What is Sickle Cell?

Sickle Cell is a defect of the haemoglobin in the red blood cells.

WHAT IS HAEMOGLOBIN (Hb)?

Haemoglobin is the red part of the blood, which binds with oxygen inside the red cell and carries it to all parts of the body. Normal haemoglobin is called Hb A. Every person has two haemoglobin genes, we inherit one from each parent. Haemoglobin can only be passed on from parent to child in their genes. It is inherited in the same way as our other natural features, e.g., eyes and hair colour. Our haemoglobin type remains the same for life; it does not change.

What Are Genes?

Genes are the ways in which parents pass on their natural characteristics to their children e.g. appearance and height as well as hair and eye colour etc.

Red Blood Cells

Normal red blood cells are round, soft and spongy like a doughnut. They can change shape to squeeze through the smallest blood vessels and then spring back into their natural round shape again.

Sickle Cell Trait (HbAS)

People with sickle cell trait have inherited normal haemoglobin A from one parent and an abnormal sickle haemoglobin S from the other parent (HbAS).

Sickle cell trait is not an illness and will never become sickle cell anaemia. People with sickle cell trait are also called "healthy carriers".

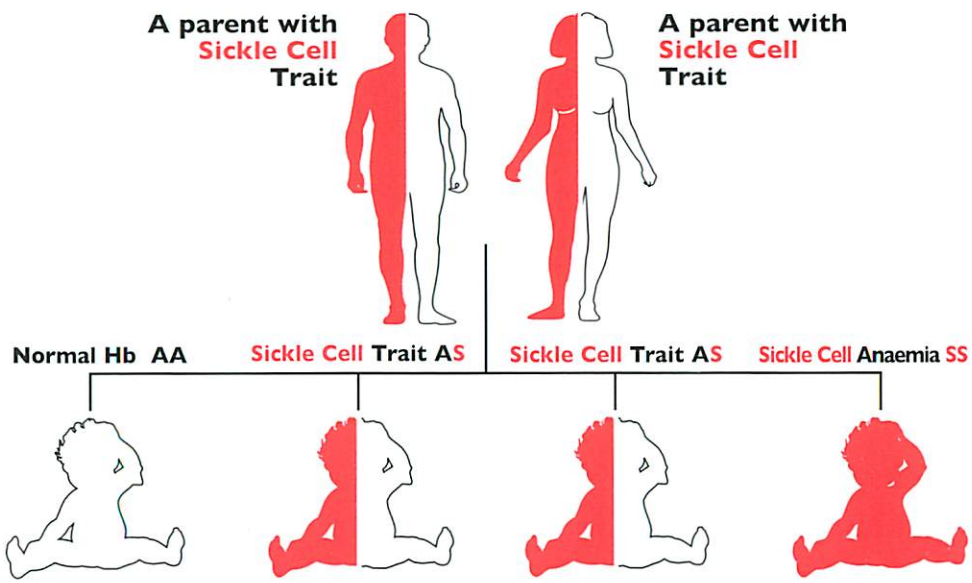
Most people with sickle cell trait are unaware of their haemoglobin type.

Why is Sickle Cell Trait Important?

Although people with sickle cell trait are healthy and will never develop sickle cell anaemia, it is still very important for them to know their haemoglobin type.

If both partners have Hb AS, there is a risk that they can have children with sickle cell anaemia.

This diagram shows the possible haemoglobin types that a couple who both have sickle cell trait (HbAS) could pass onto their children.



With EVERY pregnancy:

There is a 1 in 4 chance that the child will inherit the sickle cell gene from both parents and have sickle cell anaemia (Hb SS).

There is a 1 in 2 chance that the child will inherit a sickle gene from one parent and a normal gene from the other and have sickle cell trait (Hb AS).

There is a 1 in 4 chance that the child will inherit a normal haemoglobin gene from both parents and have a normal adult haemoglobin (Hb AA).

Sickle Cell Disorders

Sickle Cell disorders are a group of illnesses caused by the inheritance of two abnormal haemoglobin genes. The commonest of them is Sickle Cell Anaemia Hb SS

When oxygen levels are low the red blood cells change into a sickle or banana shape. Sickled red cells are hard and sticky, and cannot squeeze easily through blood vessels, especially the smallest ones. They become trapped, and so block the blood flow, this causes severe pain.

- People with Sickle Cell Anaemia, have **sickle cell crises**
- **Have a low haemoglobin**
- **Tire easily**
- **Have a low resistance to infection**

Other Sickling Conditions

There are many different haemoglobin types, such as Hb C, D, E and Beta Thalassaemia. When any of these combines with the sickle haemoglobin S they cause other serious Sickling Disorders, e.g. Hb.SC, SE, SD and Sickle Beta Thalassaemia.

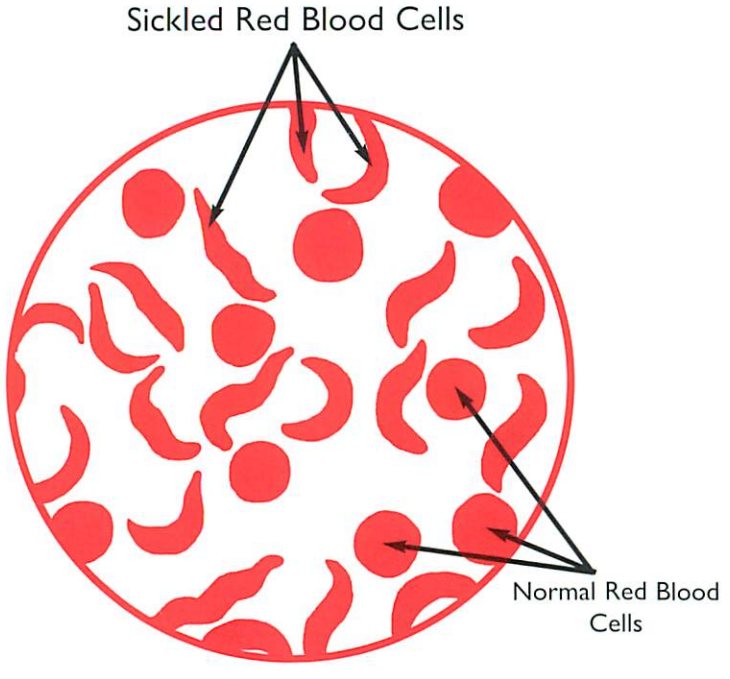
Who Is Most At Risk?

Sickle cell is found mainly in people who come from regions where Malaria is common. These are Africa, the Caribbean, Asia, the Middle East and the Mediterranean. Sickle cell can also be found among other races.

People whose parents or who themselves come from these regions should have a blood test to find out if they have a trait. It is particularly important before and in very early pregnancy. Tests to find out if a baby is affected can be carried out in early pregnancy.

Anyone, who wishes to know more, or to be tested, can call into the Sickle Cell Unit or telephone for an appointment. Blood tests can be arranged for anyone who requests it, and their families.

Sickle Cell .....*What Is It?*



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