



Newborn Screening FACT Sheet

Propionic Acidemia (PROP or PA)

What is PROP?

Propionic acidemia (PROP) is an inherited condition in which the body is unable to break down certain proteins and fats. It is considered an organic acid condition because it can lead to a harmful amount of organic acids and toxins in the body. If left untreated, it can cause brain defects or even death. However, if the condition is identified early in life and proper treatment is begun, children with PROP can minimize some of the early complications of the condition.

What Causes PROP?

Enzymes help start chemical reactions in the body. PROP happens when an enzyme called “propionyl CoA carboxylase” (PCC) is missing or not working. This enzyme changes certain amino acids so the body can use them. Glycine and propionic acid build up in the blood and cause problems when PCC doesn’t work.

What Symptoms or Problems Occur with PROP?

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

PROP causes periods of illness called Metabolic Crises. Early symptoms of a Metabolic Crisis are:

- poor appetite
- low muscle tone (floppy muscles and joints)
- too much sleepiness or lack of energy
- vomiting

If a Metabolic Crisis isn’t treated, a child with PROP can develop:

- breathing problems
- seizures
- swelling of the brain
- stroke
- coma, sometimes leading to death

Later problems can include:

- intellectual disabilities
- low ability to fight illnesses
- osteoporosis (weak bones)
- inflamed pancreas gland
- skin rashes
- poor growth

What is the Treatment for PROP?

The following treatments are often used for children with PROP:

1. Low-protein diet, medical foods and medical - formula – The best treatment for PROP is a diet low in protein. Most of the low-protein food will be carbohydrates (such as bread, cereal, noodles, fruits, vegetables). High-protein foods that should be limited or not eaten at all include:

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

The doctor may prescribe a special medical formula with the right amount of protein. There are also medical foods available for people with PROP.

2. Do not go a long time without food – Some babies and young children can have a Metabolic Crisis if they don’t eat often enough. They shouldn’t go without food for more than 4 to 6 hours. Some children may need to eat even more often. Your dietitian can give ideas for suitable snacks and knows the right foods for your child to eat.

3. Medication – The doctor may prescribe L-carnitine for your child. This is safe and natural and helps the body make energy.

Things to Remember

Even minor illness such as a cold or flu can cause a Metabolic Crisis. Call your doctor right away when your child has any of the following:

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

Many children with PROP must be treated in the hospital during illness to avoid serious health problems.