Very Long Chain Acyl-CoA Dehydrogenase Deficiency (VLCAD)

What Is VLCAD?
VLCAD is a type of fatty acid oxidation disorder. People with VLCAD can’t break down certain types of fat into energy for the body.

What Causes VLCAD?
Enzymes help start chemical reactions in the body. VLCAD happens when an enzyme called “very long chain acyl-CoA dehydrogenase” is 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 VLCAD?
[Symptoms are something out of the ordinary that a parent notices.]
There are three forms of VLCAD — infant, childhood, and adult. Symptoms can be mild or serious. Infant and childhood types of VLCAD may cause periods of illness called Metabolic Crises, or low blood sugar. Some of the first signs of a Metabolic Crisis are:

- Too much sleepiness
- Behavior changes (such as crying for no reason)
- Irritable mood
- Poor appetite

If a Metabolic Crisis is not treated, a child with VLCAD can develop:

- Breathing problems and seizures
- Coma, sometimes leading to death

Other problems include enlarged liver, enlarged heart, and muscle problems.

What is the Treatment for VLCAD?
The following treatments are often used for children with VLCAD:

- **Do not go a long time without food** – Babies and young children with VLCAD should eat often to avoid low blood sugar or a Metabolic Crisis. They shouldn’t go without food for more than 4 to 6 hours. Some babies may need to eat even more often. Children with VLCAD should have a starchy snack (such as bread, cereal, and 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.

- **Diet** – Sometimes your child will need to eat a diet low in fat (lean meat and low-fat dairy foods) and high in carbohydrates (such as bread, noodles, fruits, vegetables). Your dietitian will make any needed diet changes. Dietitians know the right foods your child should eat.

- **MCT oil and L-carnitine and other supplements** – Your doctor may prescribe MCT oil. This special oil has medium chain fatty acids that can be used in small amounts for energy. Sometimes the doctor will prescribe L-carnitine. This is safe and natural and helps the body make energy.

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

- Poor appetite
- Too much sleepiness
- Vomiting
- Diarrhea
- An infection
- A fever
- Continued muscle pain or weakness
- Reddish-brown color to the urine

Children with VLCAD need to eat extra starchy food (such as bread, cereal, and rice) and drink more fluids during any illness. When they become sick, they often need to be treated in the hospital to prevent serious health problems.