

Treatment - Nutrition Support

Since the late 1950's, a diet low in phenylalanine has been used to prevent mental retardation and other problems associated with untreated PKU. This diet, which may be slightly different for each person with PKU, can lower the blood Phe level to a range that permits normal growth and development. Since Phe is found in protein, the amounts of foods containing protein must be limited. This is often done using a medical food (formula) supplemented with foods that are allowed and foods that may be eaten in limited amounts.



Even though they need less Phe, children with PKU still need a certain amount of protein. The medical food gives babies and children with PKU the nutrients and protein they need while helping keep their Phe levels within a safe range. It has a taste and smell that may seem objectionable to someone not used to it, but children with PKU acquire the taste for the drink at an early age and have grown up with it. Your child's metabolic doctor and dietician will carefully calculate the amount of nutrients needed by your child. They will tell you what type of formula is best and how much to use, although frequent diet changes will be necessary as your child grows and develops. The amount of medical formula and food your child consumes needs to be carefully calculated, and all foods must be measured out to control the amount of Phe eaten.

Many high protein foods are also high in Phe. This means your child must not have cow's milk, regular formula, meat, fish, eggs or cheese, which are very high in Phe. Regular flour, dried beans, nuts, and peanut butter also have Phe and must be avoided or *strictly* limited, though it is usually easier for a child to avoid these foods completely. It is very important that your child avoid the sugar substitute aspartame (sold under the brand names "Equal", Nutrasweet, "Sweetmate", "Canderol"). Aspartame contains high amounts of Phe. It can quickly raise the blood levels of Phe in people with PKU. Your child must not have any diet foods or drinks that contain aspartame. Some medicines and vitamins also contain aspartame. If you are not sure, ask your pharmacist, metabolic doctor or dietician.



You will feed your baby with PKU the same way you would feed any baby...with formula in a bottle. The only difference is the *kind* of formula fed. Most children taking medical foods for PKU like them if the medical food is started early and if their family has a positive attitude. NEVER allow your baby even a taste of prohibited foods. You don't need to feel sorry for your child; he or she cannot miss something that has never been tasted.

The low-Phe diet is supplemented with foods that are very low in Phe. Many vegetables and fruits, including juices, and some breads, cereals, crackers, potato chips and popcorn have only small amounts of Phe and may be eaten in carefully measured amounts. In addition, there are other medical foods such as low protein flours, baking mixes, breads, and pastas that are made especially for people with PKU and other protein metabolism disorders. These foods are an important addition to a child's PKU diet because they add variety and may easily be sent to school as part of a meal or snack without providing too much Phe.

"Free" foods are foods that contain little or no Phe and may be eaten in any amount, with regard to Phe content. However, many times these foods are high in fat and sugar and must be regulated by parents as they would for any other child. These foods include soda, Kool-Aid, lemonade, popsicles, jelly, gumdrops, suckers, and hard candy.

Your child's food plan will depend on many things such as his or her age, weight, general health, and blood test results. Your dietician will fine-tune your child's diet over time.