*Elevated C4-0H Acylcarnitine
3-Hydroxyacyl-Coenzyme A Dehydrogenase Deficiency (HADH)
Previously Named Medium/Short Chain Acyl-CoA Dehydrogenase (M/SCHAD)

What is HADH?

3-Hydroxyacyl-Coenzyme A Dehydrogenase Deficiency (HADH) previously named Medium/short-chain acyl-CoA dehydrogenase deficiency (M/SCHAD) is a condition in which the body is unable to break down certain fats. HADH is considered a fatty acid oxidation condition because people affected with the condition are unable to change some of the fats they eat into energy the body needs to function. Most individuals who are identified as having HADH never experience symptoms, but some individuals experience serious health effects from HADH.

What Causes HADH?

When we eat food, enzymes help break it down. Some enzymes help break down fats into their building blocks, called fatty acids. Fatty acids are built like chains, and they come in a variety of lengths. They are classified as either short, medium, long, or very long. Different enzymes help break down different lengths of fatty acid chains. The enzyme 3-hydroxyacyl-CoA dehydrogenase is one enzyme that breaks down medium and short-chain fatty acids for energy.

In HADH, the enzyme 3-hydroxyacl-CoA dehydrogenase is not working correctly. If your baby has HADH, his or her body makes non-working or does not make enough 3-hydroxyacyl-CoA dehydrogenase. When this happens your baby's body cannot break down medium and short chain fatty acids for energy.

Fatty acids are important sources of energy for the heart, especially when sugar is low (such as between meals). When fatty acids cannot be broken down, harmful substances can build up in the body.

What Symptoms or Problems Occur with HADH?

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

Signs of HADH usually begin during infancy or early childhood.

These signs are highly variable from person to person. Your baby may have many of them or none at all.

- sleeping longer or more often
- behavior changes
- irritability
- poor appetite
- fever, diarrhea, vomiting
- low blood sugar (called hypoglycemia)
- trouble breathing
- weak muscle tone (known as hypotonia)
- seizures
- high insulin level

Many of these signs may occur when your baby eats foods that his or her body cannot break down. They can be triggered by long periods of time without eating, illnesses, and infections.

If your baby shows any of these signs, be sure to contact your baby's doctor immediately.

What is the Treatment for HADH?

Dietary Treatment - Your baby may need to be on a restricted diet in order to avoid foods that contain fats his or her body cannot break down. A dietician or nutritionist can help you plan a healthy diet for your baby. Eating often can also help your baby avoid many of the signs mentioned in the "Symptoms or Problems" section.

Supplements and Medications:

Some children with HADH take prescription L-carnitine supplements. L-carnitine is a substance naturally made by the body, but your baby's body might not make enough. Taking L-carnitine supplements can help the body break down fat for energy and remove harmful substances. Your baby's doctor will need to write a prescription for these supplements. HADH can cause dangerously high insulin levels in some children. If your baby has levels of insulin that are too high, your baby's doctor may prescribe medications to help your baby's body regulate insulin levels.

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Things to Remember

If HADH is treated early, children may have healthy growth and development. This is why newborn screening is so important. We do not yet know how effective the treatments for HADH are. Children may still be at risk for some signs of the condition, such as high blood sugar, high insulin levels, enlarged heart, and irregular heartbeat, or liver trouble.

Babies who do not receive treatment for HADH are at risk of life-threatening heart and breathing problems, brain damage, or coma.