmaceutical Inc http://www.pku.com/home.php

      Med Help http://www.medhelp.org/lib/pku.htm

   b. Government Websites

      http://www.nlm.nih.gov/medlineplus/ency/article/001166.htm
      http://www.nichd.nih.gov/health/topics/phenylketonuria.cfm
      www.FDA.gov
      www.CDC.gov

7. References

a. Journal articles references

Levy, H., & Waisbren, SE. (2008). Pku in adolescents: rationale and psychosocial factors in diet continuation. *ACTA PAEDIATRICA*, *83*(407), Retrievedfromhttp://www3.interscience.wiley.com/journal/121368311/abstract?CRETRY=1&SRETRY=0

http://www.fda.gov/NewsEvents/Newsroom/PressAnnouncements/2007/ucm109039.htm