

Key Words to Know

Amino Acids are the building blocks of protein. They come in two forms: essential and non-essential.

Essential Amino Acids are the amino acids that the body can't make for itself. We need to get them from food to make all non-essential amino acids, and build protein in our bodies. There are nine essential amino acids, and they are found in almost all foods, especially foods high in protein.

Enzymes are what help break down nutrients like protein so our bodies can use them. In PKU, the liver doesn't make enough of the enzyme phenylalanine hydroxylase (PAH).

Phenylalanine is the essential amino acid that a person with PKU cannot break down.

Tyrosine is the non-essential amino acid that isn't produced when PHE can't be broken down. Tyrosine is needed for the brain to work normally.

Free Foods are foods that contain less than 3 mg of PHE and don't need to be counted.

What is PKU?

PKU stands for phenylketonuria. PKU is an inherited condition causing a deficiency of the liver enzyme needed to break down the amino acid phenylalanine (PHE) into another amino acid, tyrosine. When more PHE than is needed by the body is consumed in the diet, it builds up in the blood and becomes toxic to the brain. Treatment of PKU includes elimination of foods high in PHE and the use of medical formulas.

What foods can a person with PKU incorporate into their diet?

ALWAYS	MODERATE	ELIMINATE
Low PHE Fruits & Vegetables	Grains	Meat & Poultry
Low Protein Foods	Rice	Eggs
Medical Food (formula)	Pasta	Dairy
Healthy Fats	Cereals	Nuts & Seeds
Sugar	High PHE Vegetables	Legumes
"Free Foods"	High PHE Fruits	Soy products

Examples of Low-PHE Fruits and Vegetables

Remember: everything in moderation. Keep a good track of your PHE intake and monitor your foods. All fruits & vegetables listed contain 50mg or less of PHE per 100g serving.

Fruits: acai, apple, avocado, figs, grapes, jackfruit, kiwi, mango, papaya, peaches, pears, pineapple & tangerines*

Vegetables: cabbage (white & red), celery, chard, lettuce, cucumber, carrots, choyote, eggplant & sweet green peppers*

Apps and Websites you can use to track your PHE intake:



*Information found from the Journal of Inherited Metabolic Disease Araújo ACMF, Araújo WMC, Marquez UML, Akutsu R, Nakano EY. Table of Phenylalanine Content of Foods: Comparative Analysis of Data Compiled in Food Composition Tables. *JIMD Reports*. 2017;34:87-96. doi:10.1007/8904_2016_12.