

Chronic Benign Neutropenia of childhood

What is chronic benign neutropenia?

Chronic benign neutropenia is a common type of neutropenia seen in childhood and lasts at least two months with more than half the children having neutropenia for 18 to 20 months. It appears as early as six to 12 months of age with frequent ear infections, sore throat, diarrhea and minor infections but may also be found only by chance. The rest of the blood count is normal and the child appears healthy with nothing abnormal on the physical exam. The ANC is between 0-500/mm³.

What causes chronic benign neutropenia?

The cause of this condition is not well known. It has been found that over half (40-80%) of the children with this condition have *autoimmune neutropenia* and their bodies produce *anti-neutrophil antibodies* that attach to the neutrophils and cause them to be destroyed. The reason why these antibodies form is not known. It does not seem to be associated with a family history of allergic problems or other types of allergies or serious disorders.

What is the treatment for chronic benign neutropenia?

Most of the time “watch and wait” is followed with no specific treatment since the infections are mild and the child’s bone marrow is able to produce enough neutrophils to respond to infection. However, if a child with chronic neutropenia develops high fever, he or she should see a doctor for a physical exam and cultures for specific germs and oral or injected antibiotics. Rarely, children with chronic benign neutropenia may develop pneumonia or other serious infections and will need to be hospitalized. The ANC may normalize during an infection but this is temporary.

If infections are severe and repeated then several treatments may be tried but are not very effective for long-term:

- *Prednisone*, a steroid drug, can suppress the immune system, large doses are often required and widespread side effects are common.
- *Intravenous gamma globulin (IVIG)*, a concentrated form of antibodies taken from blood plasma is very costly, and has side-effects that include allergic reactions and meningitis
- *Granulocyte colony stimulating factor (G-CSF)*, a hormone-like substance given under the skin by injection, stimulates production of neutrophils in the marrow. Severe side effects are few, so it is most commonly used if needed.

What happens to children with chronic benign neutropenia?

Children with chronic benign neutropenia rarely develop life-threatening infections and usually outgrow their disease by 3-5 years of age, when the ANC begins to rise. There is no evidence that these patients develop other blood disorders, leukemias, or other types of conditions.