FATTY ACID OXIDATION DISORDERS [FAODs]

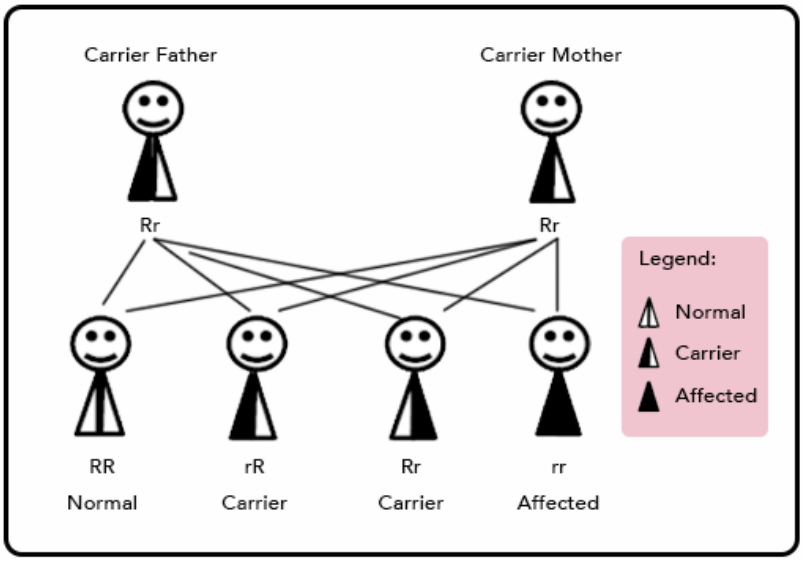
What are FAODs?

Fatty Acid Oxidation Disorders are a group of conditions due to a deficiency in an enzyme or chemical scissors that breaks down fats in the body. Children with this condition cannot properly breakdown fat in their body or from the food they eat. Children born with this condition appear normal at birth but if treatment is not given early, they may present with low blood sugar and lead to seizures, coma and death. This group of disorders can be detected through newborn screening. Early detection can prevent the complications of this condition such as low blood sugar, increased sleepiness, muscle weakness, heart problems among others.

What causes FAOD?

To efficiently use the food we eat, our body breaks it down to smaller units. Due to a lack of an enzyme or chemical scissor, children with this condition cannot effectively utilize fat from their body or from the food they eat.

FAODs are an inherited condition. The gene for the different enzymes is contained in the genetic material that we inherit from our parents. Because one part of the genetic material comes from the father and the other from the mother, the gene comes in pairs. In order to work correctly, at least one of the pairs should be working.



Parents of children with FAOD have one working and one non-working gene coding for a particular enzyme needed in the breakdown of fat. They do not manifest the disease but can pass them on to their children. They are known as carriers.

If the child inherits the non-working gene from both parents, he or she will have a FAOD. Thus, in each pregnancy, there is a 25% chance that the child will have the disorder, 50% chance of being a carrier and 25% chance of having two working genes.

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WHAT ARE THE SIGNS AND SYMPTOMS OF FAOD?

Children born with this condition appear normal at birth but if treatment is not given early, they can present with low blood sugar which can lead to seizures, coma and death. Having an enlarged heart or muscle weakness may be among the complications too.

WHAT IS THE TREATMENT OF FAOD?

The main treatment of a FAOD is through control of the diet. If the child is well or does not have any illness, he/she should eat regular meals and avoid fasting more than 6 hours. One type of FAOD called VLCAD (or very long chain acyl- CoA dehydrogenase deficiency), will require a special milk formula called MCT (medium chain triglycerides).

Please consult with your attending physician prior to starting treatment.

WHAT SHOULD I DO WHEN MY BABY IS UNWELL?

Children with FAOD may have a "metabolic crisis" which is a serious health condition caused by the build-up of toxic substances in the blood. A metabolic crisis occurs when a child is sick, has not eaten or drank well or during stressful events (such as surgery and severe infection). Your child may present with lethargy, seizures or convulsions, irritability and vomiting. If not treated properly and immediately, it might lead to serious brain damage and death. Once these signs and symptoms are present, please bring your child to the hospital for management and alert your pediatrician or metabolic physician.