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Information for the U.S. products. Please refer to the Canadian product label for the list of ingredients and nutritional information for products available in Canada.

A GUIDE FOR FAMILIES WHO HAVE A CHILD WITH

Phenylketonuria

CONTENTS

Abbott provides this booklet to health care professionals to help them counsel families, and to families to help them learn about PKU.

INTRODUCTION TO PHENYLKETONURIA

Your child has a condition called phenylketonuria (fen-il-ke-ton-u-re-ah), or PKU for short. Children who have inherited this condition can't use the amino acid phenylalanine (fen-il-al-a-neen) (PHE) in a normal way. PHE is an amino acid, which is part of protein in food. You will need to feed your child all the foods necessary for normal growth and development, but only the amount of PHE he can safely use.

Learning some medical terms in nutrition and genetics will help you understand and manage your child's diet better. If you have any questions, write them down and ask the nutritionist (dietitian), nurse, or doctor at the metabolic clinic.

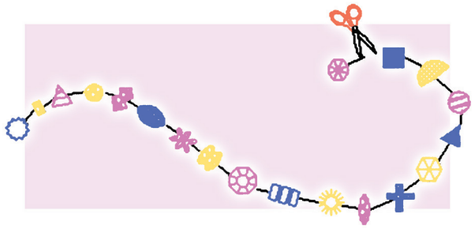


Figure 1. Amino acids are joined together like beads on a necklace to form protein. Enzymes act like scissors to remove amino acids from protein.

WHAT IS PKU?

PKU is an inherited disorder of amino acid metabolism. Proteins, which are made up of amino acids, are found in many parts of the human body, including hair, blood, skin, and muscles. Most foods contain protein. When we eat foods containing protein, this protein is split into amino acids during digestion. The amino acids are later put back together, like beads on a necklace, to form new protein. These new proteins are used to build and repair the body's tissues.

Twenty amino acids occur commonly in the human body and in the foods we eat. One of these amino acids is PHE. PHE is an essential amino acid that is important for growth.

All foods with protein contain PHE. High-protein foods are dairy products, beans and peas, eggs, meat, poultry, nuts, soy products, seafood, seeds, and nut butters. Fruits, grains, and vegetables have less protein, and therefore, have less PHE. These are allowed in the diet in measured quantities.

Splitting protein into amino acids requires a special substance that does the actual work. Think of the splitting substance as a pair of scissors snipping beads off a necklace (Figure 1). The "scissors" are called enzymes (n-simes).

Once PHE is split off from the food protein, it is absorbed, changed, and used to help form many other useful substances in the body. For example, PHE may be changed to become another amino acid called tyrosine (ti-ro-seen) (TYR). TYR is used to make hormones, skin coloring, and compounds that help nerves work.

To form TYR, an enzyme called phenylalanine hydroxylase (fen-il-al-a-neen hi-drox-il-ace) (PAH) is required. Some people, such as your child, do not have any normally working PAH, or do not have enough to handle all the PHE that is in the protein foods they eat. In either case, PHE cannot be changed to TYR, and PKU occurs. Figure 2 below shows what happens in PKU.

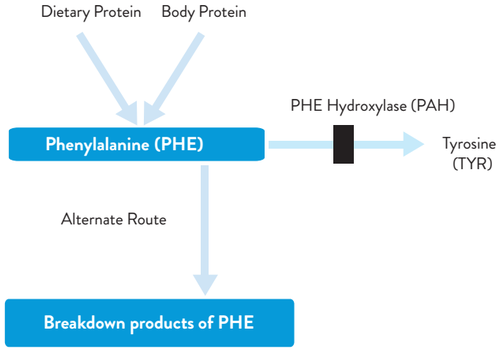
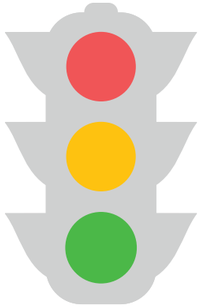


Figure 2. PHE metabolism in PKU. Compounds in blue boxes accumulate in untreated PKU.



PAH not working.
PHE cannot be broken down. Breakdown products of PHE build up and PHE not changed to TYR.

PAH not working well.
PHE may not be changed to TYR efficiently.

PAH working.
Normal PAH allows PHE to be changed to TYR. Breakdown products do not build up.

Figure 3. The phenylalanine hydroxylase (PAH) traffic light.

Think of the situation as a traffic intersection. A green light (normal PAH) allows PHE to be changed to TYR. A red light (no or too little PAH) keeps PHE from being changed to TYR. If the light is stuck on red, a traffic jam occurs—PHE increases and TYR decreases. Traffic (PHE) takes an alternate route and other breakdown products of PHE are formed (Figure 3).

If a person is not treated, these products build up in the blood and spill into the urine and perspiration. Some of these products are called ketones. People who have untreated PKU have too much PHE and ketones in their blood. In a way that we do not understand yet, the excess PHE and ketones cause brain damage.

PKU: AN INHERITED DISORDER

PKU is a genetic disease inherited from both mother and father just like other features, such as eye and skin color. Genetic information, which determines each person's characteristics, is carried on pairs of genes in every cell in the body. These genes serve as blueprints, or patterns.

Each parent of a child with PKU has one normal (●) and one altered (PKU) (●) gene. Each of their offspring will have one gene from each parent and could have one of four gene sets (Figure 4). A child who receives gene set A inherits two normal genes (●●). Her body will make enough PAH to use PHE normally. She will pass a normal gene on to each of her offspring.

A child who receives gene set B or C inherits one normal (●) and one PKU (●) gene. His body will make enough PAH to use PHE normally, but he can pass on the PKU gene to his offspring. A person with this gene set—one normal and one PKU—is called a carrier. Being a carrier does not affect the person's health. **You, as parents of a child with PKU, are carriers. Brothers** and sisters of your child with PKU may also be carriers. About 1 in every 50 people is a carrier for PKU.

A child with gene set D has PKU caused by the two PKU genes (●●) , one from her mother and one from her father. Her body will not be able to use the PHE in food normally. She will also pass a PKU gene on to each of her offspring. Your child with PKU has this gene set.

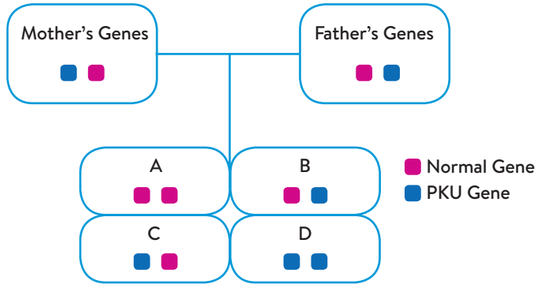


Figure 4. Genetic inheritance of PKU.

TYPES OF PKU

There are several types of PKU. The type generally refers to the amount of enzyme activity. Types include classical PKU, mild PKU, and hyperphenylalaninemia (HPA). In classical PKU there is little or no enzyme activity present. The amount of PHE is typically severely restricted. Mild PKU has a higher level of enzyme activity and the body may be able to handle a greater amount of PHE from the diet. HPA is an even milder form of the disorder and the diet can be more liberal.

DIAGNOSIS OF PKU

PKU is diagnosed by a blood test. The United States and many other countries require all babies to be tested for excess levels of PHE in the blood before they are released from the hospital.

If the initial screening test result shows that the level of PHE in the blood is high, more tests are needed immediately to learn if the baby has PKU or another disorder. In some instances, doctors hospitalize the infant to confirm the diagnosis because testing is more rapid and diagnosis and treatment can begin sooner.

SYMPTOMS OF PKU

With early diagnosis and lifelong nutrition support, a person with PKU can develop normally if blood PHE levels are kept near normal. However, if an infant's PKU has gone undetected and untreated for several months, the infant may have some or all of these symptoms:

- moderate to severe mental retardation
- skin rash
- excessive uncontrolled body movements
- convulsions
- a pungent, musty odor

In addition, the infant may be irritable and hyper active.

When PKU is diagnosed and treated, behavior and skin rash usually improve first. If diagnosis is delayed and mental retardation is already present, the special diet will result in some mental improvement; however, brain damage that has already occurred will remain. This is why diagnosing and treating PKU early is so important.

NUTRITION SUPPORT OF PKU

Since the late 1950s, a diet that reduces PHE intake has been used to prevent the mental retardation and other problems associated with untreated PKU. This diet, which is different for each person with PKU, can lower the blood PHE level to a range that permits normal mental development and growth. The special PKU diet for your child is very important. With proper nutritional management, your child will grow and develop normally.

Many foods contain protein. Those foods also contain PHE. Your child with PKU must limit the amount of foods that contain protein. Table 1 is a general guide to foods that are not allowed, foods that are limited, and foods that may be eaten freely if obesity is not a problem.

Additional Therapies. Your metabolic doctor may recommend certain vitamins or medications for your child with PKU.

Table 1. General Guide to Foods on PHE-Restricted Diets

Foods That Are Not Allowed	Foods That Are Limited	Foods That May Be Eaten Freely
Dairy products (cheese, milk, ice cream, yogurt), soy milk and soy products, beans and peas, eggs, fish and other seafood, meat, nuts, nut butters, poultry, seeds, tofu, sugar-free foods with aspartame	Breast milk, infant formulas, bread, crackers, fruit, fruit juices, low-protein cereals, popcorn, potato chips, special low-protein foods, vegetables, vegetable juices	Gumdrop candy, hard candy, jelly, Kool-Aid®, lemonade, lollipops, Popsicles®, pure sugar and fat, soda

Kuvan® (sapropterin dihydrochloride) are tablets or powder that are paired with nutrition therapy for the treatment of PKU. It has been shown to be effective in lowering plasma PHE levels and increasing PHE tolerance in a subset of patients with tetrahydrobiopterin (BH4) responsive PKU.

Large Neutral Amino Acids (LNAA) are paired with nutrition therapy for the treatment of PKU. PHE shares a common blood-brain barrier transport mechanism with several other LNAAs. Increasing levels of these LNAAs may increase competition across the blood-brain barrier or intestine and decrease brain PHE levels. LNAA are not appropriate for young children or women who are pregnant or planning to become pregnant.

Requirements for PHE, TYR, Protein, and Energy. A child with PKU who eats enough protein to grow properly gets too much PHE. Foods high in protein are cheese, eggs, meat, milk, soy milk, poultry, fish, nuts, beans and peas, seeds, and nut butters. Foods low in protein include some cereals, fruits, fats, vegetables, and sweets. On the other hand, eating these foods in the amounts needed to provide just enough PHE does not provide enough protein to meet the child's needs for growth. **To get enough protein for growth and not get too much PHE, use of a special medical food that is high in protein and free of PHE is necessary.**

To be sure your child is getting enough energy and adequate PHE, TYR, and protein for growth and development, a nutritionist carefully calculates the amount of each nutrient needed. Too little PHE, TYR, protein, or energy can result in growth failure. Frequent diet adjustments are necessary, especially during the first 6 months of life when babies grow rapidly. The nutritionist or metabolic doctor will make these diet changes based on your baby's health, growth, PHE and TYR intakes, and blood levels of PHE and TYR.

Phenex™-1 Amino Acid-Modified Medical Food With Iron is used to provide protein for infants and toddlers. Phenex-1 does not contain any PHE, so Similac® Advance® with Iron Infant Formula, breast milk, or other intact protein **must** also be fed to your baby to provide the specific amount of PHE she needs for growth and development. Breast milk is lower in PHE than infant formula or cow's milk and can be used to supply the required PHE. The decision to breastfeed should be discussed with your nutritionist and metabolic doctor. The nutritionist or metabolic doctor will tell you the exact amount of breast milk needed in addition to your

High-protein foods provide too much PHE for a child with PKU.

child's medical food. Phenex-1 is well supplied with fat, carbohydrate, minerals, and vitamins. Supplemental minerals and vitamins are not usually needed when the diet is followed as directed.

Phenex™-2 Amino Acid-Modified Medical Food is a medical food used in treating children and adults with PKU. This product contains no PHE, so PHE needs **must** be met by using other food sources. Your nutritionist will tell you which medical food is right for your child, as well as which foods and the amount your child can eat.

Phenex-1 and Phenex-2 look and taste different from milk. Phenex-2 is available both as unflavored and vanilla- flavored products. They may seem distasteful to you, but it is very important not to show this to your child, either by word or action. Your child may refuse the medical food just because you appear not to like it.

Phenex™-1 and Phenex™-2 are to be used under medical supervision.

One mother disliked the odor of the medical food so much that she made a face when she gave it to her son. Because of this, he refused the medical food for several days until she and her family realized what was wrong. She said later, "We changed our attitude to thinking this wonderful diet will make it possible for our child to have a happy life."

Aspartame (NutraSweet®) contains PHE and must not be used as a sweetener. Many foods and some medicines contain aspartame and should be avoided. Read labels carefully!

Other children in the family should be told that the Phenex is very important, and they shouldn't emphasize the difference in taste or odor between milk and medical food to the child with PKU.

Most children taking medical foods for PKU like them if the medical foods are started early and if their family has a positive attitude. Older children who start on the diet after drinking cow's milk may not like Phenex at first, but in time accept it.

Flavorings, such as Kool-Aid® Unsweetened Soft Drink Mixes, Wyler's® Unsweetened Soft Drink Mixes, and concentrated fruit juices, can be added to Phenex. Remember to count the added PHE content of the flavoring added to the formula to your child's total daily PHE allowance.

Phenex may be made into a paste and combined with some allowed fruits, such as applesauce or other fruit purees, or combined with instant pudding mixes. Aways read the label and be careful which pudding mixes you buy, as some contain more protein than others.

INTERNATIONAL SYSTEM OF MEASUREMENT (METRIC SYSTEM)

The metric system is the International System of Measurement. It is used for all medical and scientific measurements.

In the metric system, solids are weighed in grams (g) or kilograms (kg) and liquids are measured in milliliters (mL) or liters (L). A list of common conversions from the metric system to the English system used in the United States is given in Table 2. The most accurate method to be sure your child is getting the proper amount of PHE is to weigh food on scales that read in grams.

Medical Food Preparation. Mix a 24-hour supply of medical food at one time or as instructed by your nutritionist.

Tips for preparing formula for infants:

- **Always follow the instructions on the label and mix the formula according to the recipe provided by your nutritionist or metabolic doctor.**
- Wash your hands and all supplies carefully before preparing formula.
- **Do not** mix longer than indicated on the Phenex label.

Table 2. Metric to English Conversions

Metric		English
Solids		
1 g (0.001 kg)	=	0.035 oz
28 g	=	1 oz
454 g	=	1 lb
1000 g (1 kg)	=	2.2 lb
Liquids		
5 mL	=	1 tsp
15 mL	=	1 Tbsp
60 mL	=	1/4 cup
240 mL	=	1 cup
1000 mL (1 L)	=	4 1/4 cup (1.06 qt)

- Always test the temperature of heated formula before feeding by shaking a few drops on your wrist.
- Overmixing causes the fat emulsion to break. Separation of the medical food mixture then occurs. Overmixing may also add air that destroys vitamins A and C.
- Heating above 100° Fahrenheit (37.8°C) or adding hot water also may cause loss of vitamins A and C and lead to the Maillard reaction—a reaction in which some amino acids bind with carbohydrate, making them unavailable to the child.
- Mix in the approved natural protein (breast milk or infant formula) if recommended by your nutritionist and metabolic doctor.
- Refrigerate medical food after mixing it. Discard any unused medical food 24 hours after mixing, because of nutrient loss.

Feeding Your Infant. The way to feed your baby with PKU is the same as for any baby. The Phenex-1 formula will be supplemented with breast milk or infant formula such as Similac Advance. The nutritionist may have you mix the two formulas together. The Phenex-1 mixture stored in bottles in the refrigerator may be warmed before feeding.

- Shake the formula mixture and pour into a bottle.
- Set a bottle in a pan of cold water on the stove and gradually warm it or run hot tap water over the bottle.

- Never use a microwave oven to warm formula as this can result in hot spots that can burn your baby.
- Always test the temperature of heated formula before feeding by shaking a few drops on your wrist. The formula should feel lukewarm.
- If the Phenex-1 mixture drips freely, the nipple holes are the correct size. Shake the bottle well before feeding.

DO NOT WARM THE BOTTLE IN THE MICROWAVE.
Uneven warming may cause serious burns.

To feed your baby, sit in a comfortable place, hold him in the curve of your arm, and keep the nipple filled with Phenex-1 mixture so that air will not be swallowed. You should burp your baby at least once during and again at the end of each feeding. Hold him upright against your shoulder or lay him face down on your lap and gently pat his back. Once feeding begins, use within one hour or discard.

Introduction of Solid Foods. No baby is born with the ability to swallow solid foods. The swallowing reflex develops at 2 to 3 months of age. Before this time, the baby's "tongue thrust" causes the tongue to protrude, making swallowing food difficult. Wait ing to feed solid foods until the baby is developmentally ready is best.

Phenex-1 is similar to infant formula, and the amounts, as well as the kinds of cereals, fruits, and vegetables prescribed, are about the same as for any baby. Your baby's nutritionist or metabolic doctor will advise you about when your baby

Feeding time should be a pleasant experience for you and your baby.

should start eating infant cereal and strained baby foods, and which foods to introduce. Guidelines for adding various foods to your baby's diet are given in Figure 5. However, your nutritionist or metabolic doctor may suggest different ages. Follow their advice.

At 7 to 8 months of age, your child may begin trying to eat foods such as crackers, low-protein toast, or pieces of fruit without help. At about 9 months of age, your child may begin using a spoon.

If your 9- to 12-month-old child has never eaten without help, dip her fingers into the food to give her the idea of finger feeding. Later, you can teach her to pick up a spoon and help guide it to her mouth. Putting your child on your lap to guide her hand may

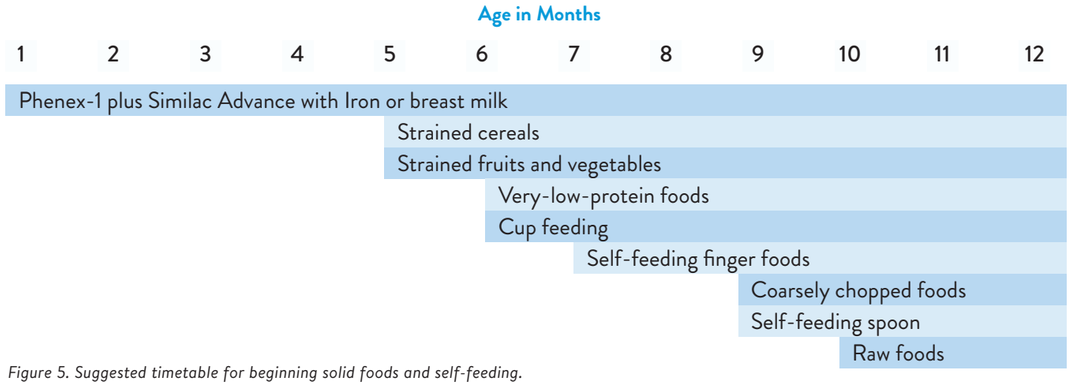


Figure 5. Suggested timetable for beginning solid foods and self-feeding.

be easier. Start with thick foods such as mashed potatoes since they do not slip off the spoon easily.

Do not worry if your child does not eat all of the foods you measure out; just estimate what was not eaten, replace the PHE with another food, and write it down.

When your child is older, the differences between the PHE-restricted diet and the diets of other children will be greater. Your child with PKU will require Phenex all his life to provide most of his protein, mineral, and vitamin needs.

Diet Guide and Food Lists. You will be given guidance at each clinic visit that spells out in detail what your child can eat. The amount and how to prepare the medical food mixture, as well as the types and amounts of food that your child is allowed, will be outlined. The nutritionist will help you work out a plan that meets your child's needs and fits into the family budget and lifestyle.

Lists of foods make meal planning easier and help you be sure your child's nutrient needs are met. You will have time to become familiar with the food lists and their nutrient content, because foods will be added slowly to your child's diet.

Be sure to check with the nutritionist before using any food that is not on the food lists provided.

When your child is older, you may need to use "free foods" to meet his energy needs. Free foods, which are high

in energy and contain little or no PHE, must not replace prescribed foods nor be used in large amounts. If your child eats too many of these free foods, he may become overweight or the extra sugar may cause dental caries. Special low-protein foods, including pasta, rice, crackers, cookies and breads, can be added to the diet. These foods will help satisfy your child's hunger.

The nutritionist can calculate the nutrient composition of the food and help you include it in the diet plan if it is not too high in PHE.

CHECKING YOUR CHILD'S PROGRESS

Blood Tests. Because your baby grows rapidly during the first year of life, blood is tested frequently for PHE content. Many doctors will ask for a blood sample once or twice a week during your baby's first 6 months, then once a week until 1 year of age. After 1 year of age, checking blood levels and diet records may be decreased to every 2 weeks if blood PHE levels are well controlled. How often your child is tested will be determined by your baby's metabolic doctor or nutritionist.

The staff at the metabolic clinic will teach you how to obtain blood samples. The procedure is quick and simple. When your child is very young, his heel is used to obtain the sample. When he is older, a finger is used. One large drop of blood is obtained by quickly pricking his heel or the side tip of his index finger with a sterile blade, or Autolet®, and then is dropped on circles drawn on filter paper or into very small tubes. Ask the health care professional at the metabolic clinic how much blood to collect. The filter paper or tubes are then either mailed or hand-carried to the clinic or laboratory. Sometimes, the clinic may obtain larger quantities of blood from a vein in your child's arm to look at all the amino acids in the blood.

Before taking a blood sample, you may be asked to accurately record your child's total food and beverage intake. On a form the clinic will provide, record the name of the food, the exact amount in household measures (cups, teaspoons, or tablespoons) or in grams that your child ate, and the PHE content based on the food lists or information given to you by the nutritionist.

Blood PHE Levels. The amount of PHE in the blood is an indirect measure of how much PHE is present in body tissues. Of most concern is the brain, because too much PHE is harmful to brain development. Because blood transports nutrients to the brain, the concentration of PHE in the blood will give the doctor and nutritionist an idea of how much PHE might be in the brain.

Recommended blood PHE levels vary, but are usually between 2 and 6 milligrams per deciliter (mg/dL) (120 and 360 micromoles per liter [µmol/L]).

Blood PHE levels that are high may indicate that your child is eating more foods that are **high** in PHE than his body needs for growth. Illness, such as colds and flu, can also cause the body to break down its own protein, releasing PHE in the blood, resulting in an elevated PHE level. When your child is not getting enough protein, because of rapid growth or inadequate intake of the medical food, the blood PHE may also rise. Generally, during rapid growth, the blood PHE level will decrease. A low PHE level usually indicates that your child is not getting enough PHE in the diet.

Clinic Visits. Because PKU is a lifelong condition that could harm growth and development, you will be asked to bring your child to the clinic frequently. If growth and development are normal and blood PHE concentrations remain within the treatment range, the frequency of clinic visits may be decreased with time.

At clinic visits, your child may be given developmental, physical, and neurological tests. Family interaction, which is important to her development, may also be evaluated. The clinic can address any questions or concerns you may have on how your child fits into your family. Diet changes will be made, if needed, and any questions you may have will be answered.

In addition to the metabolic specialist, you should have a local pediatrician or family doctor to provide required ongoing well-child care. Immunizations should be given at the usual times by this doctor, or you may obtain them from the health department.

YOUR CHILD'S GROWTH AND DEVELOPMENT

By 4 to 6 months of age, your baby's birth weight will double. The child with PKU who is kept on a controlled intake of PHE and whose diet supplies adequate nutrients should grow as well as a child without PKU.

During the second 6 months of life, the growth rate decreases. Your child may grow 1 inch (~2.5 centimeters [cm]) per month during the first 6 months and 4 inches (~10 cm) total during the second 6 months of life. This normal decline in the growth rate usually causes a decrease in appetite.

Although the requirement for energy (calories) and protein on the basis of body weight decreases, the total daily requirement for most nutrients increases with age. You will have to adjust food choices accordingly to ensure that your child has an adequate nutrient intake. The nutritionist will help you with food selections that are right for your child.

Weaning From Bottle to Cup. When the time comes to switch from the bottle, your child may need extra attention, as any child would. Weaning takes patience, especially if your child shows no interest in drinking from a cup or a glass.

Begin offering Phenex-1 from a cup when your child is between 5 and 8 months of age. Some mothers find a training cup that has a lid and a spout to be very useful.

During weaning, your child may not want to take all the prescribed Phenex in liquid form. You may have to use more of it in instant puddings, cereals, fruits, and soups. Some of the Phenex can be mixed into a paste with fruits and fed by spoon.

A child of 15 to 18 months of age may drink more medical food from a cup if she is given a small pitcher of Phenex and is encouraged to pour it into a small cup without help. Many parents have found that brightly colored straws or cups are good transitional tools.

Toddlers. Toddlers, children from 1 to 3 years of age, have a slow growth rate compared with that of infants. Toddlers may gain 4 to 5 pounds (1.8–2.3 kilograms [kg]) a year, compared with infants' gain of 12 to 22 pounds (5.5–10 kg) per year.

Growth during this period involves changes in body form. Legs lengthen and body fat decreases. Energy needs are decreased because of the slower growth rate. However, mineral and vitamin needs increase.

Toddlers seek independence and are very curious about their environment. Because toddlers want to do things for themselves, encourage your child to become self-feeders.

Preschoolers. Preschoolers also have a slow weight gain of 4 to 5 pounds (1.8–2.3 kg) per year. On the other hand, their total energy needs are greater than those of toddlers. Because your preschooler's nutrient and energy needs are greater, the nutritionist may tell you to increase foods containing high nutrient content.

Let your preschooler make some decisions. For example, permit him to choose which cereal, fruit, or vegetable to eat. Be warned that most preschoolers want to do things at their own speed. Be prepared to have your child spend so much time talking that little is eaten. This is normal behavior.

Social Interaction at Mealtime. Mealtimeis an important part of any child's social development and, whenever possible, the family should eat together. Younger children can learn how to feed themselves by watching older brothers and sisters.

Make meals for your child with PKU as similar as possible to the family's meals. Menus for him can be planned from those for the rest of the family. For example, whenever possible, use the same fruits and vegetables for everyone. The family's help and support are very important to maintaining the diet.

TEACHING YOUR CHILD DIET MANAGEMENT

Explaining the diet to your child can begin by calling allowed foods "special" or "just for you." From the time your child is very young, teach her to ask about unfamiliar foods before eating them. As your child grows older and is able to understand the concept of a missing enzyme, explain PKU to her. Some materials that you may find helpful are listed on page 17.

Toddlers and Preschoolers. Permit your preschooler to make food choices, such as what fruit to eat. Plan meals that vary in color, texture, flavor, and preparation methods. A child who is involved in food selection and preparation will be more interested in trying a new food. Involve your child in planning menus to help him become familiar with which foods are allowed and excluded. Let him help grocery shop, set the table, and prepare the food.

At about 4 years of age, children want to serve themselves. Teach your child the proper food portions. One way to do this is to use a "token" system. Tokens, symbolizing 5, 15, or 30 mg of PHE, may be used to "purchase" foods containing these amounts.

School Age. When your child reaches school age, he will become more independent in many aspects of his life, and eating is one of them. As he begins to develop logic and math skills, it is important to use these skills to help him understand the diet. Encourage your child to help prepare the medical food and calculate the amount of PHE in foods. As your child gets older, he should understand what levels of PHE are considered normal and the consequences of high PHE levels.

Adolescence. Adolescence may be a difficult time for both the child and the parents, regardless of the PKU! The influence of friends and the struggle for independence may make dietary compliance a challenge. They may feel that

PKU makes them different from their friends. Eating out with friends is part of growing up. Help your child develop the

skills for eating out, traveling, and "sticking" to the diet when not at home. Sometimes teenagers with PKU would rather tell people they are a vegetarian or vegan than explain about PKU. Help your teenager deal with her peers and not be self-conscious that she has PKU..

FEEDING PROBLEMS

Parents may be tempted to treat their child as a "sick" child and not follow their usual patterns of child rearing. The child with PKU is a normal child who just needs to manage food intake carefully. Ask your child's doctor, nutritionist, public health nurse, or social worker for support and help with problems listed below, if they occur.

Loss of Appetite. Loss of appetite can result from a variety of causes, including illness; eating too many sweet foods or desserts, which satisfy appetite and decrease the desire to eat the foods prescribed; getting too much Phenex, which may depress the appetite for other foods; or having a lower-than-normal blood PHE level.

Unusual Hunger. This may be an indication that the diet needs to be adjusted. The amount of Phenex may need to be increased because the table foods prescribed are not satisfying your child's needs. Providing low-protein foods is a great way to manage hunger without increasing protein intake.

Refusing Medical Food. A child may sometimes refuse Phenex because of normal variations in appetite, and this should not be a concern if average intake over a week is adequate. If Phenex is not offered regularly, a child may decide to refuse it. Improperly mixed Phenex also can cause refusal—too much water makes the volume too great too little water makes the medical food mixture too thick. A child may refuse Phenex as an attention-getting device, especially if he senses that his parents are anxious for him to drink the Phenex mixture. Phenex plays an important role in providing most of your child's nutrient needs. If refusal of the Phenex mixture continues to be a problem, the use of a feeding tube may need to be considered.

Refusing Solids. A child may experience normal variations in appetite or taste for certain foods. Food jags and strikes are common among young children. Or, the prescribed amount of Phenex may be too high, and the energy in it is causing her to lose her appetite.

Toddlers and preschoolers periodically have one of two characteristic feeding behaviors that cause parents concern. They may decide to stop eating—go on a "food strike"—or they may go on a "food jag."

When your child refuses to eat, offer food at the usual mealtimes, and if she refuses the food, take it away. Allow only water between meals. **Remember, Phenex supplies most of your child's nutrient needs, so her medical food should never be restricted.**

Do not give in to the food strike and offer Free Foods or foods that are not on the PHE-restricted diet. The nutritionist can help you during this trying time, so do not hesitate to call. It is also very important for parents to support each other in managing a food strike. If a child is allowed to eat foods not on her diet, blood PHE levels will not be controlled.

On a food jag, a child wants to eat the same food or foods for long periods. If the foods are nutritious and are in the diet, there's no reason for concern. Remember that most of your child's nutrient needs are supplied by Phenex.

Inappropriate Feeding Behavior. Inappropriate behaviors, such as refusing to give up the bottle and/or difficulty in eating solids, chewing, or self-feeding, may result from a variety of causes. These can include a delay in offering table foods; in teaching the child to drink Phenex by cup; or in allowing the child to self-feed, either with fingers or spoon. Always keep a positive attitude and make mealtimes a pleasurable event.

Try not to feed a child longer than necessary at mealtime to encourage self-feeding. Remember that small amounts of food are usually wasted when a child first learns to self-feed, but this is normal. Keeping food records will help the nutritionist estimate your child's intake.

A child may be using his diet as a way of getting attention or manipulating his parents. If your child has any of these problems, call the nutritionist. The nutritionist will give you support and offer suggestions to help solve the problem.

THE ROLE OF FAMILY AND OTHER CARE PROVIDERS IN MANAGING PKU

Parents carry the bulk of the responsibility for the diet. If possible, try to share the responsibility, so both parents can prepare and monitor the diet. Other children in the family, as well as the child with PKU, should learn about the diet as soon as they are old enough to understand it. Older brothers and sisters should be encouraged to be involved in feeding the child with PKU so they become familiar with which foods are allowed and excluded. Make sure they understand the importance of the diet for their brother's or sister's health. Brothers and sisters should be taught not to feel sorry for the child with PKU. Treat your child with PKU as normally as possible.

Grandparents love to spoil their grandchildren! It may be difficult for them as they sometimes feel the children are "missing out." It is important that they understand the diet and become actively involved as much as possible. A grandmother may be the ideal person to experiment with low-protein cooking and provide special PKU treats.

Explain PKU to relatives, friends, day-care providers, baby-sitters, and all teachers. They should become familiar with which foods are allowed and excluded, and understand the importance of the diet. Give a list of the foods allowed and excluded to persons who feed your child and explain the list as well as the exact menu.

Tell everyone who cares for your child that even "just a little bite" of a food is not allowed. Emphasize what can happen if your child does not stay on diet.

YOUR CHILD'S DIET DURING ILLNESS

A body temperature greater than 98.6°F (37°C) is a fever. During fever, the body's rate of using food for energy speeds up. If extra energy is not supplied during illness, the body will break down its own muscle protein and fat stores for energy. Muscle protein breakdown needs to be prevented in children with PKU, because it will release too much PHE into the blood. PHE is then carried to the brain, where it may have a harmful effect. Give your child extra low-protein food, formula and fluids during illness. This extra food will decrease the amount of muscle protein broken down for body energy.

Feeding an ill child can be very difficult. Often a child with fever is restless and loses appetite. The illness might also include stomach upset, nausea, or even vomiting. A child may become very dehydrated because of the high body temperature and a lack of adequate fluid intake.

Illness in the PKU child can cause blood PHE levels to increase or decrease. Your metabolic doctor or nutritionist will help you

decide on how to manage it. Ask about using medication such as acetaminophen (Tylenol®) to reduce fever.

Here are some suggestions of things to do when your child is ill with fever.

- Do not force-feed food or Phenex, especially if your child is nauseated or vomiting. Soda crackers may be the only food he will feel like eating. Encourage intake of any allowed foods that your child is willing to eat.
- Offer Pedialyte® Oral Electrolyte Solution with added carbohydrate, such as dextrose or table sugar (3 Tbsp sugar to 8 fl oz of Pedialyte); Pro-Phree® non-cola carbonated beverages; sugar-sweetened carbonated beverages; Kool-Aid®; Tang®; tea with sugar; vegetable. broth; or fruit juices with some sugar added.
- Dilute the Phenex mixture, or use liquid Jell-O® if tolerated.
- Freeze any of the beverages listed and make into chipped ice. Frequently feed small amounts of this chipped ice to provide energy and prevent dehydration.
- As your child's appetite improves, gradually return to the usual diet plan.

A LOOK TO THE FUTURE

Continuing the Diet. PKU is a serious health concern. Treating a child as early in life as possible helps to prevent developmental delay and neurological damage. Not following the PHE-restricted diet may cause mental and nervous system damage at any age. Children and adolescents who have elevated PHE levels may have learning and behavior problems. Adults with elevated PHE levels may have trouble concentrating at work. These problems can be reversed when they return to a PHE-restricted diet. For women with PKU, there is the strong possibility of damage to an unborn child if she is not on diet during pregnancy. Women with PKU who wish to become pregnant may have difficulty returning to the PHE-restricted diet if they have been eating an unmanaged diet.

Following a PHE-restricted diet closely and keeping PHE levels witin the treatment range, the person with PKU can lead a normal, productive life.

Family Planning. Recall that 1 of every 50 people is a carrier for PKU. The chance of two carriers marrying is 1 in 50 multiplied by 1 in 50, or 1 chance in 2,500. PKU occurs once in every 15,000 to 20,000 live births. The chance that two carriers of PKU will have a child with PKU is 1 in 4, or 25% **for each pregnancy** (Figure 6). The chance that two carriers will have a child who is a carrier is 1 in 2, or 50% for each pregnancy.

In Figure 4, you can see that a mother and father who are carriers of PKU can give their babies a normal (●) and a PKU (●) gene (B and C). These babies don't have PKU, but they are carriers for it. Each time a child is conceived by parents who are carriers of PKU, there is exactly the same probability (1 in 4, or 25%) of having a child with PKU.

Before a couple who has a child with PKU has any more children, they should take time to seriously think about the special parenting tasks that parents of a child with PKU must manage.

If you decide to have another child, give yourselves time to adjust to the special needs of the first child. Make sure you have learned diet management for your child with PKU before having another child.

Offspring of a PKU person. All children born to a parent with PKU will either be carriers of the gene or have PKU. As illustrated in Figure 6, all offspring of a parent with PKU (●●) and a parent who is normal (●●) will inherit one normal (●) and one PKU (●) gene. Each child will be a carrier.

Because the incidence of PKU carriers is approximately 1 in 50 (2%), the possibility of a person with PKU and a carrier mating and producing children is small. However, if a person with PKU does have children with a person who is a carrier for PKU, approximately one-half (50%) of their offspring will have PKU and one-half (50%) will be carriers (Figure 7).

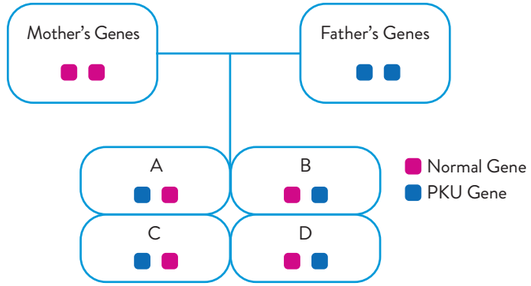


Figure 6. All children of a person with PKU and a person who doesn't have PKU will be carriers of PKU.

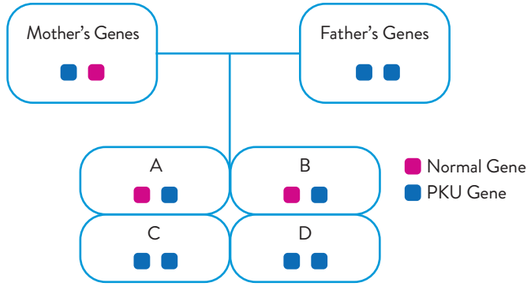


Figure 7. Approximately half of the children of a parent with PKU and a parent who is a carrier of PKU will have PKU.

Childbearing by Women with PKU. For the women with PKU, having children may cause problems. A major concern for PKU women is the stress of a pregnancy on their metabolic control. Their nutritional requirements can rapidly change during the course of a pregnancy. Maternal PKU women must be carefully monitored for their own safety, as well as for their health of the baby.

PHE is actively transported across the placenta. If a woman conceives a child while her blood PHE levels are high, mental retardation and birth defects may occur in the fetus before the pregnancy has even been confirmed. If a woman with PKU is not on a PHE-restricted diet during pregnancy, then the baby is also exposed to the increased concentrations of PHE. Prolonged high PHE levels have an increased risk of causing low-birth-weight babies, and a majority of these infants have mental retardation and/or birth defects.

If a woman with PKU who is not on a PHE-restricted diet decides to become pregnant, she should go back on diet three to six months before becoming pregnant. Reducing blood PHE levels to near normal may help prevent complications in the unborn child.

Throughout pregnancy, the mother-to-be's blood PHE levels should be maintained between 2 and 6 mg/dL (120 to 360 µmol/L) and her diet evaluated regularly. Successful pregnancies require attentive diet monitoring during prenatal, labor, delivery, and postpartum periods.

Kool-Aid®-Flavored Phenex®-1

Yield: 8 fl oz

40 g Phenex-1
3 Tbsp, **level**, sugar[1]
1/4 tsp Kool-Aid or Wyler's® **Unsweetened** Soft Drink Mix
Do not use mixes that contain NutraSweet® or aspartame.

Add water (room temperature) to ingredients to make 8 fl oz. Mix in a blender at lowest speed no more than 4 seconds. Or, shake briskly in a closed container for 10 to 12 seconds. Serve chilled.

Nutrient	*1 fl oz*	*8 fl oz*
Phenylalanine, mg	0	0
Tyrosine, mg	75	600
Protein, g	0.75	6
Energy, kcal	42	336

[1] Osmolality (concentration of particles in solution) may be too high if more sugar is added, which may cause bloating and diarrhea.
[2] The amount of drink mix may be varied according to taste preference.

Fruit Juice-Flavored Phenex®-2

Yield: 8 fl oz

20 g Phenex-2
3 fl oz concentrated apple, grape, or orange juice
Water (room temperature) to make 8 fl oz

Warm juice concentrate to room temperature. Place all ingredients in a blender at lowest speed no more than 4 seconds. Or, shake briskly in a closed container for 10 to 12 seconds. Serve chilled.

Nutrient	*Apple juice*	*Grape juice*	*Orange juice*
Phenylalanine, mg	15	15	30
Tyrosine, mg	609	603	615
Protein, g	6.5	6.7	8.6
Energy, kcal	257	275	252

1, Concentrated fruit "drinks" do not contain any protein (valine). Substitute when available.
[1]Please check with your dietitian or doctor before using this recipe in infants.

Kool-Aid®-Flavored Phenex™-2

Yield: 16 fl oz

40 g Phenex-2
3 Tbsp, **level**, sugar[1]
1/2 tsp Kool-Aid or Wyler's **Unsweetened** Soft Drink Mix[2]
Do not use mixes that contain NutraSweet® or aspartame.

Add water (room temperature) to ingredients to make 16 fl oz. Mix in a blender at lowest speed no more than 4 seconds. Or, shake briskly in a closed container for 10 to 12 seconds. Serve chilled.

Nutrient	*16 fl oz*
Phenylalanine, mg	0
Tyrosine, mg	1200
Protein, g	12
Energy, kcal	308

[1] Osmolality (concentration of particles in solution) may be too high if more sugar is added, which may cause bloating and diarrhea.
[2] The amount of drink mix may be varied according to taste preference.

Additional Tips for Flavoring Phenex Medical Food

- Add chocolate or strawberry syrup.
- Mix Phenex with fruit to make a "smoothie."
- Freeze flavored medical food into "slushies" or "popsicles."
- Add dry Phenex to pudding (lemon, tapioca, vanilla, etc) mixture. Prepare pudding with non-dairy creamer.

Use low-protein food lists to calculate protein content of flavorings.

Support Groups/Newsletters

Children's PKU Network
3306 Buman Rd.
Encinitas, CA 92024
Phone:858-756-0079
Fax:858-756-105
E-mail: pkunetwork@aol.com
Web site: www.pkunetwork.org

National PKU News
PO Box 43552
Montclair, NJ 07043
Phone: 973-619-9160
EMail: info@pkunews.org
Web site: www.pkunews.org

How Much Phe
Email: support@howmuchphe.org
Website: www.howmuchphe.org

National PKU Alliance (NPKUA)
PO Box 1872
Eau CLaire, WI 54702
Phone: 715-495-4008
Fax: 715-713-0138
Web site: www.npkua.org

Low-Protein Food Suppliers

Canbrands Specialty Foods, Inc.
3500 Laird Rd.
Mississauga, Ontario, Canada L5L 5Y4
Phone: (905) 829-6003
Email: helpdesk@canbrands.ca
Web site: www.canbrands.ca

Dietary Specialties
8 S. Commons Rd.
Waterbury, CT 06704
Phone: (888) 640-2800
Web site: www.dietspec.com

Ener-G® Foods, Inc.
5960 First Avenue South
Seattle, WA 98108
Phone: (800) 331-5222; (206) 767-3928
Fax: (206) 764-3398
E-mail: customerservice@ener-g.com
Web site: www.ener-g.com

Med-Diet™ Laboratories, Inc.
3600 Holly Lane, Suite 80
Plymouth, MN 55447
Phone: (800) 633-3438 (MED-DIET);
(763) 550-2020
Fax: (763) 550-2022
E-mail: info@med-diet.com
Web site: www.med-diet.com

PKU Perspectives
PO Box 696
Pleasant Grove, UT 84062
Phone: (866) PKU-FOOD; (801) 785-7722
Fax: (866) 701-3788
Web site: www.pkuperspectives.com

Taste Connections, LLC
Phone/Fax: (310) 371-8861
E-mail: tasteconnect@verizon.net
Web site: www.tasteconnections.com

IMPORTANT PHONE NUMBERS

Nutritionist: _____

Metabolic Doctor: _____

Pediatrician: _____

Police: _____

Fire: _____

Hospital: _____

Other: _____

_____

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Maple Syrup Urine Disease

Urea Cycle Disorders

Propionic and Methylmalonic Acidemia

Glutaric Aciduria Type I

Tyrosinemia

Isovaleric Acidemia and Disorders of Leucine Metabolism

Homocystinuria

Dietary Modification of Protein

Hypercalcemia

Ketogenic Diet Management Carbohydrate Disorders

Dietary Modification of Carbohydrate and Fat

Use under medical supervision.

Abbott Metabolic Medical Foods

Phenex® is part of an extensive
line of medical foods from
Abbott, makers of Similac®

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