Sickle Cell and Thalassaemia Centre 0121 507 6040

Sandwell and West Birmingham Hospitals NHS
NHS Trust

What is Sickle Cell Disease? (Also known as Sickle Cell Anaemia or Sickle Cell Disorder)

Sickle Cell Disease (SCD) is a genetic (inherited) blood disorder caused by the sickle mutation. This mutation results in abnormal production of haemoglobin.

It is a **quality** of haemoglobin problem; that causes features of:

1. **Haemolysis** – sickle red blood cell (rbc) have a much shorter life span (20 days compared to 120 days in normal rbc)

2. **Vasocclusion** – the sickle rbc are less flexible and more 'sticky' than normal rbc. They deform on deoxygenation and can clump and occlude small blood vessels causing painful sickle cell crises as well as vascular and organ damage.

The highest prevalence of SCD is among Black Africans and Black Caribbean, although it is also found in Mediterranean, South Asian and Middle Eastern populations.

HbSS is the most common genotype but SCD can also result from HbS/Beta Thal, HbS/Oarab, HbS/C and HbS/Dpunjab.

Sickle Cell Trait or carrier status does not cause health problems for the individual.

Acute presentations of SCD

Acute painful vasocclusive crises	Hepatic/Splenic Sequestration
Sepsis	Anaemia
Acute Chest syndrome	Gall Stones
Stroke	Priapism

Chronic problems of SCD

Leg ulcers	Avascular necrosis
Pulmonary hypertension	Retinopathy
Chronic renal failure	Chronic pain

Refer to the trust guidelines on the management of Sickle Cell Disease