Haemoglobin S (Sickle Haemoglobin):

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). When a person inherits the haemoglobin S from both parents, they are said to have sickle cell anaemia or homozygous haemoglobin S (Hb SS).

Red blood cells containing only sickle Hb are less flexible than normal and can sometimes 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.

What is the Treatment?

Sickle cell trait *is **not** an illness and requires no regular medical follow-up or treatment.*

The trait itself rarely causes health problems, though it may occasionally cause blood to appear in the urine and special care is required when having an operation, needing an anaesthetic or at high altitudes.

Sickle cell anaemia (Hb SS) is a serious condition requiring significant medical follow-up.

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 trait will pass on either a Hb S gene **or** a normal Hb gene to each of their children.

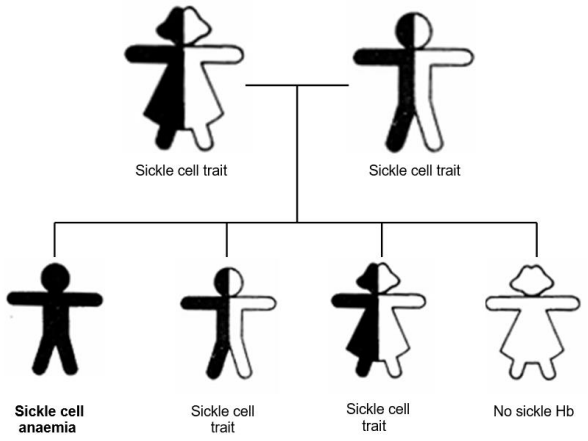
When **both** parents carry a gene for a variant Hb, there is a chance that their children could inherit the variant Hb from each parent which may cause a more severe condition.

When **two** people with sickle cell trait become parents, there are several possible outcomes:

- 25% chance that the child will inherit a Hb S gene from each parent and have the severe form of the disease (sickle cell anaemia, homozygous Hb S)
- 50% chance that the child will inherit one normal Hb gene and one Hb S gene and have sickle cell trait like its parents
- 25% chance that the child will inherit two normal haemoglobin genes from its parents and will not have Hb S

These odds are the same for each pregnancy when **both** parents have sickle cell trait.

BOTH PARENTS WITH SICKLE CELL TRAIT



Why is Knowing About Sickle Haemoglobin so Important?

The inheritance of sickle cell trait produces a mild condition.

However, the inheritance of a sickle cell gene from **both** parents produces sickle cell anaemia which is a severe condition requiring significant medical follow-up.

There are other types of variant haemoglobins or thalassaemia (another type of inherited anaemia frequent in the Mediterranean, Middle East and India) which if inherited together with sickle Hb can also produce a severe condition.

If you have sickle cell trait 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 carrier status 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 foetus.

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

Can Haemoglobin S or Thalassemia 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

Laboratory Services: Specialist Haematology Laboratory, Canterbury Health Laboratories.
Address: 524 Hagley Ave, Christchurch 8011.
Phone: (03) 364 0300.

On behalf of the Clinical Haematology Team at Christchurch Hospital