Long-Chain 3-Hydroxyacyl-CoA Dehydrogenase (LCHAD) Deficiency & Trifunctional Protein (TFP) Deficiency General Overview

Q. What are LCHAD/TFP deficiency?

A. LCHAD/TFP deficiencies are disorders that affect the way the body breaks down fats. They are treatable but can cause life-threatening illness.

Q. How does the body normally process fats?

A. The body normally uses sugars and starches from our diet for energy and uses fats as an energy reserve. When all of the sugars and starches in our bodies have been used, we break down fats for energy. LCHAD/TFP are enzymes that help break down fats.

Q. What happens to fats in a child with LCHAD/TFP deficiency?

A. In a child with LCHAD/TFP deficiency, fats cannot be broken down normally because the LCHAD/TFP enzymes do not work properly. This results in a buildup partially broken down fats, which are toxic to the body, and cause low blood sugar levels. Untreated LCHAD/TFP deficiency can be life-threatening because of extremely low blood sugar levels and heart, liver, and respiratory failure.

Q. What is the treatment for LCHAD/TFP deficiencies?

A. Treatment 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). Treatment should begin shortly after birth. Infants should have at least one nighttime feeding, or a late-night snack, to reduce the time they go without eating. Some doctors may also prescribe dietary supplements. Emergency care must be taken if a person with LCHAD/TFP deficiency becomes ill and has difficulty keeping food down. This is usually treated in the hospital with an IV. People with LCHAD/TFP deficiency require treatment through a specialty clinic with experience in treating these disorders.

Q. Why would a child have LCHAD/TFP deficiency?

A. LCHAD/TFP deficiency are inherited disorders. They result when a baby receives a double-dose of the same non-working 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 LCHAD/TFP deficiency?

A. About one in every 105,000 babies in the United States is born with LCHAD or TFP deficiency. However, parents who have a child with LCHAD/TFP 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 LCHAD/TFP deficiency.

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

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