

Tyrosinemia Type III (TYR III)

What is TYR III?

Tyrosinemia, type III (TYR III) is a condition in which the body is unable to break down certain building blocks of proteins, known as amino acids. It is considered an amino acid condition because people with this condition are unable to break down the amino acid tyrosine. TYR III can cause learning problems, seizures, and loss of balance. However, detecting TYR III early and beginning treatment can prevent some of the serious outcomes of the condition. Tyrosinemia is a condition with multiple forms, which each have different outcomes and treatments. TYR III is only one form of the condition.

What Causes TYR III?

When we eat food, enzymes help break it down. Some enzymes help break down proteins into their building blocks, called amino acids. Other enzymes break down these amino acids. In TYR III, the 4-hydroxyphenylpyruvate dioxygenase (HPD) is not working correctly.

HPD's job is to break down the amino acid tyrosine. Babies with TYR III do not make enough HPD. When HPD is not working correctly, the body cannot break down tyrosine. This causes tyrosine and other amino acids (methionine and phenylalanine) to build up in the body, which can be toxic. When HPD cannot break down tyrosine correctly, tyrosine may be converted into a harmful acid called homogentisic acid. If homogentisic acid builds up in the body, it can be dangerous. Everyone has some acid and amino acids in his or her blood, but the high levels associated with TYR III can be toxic.

What Symptoms or Problems Occur with TYR III?

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

There are three types of tyrosinemia (I, II, and III) TYR III is the least common type with less than twenty reported cases.

Signs of TYR III are highly variable and not well known.

Signs of TYR III may include:

- Poor coordination and balance
- Seizures

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 TYR III?

Although the signs and symptoms of TYR III are not well known, treatment is still recommended to try to avoid any possible health complications for your child.

Dietary Treatments

Your baby may need to be on a restricted diet in order to avoid certain proteins that his or her body cannot break down. Babies with TYR III may need to limit certain amino acids (phenylalanine, tyrosine, and methionine) in their diet. Amino acids are the building blocks of protein. These are all found in many proteins, and phenylalanine is also found in artificial sweeteners.

Special foods and formulas for children with TYR III are available. These formulas will likely need to continue through adulthood.

Things to Remember

Babies who are treated early for TYR III may never develop any signs. This is why newborn screening for TYR III is so important. Children with TYR III do have a risk of having an intellectual disability, but this risk is lower when the child receives treatment.