



Newborn Screening FACT Sheet

Biotinidase Deficiency (BIOT)

What is BIOT?

BIOT is an inherited disorder. BIOT happens when the body can't use a vitamin called biotin. People with BIOT do not have enough biotinidase activity. This condition is treatable.

What Causes BIOT?

Low biotinidase activity keeps carboxylases from using biotin. The body collects harmful matter when carboxylases can't break down nutrients in the right way. Children with BIOT need extra biotin, or health problems usually result.

What Symptoms or Problems Occur with BIOT?

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

Early symptoms of untreated BIOT are:

- ♦ seizures
- ♦ poor muscle tone
- ♦ vision problems
- ♦ poor coordination
- ♦ delay in development
- ♦ hearing loss
- ♦ skin abnormalities such as:
 - ♦ hair loss
 - ♦ rash
 - ♦ infection

Problems vary from child to child. These are some of the common ones:

- ♦ coughing
- ♦ wheezing
- ♦ lung infections
- ♦ shortness of breath
- ♦ slow weight gain and growth
- ♦ gas and stomach pain
- ♦ clubbed fingers

Problems that can occur with age are:

- ♦ motor limb weakness
- ♦ loss of body control and/or feeling
- ♦ poor vision

What is the Treatment for BIOT?

Medication – Newborns with BIOT rarely have symptoms if they are treated right away. Children with symptoms improve when they take biotin every day. People with severe BIOT should take biotin all of their lives.

Diet – Avoid raw eggs. Well-cooked eggs are safe to eat.

Things to Remember

Some infants with BIOT don't have symptoms. They often don't have problems if they get early treatment. They just need to take biotin every day. They also need to get regular medical checkups.

Children usually get better right away when treated soon after they show symptoms. Some children might continue to have problems.

These include hearing loss, trouble seeing, or developmental problems.

Your doctor can explain more about your child's biotin treatment.