Texas Department of Health

A Teacher's Guide to PKU

Hi, my name is _____

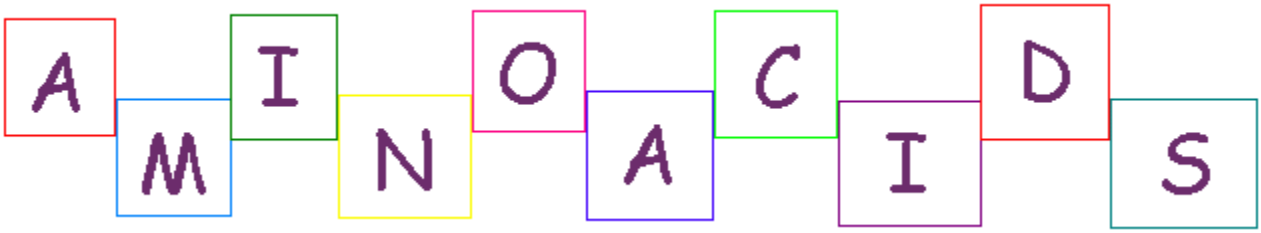
I have PKU and I want you to learn more about it so you can help me. This booklet will tell you about PKU. Except for my special diet, I am just like the other children in your classroom.

PKU Basics

These are the facts...

Phenylketonuria (or PKU) is an inherited disorder of protein metabolism. Children with PKU do not have a functioning enzyme to metabolize or break down amino acid call phenylalanine (PHE for short), which is found in all food proteins.

Protein in foods is important for building and repairing the body's tissues. Amino acids are often called the "building blocks" of protein. Twenty-two amino acids can be joined together in various combinations to form all the different kinds of proteins in foods. Enzymes are special substances in the body which work to separate the amino acids in food proteins and recombine them to form the different proteins which the body needs.



All children need a certain amount of PHE for normal growth and tissue repair. Most of the time the unused PHE is converted to another amino acid and eventually used by the body in different ways. Because the child with PKU lacks the enzyme which breaks down the extra PHE, the extra PHE builds up in the body tissues, including the blood. This extra PHE can prevent normal brain development and result in mental retardation.

The good part is...

Nearly all states check a newborn's blood during the first week of life to see if the infant has PKU. The infant with PKU can then be put on a carefully controlled diet which allows enough PHE for growth but prevents the flood of

PHE which can interfere with normal brain development. Blood tests are then done on a regular basis. With proper dietary control initiated early in life, normal physical and intellectual development can proceed in a child with PKU.

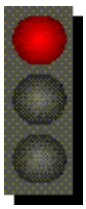
And now the diet...

Since PHE is found in all foods that contain protein, PKU children must limit their intake of protein containing foods. Foods which contain high amounts of protein are high in PHE and should not be eaten. Foods with small amounts of protein and PHE can be eaten only in controlled amounts. Some foods are "free" foods because they contain no protein and are free of PHE. They are eaten to help boost the child's intake of calories needed for energy.

A child with PKU has a special drink which has most of the protein, vitamins and minerals that a child needs for growth with little or no PHE. The special drink provides almost all of the nutrients that other children get from their food. It has a taste and smell that may seem objectionable to someone not used to it. But, PKU children acquire the taste for the drink at an early age and have grown up with it.

The amount of drink and food the child consumes in a day is carefully calculated by the child's family and nutritionist. All foods must be carefully measured out to control the amount of PHE the child eats. The amounts are adjusted to the child's changing needs as he or she grows. A child with PKU learns at an early age that his or her diet is restricted and to ask a parent if a new food is allowed. By school age, children with PKU know a good deal about their own diet.

Parents have complete, detailed lists of foods to use in feeding their child with PKU. Some examples:



NOT ALLOWED: meat, fish, chicken, turkey, milk, cheese, ice cream, yogurt, eggs, beans, nuts, peanut butter, Nutra Sweet*

ALLOWED BUT CONTROLLED: fruits, fruit juices, vegetables, vegetable juices, breads, cereals, crackers, potato chips, pop corn, special low protein foods**

FREE FOODS*: soda, Kool-Aid, lemonade, popsicles, jelly, gum drops, suckers, hard candy

*Foods, beverages, candy or gum containing NutraSweet, Equal or aspartame should be avoided by children with PKU because these sweetners are made of more than 50 percent PHE.
**Some food manufacturers use special ingredients to make low protein foods, such as pastas and baked goods, for the PKU diet. Some of these items may be sent to school as part of the child's meal or snack. These foods are an important addition to a PKU child's diet because of the added variety they allow without providing too mcuh PHE.

How you can help...

Treat the child with PKU as a normal, healthy member of the class. Remember, he/she is no different from anyone else in terms of social, emotional, physical, and academic growth and development.

Keep a good line of communication open with the child's parents to reinforce

parent teaching and monitoring of food eaten away from home. The child's parents are your closest experts on PKU.

Let the parent know

... if the child has eaten any food not allowed.

... if the child does not eat foods that are sent from home.

... if special occasions such as birthday or holiday parties are planned to make sure there is a low PHE food for the child. Parents may want to send a supply of low PHE treats that store well and can be available for special or unexpected occasions.

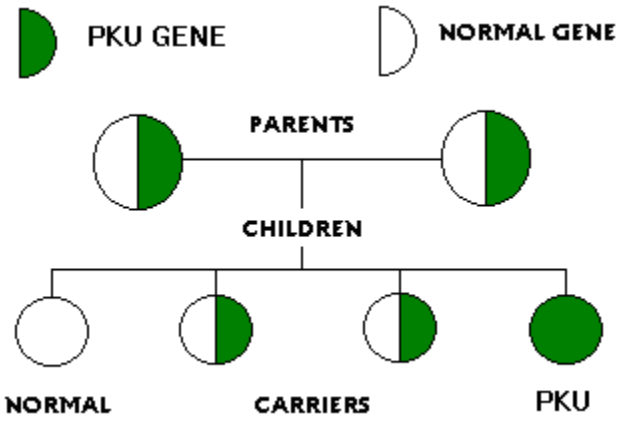
PLEASE do not feed the child with PKU *any* food not allowed on the previous list or approved by his/her parents. Even "little tastes" add up and result in an elevation of PHE in their blood. While the PKU child's curiosity may be aroused by these foods, he/she is accustomed to doing without them.

Questions & Answers

Q. How often are children with PKU born?

A. Approximately one child in every 15,000 births has PKU. This amounts to 200 to 300 new cases each year in the United Sates. Because most states have effective PKU newborn screening programs, most children with PKU start a closely controlled diet within the first month of life. When the diet is well-controlled, it is so effective that the children grow and develop normally and attend regular schools.

Q. Since PKU is inherited, do all the children in that family have PKU?



A. For a child to have PKU, each of the parents must be a "carrier" of the PKU gene. A carrier has one normal gene and one PKU gene, but can break down protein normally. A child with PKU inherited a PKU gene from each parent. When these parents have offspring, there is a 25 percent chance that the baby will be free of the PKU gene, a 50 percent chance that the baby will be a carrier, and a 25 percent chance the baby will have PKU. With each pregnancy, there is the same 25 percent chance that the newborn will have PKU. In some families, there may be only one child with PKU,

while in other families, multiple children may be affected.

Q. How do the parents know what to give the PKU child to eat and drink each day?

A. The child's diet depends on the individual child's diet prescription and on his/her preferences from the low protein foods list. The diet prescription tells how much special drink the child should have and how many milligrams of PHE the child should get from table foods each day.

Here is a typical diet for a 5-year-old boy with PKU. His diet prescription calls for 13 scoops of a special drink powder (Phenyl-Free) mixed with 24 ounces of water and 280 milligrams of PHE from table foods.

	mg. PHE
BREAKFAST:	
Kix, 6 tbs	30
Banana, 6" section	30
Orange juice, 4 oz	15
Phenyl-Free, 6 oz	0
LUNCH:	
Vegetarian vegetable soup, ½ can	60
Saltine crackers, 2	30
Lettuce, shredded, ½ cup	15
French dressing, 2 tbs.	free
Fruit cocktail, ¾ cup	15
Phenyl-Free, 6 oz.	0
SNACKS:	
Popsicle	free
Sucker, 1	free
Apple, 1 medium	8
Phenyl-Free, 6 oz.	0

DINNER:

Rice, cooked, 4 tbs.	60
Green beans, cooked, 3 tbs.	15
Jelly Gelatin, 6 tbs.	30
Phenyl-Free, 6 oz.	0
Kool-Aid, 4 oz.	free
TOTAL:	278 mg.PHE

Q. How can a child grow with so little food?

A. The PKU child's special drink contains most of the protein, vitamins and minerals needed for growth. It really is a special drink! The food the child eats gives him/her the rest of the nutrients he/she needs. "Free foods" provide additional calories.

Q. Does the child with PKU look or act differently from other children?

A. No, the child with PKU is just like other children in your classroom except that he/she has a special diet.

Q. How can I explain the PKU diet to the other children in the class?

A. Young children can understand that since cars with different engines use different fuel (gas, diesel, etc.), some children have bodies that work in different ways than others and they need different food. Older children can understand the similar concept of a "food allergy."

Don't hide the fact that the PKU child's lunch is different if asked, but no long explanation is needed. Ask the child's family for suggestions on how best to answer this question.

Q. If a child with PKU eats a high protein food, will he or she feel sick?

A. If a PKU child does eat a high protein food, he/she will probably not feel sick or different in any way. It is the elevated blood PHE level over time from the continued eating of excessive PHE that interferes with mental development. The changes may not be seen for several months to a year or more. It is this slow and subtle change which sometimes makes it difficult for a child with PKU to understand that high protein foods are harmful.

Q. What is the connection between NutraSweet and PKU that I have been hearing about lately?

A. NutraSweet is a brand name for the food sweetener, aspartame, which is being used in many presweetened foods and beverages, and in Equal, a table top sugar substitute. When aspartame is metabolized or broken down in the body, over half of it is PHE. Children with PKU should avoid products containing aspartame because it is like a protein food. This is the warning required on all food products sweetened with NutraSweet, typically found in small print near the ingredient list:

PHENYLKETONURICS: Contains Phenylalanine

Q. When can the diet be discontinued?

A. Ask the child's parents. In the past, children with PKU were frequently taken off the diet around school age, but today most doctors are now advising families to continue with the diet for a much longer time. Women with PKU who were not on the PKU diet before and during pregnancy have delivered babies with severe birth defects. Because of this, many doctors are advising women with PKU to stay on the diet through their child-bearing years.

Activities

This section has suggestions for classroom activities for all the children in your class. Remember, because the PKU child was identified early and has been on a PKU diet, he/she is like all the other children in your classroom.

Be sure to check with the parents of a PKU child in planning *any* food related activities. The parents can tell you how much of each food the child is allowed. You might even get some extra help in the classroom that day.

The following pages present some ideas for activities which you can develop to meet the needs and abilities of your students.

Differences are fun!

A PKU child has a special and, indeed, very different diet. The teacher can use this situation to teach all the children that there are differences among people and it is these very differences that make the world more interesting.

"People are unique"

- Make body tracings.
- Collect photographs of each child and whole class.
- Measure each child's height and weight.
- Describe each other (verbally for the younger child, written for the older children).
- Have each child take a turn showing the class how he/she is special.

"People eat different foods"

FOOD CUSTOMS:

Geography (seacoast, ranch lands, farms, etc.)
International (Mexican, Chinese, Italian, etc.)
Religious (Jewish, no pork; Hindu, no beef)
Seasonal (summer and winter fruits and vegetables)
Vegetarians (no foods from animal sources)

HEALTH:

- Overweight (low calories and low fat)
- High blood pressure (low salt)
- Diabetes (controlled sugar)
- PKU (special drink and low protein foods)
- Heart disease (low fat, low salt)

Skill sharpeners!

Here are several activities which could strengthen developmental and academic skills needed by all children, but especially important for the development of a PKU child's self-sufficiency. The following activities are appropriate for selective ages:

Measuring and Pouring Skills

Use cups and spoons to measure liquid and dry ingredients.

Fine Motor Skills

Use a knife to cut, chop and spread foods or playdough.

Academic skills

Follow a recipe
Keep food records
Read and collect food labels
Learn to budget money, calories, milligrams of PHE or time.

Things to do with food!

Many nutrition activities can be adapted to use allowable PKU foods so all children can participate in the same way. Remember to review the allowable PKU foods with the PKU child's parents.

USE THE SAME FRUIT OR VEGETABLE

- Slice, chop, make juice
- Prepare in different ways: raw, boiled, fried, broiled (corn can be on the cob, cutoff, popped, ground into flour)
- Trace where a food comes from or go on field trips. Oranges are grown on a tree; squeezed or canned or bottled; sold in a store; taken home (or to school) to drink.

USE DIFFERENT FRUITS OR VEGETABLES

- Compare colors, shapes, textures, tastes
- Show where grown (in ground, on a tree, bush, etc.) 

Classroom baking projects can be adapted to include the child with PKU. Special recipes have been developed which reduce the protein in baked foods by using special low protein baking mixes. Ask the PKU child's family if they have any special recipes which they would share with you and ask them how to order any special low protein products you need.

Developed by Mimi Kaufman, M.P.H., R.D. and Maria Nardella, M.A., R.D. Office of Nutrition Services. Children's Rehabilitative Services. Arizona Department of Health Services. 1740 W. Adams Room 208. Phoenix, Arizona 85007. July 1985.