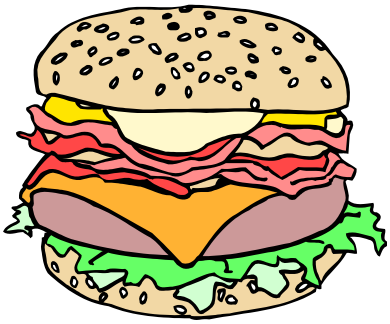
UNDERSTANDING PKU

What is PKU?

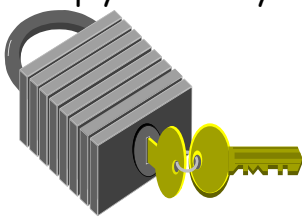
Phenylketonuria, or PKU means "phenylalanine in the urine". **Phenylalanine**, or **"phe"** for short, is an amino acid which is a building block of protein. There are 20 amino acids.

Examples of foods we eat that contain protein are meat, eggs, cheese, milk, beans, tofu, nuts and peanut butter. All food that contains protein also contains phe.

If you eat protein, a hamburger for example, the food goes into your stomach and then into your blood stream for your body to break down and use to keep you healthy and strong.

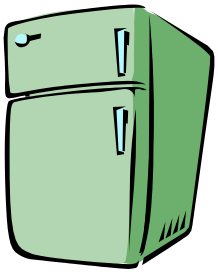


The body breaks down the protein using things called **enzymes.** The best way to think of enzymes is as keys. They "unlock" doors to help your body do the jobs it needs to do to keep you healthy.



People born with PKU are missing the enzyme that is needed to break down phe. It's like a door is locked in their body and the key is missing. The phe then builds up in the blood and spills over into the urine. That is why it is called Phenylketonuria.

Picture a refrigerator door. If you went out and bought all your groceries and you came home and couldn't get into the refrigerator, what's going to happen? All the food is going to pile up outside the refrigerator. Soon it will fill up the whole kitchen and then spill out into the next room.



In PKU, this is what happens. The key is missing to the refrigerator. Phe can't get in through the door and builds up outside. Soon it has built up so much that it spills over.

Our bodies aren't designed to cope with these high levels of phe. People can have trouble with planning and organizing, attention and processing speed, anxiety and depression. High phe levels in pregnancy can cause birth defects in the baby.

How do people get PKU?

To have PKU, you have to be <u>born</u> with it. You cannot "catch" it or get it later in life. From the moment you are conceived, you have PKU. It is passed to the baby from the mother and father. The mother and father do not have to have PKU to pass it to their baby. They are called **carriers** of the gene that causes PKU.

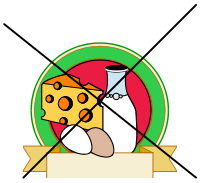
Since 1965, all babies born in Canada are tested for PKU when they are born. This way, treatment can be started right away.

How is PKU treated?

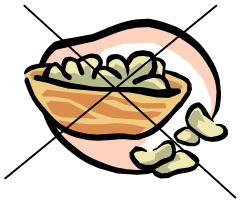
Since people with PKU are missing an enzyme (or key) to break down phenylalanine , they cannot eat high protein foods. These are foods to be avoided:

- **Meat**: chicken, turkey, fish, beef, pork, lamb

- **Dairy**: milk, cheese, yoghurt, cottage cheese, ice cream



- **Nuts and seeds** including peanut butter



- **Tofu, beans** and any other "meat substitutes" that vegetarians may eat to substitute protein in their diet

- food or drinks sweetened with **Aspartame®** such as Diet Coke®. Aspartame contains phenylalanine.

There are many other foods such as breads and pastas that contain protein. These foods are to be eaten in limited quantities only. Examples are french fries, crackers, regular pasta and bread.

Foods that can be eaten in unlimited quantities are free foods. These include fruits and vegetables and low protein products. Low protein products are special foods for people with PKU. They include cookies, pastas and breads made with special flour and contain very little phe.

Are people with PKU allergic to protein?

No, people with PKU are not allergic to protein. Remember that there are 20 building blocks of protein. We need to get all 20 to keep our bodies working right. People with PKU still need protein, but can only tolerate small amounts of one of protein's building block--phenylalanine. So, people with PKU need 19 out of 20 building blocks of protein.

Where do they get their protein if they can't eat it? They drink it! The special medical formula that people with PKU drink provides all the building blocks of protein except phe. It also gives them the vitamins and minerals they need. This special formula is a lot like the protein drinks that body builders use.

What happens if the person with PKU eats protein?

If the person with PKU eats too much protein, phe builds up in the blood. Remember the groceries and how the refrigerator was locked? Phe will build up in the blood like the groceries building up outside the refrigerator door.

High levels of phe not only make the person feel unwell, they can also damage the brain. Babies born with PKU and not treated with the special diet right from birth do not develop normally. They can be cognitively impaired.

How long do people with PKU need to stay on the diet?

It is now known that the PKU diet is "for life". Young people with PKU who have been on diet for the childhood and teen years are sometimes choosing to go off diet. **Stopping the diet is harmful**. The phe in the blood is still toxic to the brain even though the person has reached adulthood. Studies show that people off diet:

- experience more cases anxiety, depression and phobias than those on diet,
- may have slower information processing speed as well as trouble with attention and planning/organizing (also known as executive function)

Other symptoms related to high phe levels include poor judgement, skin problems (eczema), headaches, lack of energy, mood swings, lack of ability to concentrate, loss of short term memory, poor performance in school or inability to hold a job and poor personal relationships. High phe levels are not safe.

When is it too late to go on the PKU diet?

It is never too late to either start or return to the PKU diet. Although brain damage is not reversible, the "intoxication" effect of the high phe levels in the blood is reversible. By following the PKU diet, the person will feel better and reduce the symptoms related to high phe levels.

How can you tell if the phe levels are too high?

People with PKU test their blood every month by pricking their fingers and applying blood to a special blood dot card. The card is sent to the lab where the level of phe in their blood can be determined. A good phe level is less than 6.

How can friends help?

Friends play a very special role in helping the person with PKU. It is very hard to constantly be faced with food temptations when you are trying to stick to a special diet. Understanding the challenges of living with PKU and offering a listening ear to frustrations will be really helpful.

Learn to prepare some favourite special dishes. If you are having a person with PKU over for a meal, ask them what they would like to eat and offer low-protein snack options such as regular pop/juice or water and fruits or vegetables.