

Isovaleric Acidemia (IVA)

General Overview

Q. What is IVA?

A. IVA is a disorder that affects the way the body processes protein. It is treatable, but can cause life-threatening illness.

Q. What happens to proteins in a child with IVA?

A. Proteins are made up of amino acids. These amino acids are normally used to make new proteins or are broken down further for energy. Children with IVA cannot properly use the amino acid leucine found in the protein they eat. If left untreated, by-products of leucine, including isovaleric acid, build up in the bloodstream, urine, and body tissue, which can be life-threatening.

Q. What is the treatment for IVA?

A. Isovaleric acidemia is treated with a special diet that is low in leucine. Treatment should begin shortly after birth and is life-long. Some doctors may also prescribe dietary supplements. The child must continue on a medical formula that provides protein and essential nutrients but contains little or no leucine. People with IVA require treatment through a specialty clinic with experience in treating this disorder.

Q. Why would a child have IVA?

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

Q. How common is IVA?

A. About one in every 96,000 babies in the United States is born with IVA. However, parents who have a child with IVA 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 IVA.

For more information about IVA, please see the Disorders section of our website:

www.doh.wa.gov/nbs.

