



Newborn Screening FACT Sheet

Long Chain 3-Hydroxyacyl-CoA Dehydrogenase Deficiency (LCHAD)

What is LCHAD?

LCHAD is a type of fatty acid oxidation disorder. People with LCHAD have problems breaking down fat into energy for the body.

What Causes LCHAD?

Enzymes help start chemical reactions in the body. LCHAD happens when an enzyme called “long chain 3-hydroxyacyl-CoA dehydrogenase” is either missing or not working. This enzyme breaks down certain fats from the food we eat into energy. It also breaks down fat already stored in the body.

What Symptoms or Problems Occur with LCHAD?

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

LCHAD can cause mild problems in some people and more serious problems in others. Babies and children with LCHAD usually begin to show symptoms sometime from birth through age two. LCHAD causes periods of low blood sugar.

The first symptoms of low blood sugar are:

- extreme sleepiness or tiredness
- weakness
- nausea
- vomiting
- feeling irritable or jittery
- behavior changes (such as crying for no reason)

If low blood sugar is not treated, a child with LCHAD can develop:

- breathing problems
- swelling of the brain
- seizures
- coma, sometimes leading to death

Symptoms often show up after having nothing to eat for more than a few hours. They also show up when a child with LCHAD gets sick or has an infection. Nerve problems and vision problems can happen later.

What is the Treatment for LCHAD?

The following treatments are often used for children with LCHAD:

1. Do not go a long time without food – Babies and young children with LCHAD need to eat often to avoid low blood sugar. They should not go without food for more than 4 to 6 hours. Some babies need to eat even more often. It is important that babies be fed during the night.

Young children with LCHAD should have a starchy snack (such as bread, cereal, rice) before bed and another during the night. They need another snack first thing in the morning. Raw cornstarch mixed with water, milk, or other drink is a good source of long-lasting energy. Your dietitian can give you ideas for good night-time snacks. Dietitians know the right foods your child should eat.

2. Diet – Sometimes a low-fat, high-carbohydrate (such as vegetables, bread, fruits) diet is advised. People with LCHAD cannot use certain building blocks of fat called “long chain fatty acids.” A dietitian can help create a food plan low in these fats.

3. MCT oil, L-carnitine and other supplements – People with LCHAD often use MCT oil. This special oil has medium chain fatty acids. It can be used in small amounts for energy. Doctors prescribe L-carnitine for some children. This is safe and natural and helps body cells make energy. It also helps the body get rid of harmful wastes. Some doctors suggest taking DHA. This may help prevent loss of eyesight.

Things to Remember

Always call your doctor when your child has any of the following:

- poor appetite
- low energy or too much sleepiness
- vomiting
- diarrhea
- an infection
- a fever
- continuing muscle pain or weakness
- reddish-brown color to the urine

Children with LCHAD need to eat extra starchy food and drink more fluids during any illness.