
Sickle Cell Anaemia

Sickle cell anaemia is an illness in which haemoglobin molecules are misformed, and thus haemoglobin does not function as normal. The condition takes its name from the fact that abnormal red blood cells may become rigid, rough, and crescent shaped - like a sickle.

Sickle cell anaemia tends only to occur in people of African or Afro-Caribbean origin.

Cause

Sickle cell anaemia is caused by an inherited genetic disorder, believed to be the result of an evolved response to the disease malaria.

The genetic disorder leads to an incorrect amino acid being used in the structure of haemoglobin.

The disease develops when a child inherits 2 sickle cell genes - one from each parent.

Should a child only inherit one sickle cell gene, they will not then develop the disease, but will carry sickle cell trait, and may pass on the gene to their children.

Signs & Symptoms

The sickle shape of red blood cells causes the blood to be more viscous than normal, which may result in poor flow through small capillaries. The distorted haemoglobin structure is less well able to carry or release Oxygen than normal haemoglobin.

The overall results are reduced bone marrow activity and poor tissue perfusion, which are shown by:

- Chronic pain.
- Chronic fatigue.
- Swelling of tissues.
- Increased susceptibility to infection.
- Tissue destruction.
- An elevated risk of premature death through heart failure or stroke.

Treatment

There is no effective treatment for sickle cell anaemia; sufferers require life-long management to minimise the effects of their condition.

Sickle cell crisis

A sickle cell crisis develops, maybe during an infection, pregnancy, or following use of anaesthetics, when increased numbers of sickle shaped cells clump together and either block, or partially obstruct, small blood vessels.

Crises may occur occasionally, or frequently. Frequent, repeated crises may lead to other chronic conditions (eg. heart failure) through tissue damage.

Signs & symptoms

- Fever.
- Severe pain - especially in the back, abdomen, arms, and legs.
- Swelling at the joints.
- Tiredness and weakness.
- Laboured breathing.
- Pallor.

Treatment

- Administer Oxygen. Administer Entonox if required to relieve severe pain.
- Position the patient in a manner most comfortable for them.
- Transport to hospital - if possible to the patient's regular treatment centre.