

Guidelines for Issuing PKU Formula

Background

Phenylketonuria (PKU) is an inherited metabolic disorder in which the body cannot breakdown and use phenylalanine, one of the amino acids in protein. As a result, phenylalanine accumulates in the blood and other parts of the body. The excess phenylalanine prevents the brain from growing and developing normally. It also causes other problems such as skin rash, excessive restlessness, irritable behavior and musty odor.

PKU cannot be cured, but can be effectively treated by a special diet which must be started within the first few weeks of life. With proper treatment the infant with PKU will grow and develop normally both mentally and physically. Current research indicates that the special diet should be a diet for life to prevent loss of intellectual abilities. A young woman with PKU must be in strict dietary control at least 3 months before she gets pregnant and during her pregnancy to prevent devastating heart defects, microcephaly, or miscarriage.

The diet must be carefully planned and constantly monitored by nutritionists and physicians with specialized training to allow enough phenylalanine for growth of the child or fetus, but not enough to produce any harmful effects. Careful attention must also be paid to the energy needs of the person to prevent the breakdown of the body's protein. Each person's diet needs depend on the severity of the enzyme deficiency and the person's age, growth rate and current state of health. Special formula that is free of phenylalanine is the primary food for the person with PKU with very small amounts of table foods that are low in protein and contain phenylalanine added in measured amounts. The amount of phenylalanine, tyrosine, protein, and calories in the diet are routinely measured and changed depending on the person with PKU's needs and results of tests of blood phenylalanine levels.

WIC Provisions

WIC staff will now be able to provide the PKU formulas to women, infants and children with phenylketonuria. Families receiving formula from WIC must understand that they will receive no diet instructions from the WIC clinic. Families must be in routine contact with staff from a metabolic center.

Guidelines

WIC will only provide formula if a formula prescription from a Metabolic Center is provided. A copy of the formula request/prescription is attached. All metabolic centers have been provided a copy.

A current prescription must be provided at each certification. If a prescription is not brought to the clinic, the metabolic center can be contacted and a new prescription faxed for the certification record.

PKU formulas are Level III formulas and can be approved by local agency RD's, LD's, or by calling the state agency for approval.