Very Long-Chain Acyl-CoA Dehydrogenase (VLCAD) Deficiency General Overview

Q. What is VLCAD deficiency?

A. VLCAD deficiency is a treatable disorder that affects the way the body breaks down fats. If left untreated, VLCAD deficiency can cause life-threatening illness.

Q. How does the body normally process fats?

A. The body normally uses sugars from our diet for energy and uses fats as an energy reserve. When all of the carbohydrates and sugars in our bodies have been used, we break down fats for energy. One of the enzymes that helps break down fats is called VLCAD.

Q. What happens to fats in a child with VLCAD deficiency?

A. In a child with VLCAD deficiency, fats cannot be broken down normally because the VLCAD enzyme does not work properly. This can result in high levels of partially broken down fats, which are toxic to the body.

Q. What are the effects of having VLCAD deficiency if it is not treated?

A. Untreated VLCAD deficiency can be life-threatening because of extremely low blood sugar levels, serious heart problems, and possible kidney failure.

Q. What is the treatment for VLCAD deficiency?

A. Treatment for VLCAD deficiency involves avoiding long periods of time without eating and having meals that are high in carbohydrates and low in fats (a special formula may be required). Infants should have at least one nighttime feeding, or a late-night snack, to reduce the time they go without eating. Treatment should begin shortly after birth. Some doctors may also prescribe dietary supplements. Emergency care must be taken if a person with VLCAD deficiency becomes ill and has difficulty keeping food down. This is usually treated in the hospital with an IV. People with VLCAD deficiency require treatment through a specialty clinic with experience in treating this disorder.

Q. Why would a child have VLCAD deficiency?

A. VLCAD deficiency is an inherited disorder. It results when a baby receives a double-dose of a non-working VLCAD gene (one from each parent). For more information about this, contact your health care provider or a genetic counselor.

Q. What are the chances that a child will be born with VLCAD deficiency?

A. About one in every 121,000 babies in the United States is born with VLCAD deficiency. However, parents who have a child with VLCAD deficiency each carry one copy of the non-working gene. That means, with each pregnancy, there is a one in four chance of the child having VLCAD deficiency.

For more information about VLCAD deficiency, please see the Disorders section or our website: www.doh.wa.gov/nbs.

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Revised: 8/27/08