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A GUIDE FOR FAMILIES WHO HAVE A CHILD WITH

Homocystinuria

CONTENTS

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

INTRODUCTION TO HOMOCYSTINURIA

Your child has been diagnosed with a condition called *homocystinuria* (ho-mo-sis-tin-u-re-ah), or HCU for short. Children who have inherited this condition can't use the amino acid methionine (me-thi-o-neen) (MET) in a normal way. MET is 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 amounts of MET 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.

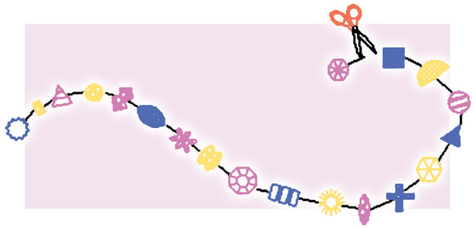


Figure 1. Amino acids are joined together like beads on a necklace to form protein. Enzymes act like scissors to remove amino acids from protein.

WHAT IS HCU?

HCU is an inherited disorder of amino acid metabolism. Proteins, which are made up of amino acids, are found in many parts of the 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. MET is one of these amino acids. All foods with protein contain MET. 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 so have less MET.

Splitting protein into amino acids requires a special substance. 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 MET is split off from the food protein and absorbed, it is used to help form many other useful substances in the body. One of these substances is called cysteine (sis-tay-een) (CYS), another amino acid. The body uses MET and CYS to make protein.

To form CYS from MET, another enzyme called cystathionine- beta-synthase (sis-ta-thi-o-neen bay-ta sin-thase) (CßS) is needed. People with HCU, such as your child, do not have any normally working enzyme (CßS) or don't have enough to handle all the MET that is in the protein foods they eat. In either case, MET can't be changed to CYS. It builds up in the body as homocystine and causes the symptoms of HCU.

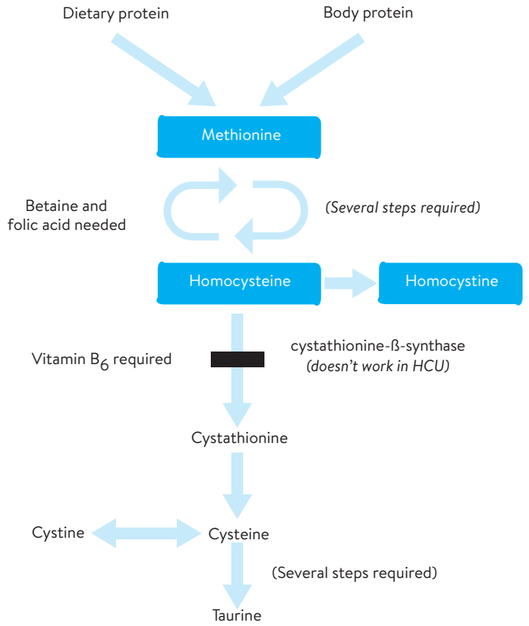
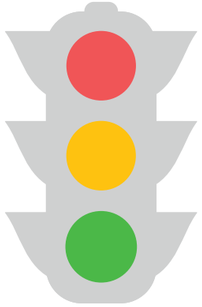


Figure 2. Metabolic pathway for MET. Shaded substances build up in the body when CβS doesn't work..

Think of the situation as a traffic light (Figure 3). A green light (normal CßS) changes MET to CYS. A red light (no or too little enzyme) keeps MET from being changed to CYS. If the light is stuck on red, a traffic jam occurs and the substances in the shaded boxes in Figure 2 build up in the blood and spill into the urine and perspiration. People who have HCU have too much MET, homocysteine, and homocystine, and not enough CYS and taurine (tor-een) (another amino acid) in their blood.



CßS not working.
MET cannot convert to CYS. MET, homocysteine, and homocystine build up in the blood.

CßS not working well.
MET may not be broken down efficiently.

CßS working.
MET changes to CYS.

Figure 3. The cystathionine-beta-synthase (CβS) traffic light.

HCU: AN INHERITED DISORDER

HCU 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, to direct the body to make enzymes such as CßS.

Each parent of a child with HCU has one normal (●) and one altered (HCU) (●) 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 CßS to use MET normally. She will pass a normal gene on to each of her offspring.

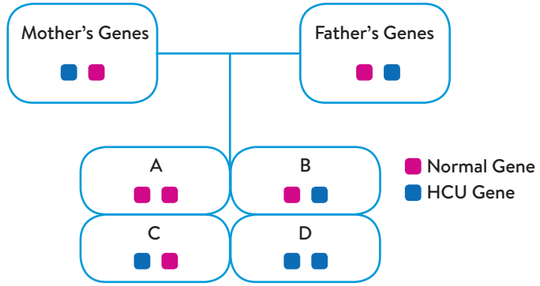


Figure 4. Genetic inheritance of HCU.

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

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

DIAGNOSIS OF HCU

Most states require all babies to be screened for HCU and other conditions before they are discharged from the hospital. In cases where HCU is not tested in newborn screening, a child may be diagnosed after showing symptoms of the condition.

If the initial screening tests show that a baby may have HCU, additional blood and urine are collected for more precise measurements that will confirm the diagnosis. Some doctors may hospitalize the infant to confirm diagnosis so that treatment can be started sooner if the baby has HCU.

TYPES OF HCU

Classical HCU has been associated with a deficiency in CßS. Most HCU cases (~80%) are this form. Classical HCU can be divided into two forms:

Pyridoxine (Vitamin B_6) responsive HCU. Vitamin B_6 is a cofactor of CßS. This means that it helps CßS work better. When people with HCU are responsive to B_6 supplementation, it means that B6 does help CßS to work better and they have reduced homocysteine levels in their body. About 50% of people with HCU are Vitamin B_6 responsive.

Pyridoxine (Vitamin B_6) non-responsive HCU. If people with HCU are B_6 non-responders, it means that supplementation with Vitamin B_6 does not help CßS work better; therefore, their homocysteine concentration does not decrease.

We now know that there are other enzyme deficiencies that can cause HCU. These other enzyme deficiencies cause problems in converting homocysteine back to methionine. This leads to a build-up of homocysteine in the body. This booklet does not go into detail of these rarer forms; consult your metabolic doctor for more information.

Table 1. General Guide to Foods on MET-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	Gumdrop candy, hard candy, jelly, Kool-Aid®, lemonade, lollipops, Popsicles®, pure sugar and fat, soda

SYMPTOMS OF HCU

If diagnosis of HCU is not made at birth, a diagnosis may be made after a child starts showing symptoms of HCU. HCU has a wide range of symptoms that may start showing at around a few months of age. HCU affects the eyes, bones, central nervous system (the brain and nerves), and vascular system (the system that carries blood in the body).

Eyes. The most common effect on the eyes is that the lens disconnects from its usual position and moves downward. When the lens relocates, the person becomes nearsighted.

Other, less commonly seen problems include glaucoma (increased pressure in the eye), retinal detachment, cataracts, and optic nerve atrophy (shrinkage).

Bones. The most consistent problem with bones is osteoporosis, which causes bones to become porous and brittle. The most common site of osteoporosis is in the spine, which develops a condition called scoliosis (crooked spine).

Thinning and lengthening of the long bones of the arms and legs may also occur, causing the child to be tall and thin. Other bone problems include an abnormally high foot arch, extra prominence or depression of the breastbone, and knock-knees.

Brain. Mental retardation causing developmental delay and learning difficulties is the most commonly seen effect on the central nervous system in untreated people with HCU. Seizures, mental illness, depression, and behavior or personality disorders also occur.

Blood. Individuals with HCU have a tendency to develop blood clots (thrombosis). Blood clots can cause other things to happen like heart attack, stroke, and deep vein thrombosis.

TREATMENT OF HCU

The treatment your child receives depends on how old she was when diagnosed, how much CßS activity is present, and how she responds to therapy. Beginning treatment as soon as possible is critical. If a child is diagnosed through newborn screening, the goal of treatment is to maintain normal blood MET levels, which promotes normal growth and development and prevents symptoms from occurring. If diagnosis is made later in life, the goal of treatment is to maintain normal blood MET levels and prevent further development of symptoms.

During early treatment, all patients are given large doses of vitamin B_6 to determine if they are responders. For B_6 responders who maintain normal levels with homocysteine, B_6 may be the main form of treatment. If blood MET levels don't decrease to normal range, the MET-restricted diet is started.

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

- **Pyridoxine (Vitamin B_6)** is a cofactor that helps optimize CßS activity to help reduce homocysteine levels in the body.
- **Folic Acid** is also a cofactor that helps optimize enzyme activity to help reduce homocysteine levels in the body as well as to help prevent deficiency.

- **Cobalamin (Vitamin B_{12})** is also a cofactor that helps optimize enzyme activity to help reduce homocysteine levels in the body.
- **Betaine** is a medication that may be given to reduce the homocystine levels in the blood.
- **Cystine** becomes an essential amino acid in HCU since MET cannot be converted to CYS.

NUTRITION SUPPORT OF HCU

A diet that reduces MET intake and supplemented with CYS and folic acid is used to help prevent the problems associated with untreated HCU. This diet, which is different for each person with HCU, lowers the level of homocysteine and the other toxic substances described earlier to a range that may allow normal mental development and growth.

To provide enough energy and adequate MET, CYS, protein, minerals, and vitamins, a diet restricted in MET is designed just for your child to maintain proper amino acid blood levels and growth.

Many clinics do not have their patients count MET anymore. Could you add a sentence here "Your nutritionist will teach you how to count the MET or grams of protein in the food your child eats.

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

To be sure your child is getting enough energy, protein, and adequate MET for growth and development, the nutritionist carefully calculates the amount of each nutrient needed. Too little MET, 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, MET intakes, blood levels of MET, and homocysteine levels.

Hominex®-1 Amino Acid-Modified Medical Food With Iron is a medical food used to provide protein for infants and toddlers with HCU. Hominex-1 does not contain any MET. Similac Advance Infant Formula with Iron, breast milk, or other intact protein **must** also be fed to provide the specific amount of MET your baby needs for growth and development. Breast milk is lower in MET than infant formula or cow's milk and can be used to supply the required MET. 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 child's medical food. Hominex-1 contains a soluble form of CYS and is well supplied with fat, carbohydrate, minerals, and vitamins. Supplemental minerals and vitamins are not usually needed when the diet is followed as directed.

Hominex®-2 Amino Acid-Modified Medical Food is a medical food used in treating children and adults with HCU. Hominex-2 contains no MET, so MET 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 how much of each your child can eat. Hominex-2 also contains a soluble form of CYS and is well supplied with fat, carbohydrate, minerals, and vitamins. **Hominex-1 and Hominex-2 must be used under the supervision of a physician.**

Hominex-1 and Hominex-2 taste different from milk. Most children and adults get accustomed to the flavors of the foods that they eat. 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.

One mother disliked the odor of the medical food so much that she made a face each time 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. As she said later, "We changed our attitude to thinking this wonderful diet will make it possible for our child to have a happy life."

Other children in the family should be told that Hominex is very important, and they should not emphasize the difference in taste or odor between milk and medical food to the child with HCU.

Most children taking medical foods for HCU like them **IF**

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

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 Hominex 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 Hominex. Hominex 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. Check 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 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 MET is to weigh foods on a scale that reads in grams.

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)

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 follow the mixing instructions provided by your nutritionist or metabolic physician.**
- Wash your hands and all supplies carefully before preparing formula.
- **Do not** mix longer than indicated on the Hominex label.
- 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 HCU is the same as for any baby. The Hominex-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 Hominex-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 Hominex-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 Hominex-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 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. Waiting to feed solid foods until the baby is developmentally ready is best.

Hominex-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 would be fed 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 suggest different ages. Follow their advice.

At about 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.

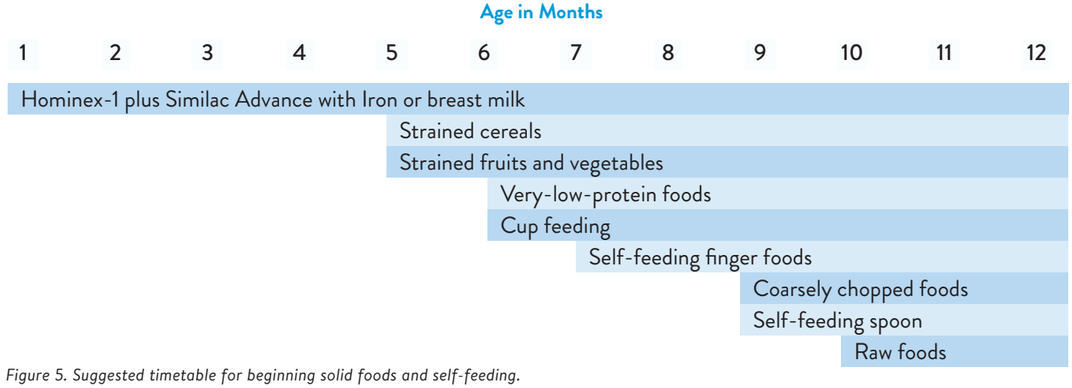


Figure 5. Suggested timetable for beginning solid foods and self-feeding.

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 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 MET with another food, and write it down.

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

Diet Guide and Food Lists. At each clinic visit, you will be given guidance 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.

When your child is older, you may need to use Free Foods (see Table 1) to meet her energy needs. Free foods, which are high in energy and contain little or no MET, must not replace prescribed foods nor be used in large amounts. If your child eats too many of these free foods, she may become overweight, or the extra sugar may cause tooth decay. 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.

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 MET.

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

CHECKING YOUR CHILD'S PROGRESS

Blood Tests. Because your baby grows rapidly during the first year of life, blood is checked frequently for MET and CYS content as well as homocysteine levels. Most doctors will ask for a blood sample once a week during the first 6 months, then every 2 weeks until 1 year of age. After 1 year of age, checking blood levels and diet records may be decreased if blood MET, CYS, and homocysteine levels are well controlled. How often your child is tested will be determined by his metabolic doctor.

For 3 days before taking the blood sample, you will be asked to accurately record your child's total food and beverage intake. In this "food diary" (which the clinic may 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 MET content based on information given to you by the nutritionist.

MET and Homocysteine Levels. The concentration of MET in the blood is an indirect measure of how much MET is present in body tissues. Of most concern are the brain and the vascular system, because when too much MET builds up, homocysteine made from MET may cause blood clots to form. Because blood transports nutrients to all body organs, the concentration of MET in the blood will give the doctor and nutritionist an idea of how much homocysteine might be in them. Blood levels of MET that are too low can also result in poor growth.

Blood MET levels that are **high** may indicate that your child is eating more foods that are high in MET than his body needs for growth. Illness, such as colds and flu, can also cause the body to break down its own protein, releasing MET into the

blood. When your child is not getting enough protein, because of rapid growth or inadequate intake of the medical food, blood MET levels may also rise. Initially during rapid growth, blood MET levels will decrease and then increase again as the body breaks down its own protein.

A **low** MET level usually indicates that your child is not getting enough MET in the diet.

Clinic Visits. Because HCU 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 MET 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 staff 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 HCU whose intake of MET is well controlled and whose diet supplies adequate nutrients should grow as well as a child without HCU.

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 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 Hominex-1 from a cup when your child is between 5 and 8 months of age. Some parents 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 Hominex in liquid form. You may have to use more of it in instant puddings, cereals, fruits, and soups. Some of the Hominex can be mixed into a paste with fruits and fed by spoon.

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

A child that is 15 to 18 months may drink more medical food from a cup if she is given a small pitcher of Hominex and is encouraged to pour it into a small cup without help. Many parents have found 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 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 HCU 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 HCU diet to your child can begin by calling allowed foods "special" or "just for you." From the time your child is very young, teach him 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 HCU. 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 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 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, 10, or 20 mg of MET, or 1 gram of protein, may be used by your child to "purchase" foods containing these amounts.

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 MET in foods. As your child gets older, she should understand what levels of MET are considered normal and the consequences of high MET levels.

Adolescence. Adolescence may be a difficult time for both you and your child, regardless of HCU! The influence of friends and the struggle for independence may make dietary compliance a challenge. Teens may feel that HCU 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 HCU would rather tell people they are vegetarian or vegan than explain HCU and the diet. Help your teenager deal with her peers and not be self-conscious that she has HCU. Finding a peer with HCU 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 HCU is a normal child who needs to manage food intake carefully. Ask your child's health care team for support and help with problems listed here if they occur.

Loss of Appetite. Loss of appetite can result from a variety of causes including poor metabolic control; illness; eating too many sweet foods or desserts that satisfy the appetite and decrease the desire to eat the foods prescribed; getting too much Hominex, which may depress the appetite for other foods; or having lower than normal blood MET levels. Medications may also decrease appetite.

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

Refusing Medical Food. A child may refuse Hominex because of normal variations in appetite, and this should not be of concern if average intake over a week is adequate. If Hominex is not offered regularly, a child may decide to refuse it. Improperly mixed Hominex also can cause refusal— too much water makes the volume too great; too little water makes the Hominex mixture too thick. A child may refuse Hominex as an attention-getting device, especially if he senses that his parents are anxious for him to drink the Hominex mixture. Hominex plays an important role in providing most of your child's nutrition needs. If refusal of the Hominex 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. Your child may be asserting independence by holding out for sweet foods or the prescribed amount of Hominex 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 by going on a "food strike," or they may go on a "food jag." Food jags and strikes are common among young children.

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 Hominex supplies most of your child's nutrient needs, 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 MET-restricted diet. The nutritionist can help you during this trying time, so do not hesitate to call. It is also very important for both parents to support each other in managing a food strike. If a child is allowed to eat foods not on her diet, MET 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 is no reason for concern. Remember that most of your child's nutrient needs are supplied by Hominex.

Inappropriate Feeding Behavior. Inappropriate feeding 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, delay in teaching the child to drink Hominex 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 self-feed, 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 HCU

Parents carry the bulk of the responsibility for managing their child's HCU. If possible, try to share the responsibility in preparing meals and monitoring your child's diet. Other children in the family, as well as the child with HCU, 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 HCU 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 not feel sorry for the child with HCU because she is on a special diet. Treat your child with HCU as normally as possible.

Grandparents love to spoil their grandchildren! It may be difficult for them as they sometimes feel the child with HCU is "missing out." It is important 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 HCU to relatives, friends, day-care providers, baby-sitters, 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 breakdown needs to be prevented in children with HCU because it will release too much MET into the blood. MET is changed to homocysteine, which is carried to the brain and other body organs where it may have a harmful effect. Give your child extra low-protein food, formula, and fluids during illness. The 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 has a loss of appetite. The illness might also include stomach upset, nausea, or vomiting. A child may become very dehydrated because of the high body temperature and a lack of adequate fluid intake.

If you suspect a cold or virus, or if your child has a fever, it is important to call your pediatrician or metabolic doctor immediately. Illness in a child with HCU can be very serious. Your metabolic doctor or nutritionist will help you decide

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 Hominex, 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 Hominex 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 HCU 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

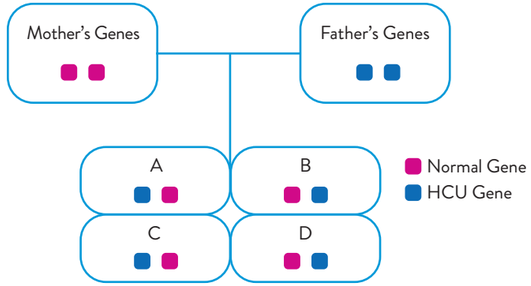


Figure 6. All children of a person with HCU and a person who doesn't have HCU will be carriers of HCU.

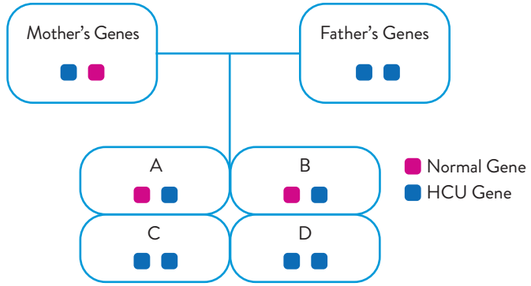


Figure 7. Approximately half of the children of a parent with HCU and a parent who is a carrier of HCU will have HCU.

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 HCU Diet. HCU 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 HCU diet may lead to osteoporosis, blood clots, eye damage, and nervous system damage at any age. Your metabolic doctor will tell you that your child must be on diet for life.

Following a MET-restricted diet closely and keeping MET levels within treatment range enable the person with HCU to lead a normal, productive life.

Family Planning. The chance that two carriers of HCU will have a child with HCU is 1 in 4, or 25% **for each pregnancy** (Figure 4). 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 with a child who has HCU plan to have any more children, they should take time to seriously think about the special parenting tasks that parents of a child with HCU 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 have HCU *before* pregnancy, prenatal testing for HCU may be possible during the early part of pregnancy. These options should be discussed with your doctor before you try to become pregnant.

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 HCU before having another child.

Offspring of a Person with HCU. As shown in Figure 6, all offspring of a parent with HCU (●●) and a parent who has two normal genes (●●) will inherit one normal (●) and one HCU (●) gene. Each one of their children will be a carrier.

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

Childbearing by Women with HCU. For the woman with HCU, having children may cause problems. A major concern for women with HCU is the stress of the pregnancy on her metabolic control. For women with HCU, there is an increased risk of blood clots during and after pregnancy. This tendency is greater than that in pregnant women without HCU. Maintaining blood homocysteine levels in the normal range by strict diet control of MET may help decrease this tendency.

The 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. HCU is a serious health concern. Successful pregnancies require attentive diet monitoring during the prenatal, labor, delivery, and postpartum periods.

Kool-Aid®-Flavored Hominex®-1

Yield: 8 fl oz

40 g Hominex-1
3 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*
Methionine, mg	0	0
Cystine, mg	22.5	180
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 Hominex®-2

Yield: 8 fl oz

30 g Hominex-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*
Methionine, mg	6	3	12
Cystine, mg	6	NA	18
Protein, g	6.5	6.7	8.6
Energy, kcal	257	275	252

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

Kool-Aid®-Flavored Hominex™-2

Yield: 16 fl oz

40 g Hominex-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*
Methionine, mg	0
Cystine, mg	360
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 Hominex Medical Food

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

HCU Network America
Danae' Bartke,
CFO15 South Mallory Avenue
Batavia, Illinois, 60510
website:www.hcunetworkamerica.org
Email:info@hcunetworkamerica.org
Phone:630-360-2087

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:	_____	Fire:	_____
Metabolic Doctor:	_____	Hospital:	_____
Pediatrician:	_____	Other:	_____
Police:	_____		_____

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STEP 2 — **We Help** determine available coverage options

STEP 3 — **We Connect** your covered patients to a supplier

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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.

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