4 Phenylketonuria (PKU)

Patient name: _____ **Admission:** _____

DATE | NRS INITIAL

I. The client/caregiver can define phenylketonuria (PKU).

A. It is a rare condition in which the body does not properly break down an amino acid called phenylalanine.
B. PKU is an inherited disorder.
C. High levels of phenylalanine are harmful to the central nervous system.
D. PKU can lead to mental retardation.

II. The client/caregiver can list signs and symptoms of PKU.

A. Symptoms are as follows:
- Skin rashes
- Tremors
- Jerking movements of arms or legs
- Seizures
- Mental retardation
- Attention deficit hyperactivity disorder
- Light complexion, hair, and eyes
- "Mousy" odor to the urine, breath, and sweat

III. The client/caregiver can list how to test for PKU and standard treatments.

A. Testing for PKU is detected with a simple blood test. Most states require a PKU screening test for all newborns. Ask your health care provider if this is done at the time of your baby's birth.
B. PKU is a treatable disease.
C. Treatment is a diet that is extremely low in phenylalanine. This diet is necessary to prevent or reduce mental retardation.
D. Lofenalac is a special infant formula made for infants with PKU. It can continue to be used throughout life as a source of protein.

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IV. The client/caregiver can list measures to manage this disorder.

A. Foods that are high in phenylalanine should be avoided:
1. High-protein foods, such as milk, ice cream, eggs, nuts, beans, chicken, steak, and fish
2. Foods, medicine, and beverages containing NutraSweet (aspartame)

B. Lofenalac is a special infant formula made for infants with PKU. It can continue to be used throughout life as a source of protein.
C. Seek genetic testing to determine if you are a carrier of this defective gene.
D. Seek help from dietician because diet can be very restricted. Some suggestions they may make are as follows:
1. Use diluted nondairy creamer for cereal.
2. Corn flakes or puffed rice are suggested cereals.
3. A packed lunch could include rice cakes, grapes, applesauce, lemonade, and jelly beans.
4. Safe seasonings and herbs to use in cooking are basil, cilantro, lemon juice, sesame oil, maple syrup, or honey.

E. Read labels carefully for PKU listed as an ingredient.
F. Work with child's teachers, daycare providers, and so forth by explaining disorder and proper choices.
G. Be informed. Start children in planning food choices early.
H. Join support groups and learn from others about coping skills and foods.

(Continued)

RESOURCES

March of Dimes
www.marchofdimes.com/

National PKU News
www.pkunews.org/

National Institute of Child Health and Human Development
www.nichd.nih.gov/

Dietician

Support groups

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