

Glutaric Acidemia Type I (GA I) (Glue-tar-ick a-sid-EE-me-ah Type I)

What is GA I?

Glutaric Acidemia Type I (GA I) is a metabolic disorder. This means the body has a chemical condition caused by a problem in an enzyme called glutaryl CoA dehydrogenase. This enzyme is involved in the break-down of two parts of protein called lysine and tryptophan. Proteins are the building blocks of our body's chemical function. Because these parts of protein cannot be broken down completely, high levels build up in the blood and tissues and are passed in the urine. GA I is inherited. This means it is present at birth.

What type of problems occurs with GA I?

A baby who has this condition may have an enlarged head size, low blood sugar, and uncontrolled muscle movement and muscle contractions. If untreated, this condition can lead to vomiting, poor feeding, seizures, and poor muscle tone. Early diagnosis and treatment will help prevent these problems.

What is the chance my baby will have GA I?

There are less than 100 cases of GA-I known in the United States. This condition occurs more often in certain populations, such as the Amish and people of Swedish descent. Babies born with this condition have a changed gene from each parent. A person who has one changed gene is called a carrier. A person who is a carrier does not have symptoms. If both parents are carriers, either parent can pass on the changed gene to their baby. If both parents pass on the changed gene, the baby will have the condition. If both parents are carriers, for each pregnancy:

- There is a 25% chance that the baby will be born with this condition.
- There is a 50% chance that the baby will be a carrier for this condition.
- There is a 25% chance that the baby will not be born with this condition and will not be a carrier.

What is the treatment of GA I?

The treatment of this condition includes a low protein diet and medicine called L-carnitine. If your child becomes ill you should seek medical care quickly. Your baby's metabolic doctor will help you make sure that your baby gets the right diet and medical care.

Where in Virginia can I take my baby for care?

Please speak to your baby's pediatrician about obtaining a referral to a pediatric metabolic specialist in your area. If you want to know more about this condition, please contact Virginia Newborn Screening Services, Virginia Department of Health. The Web site is <http://www.vahealth.org/gns>.



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