

NEUTROPENIA – THE FACTS



Barth Syndrome
Trust



What is neutropenia?

If you are told that you have Neutropenia it means that you have an abnormally low level of neutrophils, a type of white blood cell. All white blood cells help the body fight infection. Neutrophils fight infection by destroying harmful bacteria and fungi (such as yeast) that invade the body.

If you have neutropenia it means that you are more likely to get a serious infection because you do not have enough neutrophils to destroy these harmful germs that cause disease.

Nine out of every ten males with Barth Syndrome get neutropenia at some time; some are neutropenic some or all of the time, but others may only be neutropenic occasionally. The neutrophil count should be more than $1.5 \times 10^9/L$ most of the time. Neutropenia is said to be severe when the neutrophil count falls to less than $0.5 \times 10^9/L$.

How will I know I have it?

Neutropenia itself does not cause any symptoms. You may only find out you have neutropenia by having a blood test or when you get an infection. If you spot any of the following signs, it may mean that you are becoming neutropenic, so please tell your parents or your medical team.

- Mouth ulcers
- Excessive skin spots or infection
- Redness, swelling, or pain, particularly around a cut or wound
- A fever (temperature of $38^\circ C$ or higher)
- Chills or sweating
- Diarrhoea
- A cough or chest infection
- Pain or sores around the anus



Treating Neutropenia

If the number of your neutrophils gets too low or you have a temperature of 38 degrees or higher you may need to go to hospital to get your blood checked and have other tests. You may need to stay in hospital for a few days and be given antibiotics.

What is GCSF?

If you have low neutrophil counts it is often better to help your bone marrow make new neutrophils by giving GCSF (granulocyte colony stimulating factor). Bone marrow is the material inside the bones where blood cells are made. G-CSF is a special protein made in the bone marrow that stimulates bone marrow to make neutrophils. G-CSF is given by injection under the skin, the injections will usually make you feel much better, and often stop mouth ulcers developing.

You may have injections 2 -3 times a week and you will need to have your blood checked regularly to see if you are on the right dose. We usually ask you to do this either just before a dose of GCSF or on the day after. This way we can see if your neutrophil count gets too high or too low. Sometimes we will ask you to take an antibiotic called Penicilin just in case the GCSF has not helped you make enough neutrophils.