This booklet is intended by Mead Johnson to be provided to you by your healthcare professional as part of his or her care plan in the dietary management of PKU. Your healthcare professional is your primary source of information and advice. Always discuss with your healthcare professional any information you receive and any changes you wish to make.
PKU is the common term for phenylketonuria, an inherited disorder in which the enzyme phenylalanine hydroxylase in the liver is inactive or missing (see Figure 1). About one in every fifteen thousand babies born in the United States has PKU.

Phenylalanine is only one of the many amino acids which are joined together to form proteins. Usually, when a person eats foods containing protein, the body uses the amino acids from that food protein for growth and repair of body tissues. Often amino acids from foods are eaten in excess of the body's needs. These excess amino acids are chemically changed by enzymes into other compounds or used for energy. Since people with PKU are missing the enzyme that breaks phenylalanine into tyrosine, the excess phenylalanine eaten in foods accumulates in the blood and begins to damage the brain.

What Are the Effects of PKU?

If blood phenylalanine levels stay too high for a long time, the damage to the brain is severe and irreversible. The harmful effects of PKU can be prevented if a diet low in phenylalanine is started in early infancy and maintained throughout life.

How Are the Effects of PKU Prevented?

A phenylalanine-restricted diet is the only way to bring blood phenylalanine levels down to a “safe” range. At these “safe” levels, levels of less than 10 mg/dL (some clinics would encourage blood phenylalanine levels between 2 and 6 mg/dL as a “safer” range), the brain can function normally, and the person with PKU can learn easily and have stable emotions.
How Long Must A Person with PKU Follow This Special Diet?

In the past, children with phenylketonuria were sometimes advised to discontinue their phenylalanine-restricted diet as they grew older. It was not known then that this recommendation would have harmful effects. Most young people with PKU who were taken “off diet” didn’t monitor their blood phenylalanine levels and weren’t given any reason to be concerned about them. These young people began to experience the same kinds of difficulties, such as a reduced attention span, poor concentration, and poor memory. Recently, many of these people have decided to go back “on diet” hoping to feel better. In order to go back “on diet,” a person must consume a diet that is low in phenylalanine so that blood phenylalanine levels are in the “safe” range.

The most important thing to remember is it is never too late to go back “on diet.” For most young adults with PKU, a phenylalanine-restricted diet not only helps them feel better, but also improves their attention span, concentration, and memory. In general, young adults who have made these changes report they think and feel better. The effort it takes to bring down blood phenylalanine levels is well worth it for everyone no matter how long they have been “off diet.”

There are compelling reasons for each person with PKU to continue to manage his or her blood phenylalanine levels in the safe treatment range during their entire lifetime.

Possible health problems caused by high blood phenylalanine levels:

- Headaches
- Body odor
- Hair loss
- Mild to severe skin rash
- Irritability
- Impairment of short-term memory
- Greater prevalence of thought disorders and mood disturbances, such as depression, impulse control, violent behavior, or fear of leaving the house
- Blackouts or periods of loss of consciousness
- Seizures
- Spasticity, ie, involuntary muscular contractions and/or rigidity
- Damage to the nervous system and the brain
What Is the Diet for PKU?

The diet for PKU consists of a milk substitute, such as Phenyl-Free® 2 or Phenyl-Free 2HP dietary powder and measured amounts of fruits, vegetables, bread, pastas, and cereals.

Many foods must be eliminated from a low-phenylalanine diet. These foods are high-protein foods such as milk and dairy products, meat, fish, chicken, eggs, beans, and nuts. All contain large amounts of phenylalanine. Eating these foods will cause high blood phenylalanine levels. Figure 2 indicates the relative phenylalanine values of a variety of foods.

It is not unusual for someone on a phenylalanine-restricted diet to have two kinds of vegetables and a small baked potato for dinner. However, if these foods were all a person on a phenylalanine-restricted diet consumed, their diet would be lacking in protein, vitamins, and minerals. This is where the special formula comes in. Phenyl-Free 2 and Phenyl-Free 2HP provide protein (without phenyl-alanine), energy, vitamins, and minerals that are essential for good health and nourishment. The blood phenylalanine level of the person with PKU is a direct reflection of the amount of phenylalanine consumed from food. Figure 3 illustrates this relationship.

Target Your Food Choices

Figure 2

A target is an easy way to visualize the foods allowed on the diet for PKU. Phenyl-Free 2 and Phenyl-Free 2HP are at the center of the target. As foods get farther from the “bull’s eye” center, they are higher in phenylalanine. The foods outside the target are not allowed on the low-phenylalanine meal plan at all.
How Is PKU Monitored?

Monthly blood phenylalanine levels help people with PKU monitor their progress with the diet. These blood tests check for phenylalanine buildup in the blood, which is from eating too much phenylalanine. People with PKU should keep their blood phenylalanine levels in the “safe” range, between 2-10 mg/dL.

Regular measurement of blood phenylalanine levels can be done in two ways: the first method is a blood draw at a hospital or clinic; the second method involves collection of a blood sample at home on a filter paper that is mailed to the laboratory for analysis. Mailing in blood tests is a great way for people with PKU to stay in touch with how they are doing on food choices during the month. It is also important to regularly visit the PKU clinic and talk with the PKU team.

During these visits, everyone on the team works together to give the best possible care and guidance for people with PKU. The visit should include a blood draw, a neurological exam, and a chance to discuss ways to more effectively manage the low-phenylalanine food pattern.

Figure 3

As the amount of phenylalanine eaten is increased, so is the blood phenylalanine level.
Food Records

The nutritionist will request a diet record to accompany each blood phenylalanine sample. Generally, a three-day diet record is requested of all foods and beverages eaten and the amounts consumed. Try not to change eating patterns just to make the diet record look good. It is a good idea to monitor the intake of food and formula on a daily basis. Here is an example of a one-day record:

**Monday**

<table>
<thead>
<tr>
<th>Time</th>
<th>Breakfast</th>
<th>Lunch</th>
<th>Dinner</th>
<th>Snack</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>1 cup puffed rice</td>
<td>2 cups vegetarian veg. soup</td>
<td>8 oz. Phenyl-Free 2 or Phenyl-Free 2HP</td>
<td>8 oz.</td>
</tr>
<tr>
<td></td>
<td>1/4 cup Rich’s Coffee Rich</td>
<td>2 low-protein crackers</td>
<td>Phenyl-Free 2HP</td>
<td>Phenyl-</td>
</tr>
<tr>
<td></td>
<td>1 peach (80 g)</td>
<td>1 apple (100 g)</td>
<td>1 cup cauliflower</td>
<td>Free 2</td>
</tr>
<tr>
<td></td>
<td>8 oz. Phenyl-Free® 2 or Phenyl-Free 2HP</td>
<td>12 oz. Coke</td>
<td>1 cup broccoli</td>
<td>HP</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>1 baked potato with 2 Tbsp. Nucoa margarine</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>12 oz. cranberry juice</td>
<td></td>
</tr>
<tr>
<td>Snack:</td>
<td>8 oz. Phenyl-Free 2 or Phenyl-Free 2HP</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
| Total Phenylalanine: ____mg
Friends Have a Special Role

Every person is unique, so food patterns are designed for each individual's taste and body size. You may not be able to make perfect choices every day. There will be times when an understanding friend can provide the support needed to help with choosing low-phenylalanine foods. Every day, people with PKU are faced with the challenge of correctly choosing foods within these limitations to meet their dietary needs. The more support they receive from family and friends, the easier their task becomes.

What is Maternal PKU?

Young women with phenylketonuria need to understand the risks of pregnancy. Effects of the mother's high blood phenylalanine levels may damage the baby before it is born. All women with PKU, along with their families and partners, should talk with PKU clinic team members to understand these risks. This information will allow them to make a knowledgeable decision about family planning. Clinic team members also have information on adoption as a family planning option.

For more information:
See Mead Johnson's Maternal PKU, Your Diet and You.
For ordering information, call 800-BABY123 or order online at www.store.enfamil.com