

Isovaleric Acidemia (IVA)

What is IVA?

Enzymes help start chemical reactions in the body. IVA happens when an enzyme called “isovaleryl-CoA dehydrogenase” is missing or not working well. This enzyme helps break down harmful “isovaleric acid.” This acid builds up in the blood and causes problems when a child with IVA eats food with leucine. Leucine is in all foods that have protein (such as meat, beans, peanut butter, milk).

What Causes IVA?

People with IVA have a pair of genes that don't work as they should. These genes cause the “isovaleryl-CoA dehydrogenase” enzyme to not work well or not be made at all.

What Symptoms or Problems Occur with IVA?

[Symptoms are something out of the ordinary that a parent notices.]

Babies with IVA seem healthy at birth. Symptoms often start between one day and two weeks of age. IVA causes periods of illness called Metabolic Crises. Some of the first signs are:

- ◆ poor appetite
- ◆ too much sleepiness, low energy
- ◆ vomiting
- ◆ feeling cold
- ◆ “sweaty feet” odor

If a Metabolic Crisis is not treated, a child with IVA may develop:

- ◆ breathing problems
- ◆ seizures
- ◆ strokes
- ◆ mental retardation
- ◆ coma, sometimes leading to death

The less severe kind of IVA shows up later in childhood. Some problems may include:

- ◆ poor growth
- ◆ learning difficulties

What is the Treatment for IVA?

Early treatment prevents Metabolic Crises and related problems. Treatment should start as soon as you know your child has IVA. Treatment

usually lasts all life long. Treatment often includes:

1. Low-leucine diet, medical foods and formula –

Most children need to eat foods low in leucine (such as vegetables and fruit). Special medical foods and formulas are usually part of the diet. You will get a food plan that has the right amount of protein and nutrients to keep your child healthy. Your child should continue a special food plan for life. High-protein foods your child should limit or not eat include:

- ◆ milk and milk products
- ◆ meat and poultry
- ◆ fish
- ◆ eggs
- ◆ dried beans and peas
- ◆ nuts and peanut butter

2. Medications – The doctor may prescribe the amino acid Glycine to help the body get rid of isovaleric acid. This can help prevent Metabolic Crises in children with IVA. L-carnitine may also help some children. This is safe and natural and helps the body make energy. Only use the kind your doctor tells you to use. Do not use any medication or supplement without checking with your doctor.

Things to Remember

Even minor illnesses such as a cold or the flu can cause a Metabolic Crisis. Call your doctor right away when your child has any of the following:

- ◆ loss of appetite
- ◆ vomiting
- ◆ diarrhea
- ◆ infection or illness
- ◆ fever

Children with IVA need to eat more starchy foods (bread, cereal, rice, noodles) and drink more fluids when they are ill - even if they're not hungry – or they could have a Metabolic Crisis. They also need to not eat protein foods when they are sick. If they can't eat, or if they show signs of a Metabolic Crisis, they may need to be treated in the hospital.