

How SAM can help you with your diet?

Findings from research and individuals' with PKU experiences have created SAM, three factors that can help you to manage your PKU. You should use SAM to return or stay on the special PKU diet.

SAM stands for:

Social Support: You are not alone in your effort to control your PKU. It is easier to achieve your metabolic goals if you have the help and support of others: family, health care providers, social workers, others with PKU from support networks, discussion groups and camps.

Attitude: Having a positive attitude about PKU and its treatment is an important step toward metabolic control. Having PKU and having to follow the special diet is hard and may make you angry. But, PKU is a disorder you can control. People that strictly follow the diet often realize that it PKU diet has helped them have healthier eating habits, have self-control and be more creative.

Manageability: Manageability is your ability to implement the special PKU diet in your life. There are many new choices in formula and diet. Take your time, work with a dietitian and find what diet plan works for you. Building habits and organizing your diet to fit your lifestyle will make following the diet easier. You have the knowledge and the skills to follow through and manage your PKU!

Do people ask you why you are such a “picky” eater?

Sometimes the best way to deal with this question is to be honest. However, sometimes you don't want to be open about your PKU. answers you could give are:

I am vegetarian.

I like eating healthy!

However, by telling your friend the truth, you have an ally who may help you!

Transition to Adult Health Care

PKU Controlling Phe Throughout Life



Children's Hospital Boston Transition to Adult Care Education Initiative



Children's Hospital Boston

You may know by now all about PKU, its causes, implications and treatments. However, we have gathered some basic information for you to refresh your knowledge or answer any questions you may still have.

Also, what you may not know is that research has shown that people with PKU should stay on the special diet and formula throughout their lives.

But, let's start with some useful information.

What is PKU?

Phenylketonuria or PKU is a rare inherited biochemical disorder. Approximately one in every 15,000 infants in the United States is born with PKU.

Being born with PKU means that to stay healthy you need to follow a treatment. This treatment limits phenylalanine and consists of a diet and a special formula.

Specifically, PKU prevents your body from metabolizing phenylalanine, which is one of the amino acids that are joined together to form proteins. A person with PKU does not have the enzyme needed to change phenylalanine into another amino acid, tyrosine. As a result, when the infant or adult is untreated or poorly treated, phenylalanine rises to high levels in the blood and can damage the brain by causing mental retardation, seizures, learning disabilities and emotional problems. Controlling PKU through diet keeps your brain phenylalanine levels low and allows you to be healthier.

Maternal PKU is when a woman with PKU is pregnant and must follow strict control of her diet to protect her baby. Diet helps to avoid severe brain damage to the unborn child caused by high phenylalanine levels.

What causes PKU?

PKU is an inherited condition. That means that it is passed through families and people who have PKU are born with it. Both parents need to have a PKU gene for you to inherit PKU. If you inherit only one gene of PKU that means you are a carrier, you have no symptoms and you may not even know that you are a carrier. However, there is a chance that you pass the PKU gene to your children.

An individual cannot cause or prevent having PKU. We have no control over genes that are in our body, or are inherited by our parents. However, you *have* control over managing PKU. You *can* control your phenylalanine levels through diet and formula and be healthy.

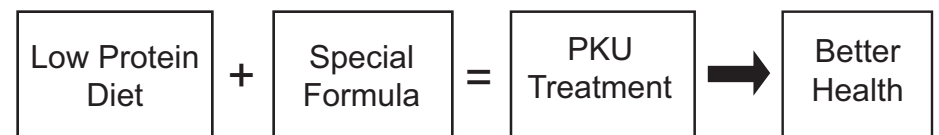
(A place of our own: <http://www.peds.umn.edu/pku/Basics.htm> and medhelp)

How can PKU be treated?

Protein is composed of amino acids joined together like beads on a string. One of these amino acids is phenylalanine. To avoid a build-up (high levels) of phenylalanine, people with PKU must remove most forms of protein from their diet. So, to control your PKU you need to follow a diet that is low in protein. However, a diet without protein would not allow you to grow and develop normally, because amino acids are necessary for life. Therefore, to control your PKU and receive proper nutrition, you must also take a special formula that contains all the necessary amino acids except phenylalanine.

Following the diet and taking the formula in an every day basis may be hard and frustrating. But if you think how PKU affects your health if you don't control it, you realize that it is worth it.

Sticking to a low protein diet and consuming the formula will help you manage your PKU and feel healthier.



What are some of the implications of PKU?

If strict diet is not followed:

Almost all individuals with PKU may “cheat” once in a while in their dietary control and have only few negative effects. However, high phenylalanine levels for long periods of time can cause emotional and psychological problems. Think of the implications, you yourself may have experienced when not following the diet and taking the formula:

- agoraphobia
- anxiety, panic attacks and depression
- trouble concentrating
- headaches
- sleep disorders
- short temper, mood swings and antisocial behavior
- memory loss
- lethargy

Controlling your phe levels may be a challenge, but it prevents your PKU from affecting your progress in school, college or work, as well as your social and personal life.

In rare cases, if the diet is terminated permanently, seizures or other severe neurological symptoms can occur.

In cases of Maternal PKU, it is extremely important to resume or maintain the diet, because you are protecting both your own and your unborn child’s health.

If strict diet is followed:

It is true, that even if you do control your phenylalanine levels, you may still have some effects, for instance in arithmetic or other analytical activities, or in memory, problem solving and attention, but in much lower levels. Maintaining dietary control helps you improve academic, professional and personal life. It makes you feel better and be healthier.

How long must a person with PKU stay on the diet?

It is now recommended that people with PKU must maintain diet for life.

In the past, it was advised to discontinue their special diet when they were children. However, studies have suggested that the diet should be continued indefinitely into adulthood to avoid the harmful effects of high phenylalanine levels.

It is important to understand that even if you have been off the diet, it is never too late to go back on the diet. By controlling your phe levels, you will soon feel and think better and be able to better remember things and concentrate.

How is PKU monitored?

PKU is monitored with blood tests that check for phenylalanine levels in the blood caused by individuals’ phenylalanine intake through food. The more phenylalanine is in your food, the higher the levels in your blood.

As an adult with PKU you should keep your blood phe levels between 2 and 10 mg/dl.

There are two methods of measuring your phe blood levels:

- 1) Going to a hospital or clinic and having a blood draw, which directly measures phenylalanine in the blood.
- 2) Collecting a blood sample on a filter paper at home and mailing it to the laboratory for analysis.

Monitoring your phe blood levels regularly is very important and helps you better control your diet. Your phe levels are an indicator of how well you are managing your diet, whether you are making the right food choices and what you may need to change in your diet.

Knowing your phe levels encourages you to keep going with your diet: Knowing that your phe is in an acceptable level encourages you to keep it that way! Finding out that your phe levels should be lower, motivates you to make the effort to lower them!