

Chronic Benign Neutropenia

Chronic benign neutropenia of childhood is a medical term for a condition which affects the neutrophils.

What are neutrophils?

Blood contains 3 types of cells: white blood cells, red blood cells, and platelets. Neutrophils are one type of white blood cell, and they are an extremely important component of the immune system. They are one of the body's main defenses against bacterial infections.

Neutrophils, like other white cells, red cells, and platelets, are formed in the bone marrow. Neutrophils are released from the bone marrow into the bloodstream, circulate in the blood for a few hours, and eventually enter body tissues (skin, mouth, intestines, lungs, etc), where they fight bacteria.

What is neutropenia?

A low neutrophil count is called '*neutropenia*'. A blood test is used to estimate the number of neutrophils in the bloodstream (this is called the absolute neutrophil count or ANC). The normal neutrophil count varies by age and ethnicity, but in general the lower limit of normal before age 1 is $1.0 \times 10^9/L$, and $1.5 \times 10^9/L$ after age 1.

After the first year of life neutropenia is defined as:

- Mild if the ANC is $1.0-1.5 \times 10^9/L$
- Moderate if the ANC is $0.5-1.0 \times 10^9/L$
- Severe if the ANC is $<0.5 \times 10^9/L$

What are the signs and symptoms of neutropenia?

Children often have no symptoms, and neutropenia is found incidentally.

However, a low neutrophil count may cause an increased frequency of minor bacterial infections, such as ear infection (otitis media), throat infection (pharyngitis), skin boils, mouth sores, inflamed gums and urinary tract infections.



When the ANC is very low ($<0.5 \times 10^9/L$), serious and even life-threatening infections may occur. These include lung infection (pneumonia), infection of the spinal fluid (meningitis), and blood infection (bacteremia). Children with these infections must be hospitalized and given intravenous antibiotics.

What causes neutropenia?

Neutropenia has many causes. In the majority of cases, neutropenia is related to a recent illness, infection, or medication. This type of neutropenia is temporary and resolves within 3-6 weeks.

If the neutropenia does not improve after 8 weeks, it is considered *chronic*. The most common cause of chronic neutropenia in childhood is chronic benign neutropenia.

Less common causes are described below:

Neutropenia can be one part of a disease process that is affecting other parts of the body. These diseases can be differentiated from other causes of neutropenia by medical evaluation and further blood tests.

The most serious cause of neutropenia occurs when the bone marrow is having problems making neutrophils, usually because of damage to the bone marrow or the presence of abnormal cells. These diseases are usually accompanied by abnormalities of the other blood cells, namely the red cells and platelets. Sometimes a bone marrow test is needed to look for these causes.

Rarely, the neutropenia is present from birth (*congenital*) or passed down from one or both parents (*inherited*). In these cases, infections are often very severe.

What is chronic benign neutropenia?

Chronic benign neutropenia is the most common type of neutropenia in childhood. It can appear as early as 6-12 months of age, or later in childhood. In the majority of children the neutropenia persists for a few years and then spontaneously resolves. Children with chronic benign neutropenia are generally healthy and have

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normal physical exams. Their ANC can go up and down but is usually between $0.00 - 1.5 \times 10^9/L$.

What causes chronic benign neutropenia?

An antibody is present that attaches to neutrophils and causes them to be destroyed. Around 50% of children will test positive for *anti-neutrophil antibodies*. The reason why these antibodies form is not fully understood, but there is often a history of preceding viral infection.

What is the treatment for chronic benign neutropenia?

Most of the time, no specific treatment is necessary. This is because children with chronic benign neutropenia have mild infections, and their bone marrow is able to produce enough neutrophils to respond to an infection.

However, when the neutrophil count is low (and especially when it is severely low $<0.5 \times 10^9/L$), the body has difficulty fighting bacterial infections.

When to seek help

If a child with neutropenia develops a fever ($>38.5^\circ\text{C}$ or $>38^\circ\text{C}$ x 2 readings taken 1 hour apart), the child should see a doctor. Depending on the assessment, your physician may choose:

- Observation with close follow-up (within 24 h)
- Oral antibiotics +/- lab tests
- Intravenous or intramuscular antibiotics as an out-patient + lab tests
- Admission to hospital for intravenous antibiotics

How can I help?

It is vital to ensure that your child is living a healthy lifestyle. This includes providing them with a well-balanced diet and ensuring that they engage in regular daily physical activity.

It is important for you to understand your child's condition, so please ask your healthcare provider for any information that may help you and your child.