



ISOVALERIC ACIDEMIA (IVA)

FAMILY FACT SHEET

What is a positive newborn screen?

Newborn screening is done on tiny samples of blood taken from your baby's heel 24 to 36 hours after birth. The blood is tested for rare, hidden disorders that may affect your baby's health and development. The newborn screen suggests your baby might have a disorder called IVA.

A positive newborn screen does not mean your baby has IVA, but it does mean your baby needs more testing to know for sure.

You will be notified by your primary care provider or the newborn screening program to arrange for additional testing.

What is IVA?

IVA affects an enzyme needed to break down proteins from the food we eat, so they can be used for energy and growth. In IVA, the enzyme used to break down proteins is missing or not working properly.

A person with IVA doesn't have enough enzymes to break down protein containing leucine, which causes a substance called isovaleric acid to build up in the body. Isovaleric acid is harmful to the body and causes health problems if it builds up.

IVA is a genetic disorder that is passed on (inherited) from parents to a child. The mother and father of an affected child carry a gene change that can cause IVA. Parents usually do not have signs or symptoms, or even know they carry the gene change.

What problems can IVA cause?

IVA is different for each child. Some children with IVA have few health problems, while other children may have very serious complications.

If IVA is not treated, a child might develop:

- Sleepiness
- Feeding problems
- "Sweaty feet" odor
- Seizures
- Stroke
- Brain damage

It is very important to follow the doctor's instructions for testing and treatment.

What is the treatment for IVA?

IVA can be treated. Treatment is life-long and can include:

- Diet low in leucine – a dietitian will help you set up the best diet for your child.
- Medications to help the body get rid of isovaleric acid and make energy.

Children with IVA should see their regular doctor, a doctor who specializes in IVA, and a dietitian.

Prompt and careful treatment helps children with IVA live the healthiest lives possible.

Michigan Resources and Support

Michigan Newborn Screening

Nurse Consultant

Toll-free: 1-866-673-9939

newbornscreening@michigan.gov

Michigan Metabolic Coordinating Centers

Children's Hospital of Michigan: 1-313-832-9343

Michigan Medicine: 1-734-764-0579

Children's Special Health Care Services

Toll-free: 1-800-359-3722