Sickle Cell Anaemia (Hb SS): Information Sheet

Produced by Specialist Haematology, Canterbury Health Laboratories

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What is Haemoglobin?

Haemoglobin (Hb) is a protein found in red blood cells; it carries oxygen from the lungs to all parts of the body and gives blood its red colour. Hb is made up of haem groups, which contain iron and globin chains which are made of protein.

The combination of different globin chains with the haem groups produces the different types of haemoglobin present in people. The normal adult haemoglobin is Hb A.

Where a person has inherited Hb A from both parents, they will have the usual Hb combination, Hb AA.

What is Sickle Haemoglobin (Hb S)?

Sickle haemoglobin (Hb S) is a variant Hb and occurs when the structure of the globin chain is altered.

Sickle cell trait (heterozygous Hb S) occurs when a person inherits the normal Hb from one parent and the sickle Hb from the other (Hb AS).

Sickle cell anaemia or homozygous Hb S (Hb SS) occurs when a person inherits sickle Hb from both parents.

In sickle cell anaemia the red blood cells contain mainly sickle Hb (Hb S).

The presence of large amounts of Hb S in the red cells causes them to become very rigid, taking on a sickled shape. This makes the red cells less flexible than normal cells causing them to pile together and block small blood vessels.

Who does it affect?

Sickle Hb occurs most frequently in people of African, Caribbean, Indian, Pakistani, Middle Eastern or Eastern Mediterranean ancestry.

How Does it Present?

Because of their rigid shape, sickled red blood cells can't squeeze through small blood vessels as easily as normal red cells. This can cause blockages of small blood vessels which then stop the oxygen from getting through to where it is needed. This in turn can lead to severe pain and damage to organs.

Symptoms are rarely seen before of the age of six months. The main symptoms of sickle cell anaemia are episodes of anaemia, pain or infection and are called crises. The frequency of these crises varies. Most serious acute complications occur during childhood.

Anaemia can be moderate to severe and may worsen during infection, pregnancy or during a crisis.

A pain crisis due to the blockage of small vessels can affect the chest, abdomen, back, jaw, legs and arms and in infancy the hands and feet. If these pain crises are severe, they may require hospitalisation.

Both minor infections and serious life-threatening infections like septicaemia, pneumococcal meningitis and osteomyelitis can occur.

Pregnancy can be associated with serious complications for people with sickle cell anaemia.

What is the Treatment?

Sickle cell anaemia is a serious condition requiring significant medical follow-up.

- Patients should try to maintain good general health and nutrition.
- Folic acid supplements may be suggested.
- Sometimes prophylactic antibiotics are beneficial.
- Special care is needed when having an operation, needing an anaesthetic or at high altitudes.
- Infections must be treated promptly, with severe infections requiring hospitalisation.
- Minor pain crisis can be treated at home with pain killers but do often require a patient to be admitted to hospital.
- If the anaemia becomes severe during a crisis a blood transfusion may be needed.
- Administration of a drug to increase the level of foetal haemoglobin (Hb F) can often improve clinical symptoms.

How is Sickle Haemoglobin inherited?

Haemoglobin S is inherited from either one or both parents who carry the Haemoglobin S gene on the DNA in their cells.

If a person is born with Haemoglobin S, they will have it all their life. It is **not** contagious or infectious.

How Will This Affect My Children?

Every child inherits haemoglobin genes from each parent. The type of haemoglobin gene inherited from the parent determines the type of haemoglobin the child will have.

A person with sickle cell anaemia **will** pass on a Hb S gene to each of their children.

When **both** parents carry a gene for a variant Hb, there is a chance that any child could have a severe form of the variant haemoglobin.

Why is Knowing About Sickle Haemoglobin so Important?

Sickle cell anaemia is a serious condition requiring significant medical intervention.

If your partner has a variant Hb or thalassaemia, future children are at risk of inheriting a serious sickling disorder.

If you have sickle cell anaemia it is important to have your partner tested for the presence of a variant haemoglobin or thalassaemia before you have children.

Is There a Test for Sickle Haemoglobin?

YES ! Blood tests can show whether an individual has sickle haemoglobin.

It is important to have your partner tested for a variant Hb or thalassaemia before you have children. Other members of your family who are of reproductive age should also be tested for haemoglobin S.

The documentation of sickle Hb is important as:

1. It avoids the need for repeat testing in the future.
2. Extra care needs to be taken with the administration of anaesthetics.
3. Haemoglobin S is an inherited disorder which if passed on to a child by **both** parents could result in a severe condition.

However, if two people who are at risk of having a child with a serious sickling disorder do become parents, there are special prenatal tests that can be done to detect or rule out sickle cell disease in the fetus.

Early diagnosis of a possible severe form of sickle haemoglobin is important so that treatment can prevent as many complications as possible.

Can Sickle Haemoglobin Be Prevented?

Sickle haemoglobin cannot be prevented at this time, but a programme of health education, testing for the trait, genetic counselling and prenatal diagnosis can provide families with full medical information to help them have healthy children.

People who think they may have sickle haemoglobin should go to their family doctor for advice and testing.

Your family doctor can contact a Haematologist at the Community Laboratory or the Haematology Department at Canterbury Health Laboratories on (03) 364 0300 if further clinical input is required.

The Specialist Haematology Laboratory at Canterbury Health Laboratories provides a comprehensive diagnostic service in conjunction with the family doctor.

Contact for Canterbury Health Laboratories

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On behalf of the Clinical Haematology Team at Christchurch Hospital