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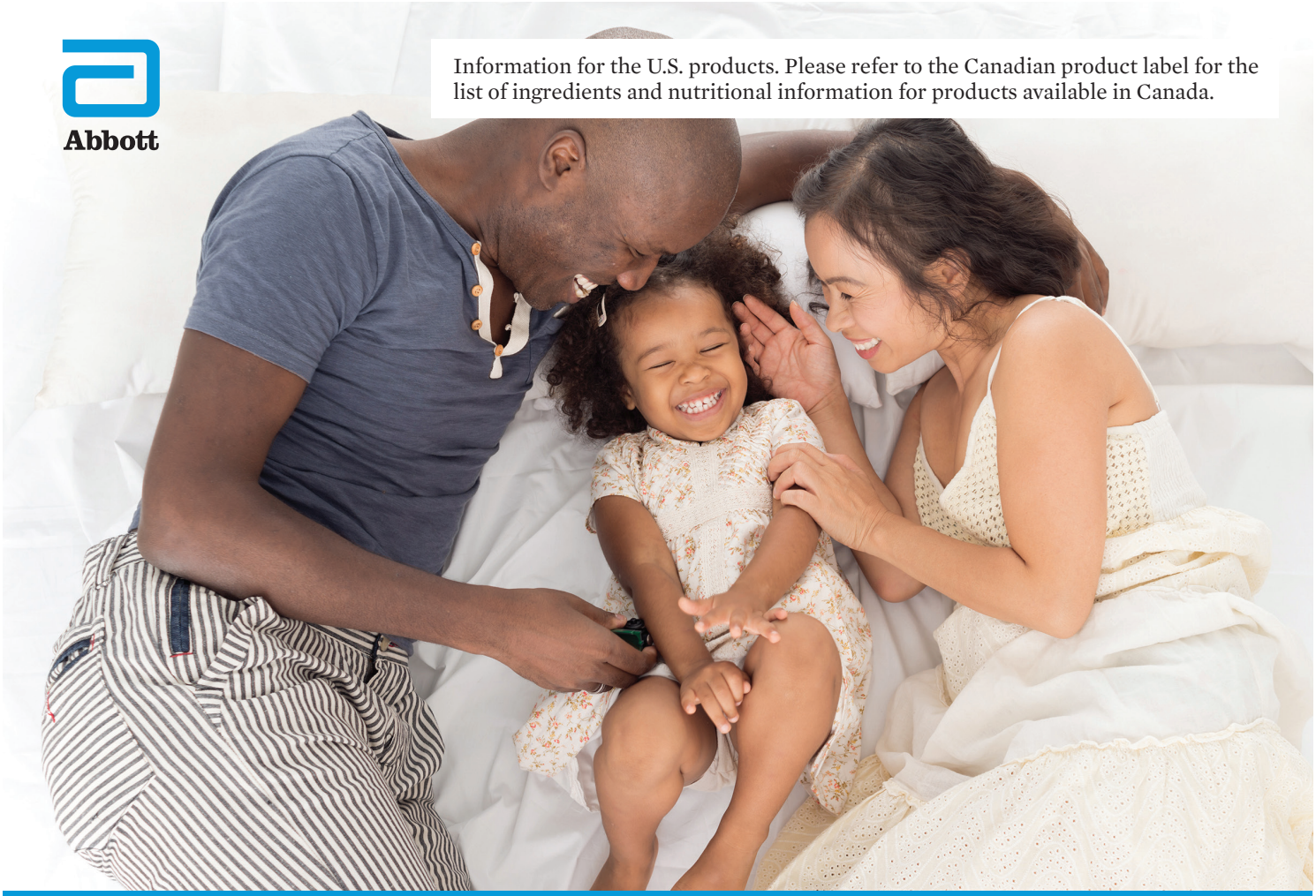
Information for the U.S. products. Please refer to the Canadian product label for the list of ingredients and nutritional information for products available in Canada.



A GUIDE FOR FAMILIES WHO HAVE A CHILD WITH

Glutaric Aciduria Type I

CONTENTS

Abbott provides this booklet to health care professionals to help them counsel families, and to families to help them learn about Glutaric Aciduria Type I.

INTRODUCTION TO GLUTARIC ACIDURIA TYPE I

Your child has been diagnosed with a condition called *Glutaric Aciduria* (glu-tar-ic a-sid-ur-ee-ah) Type 1 (GA-1). Children who have inherited this disorder cannot use the essential amino acids lysine (lie-seen) (LYS) and tryptophan (trip-toe-fan) (TRP) in a normal way. LYS and TRP 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 amounts of LYS and TRP 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.

WHAT IS GA-1?

GA-1 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, bones, and muscles. Most foods also contain protein.

Twenty amino acids occur commonly in the human body and in the foods we eat. Nine of these are called "essential amino acids" because they are essential for growth, and the only way to obtain them is through the diet. LYS and TRP are two of these amino acids.

All foods with protein have LYS and TRP. Common high-protein foods include dairy products, beans and peas, eggs, meat, poultry, nuts, seeds, nut butters, soy products, and seafood. Fruits, grains, and vegetables contain less protein, therefore have less LYS and TRP.

When we eat foods containing protein, this protein is split into amino acids during digestion. After absorption, the amino acids are 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.

Splitting protein into amino acids requires a special substance called an enzyme (en-sime). Think of the enzyme as a pair of scissors snipping beads off a necklace (Figure 1).

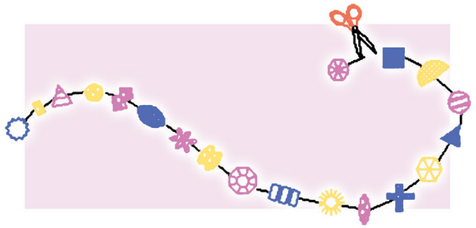


Figure 1. Amino acids are joined together like beads on a necklace to form protein. Enzymes act like scissors to remove amino acids from protein.

Once LYS and TRP are split off from food protein, they are absorbed, changed, and used to help form many other useful substances in the body. For the body to rid itself of LYS, TRP, and their breakdown products, or to use excess LYS and TRP for energy, an enzyme called glutaryl-CoA dehydrogenase (glu-tar-il Co-A de-hidro-gin-ace) (GDH) is required. People with GA-1, such as your child, do not have any normally working GDH or not enough to manage all the LYS and TRP that is in the protein foods they eat. In either case, LYS and TRP cannot be used normally, and they or the products from their breakdown build up in the body as glutaric acid and 3-hydroxy (3-OH) glutaric acid.

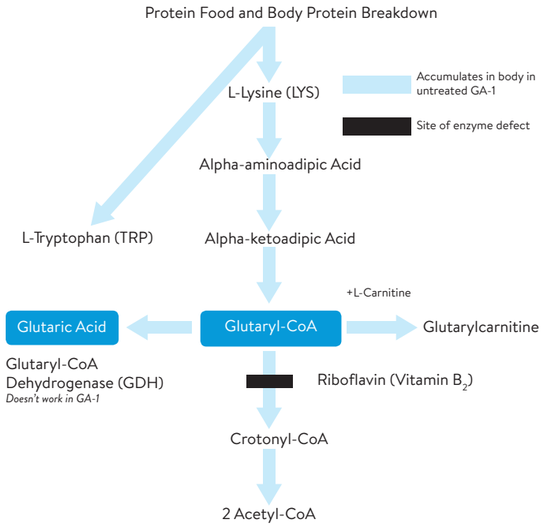
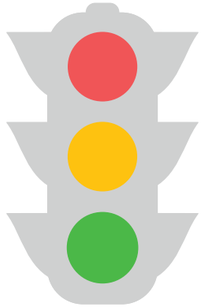


Figure 2. Metabolic pathway for LYS and TRP. Substances in dark blue boxes build up when GDH does not work.

Think of the situation as a traffic light (see Figure 3). A green light (normal GDH) allows LYS and TRP to be used normally. A red light (no or too little GDH) keeps LYS and TRP from being used normally. If the light is stuck on red, a traffic jam occurs—glutaric acid and 3-OH glutaric acid build up in the blood and tissues, spill into the urine, and may cause the symptoms of GA-1.

If a person is not treated, glutaric acid and 3-OH glutaric acid build up in the blood and urine and affect the nervous system and the body's ability to maintain other normal functions.



GDH not working.
LYS and TRP cannot be broken down. Glutaric acid and 3-OH glutaric acid build up in the blood and urine.

GDH not working well.
LYS and TRP may not be broken down efficiently.

GDH working.
LYS and TRP used normally. Breakdown products do not build up.

Figure 3. The glutaryl-CoA dehydrogenase (xwfGDH) traffic light.

GA-1: AN INHERITED DISORDER

GA-1 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, for making body tissues and enzymes such as GDH.

Each parent of a child with GA-1 has one normal (●) and one altered GA-1 (●) gene. Each of their offspring will have one gene from each parent and could have one of four gene sets (Figure 4). A child who receives gene set A inherits two normal genes (●●). Her body will make enough GDH to use LYS and TRP normally. She will pass a normal gene on to each of her offspring.

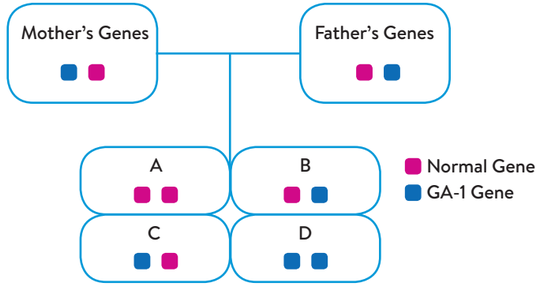


Figure 4. Genetic inheritance of GA-1.

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

A child with gene set D has GA-1 caused by the two GA-1 genes (●●), one from the mother and one from the father. Her body will not be able to use the LYS and TRP in food normally. She will also pass a GA-1 gene on to each of her offspring. Your child with GA-1 has this gene set.

DIAGNOSIS OF GA-1

Most states require all babies to be screened for GA-1 and other conditions before they are discharged from the hospital. In cases where GA-1 is not tested in newborn screening, an infant may be diagnosed after showing symptoms of the condition.

If the initial screening tests show that a baby may have GA-1, 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 GA-1.

SYMPTOMS OF GA-1

With early diagnosis and lifelong nutrition support, a person with GA-1 can have normal growth and development if blood organic acid levels are controlled. Symptoms of GA-1 usually don't appear in the first few weeks of life. Typically, an illness or infection in the first few years of life triggers symptoms that lead doctors to test for GA-1. Signs and symptoms of GA-1 include:

- Degenerative or permanent brain injury (encephalopathy)
- Weakness or lack of muscle control (hypotonia)
- Excessively large head (macrocephaly)
- Build-up of ketones (substances resulting from the breakdown of fat)
- Vomiting
- Enlarged liver (hepatomegaly)
- Convulsions and coma
- Failure to coordinate muscle movement (ataxia)
- Involuntary movements (spasticity)
- Kidney cysts (sacs that hold abnormal amounts of fluid)
- Low blood sugar (hypoglycemia)
- Metabolic acidosis (excess acid in the blood)
- Abnormal brain function

Table 1. General Guide to Foods on LYS- and TRP-Restricted Diets

Foods That Are Not Allowed	Foods That Are Limited	Foods That May Be Eaten Freely
Dairy products (cheese, milk, ice cream, yogurt), soy milk and soy products, beans and peas, eggs, fish and other seafood, meat, nuts, nut butters, poultry, seeds, tofu	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

NUTRITION SUPPORT OF GA-1

A diet that reduces LYS and TRP intake is used to help prevent most of the symptoms of GA-1. This diet, which is different for each person with GA-1, can lower the blood level of glutaric acid and 3-OH glutaric acid to a range that may allow normal mental development and growth.

The special diet for your child is designed for his individual needs, and following it is very important. With proper nutritional management, your child may grow and develop normally.

Many foods contain protein. Those foods also contain LYS and TRP. Your child with GA-1 must limit the amount of foods that contain protein. Table 1 is a general guide of foods that aren't allowed, foods that are limited, and foods that may be eaten freely if obesity is not a problem.

Additional Therapies. Your metabolic doctor may recommend certain vitamins, minerals, and medications for your child with GA-1. Common ones include:

- **Carnitine** is a nutrient that helps energy enter cells in the body. Carnitine can also bind with toxic substances and help the body get rid of them.
- **Riboflavin** helps GDH work better.

Requirements for LYS, TRP, Protein, and Energy. A child with GA-1 who eats enough protein to grow properly gets too much LYS and TRP. 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 LYS and TRP does not provide enough protein to meet your child's needs for growth. **To get enough protein for growth and not get too much LYS and TRP, a special medical food that is high in protein and free of LYS and TRP is required.**

To be sure your child is getting enough energy, protein, and adequate LYS and TRP for growth and development, the nutritionist carefully calculates the amount of each nutrient needed. Too little LYS and TRP, 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, LYS and TRP intakes, blood levels of LYS and TRP, and urinary excretion of organic acids.

Glutarex®-1 Amino Acid-Modified Medical Food With Iron is a medical food used to provide protein for infants and toddlers with GA-1. Glutarex-1 does not contain any LYS and TRP. Similac Advance Infant Formula with Iron, breast milk, or other intact protein must also be fed to provide the specific amount of LYS and TRP your baby needs for growth and development. Breast milk is lower in LYS and TRP than infant formula or cow's milk and can be used to supply the required LYS and TRP. 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. Glutarex-1 contains carnitine 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.

Glutarex®-2 Amino Acid-Modified Medical Food is a medical food used in treating children and adults with GA-1. Glutarex-2 contains no LYS and TRP, so LYS and TRP 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. **Glutarex-1and Glutarex-2 must be used under the supervision of a physician.**

Glutarex-1 and Glutarex-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 every 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 Glutarex is very important, and they should not emphasize the difference in taste or odor between milk and medical food to the child with GA-1.

Most children taking medical foods for GA-1 like them **IF** the medical foods are started early and **IF** their family has a positive attitude. Older children who start on the diet after drinking cow's milk may not like Glutarex 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 Glutarex. Glutarex 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.

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 LYS and TRP 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.

Tips for preparing formula for infants:

- **Always follow the instructions on the label and mix formula according to the recipe provided by your nutritionist or metabolic doctor.**
- Wash your hands and all supplies carefully before preparing formula.

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

- **Do not** mix longer than indicated on the Glutarex 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 GA-1 is the same as for any baby. The Glutarex-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 Glutarex-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 Glutarex-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 Glutarex-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.

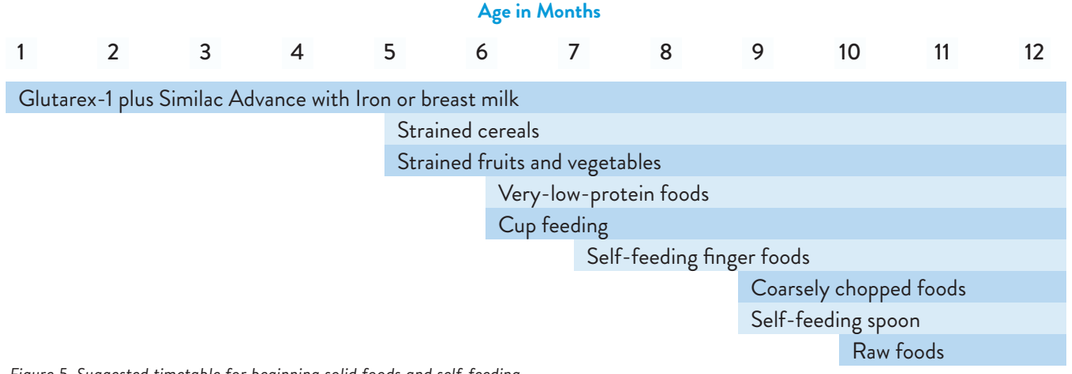


Figure 5. Suggested timetable for beginning solid foods and self-feeding.

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.

Glutarex-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.

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 LYS and TRP with another food, and write it down.

When your child is older, the differences between the LYS and TRP-restricted diet and the diets of other children will be greater. Your child with GA-1 will require Glutarex 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 to 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 LYS and TRP, 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, 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 LYS and protein.

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

CHECKING YOUR CHILD'S PROGRESS

Laboratory Tests. Because your baby grows rapidly during the first year of life, blood and urine samples may be tested frequently to monitor how well his GA-1 is controlled, and to indicate if changes in treatment are needed. Your doctor may ask for a blood and/or urine test frequently during your baby's first year of life. After 1 year of age, blood and/or urine may be tested less frequently if blood LYS, TRP, and glutaric acid levels are well controlled. Various methods are used to monitor the amino acids and glutaric acid levels in the blood and urine. Your baby's metabolic doctor or nutritionist will determine how often your child is tested.

Before taking a blood sample, you may be asked to accurately record your child's total food and beverage intake. In this "food diary," record the name of the food, the exact amount in grams or household measures (cups, teaspoons, or tablespoons) that your child ate, and the LYS and TRP 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.

LYS, TRP, and Glutaric Acid Levels. The amount of LYS and TRP in the blood is an indirect measure of how much LYS and TRP are present in body tissues. Of most concern is the brain, because too much LYS, TRP, and glutaric acids are harmful to brain development. Because blood transports nutrients to the brain, the concentration of LYS, TRP, and glutaric acids in the blood will give the doctor and nutritionist an idea of how much of these substances might be in the brain.

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

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

Clinic Visits. Because GA-1 is a lifelong condition that could harm your child's growth and development, you will be asked to take your child to the clinic frequently. If growth and development are normal and laboratory concentrations remain within 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 your child's development, may also be evaluated. Diet changes will be made, if needed, and any questions you may have will be addressed.

In addition to the metabolic specialist, you should have a local pediatrician or family doctor to provide required ongoing well-child care. This doctor should give immunizations at the usual times, 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 GA-1 whose LYS and TRP intakes are well controlled and whose diet supplies adequate nutrients should grow as well as a child without GA-1.

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

Although the requirement for energy (calories) and protein based on 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 Glutarex-1 from a cup when your child is between 5 and 8 months of age. Because taste buds vary on different parts of the tongue, Glutarex may taste different when taken from a cup instead of a bottle. Also, the smell of Glutarex 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 Glutarex at a cold temperature may also increase your child's willingness to drink from a cup.

During weaning, your child may not want to take all the prescribed Glutarex 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 GLUTAREX® IS FED AS A PASTE.
Consult your child's nutritionist.

A child that is 15 to 18 months may drink more medical food fA child of 15 to 18 months of age may drink more medical food from a cup if she is given a small pitcher of Glutarex 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 GA-1 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 can begin by calling allowed foods "special" or "just for you." From the time your child is very young, teach her to ask about unfamiliar foods before eating them. As your child grows older and is able to understand

the concept of a missing or nonworking enzyme, explain GA-1. 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. One way to do this is to use a "token" system. Tokens symbolizing 1 gram of protein may be used 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 protein in foods. As your child gets older, she should understand what levels of LYS and TRP are considered normal and the consequences of high LYS and TRP levels.

Adolescence. Adolescence may be a difficult time for both you and your child, regardless of GA-1! The influence of friends and the struggle for independence may make dietary compliance a challenge. Teens may feel that GA-1 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 GA-1 would rather tell people they are vegetarian or vegan than explain about GA-1. Help your teenager deal with her peers and not be self-conscious that she has GA-1. Finding a peer with GA-1 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 GA-1 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 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 Glutarex, which may depress the appetite for other foods; or having lower than normal blood LYS and TRP level. Medications may also decrease appetite.

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

Refusing Medical Food. A child may refuse Glutarex because of normal variations in appetite, and this should not be of concern if average intake over a week is adequate. If Glutarex is not offered regularly, a child may decide to refuse it. Improperly mixed Glutarex also can cause refusal—too much water makes the volume too great; too little water makes the Glutarex mixture too thick. A child may refuse Glutarex as an attention-getting device, especially if he senses that his parents are anxious for him to drink the Glutarex mixture. Remember, Glutarex 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. The prescribed amount of Glutarex 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 Glutarex 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 TRP- and LYS-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, blood LYS and TRP 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 Glutarex.

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 Glutarex 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 GA-1

Parents carry the bulk of the responsibility for managing their child's GA-1. 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 GA-1, 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 GA-1 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 GA-1 because she is on a special diet. Treat your child with GA-1 as normally as possible.

Grandparents love to spoil their grandchildren! It may be difficult for them as they sometimes feel the child with GA-1 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 GA-1 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 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 GA-1 because it will release too much LYS, TRP, and organic acids into the blood. LYS, TRP, and organic acids are carried to the brain and other body organs where they 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 GA-1 can be very serious and may require a visit to the emergency room. Your metabolic doctor or nutritionist will help you decide on how to manage it. Ask your nutritionist for a "sick-day plan" to use during illness. A sick-day plan provides adequate calories to meet energy needs and helps keep your child out of the hospital. 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 Glutarex, 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 Glutarex 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 GA-1 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.

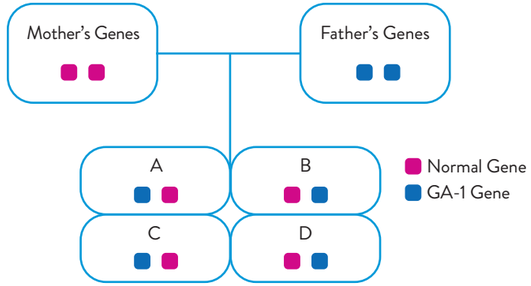


Figure 6. All children of a person with GA-1 and a person who doesn't have GA-1 will be carriers of GA-1.

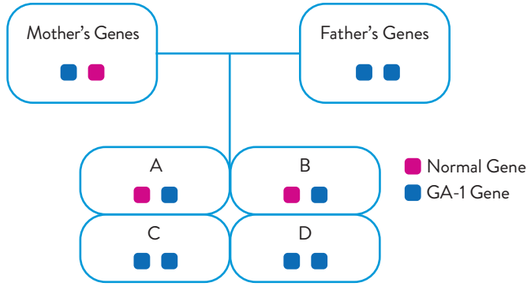


Figure 7. Approximately half of the children of a parent with GA-1 and a parent who is a carrier of GA-1 will have GA-1.

A LOOK TO THE FUTURE

Continuing the GA-1 Diet. GA-1 is a serious health concern. Treating a child as early in life as possible may prevent developmental delay and severe neurological damage. Not following the LYS- and TRP-restricted 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.

Family Planning. The chance that two carriers of GA-1 will have a child with GA-1 is 1 in 4, or 25%, for each pregnancy (Figure 4). Because you have a child with GA-1, you know that both parents are carriers. That means you have a 25% risk with each pregnancy of having another child with GA-1. 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 GA-1 plan to have any more children, they should take time to seriously think about the special parenting tasks that parents of a child with GA-1 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 GA-1 before a pregnancy, prenatal testing for GA-1 is possible during the early part of the pregnancy. A couple should consider and discuss all their options with their metabolic doctor before 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 GA-1 before having another child.

Offspring of a Person with GA-1. All children born to a person with GA-1 will be carriers of the gene or have GA-1. As shown in Figure 6, all offspring of a parent with GA-1 (●●) and a parent who is normal (●●) inherit one normal (●) and one GA-1 (●) gene. Each one of their children will be a carrier. If a person with GA-1 has children with a carrier for GA-1, approximately one-half (50%) of their offspring will have GA-1 and one-half (50%) will be carriers (Figure 7).

Childbearing by Women with GA-1. For the woman with GA-1, having children may cause problems. A major concern for GA-1 women is the stress of the pregnancy on her metabolic control. 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.

GA-1 is a serious health concern. However, by following an LYS and TRP-restricted diet closely and keeping blood levels of LYS, TRP, and glutaric acids within the treatment range, the person with GA-1 can lead a normal, productive life. Successful pregnancies require attentive diet monitoring during the prenatal, labor, delivery, and postpartum periods.

Kool-Aid®-Flavored Glutarex®-1

Yield: 8 fl oz

40 g Glutarex-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*
Lysine, mg	0	0
Tryptophan, mg	0	0
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 Glutarex®-2

Yield: 8 fl oz

30 g Glutarex-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*
Lysine, mg	33	12	33
Tryptophan, mg	6	NA	9
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 Glutarex™-2

Yield: 16 fl oz

40 g Glutarex-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*
Lysine, mg	0
Tryptophan, mg	0
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 Glutarex Medical Food

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

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

Support Groups/Newsletters

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: _____

_____

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Hypercalcemia

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