

Isovaleric Acidemia (IVA)

(ice-o-va-lair-ic acid-ee-me-ah)

What is a positive newborn screen?

Newborn screening is done on tiny samples of blood taken from your baby's heel 24 to 48 hours after birth. Newborn screening tests 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 (eye-vee-ay).

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

Your baby's doctor will help arrange for more testing by specialists in disorders like IVA.

What is isovaleric acidemia?

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 enzyme 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 disorder that is passed on, or inherited, from a child's mother and father. Because IVA is a genetic disease, family members are at risk of having IVA too, even if no one in the family has had it before.

What problems can isovaleric acidemia 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
- Mental retardation

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

What is the treatment for isovaleric acidemia?

IVA can be treated. The treatment is life-long. Treatment for children with IVA can include:

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

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

Children with IVA can benefit from prompt and careful treatment.

Resources for parents**Genetics Home Reference**

<http://ghr.nlm.nih.gov>

Organic Acidemia Foundation

www.oaanews.org

MN Children & Youth with Special Health Needs

(651) 201-3650 or (800) 728-5420

MN Newborn Screening Program

www.health.state.mn.us/newbornscreening