PKU.com

What you should know about PKU

»Information that can make a real difference in a child's life

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You know a child with unique dietary needs

Dear Reader,

You are likely a teacher, a babysitter, a child-care provider, or a parent. Whatever your connection is, you happen to know a child with a condition inherited from both parents known as PKU, or **P**henyl**K**eton**U**ria (fen-il-key-ten-U-re-a).

Children with PKU do not generally stand out. They are like other children with one exception: they're on a special diet that is very low in an amino acid called phenylalanine (or Phe for short). This Phe-restricted diet is essential for normal growth and mental development. It is critical that this diet be maintained at all times. Any change or addition to the Phe-restricted diet can be harmful.

Since you are an adult who will be responsible for this child for some period of time, you should understand what simple but important measures you can take to help this child. Your school may have a policy applicable for children with PKU.

This booklet has been designed with input from medical professionals with expertise in PKU. In it you will learn:

- What PKU is
- How PKU is managed
- How you can help

The more you know about PKU, the better you can help this child. After you've read this brochure, you can learn more by going to PKU.com, a new website committed to the needs of the PKU community.

Just by reading this brochure, you are making a difference in the life of a child with PKU.

Thank you.

Regards,

The people @ PKU.com

About 20,000 people in the US have PKU[1,2]

What is PKU?

PKU, which stands for **P**henyl**K**eton**U**ria (fen-il-key-ten-U-re-a), is a rare condition inherited from both parents.[1]

A problem with amino acid metabolism

People with PKU are unable to process a certain amino acid called **phenylalanine** (**Phe** for short). Phe is found in high amounts in all protein-containing foods (eg, meat, eggs, dairy, nuts), and in lower amounts in many other foods such as wheat and flour products like pasta and bread, and all fruits and vegetables. Their bodies do not make enough of an enzyme called **phenylalanine hydroxylase** (**PAH**) that breaks down Phe.[1] If people with PKU are not careful about what they eat, they will eat too much Phe, and their bodies will not be able to use the Phe fast enough. Instead, the Phe will build up in their blood and can be toxic.[3]

An inherited condition, identified at birth

Newborn screening for PKU began in the 1960s[1]; as a result, children are diagnosed at birth. Currently, all children in the United States are screened for PKU at birth by taking a drop of blood from the heel. Since then, about 20,000 people[1] in the United States are living with PKU and managing their condition with a special diet low in the amino acid phenylalanine.[1] Children with PKU are like other children, except for what they eat.

A condition that can affect brain health

High-blood Phe levels over a long period of time can be bad for the brain.[3] If blood Phe levels are kept within the recommended range, children can grow and develop normally.[1,3]

A condition that is currently managed with a Phe-restricted diet[1]

Currently, the only way to successfully manage PKU is by carefully following a **Phe-restricted diet**, in order to limit the amount of Phe in the body. This approach has been successful, and people with PKU can lead relatively normal lives, if they follow the diet very strictly.[1]

How is PKU managed?

Not too much Phe

Because Phe is an important amino acid for growth, development, and brain function, people with PKU must eat some Phe, but only in *prescribed amounts*. This means they eat foods that contain Phe, but only as much as their dietitian allows.[1] The amount of Phe they're allowed to eat is called their **Phe tolerance** and is dependent on age, gender, and growth rate.[1]

The right amount of protein[1]

Foods that are high in protein have too much Phe, so they are avoided in the Phe-restricted diet. To make up for the missing protein, the diet includes a prescription of medical food—that often comes in the form of a powder formula—containing everything else found in protein, except Phe. A majority of the protein that people with PKU consume comes from this prescription medical food. People with PKU drink this medical food every day with every meal.

Remaining Calories from fats, sugars, and starches

Prescribed amounts of low-protein foods and prescription medical food alone do not provide enough daily calories for a child with PKU. The remaining calories in the Phe-restricted diet come from foods that are high in fat, sugar, or starch (which are typically lower in protein). Of course, these foods must be eaten in moderation to reduce the risk of obesity.

A balancing act

The balance is tricky to maintain. Over time, prolonged high-blood Phe levels can be bad for the brain.[3] While you may not be able to see the effects of high Phe levels, there are changes happening in the body and brain. This is why people with PKU work hard every day to keep the balance just right.

The fundamentals of PKU

The three R's

3R

To help maintain consistently low blood Phe levels, PKU management involves the **3R** approach: **Restrict, Replace,** and **Record.** People with PKU must **Restrict** their diet to the right amount of low-Phe foods, **Replace** high-Phe foods with low-Phe alternatives, and **Record** all the Phe that is consumed.

Restrict:

- Avoid high-Phe foods entirely
- Eat low-Phe foods in prescribed amounts
- Adjust the diet according to the Phe tolerance of the individual child

Replace:

- Obtain most protein from prescription medical food, such as the formula, in place of high-protein foods
- Supply remaining calories from fats, sugars, and starches
- Enjoy specialty low-Phe alternatives

Record:

- Track food consumed every day
- Keep a record of foods eaten, so a registered dietitian can make informed diet decisions
- Obtain frequent blood tests to ensure blood Phe levels are within the recommended range

Most of this responsibility lies with the parent, the doctor, and a registered dietitian.

Read on to learn about the things you can do to help while the child is under your care.

What is your responsibility?

Learn about foods that people with PKU may be able to eat

Examples of no-Phe foods:

- Phe-free formula
- All oils (olive, peanut, vegetable, etc.)
- "Pure sugar" candies not containing aspartame

Examples of low-Phe foods allowed in prescribed amounts:

- All fruits and fruit juices
- All vegetables
- Starch, including most cereal and grain products
- Toppings and condiments

This list is not complete, and you do not need to memorize it. It is meant to provide examples of the kinds of **foods people with PKU can eat.**

What is your responsibility?

Learn about foods that people with PKU may NOT be able to eat

- Meat (and foods containing)
- Nuts (and foods containing)
- Dairy (and foods containing)
- Legumes (and foods containing)
- Eggs (and foods containing)
- Baked goods high in flour
- Foods containing aspartame (sold as Equal® or NutraSweet®)

This list is not complete, and you do not need to memorize it. It is meant to provide examples of the kinds of **foods people with PKU CANNOT eat.**

A day in the PKU life

PKU is a daily commitment

Everything a person with PKU eats must be counted. This information helps a registered dietitian keep track of growth and development and ensure that each person is following a healthy diet. Food is tracked in a **"Phe diary."** The following is an example of what a person with PKU might eat on a given day:

14-year-old allowed 8 mg Phe/kg (total 406 mg Phe/day)[4]		mg Phe
Breakfast	1 cup corn- or rice-based cereal	**40**
	6 oz orange juice	**20**
	8 oz phenylalanine-free formula	**0**
Lunch	2 slices low-protein bread (tapioca)	**76**
	1 slice low-protein imitation American cheese	**29**
	margarine	**0**
	6 potato chips	**30**
	1 medium peach	**38**
	12 oz sweetened beverage	**0**
Snacks	8 oz phenylalanine-free formula	**0**
	1 apple	**10**
	1 cup fruit ice	**0**
Dinner	8 oz phenylalanine-free formula	**0**
	1 cup low-protein pasta	**40**
	1/4 cup marinara sauce	**32**
	1/2 cup green beans	**34**
	1/2 cup iceberg lettuce	**12**
	5 cherry tomatoes	**13**
	2 tbsp Italian dressing	**0**
Snacks	8 oz phenylalanine-free formula	**0**
	1/2 rice and marshmallow treat	**32**
Total		**406**

Different people tolerate different amounts of Phe. Every person's diet varies, and the diet presented above may not be typical. Children with PKU might enjoy low-Phe ethnic foods specific to their culture.

What is consistent among all children with PKU is the importance of following the diet closely. All children with PKU face the risks if they don't follow the diet. Fortunately, your help can be an important part of this combined effort of parents, doctors, and dietitians to manage PKU.

What role can a teacher play?

Here are a few suggestions for helping to manage children with PKU

It is important that children with PKU only eat foods provided by their parents

- Parents of children with PKU know best what a child with PKU can eat

It is helpful to notify parents of children with PKU of class parties where food will be served

- The parent can prepare special food to be taken to school, so the child does not feel left out

You may think about enforcing a class policy against any student trading food

- Children with PKU should not trade food with other children. A class policy helps keep them from feeling singled out

It is important that the child take home all uneaten foods from lunch

- The parents need to know what was uneaten, so they can record it in their child's Phe calendar

The more you know about PKU, the better you can help

PKU.com

Welcome to the PKU community!

Thank you for taking the time to learn about PKU. By understanding **What PKU is, How PKU is managed,** and **How you can help,** you are already making a big difference in a child's life.

For more information, visit PKU.com

As part of its commitment to the PKU community, PKU.com informs people about PKU, so we can all better work together to help people with PKU. At PKU.com, you can learn more about the science and current treatment of PKU, and read about real experiences from people with the condition. When it comes to a child's health, the more you know, the better.

References: 1. Phenylketonuria (PKU): screening and management. *NIH Consensus Statement* [serial online]. October 16–18, 2000;17(3):1–27. Available at: http://consensus.nih.gov/2000/2000Phenylketonuria113html.htm. Accessed July 12, 2006. **2.** US Census Bureau data. Available at: http://www.census.gov. Accessed July 19, 2006. **3.** Centerwall SA, Centerwall WR. The discovery of phenylketonuria: the story of a young couple, two retarded children, and a scientist. *Pediatrics.* 2000;105:89–103. **4.** Scheutt VE. *Low protein food list for PKU*. Burnaby, British Columbia, Canada: 2002, Hemlock Printers.

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