

Neutropenia

Neutrophils are white blood cells which protect against bacterial and fungal infection. A low neutrophil count can put you at serious risk of bacterial infection. If such a low count is observed on multiple occasions, the patient is said to suffer from severe chronic neutropenia.

Although neutropenia is a major feature of Barth syndrome, there are some (around 10%) of children who do not have it at all. It can also present later in life.

Signs of neutropenia to watch out for include:

Mouth ulcers, sore or bleeding gums, skin spots or perianal rashes in the nappy area or repeated infections.

Perianal Streptococcal Dermatitis: this perianal infection is easily misdiagnosed as thrush (fungal infection). It shows up as a bright red area around the anus, sometimes with spots spreading out onto the buttocks or thighs. There may be diarrhoea, blood streaking of stools or passage of mucus on stools due to infection within the rectum. Children may be lethargic and generally unwell with this infection and can have symptoms over many weeks or months. The infection is easily proven by an anal swab showing growth of Streptococci and will usually resolve within 1-2 days on treatment with penicillin. It is very important to recognise perianal Streptococcal dermatitis in neutropenic patients since this can be an important source of bacteria spreading throughout the body and bloodstream.

A child may develop a serious bacterial infection whilst his count is very low or even zero but once in hospital, his count may creep up to a normal range, tricking doctors into thinking that everything is normal. The reason for this increase of neutrophils into a low normal range is simply because his body is trying to mount a defence against the infection.

Most children have a lower than normal baseline temperature (around 36.5°C), probably due to their low muscle mass, so any persistent increase in temperature to 38°C or above should be taken seriously.

Medications for neutropenia

- **Prophylactic antibiotics:** a daily dose of a broad-spectrum antibiotic like Pen V or Septrin is often enough to provide protection against bacterial infections and reduce episodes of illness.
- **Granulocyte Colony Stimulating Factor (G-CSF)** can normalise neutrophil counts and prevent infections. G-CSF is given via a subcutaneous injection just under the skin.