

3-Methylcrotonyl CoA Carboxylase Deficiency (3MCC)

What is 3MCC?

3-methylcrotonyl-CoA carboxylase deficiency (3-MCC) is a condition in which the body is unable to break down certain proteins properly. 3MCC is considered an organic acid condition because it can lead to harmful amounts of organic acids and toxins in the body. The symptoms of 3MCC vary. Early detection and treatment can often help children with 3MCC lead healthy lives.

What Causes 3MCC?

Enzymes help start chemical reactions in the body. 3MCC happens when an enzyme called “3-methylcrotonyl CoA carboxylase” is missing or doesn’t work right. This enzyme helps break down leucine. All foods with protein contain leucine. Harmful matter can build up in the blood and cause problems when someone with 3MCC eats protein.

What Symptoms or Problems Occur with 3MCC?

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

Symptoms of 3MCC vary. Many people with 3MCC have no symptoms. Others have periods of illness called Metabolic Crises. Some of the first signs are:

- poor appetite
- sleeping longer or more often
- muscle weakness
- irritable mood
- other behavior changes (such as crying for no reason)
- vomiting

If a Metabolic Crisis is not treated, a child with 3MCC might develop:

- breathing problems
- seizures
- liver failure
- coma, sometimes leading to death

Later problems can include:

- low muscle tone
- weakness
- lack of energy
- poor growth

What is the Treatment for 3MCC?

These treatments are sometimes used for babies and children with 3MCC:

1. Low-leucine diet - Most foods in a low-leucine diet will be carbohydrates (such as bread, cereal, noodles, fruits, vegetables). High protein foods to limit or avoid include:

- milk and milk products
- meat and poultry
- fish
- eggs
- dried beans and peas
- nuts/peanut butter

2. Medical foods and formula – There are special medical foods available for people with 3MCC. These foods include special low-protein flours, noodles, and rice. Some children also need a special leucine-free formula. A dietitian will tell you how to use these foods and formula. Dietitians know the right foods for your child to eat.

3. Medications – The doctor may prescribe L-carnitine for your child. This is safe and natural and helps cells make energy. It also helps the body get rid of harmful wastes.

Things to Remember

Minor illnesses such as a cold or flu can cause a Metabolic Crisis in some children with 3MCC. You may need to call your doctor right away when your child has any of the following:

- loss of appetite
- vomiting
- infection or other illness
- fever
- diarrhea

Some children with 3MCC need to eat more starchy foods (such as bread, cereal, and rice) and drink more fluids when they are sick - even if they are not hungry - to avoid a Metabolic Crisis.