



## Glutaric Aciduria Type 1

An experienced healthcare professional should be consulted for the management of Glutaric Aciduria Type 1

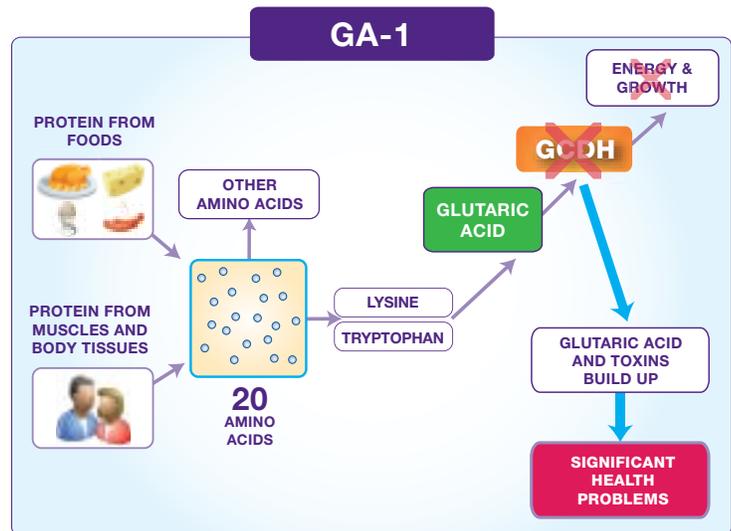
### What is Glutaric Aciduria Type 1?

Glutaric Aciduria Type 1 (GA-1) is an inherited, genetic disorder. This disorder can also be called Glutaric Acidemia Type 1. GA-1 occurs when there is a defect in an enzyme that helps break down protein from food. The name of the enzyme is *glutaryl-CoA dehydrogenase (GCDH)*.

Protein is made up of smaller units called amino acids. Amino acids are important to build muscle and other body tissues for normal growth. Any extra amino acids are normally broken down to produce energy. In GA-1, two of the amino acids cannot be broken down in the body. These amino acids are **lysine** and **tryptophan**. When someone with GA-1 eats too much protein from food, does not get enough calories or becomes ill, blood levels of lysine and tryptophan increase and so do levels of **glutaric acid**, a toxic byproduct. Glutaric aciduria gets its name because of the build-up of glutaric acid in blood and urine of people with this disorder.

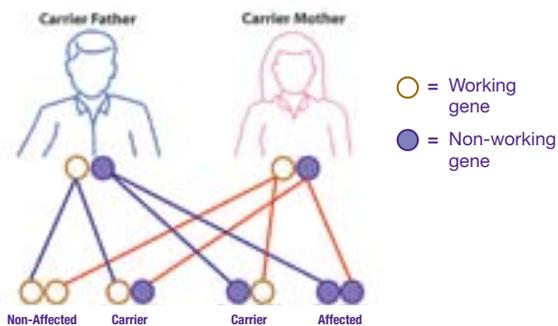
High levels of glutaric acid are especially dangerous to an infant's brain. The area of the brain that is most affected, which is called the basal ganglia, controls movement. Thus, affected infants and children can develop spasms and jerking movements with a rigid body posture, especially after a serious illness. Long-term problems can include poor growth, abnormal motor skills, slow development and learning problems.

Finding out that your child has GA-1 can be overwhelming. Fortunately, nutrition management is available to help prevent many of these problems. Take comfort in knowing that your metabolic healthcare team can provide you with the best advice to help manage GA-1.



### Autosomal Recessive Inheritance

Chances for each child when both parents are carriers



### Incidence, Genetics and Newborn Screening

GA-1 is a rare disorder that occurs in about 1 in every 40,000 births. It occurs more often in the Amish community and Ojibwa population of Canada, affecting about 1 in every 300 births.

GA-1 is inherited as a "recessive disorder." In recessive disorders, both the mother and father are "carriers," and they do not have any symptoms of the disorder (see diagram). With each pregnancy there is a 1 in 4 (25%) chance that the infant will be affected. A genetic counselor can help explain the inheritance of GA-1 and the risks to future infants.

GA-1 can be screened for at birth through a simple blood test. For more details on newborn screening (NBS) in the US, visit <http://genes-r-us.uthscsa.edu>; for information on NBS in Canada, see <http://www.cadth.ca>.

## Management of Glutaric Aciduria Type 1

There is no cure for GA-1, but it can be managed with a modified diet, medication and special medical formulas specifically designed for persons with GA-1. The modified diet for GA-1 is low in lysine and tryptophan. It is important that an individual with GA-1 remains on treatment for life.

A medical formula is a very important part of the diet for GA-1. The protein source in GA-1 medical formulas is individual amino acids, but lysine and tryptophan are left out. This allows a person with GA-1 to get enough protein without the parts of protein that can be harmful. Medical formulas may also provide calories, vitamins and minerals the body needs for normal growth. For some, a small amount of tryptophan may need to be added to the formula to prevent tryptophan deficiency.

To provide just enough lysine and tryptophan that a person with GA-1 needs, a limited amount of natural protein-containing foods are allowed in the diet. For infants, breast milk or regular infant formula is given in precise amounts. As the baby grows and can eat solid foods, the breast milk or regular infant formula will be removed from the diet and lysine and tryptophan will come from foods instead.

Since all foods with protein contain lysine and tryptophan, individuals with GA-1 must limit their intake of foods high in protein. These include milk and dairy products, meat, poultry, fish, eggs, beans, nuts and peanut butter. Regular breads and pastas may be allowed in small amounts, but special low-protein versions of these foods are often used instead to allow for more choices in the diet and less concern for eating too much protein. Vegetables and fruits are allowed. Typically, the amount of lysine in the diet is counted. Foods are usually weighed or measured to ensure that extra lysine is not eaten. A metabolic dietitian will work closely with people with GA-1 to prescribe the best diet plan and help with needed changes in the future.



Nutricia North America provides a range of medical formulas as well as low protein foods. Please contact us for more information. Your dietitian will help you decide which products are best.

In addition to a modified diet, those with GA-1 may also be treated with L-carnitine. This medication can help reduce the amount of glutaric acid in the blood.

### If you or your child is sick

During illness or after an injury, the body increases the breakdown of protein stores. This can raise the level of glutaric acid in the blood and lead to serious medical problems. Illness with a fever can be especially harmful. Early signs that require immediate medical attention include vomiting, excessive sleepiness, coordination problems and/or changes in mental status. During any illness, it is very important to notify your metabolic clinic immediately. Often, the diet is adjusted to either remove or decrease protein and increase calories. This can help slow down the breakdown of protein stores. Medications may also be adjusted during illness. Your clinic will give you an emergency letter - if you notice symptoms of high glutaric acid levels, take this letter with you to the emergency room. During an illness, a hospital stay may be needed.

### Resources

- Star-G website: <http://www.newbornscreening.info>
- Genetics Home Reference website: <http://www.ghr.nlm.nih.gov/condition/glutaric-acidemia-type-1>

*Nutricia North America would like to thank Sandy van Calcar, PhD, RD; University of Wisconsin, Madison for her consultation.*

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