

Glutaric Acidemia Type I (GA-I)

General Overview

Q. What is GA-I?

A. GA-I is a treatable disorder that affects the way the body processes protein. It is treatable, but can be life-threatening.

Q. How does the body normally process proteins?

A. Proteins are made up of amino acids. These amino acids are normally used to make new proteins or broken down further by enzymes to make energy.

Q. What happens to protein in a child with GA-I?

A. In a child with GA-I, one of the enzymes needed to break down the amino acids lysine and tryptophan does not work properly. This results in a build up of glutaric acid, which, if untreated, is toxic to the brain and other body systems.

Q. What is the treatment for GA-I?

A. GA-I is treated with a special diet that is low in lysine and tryptophan. Treatment should begin shortly after birth and is life-long. Since protein is essential for normal growth and development, the child must continue on a medical formula that provides protein and essential nutrients but contains little or no lysine and tryptophan. Some doctors may also prescribe dietary supplements. Other medications may be given by IV if the child becomes ill. People with GA-I require treatment through a specialty clinic with experience in treating this disorder.

Q. What are the effects of having GA-I if it is not treated?

A. Untreated GA-I can result in failure to thrive, developmental delay, long-term loss of motor function, and possibly coma or death.

Q. Why would a child have GA-I?

A. GA-I is an inherited disorder. It results when a baby receives a double-dose of a specific non-working gene involved in breaking down lysine and tryptophan (one from each parent). For more information about this, contact your health care provider or a genetic counselor.

Q. How common is GA-I?

A. About one in every 137,000 babies in the United States is born with GA-I. However, parents who have a child with GA-I each carry one copy of the non-working gene. That means, with each pregnancy, there is a one in four chance of the child having GA-I.

For more information about GA-I, please see the Disorders section of our website: www.doh.wa.gov/nbs.



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