Abbott

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

Propionic Acidemia

CONTENTS

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

INTRODUCTION TO PROPIONIC ACIDEMIA

Your child has a condition called propionic acidemia (pro-pee-on-ic a-cid-em-ia), or PA for short. Children who have inherited this condition cannot use the amino acids isoleucine (i-so-lu-seen) (ILE), methionine (me-thi-o-neen) (MET), threonine (three-o-neen) (THR), valine (vay-leen) (VAL), and some fats in a normal way.

These amino acids are found in all foods that contain protein. You will need to feed your child all the foods necessary for normal growth and development, but only the amount of ILE, MET, THR, VAL, and specific fats he can safely use.

Learning some medical terms in genetics and nutrition 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.

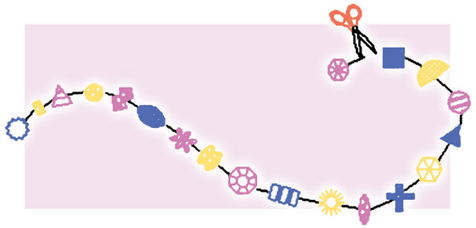


Figure 1. Amino acids are joined together like beads on a necklace to form protein. Enzymes act like scissors to remove amino acids from protein.

WHAT IS PA?

PA is an inherited disorder of amino acid and odd-chain fatty 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. Nine of these amino acids are called "essential amino acids" because the body must have them to live, and the only way to obtain them is from food. Four of these essential amino acids are ILE, MET, THR, and VAL.

All foods with protein contain ILE, MET, THR, and VAL. High-protein foods are dairy products, beans and peas, eggs, meat, poultry, nuts, soy products, seafood, seeds, and nut butters. Fruits, grains, and vegetables contain less protein and so have less ILE, MET, THR, and VAL.

Splitting protein into amino acids requires special substances called enzymes (n-simes) Think of an enzyme as a pair of scissors snipping beads off a necklace (Figure 1).

Once ILE, MET, THR, and VAL are split off the food protein, they are absorbed, changed, and used to help form many other useful substances in the body.

In PA, the enzyme called propionyl-CoA carboxylase (pro-peeo-neel Co-A car-box-ill-ase), or PCC, is absent or not working properly. People with PA, such as your child, cannot make enough PCC to handle all the ILE, MET, THR, and VAL in the protein foods they eat, and ILE, MET, THR, VAL, and some fats cannot be used normally. They are changed and build up in the body as the organic acids propionylCoA, propionic acid, and other toxic (poisonous) substances, such as methylcitrate (meth-il-sit-rate) and hydroxypropionate (hi-drox-ee- pro-pee-o-nate), and cause the symptoms of PA (Figure 2).

Think of the situation as a traffic light (Figure 3). A green traffic light (normal PCC) allows ILE, MET, THR, VAL, and odd-chain fatty acids to be used normally. A red light (too little PCC) keeps ILE, MET, THR, VAL, and some fats from being used. If the light is stuck on red, a traffic jam occurs—ILE, MET, THR, VAL, odd-chain fatty acids, and the organic acids they create increase and cause the symptoms of PA.

If a person is not treated, these products build up in the blood and spill into the urine and perspiration (sweat). People who have untreated PA have too much ILE, MET, THR, VAL, odd-chain fatty acids, and organic acids in their blood and urine.

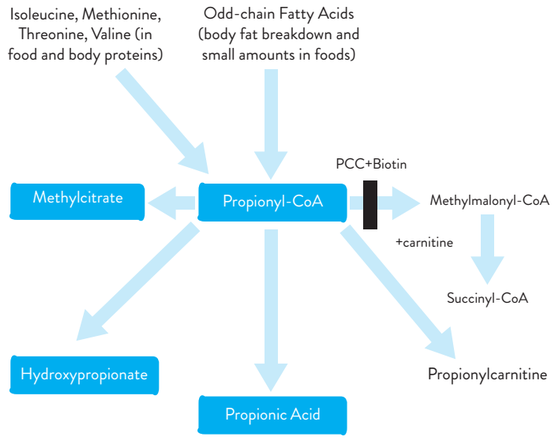
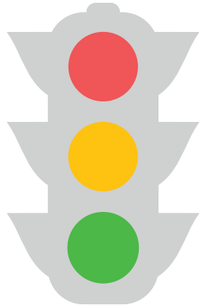


Figure 2. Metabolic pathway for ILE, MET, THR, VAL, and some fats in PA. Substances in the shaded boxes are organic acids that build up when PCC does not work.



PCC not working.
ILE, MET, THR, VAL, and certain fats cannot be broken down. Toxic metabolites build up.

PCC not working well.
ILE, MET, THR, VAL, and certain fats may not be broken down efficiently.

PCC working.
ILE, MET, THR, VAL, and certain fats used normally.

Figure 3. The propionyl-CoA carboxylase (PCC) traffic light..

PA: AN INHERITED DISORDER

PA is a genetic disorder 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 body cell. These genes serve as blueprints, or patterns, for making body tissues.

Each parent of a child with PA has one normal (●) and one altered PA (●) gene. Each one 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 PCC to use ILE, MET, THR, VAL, and odd-chain fatty acids 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 PA (●) gene. His body will make enough PCC to use ILE, MET, THR, VAL, and odd-chain fatty acids normally, but he can pass on the PA gene to his offspring. A person with this gene set—one normal and one PA—is called a carrier. Being a carrier does not affect the person's health. **You, as parents of a child with PA, are carriers.** Brothers and sisters of your child with PA may also be carriers.

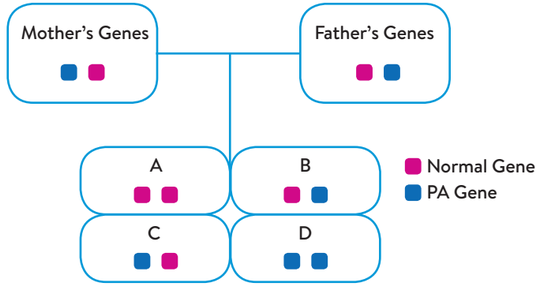


Figure 4. Genetic inheritance of PA.

A child with gene set D has PA caused by inheriting the two PA genes (●●), one from her mother and one from her father. Her body will not be able to use the ILE, MET, THR, VAL, and odd-chain fatty acids in food normally. She will also pass a PA gene on to each of her offspring. Your child with PA has this gene set.

DIAGNOSIS OF PA

Most states require all babies to be screened for PA and other conditions before they are discharged from the hospital. In the states that do not include PA in their newborn screening, many infants are diagnosed after they start showing symptoms of the condition.

If the initial screening tests show that a baby may have PA, additional blood and urine are collected for more precise measurements that will confirm the diagnosis. The doctor may recommend hospitalization while this testing is done so that treatment can be started sooner if the baby has PA.

SYMPTOMS OF PA

Excessive ILE, MET, THR, VAL, and propionic acid in blood and tissues affect the nervous system. Newborns with severe PA typically show symptoms within the first days of life, sometimes before the screening result is available. Children with mild or moderate PA may not show symptoms until early childhood. These symptoms may be brought on by illnesses or even exhaustion. An infant or child who is untreated, ill, or in poor control may have some or all of these symptoms:

- Anorexia (refusal to eat)
- Irregular muscle movement, poor sucking ability
- Vomiting
- Dehydration
- Lethargy (excessive tiredness or sleepiness)
- Acidosis (excess acid in the blood)
- Respiratory difficulty (unusually rapid breathing)
- Hypotonia (poor muscle tone)
- Seizures

Poor control or late diagnosis of PA may also cause some delay in development.

Table 1. General Guide to Foods on Protein-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 seafood, meat, nuts, nut butters, poultry, seeds, tofu	Breast milk, infant formulas, bread, crackers, fruit, fruit juices, low-protein cereals, popcorn, potato chips, special low-protein foods, vegetables, vegetable juices, bread with added calcium propionate or sodium propionate, fish oils, butter, lard, olive oil, excess vegetable oils	Gumdrop candy, hard candy, jelly, Kool-Aid®, lemonade, lollipops, Popsicles®, pure sugar, soda, coconut oil, palm oil, peanut oil

NUTRITION SUPPORT OF PA

Since the late 1960s, a diet that reduces protein intake has been used to prevent the acidosis, developmental delay, and other problems associated with untreated PA. This diet controls the blood levels of propionic acid and other toxic substances described earlier within a range that may allow normal mental development and growth. The special PA diet for your child is designed for his individual needs, and following it is very important.

Many foods contain protein, which contains ILE, MET, THR, and VAL. Therefore, the amounts of foods that contain protein must be limited in your child's diet. 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).

Odd-Chain Fatty Acids. Food fats that contain fatty acids with an uneven number of carbons are called odd-chain fatty acids. These fats produce some propionic acid when they are used in the body. Food fats that contain a small amount of odd-chain fatty acids include butter, chicken fat, cream, some fish oils, lard, and olive oil. Therefore, these foods may be restricted for your child. Avoidance of fasting is very important as fasting leads to a build-up of odd-chain fatty acids. The higher level of propionyl-CoA acts as a "starter" for the production of odd-chain fatty acids.

PUFAs. Vegetable oils such as canola, corn, safflower, and soy contain large amounts of polyunsaturated fatty acids (PUFAs). According to some reports, when large amounts of PUFAs are eaten, a small portion of each PUFA is broken down to propionic acid. Not everyone agrees with these reports. However, until more is known on this topic, the best approach may be to not consume excess amounts of vegetable oils that are high in PUFAs. Coconut oil, palm oil, and peanut oil are low in PUFAs and may be used as energy (calorie) sources. Ask your metabolic doctor or dietitian what is best for your child.

Requirements for ILE, MET, THR, VAL, Protein, and Energy. A child with PA who eats enough protein to grow properly gets too much ILE, MET, THR, and VAL. Foods high in protein are cheese, milk, soy milk, eggs, meat, poultry, fish, nuts, tofu, beans and peas, seeds, and nut butters. Foods low in protein include some grains, fruits, fats, vegetables, and sweets. Eating these foods in only small amounts to provide just enough ILE, MET, THR, and VAL will not meet your child's needs for growth. **A special medical food that is high in protein but contains no MET or VAL and is low in ILE and THR is also necessary.**

To be sure your child is getting enough energy and protein (as well as adequate amounts of ILE, MET, THR, and VAL) for growth and development, the nutritionist carefully calculates the amount of each nutrient needed. Too little ILE, MET, THR, VAL, protein, or energy can result in growth failure. Frequent diet adjustments are necessary, especially during the first years of life when children grow rapidly. The nutritionist or metabolic doctor will make these diet changes based on your child's health, growth, dietary intake, and laboratory tests.

Propimex®-1 Amino Acid-Modified Medical Food With Iron is a medical food used to provide protein for infants and toddlers with PA. Propimex-1 contains very little ILE and THR and no MET or VAL. Formulas such as Similac® Advance® Infant Formula with Iron, breast milk, or other intact protein, **must** also be fed to provide the specific amounts of the amino acids your child needs for growth and development. Breast milk is lower in ILE, THR, MET, and VAL than infant formula or cow's milk and can be used to supply the required amino acids. 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 formula or breast milk your child will need in addition to his medical food. Propimex-1 is well supplied with fat, carbohydrate, minerals, and vitamins. Supplemental minerals or vitamins are not usually needed when the diet is followed as directed.

Propimex®-2 Amino Acid-Modified Medical Food is a medical food used in treating children and adults with PA. This product contains little ILE and THR and no MET and VAL, so some whole protein containing these amino acids **must** come from food sources. Your nutritionist will tell you which medical food is right for your child. **Propimex-1 and Propimex-2 are to be used under medical supervision.**

Propimex-1 and Propimex-2 taste different from milk. Most children and adults get accustomed to the flavors of the foods they eat. They may seem distasteful to you, but it is important not to show this to your child, either by word or action. Most children taking medical foods for PA like them **IF** they are started early and **IF** their family has a positive attitude.

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

Flavorings, such as Kool-Aid® Unsweetened Soft Drink Mixes, Wyler's® Unsweetened Soft Drink Mixes, Mio Liquid water enhancers, and concentrated fruit juices can be added to Propimex. Remember to count the added ILE, THR, MET, and VAL if present in any of the flavorings. Propimex may also be made into a paste and combined with some allowed fruits, such as applesauce or other fruit purees, or combined with instant pudding mixes. Check the label and be careful which pudding mixes you buy, as some contain more protein than others.

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)

INTERNATIONAL SYSTEM OF MEASUREMENT (METRIC SYSTEM)

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

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 protein is to weigh foods on a scale that reads in grams.

Medical Food Preparation. Mix a 24-hour supply of medical food at one time, or as instructed by your nutritionist. Your nutritionist will tell you exactly how much Propimex and other formulas or breast milk to use.

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 Propimex label.

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

- 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° F (37.8° C) or adding hot water 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 or metabolic doctor.
- Refrigerate the medical food after mixing. Discard any unused medical food 24 hours after mixing because of nutrient loss.

Feeding Your Infant. The way you feed your baby with PA is the same as for any baby. The Propimex-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 Propimex-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 Propimex-1 mixture drips freely, the nipple holes are the correct size. Shake the bottle well before feeding.

To feed your baby, sit in a comfortable place, hold her in the curve of your arm, and keep the nipple filled with the Propimex-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 her upright against your shoulder or lay her face down on your lap and gently pat her back.

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

Introduction of Solid Foods. No baby is born with the ability to swallow solid foods. The swallowing reflex usually develops at 2 to 3 months of age. Before this time the baby's "tongue thrust" reflex, which he needs to suck from a nipple, causes his tongue to protrude, making swallowing food difficult. Waiting to feed solid foods until the baby is developmentally ready is best.

The amounts and kinds of cereals, fruits, and vegetables that are appropriate to feed your child are about the same as for any baby. Your baby's nutritionist or metabolic doctor will advise you about when 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 have different suggestions. Follow their advice.

As your child begins to eat solid foods, the protein they contain must be calculated into his diet. At the same time, your nutritionist may begin to reduce the amount of baby formula or breast milk in the Propimex mixture so that his intake of ILE, MET, THR, and VAL remains the same. You may be given a prescribed amount of protein your child's diet must be restricted to each day, and his food must be chosen so that it provides this amount.

At about 7 to 8 months of age, your child may begin to eat finger foods such as crackers, low-protein toast, or pieces of fruit without help. At about 9 months of age, your child may begin to feed herself 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 be easier. Start with thick foods such as mashed potatoes because they don't slip off the spoon easily.

Don't worry if your child doesn't eat all of the foods you measure out; just estimate what was not eaten, replace the protein with another food, and write it down.

When your child is older, the differences between the ILE-, MET-, THR-, and VAL-restricted diet and the diet of other children will be greater. Your child with PA will require Propimex all her life to provide most of her protein, mineral, and vitamin needs.

Some children with PA have poor appetites. These children sometimes eat so little food that they break down their own body fat and protein for energy, or they don't grow well. Poor appetite and inadequate food intake can make the child with PA very sick.

If your child isn't eating all of his prescribed diet every day, talk with his metabolic doctor or nutritionist immediately.

Diet Guide and Food Lists. You will be given guidance at each clinic visit that spells out in detail what your child can eat, including how to prepare the medical food mixture, and the types and amounts of food that he is allowed. The nutritionist will tell you the daily amount of protein that meets your child's needs. She will also help you learn to choose his food so that it provides the amount that is needed 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.

When your child is older, you may need to use Free Foods (see Table 1) to meet his energy needs. Free foods, which are high in energy and contain little or no protein, 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 tooth decay. Special low-protein foods, including pasta, rice, crackers, cookies, breads, and other products can be added to the diet to increase the amount and variety of food allowed. These foods will help satisfy your child's hunger.

If you have questions about the content of certain foods, 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 protein.

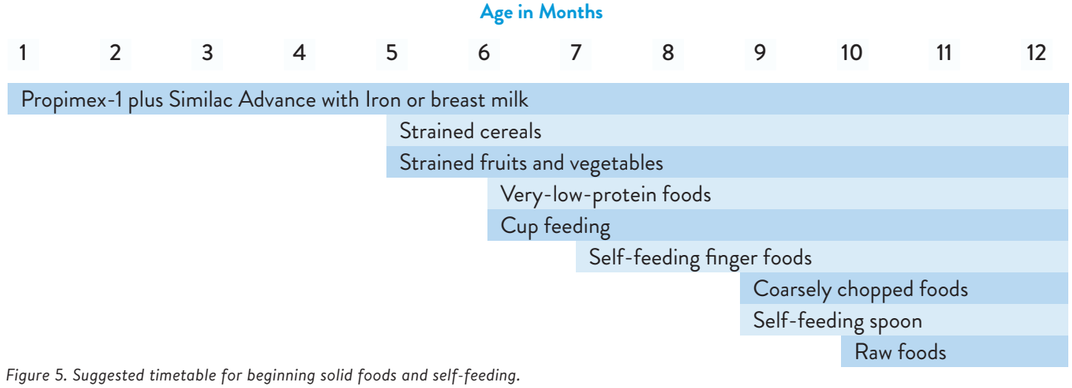


Figure 5. Suggested timetable for beginning solid foods and self-feeding.

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

Additional Therapies. Certain vitamins or medications for your child with PA may be recommended by your metabolic doctor.

Biotin is a vitamin cofactor that may help the enzyme PCC work better. Biotin may be prescribed on a trial basis to see if it is beneficial.

Carnitine is a nutrient that helps energy enter cells in the body. Carnitine can also bind with toxic metabolites and help with their excretion.

Metronidazole (me-troe-ni-da-zole) (Flagyl®) is an antibiotic that may be given to reduce the amount of propionate that is produced by bacteria in the gut. Other antibiotics may be used for this purpose as well.

Constipation can be a common problem in PA, and medications to increase gut functioning are often used to treat it. Your metabolic doctor will decide what medications are appropriate and prescribe them in doses based on your child's individual needs.

CHECKING YOUR CHILD'S PROGRESS

Laboratory Tests. Because your baby grows rapidly during the first year of life, blood and urine samples will be tested frequently to monitor how well his PA is controlled, and to indicate if changes in treatment are needed. Your doctor may ask for blood and urine tests frequently in the first year of life. After 1 year of age, blood and urine will be checked less frequently if blood ILE, MET, THR, VAL, and organic acid levels are well controlled. Your baby's metabolic doctor will determine how often your baby is tested.

Urine may be monitored for the presence of organic acids and their toxic substances. When a child is not getting enough calories or is ill, his body fat is broken down for energy. Ketones, made from excess use of body fat for energy may be detected in the urine. The clinic may provide you with test strips (Ketostix®) that measure the level of ketones in the urine. The presence of ketones in the urine may indicate that your child is not getting enough calories.

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 protein content based on the food lists or information given to you by the nutritionist. This will help the metabolic doctor and nutritionist evaluate your child's laboratory test results.

ILE, MET, THR, and VAL Levels. The amount of ILE, MET, THR, and VAL in the blood is an indirect measure of the amount of these amino acids present in body tissues. Of most concern is the brain, because too much ILE, MET, THR, VAL, and propionic acid are harmful to brain development. Because blood transports nutrients to the brain, the concentration of ILE, MET, THR, VAL, and propionic acid in the blood will give the doctor and nutritionist an idea of the amount of ILE, MET, THR, and VAL that the brain is exposed to. Blood glycine and carnitine levels may also be monitored. Recommended blood ILE, MET, THR, and VAL levels vary, and your metabolic doctor or nutritionist will discuss what your child's laboratory results mean.

Blood ILE, MET, THR, and VAL levels that are **high** may indicate that your child is eating more foods that are high in protein than his body needs for growth. Illness, such as colds and flu, can also cause the body to break down its own protein, releasing ILE, MET, THR, and VAL into the blood. When your child is not getting enough protein, because of rapid growth or inadequate intake of the medical food, the blood ILE, MET, THR, and VAL levels may also rise. Initially during rapid growth, the blood ILE, MET, THR, and VAL levels will decrease and then increase again as the body breaks down its own protein.

Low ILE, MET, THR, and VAL levels usually indicate that your child is not getting enough of these amino acids from whole protein in the diet.

Clinic Visits. Because PA 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 laboratory testing shows good control of PA, the frequency of clinic visits may be decreased with time.

At clinic visits, your child may be given developmental, physical, and neurologic tests. Family interaction, which is important to her development, may also be evaluated. Diet changes will be made, if needed, and any questions or concerns you may have will be addressed.

In addition to the metabolic specialist, you should have a local pediatrician or family doctor to provide 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 infant's birth weight will double. The child with PA whose intake of ILE, MET, THR, and VAL is well controlled and whose diet supplies adequate nutrients should grow as well as a child without PA.

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 and protein on the basis of body weight decreases, the total daily requirement for most nutrients increases with age. You will need 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 Propimex-1 from a cup when your child is between 5 and 8 months of age. Because tastebuds vary on different parts of the tongue, Propimex may taste different when taken from a cup instead of a bottle. Also, the smell of

Propimex may be more pronounced in an open cup. Some parents find a training cup that has a lid and a spout to be very useful. Offering the Propimex at a cold temperature may also increase your child's willingness to drink it from a cup.

During weaning, your child may not want to take all the prescribed Propimex in liquid form. You may offer some of it in instant puddings, cereals, fruits, and soups; or it can be mixed into a paste with fruits and fed by spoon.

ADDITIONAL WATER MUST BE OFFERED WHEN PROPIMEX® IS FED AS A PASTE.
Consult your child's nutritionist.

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 Propimex and is encouraged to pour it into a small cup without help. Many parents have found using brightly colored straws, special cups, or sports bottles to be good transitional tools to help wean a child from the bottle.

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 the 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 feed himself.

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 with a high nutrient content. These foods are packed with vitamins and minerals and are energy dense.

Let your preschooler make some decisions. For example, permit him to choose which cereal, fruit, or vegetable to eat. Be aware 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. Mealtime is an important part of every 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 PA 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. You can also prepare a low-protein pasta or meatless dish that is similar to the one served to other family members. 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 she 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 or nonworking enzyme, explain PA. 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 he wants to eat. Plan meals that have variety 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 become familiar with foods allowed and excluded. Let him help grocery shop, set the table, and prepare the food.

At about 3 to 4 years of age, children want to serve themselves. Teach your child the proper food portions. Your dietitian may provide you with a technique or way to count protein that you can teach your child.

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

Adolescence. Adolescence may be a difficult time for both you and your child, regardless of the PA! The influence of friends and the struggle for independence may make dietary compliance a challenge. Teens may feel that PA 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 PA would rather tell people they are vegetarian or vegan than explain about PA. Help your teenager deal with her peers and not be self-conscious that she has PA. Finding a peer with PA through support groups can be a great comfort for a teen. Ask the clinic for resources and suggestions.

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 PA is a normal child who needs to manage food intake carefully. Ask your child's doctor, nutritionist, public health nurse, or social worker for support and help if any of the following problems should occur.

Loss of Appetite. Loss of appetite can result from a variety of causes, including illness; eating too many sweet foods or desserts that satisfy the appetite and decrease the desire to eat the foods prescribed; getting too much Propimex, which may depress the appetite for other foods; or having lower than normal amino acid blood levels. Medications may also decrease appetite. Some children have a poor appetite despite adequate treatment and dietary management. In such cases, it may be necessary to use a feeding tube to ensure nutrition needs are met.

Unusual Hunger. Unusual hunger may indicate that your child's diet needs to be adjusted—the amount of Propimex may need to be increased because the table foods prescribed are not satisfying her needs. Using low-protein foods is a great way to manage hunger without increasing protein intake.

Refusing Medical Food. A child may sometimes refuse Propimex because of normal variations in appetite, and this should not be of concern if his average intake over a week is adequate. However, if Propimex is not offered regularly, a child is more likely to refuse it. Improperly mixed Propimex can also be a cause—too much water makes the volume too great; too little water makes the medical food mixture too thick. Or, a child may refuse Propimex as an attention-getting device, especially if he senses that his parents are anxious for him to drink the Propimex mixture. Remember, Propimex plays an important role in providing most of your child's nutrition needs. If refusal of the medical food 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. Or, the prescribed amount of Propimex may be too high and the energy it provides may cause 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."

During a food strike when your child refuses to eat, offer food at usual mealtimes and if she refuses the food, take it away. Allow only water between meals. She will become hungry and then eat. **Remember that most of your child's nutrient needs are supplied by Propimex, so her medical food should never be restricted.**

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

During a food jag, a child wants to eat the same food or foods for long periods. If the foods are nutritious and are appropriate for the PA diet, there is no reason for concern.

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, difficulty in teaching the child to drink Propimex by cup, or not allowing the child to feed himself either with fingers or spoon. Always keep a positive attitude and make feeding 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 feed himself, but this is normal. Keeping food records will help your nutritionist estimate your child's intake.

A child may be using his diet as a way of getting attention or manipulating 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 A PA

Parents carry the bulk of responsibility for their child's diet. If possible, try to share the responsibility of preparing and monitoring the diet. Other children in the family, as well as the child with PA, should learn about the diet as soon as they are old enough to understand it. Older brothers and sisters should be encouraged to help feed the child with PA so they become familiar with foods 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 PA because he is on a special diet. Everyone should treat the child with PA as normally as possible.

Grandparents love to spoil their grandchildren! It may be difficult for them as they sometimes feel the child with PA is "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 low-protein treats.

Explain PA to relatives, friends, day-care providers, babysitters, and all teachers. They should become familiar with foods allowed and excluded and understand the importance of the diet. Give a list of the foods allowed and not allowed to anyone who feeds 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 high-protein food is not allowed. Emphasize what can happen if your child does not stay on the diet.

YOUR CHILD'S DIET DURING ILLNESS

A body temperature greater than 98.6° F (37° C) or a rectal temperature over 100° F (37.8° 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 and body fat breakdown needs to be prevented in children with PA because it will cause blood levels of ILE, MET, THR, VAL, and some fatty acids that create propionic acid to rise (see Figure 2). These are carried to the brain where they may have a harmful effect. Give your child extra low-protein foods, formula, and fluids during illness. This extra food energy 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 cause 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.

If you suspect your child has a cold or virus, or if he has a fever, it is important to call your pediatrician or metabolic doctor immediately. Illness in a child with PA can be very serious. Your metabolic doctor or nutritionist will help you decide how to manage it. Ask your nutritionist for a "sick day plan" to use during illness. A sick-day plan provides adequate calories and amino acids to meet protein needs and may involve more medical food and less ILE, MET, THR, and VAL. A sick-day plan helps keep your child out of the hospital. Ask about beginning medication such as acetaminophen (Tylenol®) to reduce fever and fluids to prevent dehydration.

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

- Do not force-feed food or Propimex, 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.
- Encourage your child to continue drinking his medical food if he tolerates it.
- 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 Propimex mixture, or use liquid Jell-O® if it is 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.

Emergency Letter. Individuals with PA should have an emergency letter with them at all times. This letter provides important information such as the name of the condition, explanation of symptoms, the importance of timely treatment and treatment strategies, and your metabolic doctor's contact information. In times of illness or stress that may require hospitalization, this letter can be presented. This letter can be provided by your metabolic team.

A LOOK TO THE FUTURE

Continuing the Diet. PA 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 PA diet may cause mental and nervous system damage at any age. Lifelong nutrition support must be adapted to each person's needs. Metabolic doctors and nutritionists provide support that is essential for helping your child have a normal, productive life.

Current research is looking at alternative treatments for PA, such as enzyme replacement, gene therapy, and liver transplantation. However, until these therapies can be proven effective and safe, dietary treatment for life is essential.

Family Planning. The chance that two carriers of PA will have

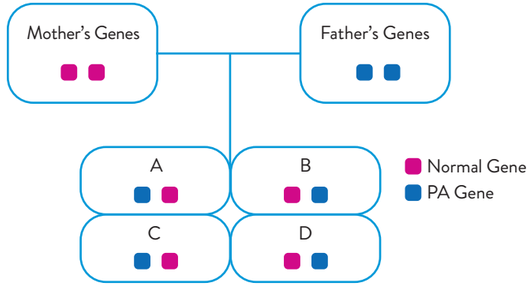


Figure 6. All children of a person with PA and a person who doesn't have PA will be carriers of PA.

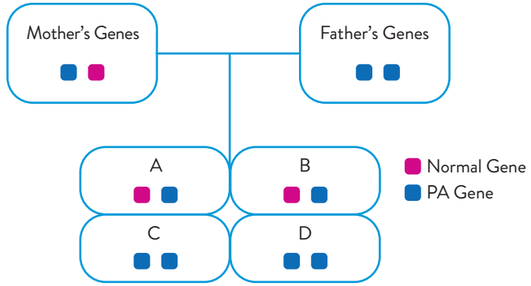


Figure 7. Approximately half of the children of a parent with PA and a parent who is a carrier of PA will have PA.

a child with PA is 1 in 4, or 25%, **for each pregnancy** (Figure 4). Because you have a child with PA, you know that both parents are carriers. That means you have a 25% risk with each pregnancy of having another child with PA. The chance that two carriers will have a child who is a carrier is 1 in 2, or 50% for each pregnancy.

Before a couple who has a child with PA plan to have any more children, they should take time to seriously think about the special parenting tasks that parents of a child with PA must manage. Genetic counseling is recommended to review the risks and to discuss several reproductive options that are available before and during pregnancy.

While there is no test that can determine if another child will be affected with PA before a pregnancy, prenatal testing may be possible during the early part of the pregnancy. A couple may want to discuss and consider all their options with their metabolic doctor and genetic counselor before having another pregnancy.

If you decide to have another child, give yourselves time to adjust to providing for the special needs of the first child. Parents will want to be skilled in diet management for PA before having another child.

Offspring of a Person with PA. All children born to a person with PA will be carriers of the gene. As shown in Figure 6, all offspring of a parent with PA (●● ●●) and a parent who is normal (●● ●●)) inherit one normal (●●)) and one PA (●●) gene. Each one of their offspring will be a carrier.

If a person with PA has children with a carrier for PA, approximately one-half (50%) of their offspring will have PA and one-half (50%) will be carriers (Figure 7).

Childbearing by Women with PA. For the woman with PA, having children may cause problems. A major concern for women with PA is the stress of pregnancy on her metabolic control. Nutritional requirements can rapidly change during the course of a pregnancy. Pregnant women must be monitored carefully for their own safety, as well as the health of their baby. Successful pregnancies require careful treatment and monitoring during the prenatal, labor, delivery, and postpartum periods.

Kool-Aid®-Flavored Propimex®-1

Yield: 8 fl oz

40 g Propimex-1
2 Tbsp, **level**, sugar[1]
1/4 tsp Kool-Aid or Wyler's® **Unsweetened** Soft Drink Mix

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*
Isoleucine, mg	6	48
Methionine, mg	0	0
Valine, mg	0	0
Threonine, mg	5	40
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 Propimex®-2

Yield: 8 fl oz

30 g Propimex-1
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*
Isoleucine, mg	48	57	76
Methionine, mg	0	1	13
Threonine, mg	40	60	69
Valine, mg	6	20	20
Protein, g	6.5	6.7	8.5
Energy, kcal	257	276	251

[1]Please check with your dietitian or doctor before using this recipe in infants.

Kool-Aid®-Flavored Propimex™-2

Yield: 16 fl oz

40 g Propimex-2
3 Tbsp, level, sugar[1]
1/2 tsp Kool-Aid or Wyler's **Unsweetened** Soft Drink Mix[2]

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*
Isoleucine, mg	96
Methionine, mg	0
Valine, mg	0
Threonine, mg	80
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 Propimex Medical Food

- Add chocolate or strawberry syrup.
- Mix Propimex with fruit to make a "smoothie."
- Freeze flavored medical food into "slushies" or "popsicles."
- Add dry Propimex 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

Propionic Acidemia Foundation
Jill Chertow Franks, Director
1963 McCraren Rd.
Highland Park, IL 60035
Email: paf@pafoundation.com
U.S. Toll Free 1-877-720-2192

Organic Acidemia Association
Kathy Stagni, Executive Director
9040 Duluth St.
Golden Valley, MN 55427
Email: mkstagni@gmail.com
Phone: (763) 559-1797 (Central Time)
Fax: (866) 539-4060 (Toll Free)

Menta Pitre, Director
201 E. 14th Place
Larose, LA 70373
E-mail: menta@ooanews.org
Phone: (985) 856-5631 (Central Time)

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: _____

_____

Your Path to Insurance Reimbursement Support

pathway plus

Abbott offers a reimbursement support program to help determine your coverage options for Abbott Metabolic products and will connect you with a supplier who can help get it delivered right to you door.

How it works

STEP 1
You Call with patient's information, insurance and diagnosis

STEP 2
We Help determine available coverage options

STEP 3
We Connect your covered patients to a supplier

Call To Get Started: 1-855-217-0698 (M-F, 8:30 a.m. to 5:00 p.m. EST)

To learn more, visit **www.pathway-plus.com**

Abbott Metabolic Medical Foods

21 products to meet a wide range of nutrition needs in more than 40 inborn errors of metabolism.

Phenylketonuria (PKU)

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

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

20202995/July 2020
LITHO IN USA

Abbott