Haemoglobin S – Delta/Beta Thalassaemia:

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 Haemoglobin S?

Red blood cells containing only sickle Hb are less flexible than normal and can sometimes pile together and block small blood vessels.

This occurs if a person inherits sickle haemoglobin from both parents, this condition is called **sickle cell anaemia** or homozygous haemoglobin S (Hb SS).

This can also occur if a person inherits sickle Hb from one parent and another variant haemoglobin or thalassaemia from the other parent.

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

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

What is Thalassaemia?

Thalassaemia is a type of inherited anaemia (red blood cell deficiency). People with thalassaemia don't produce enough globin chains.

The two main types are called alpha (α) and beta (β) thalassaemia, depending on which globin chains are affected. Less commonly, two globin chains are affected as in delta-beta (δβ) thalassaemia.

Delta-Beta Thalassaemia

δβ thalassaemia occurs when a person inherits one normal Hb gene and one thalassaemia gene which produces no delta or beta globin chains.

Most people with δβ thalassaemia have a mild form, called **δβ thalassaemia trait** (sometimes called heterozygous δβ thalassaemia).

When a person inherits a δβ thalassaemia gene from both parents, they will have homozygous δβ thalassaemia.

δβ thalassaemia occurs most frequently in people of the

- **Central Mediterranean:** Italy, Sicily, and Malta
- **Southeastern Europe:** The Balkans, Greece, and Hungary
- **Eastern Mediterranean & Near East:** Turkey, Israel, and Egypt

What is Haemoglobin S / Delta-Beta Thalassaemia?

Haemoglobin S / delta-beta thalassaemia (Hb S/δβ thalassaemia) occurs when a person inherits a haemoglobin S (Hb S) gene from one parent and a delta-beta (δβ) thalassaemia gene from the other parent. Delta-beta thalassaemia is a rare inherited blood disorder caused by reduced production of both delta and beta globin chains, often resulting in increased levels of fetal haemoglobin (Hb F).

If a person is born with Hb S/δβ thalassaemia, they will have it for life. It is **not** contagious or infectious.

The severity of Hb S/δβ thalassaemia varies from person to person. Many individuals have a relatively mild anaemia because the increased fetal haemoglobin (Hb F) can reduce red blood cell sickling. However, some people may develop complications similar to those seen in sickle cell disease including painful vaso-occlusive crises, acute chest syndrome, gallstones, or other sickle cell-related complications.

What is the Treatment?

Haemoglobin S/detla-beta thalassaemia usually requires ongoing medical follow-up.

Treatment may include:

- 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.

Children require regular monitoring to ensure normal growth and development.

How Will This Affect My Children?

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

A person with Hb S/delta-beta thalassaemia will pass on **either** the Hb S gene or the delta-beta thalassaemia gene to each of their children.

When **both** parents carry a gene for a variant haemoglobin or thalassaemia, there is a chance that their children could inherit a severe haemoglobin disorder.

Why is Knowing About Haemoglobin S/ Delta-Beta Thalassaemia so Important?

Haemoglobin S/delta-beta thalassaemia can range from a moderate to severe inherited blood disorder.

A person with Hb S/delta-beta thalassaemia will pass on **either** a haemoglobin S gene or a delta-beta thalassaemia gene to each of their children.

Inheritance of significant haemoglobin variants or thalassaemia genes from **both** parents may result in a child being born with a severe haemoglobin disorder requiring lifelong medical care.

If you have Hb S / delta-beta thalassaemia it is important to have your partner tested for the presence of thalassaemia or a variant haemoglobin before you have children.

Is There a Test for Haemoglobin S and/or Thalassemia?

YES! Blood tests can determine whether an individual has haemoglobin S, thalassaemia, or a combination of the two.

If you have Hb S/delta-beta thalassaemia, it is important to have your partner tested before starting a family. Other relatives of reproductive age may also wish to consider testing.

Documentation of carrier status or of a condition requiring medical follow-up is important because:

- Patients with Hb S/delta-beta thalassaemia may require regular lifelong monitoring and specialist medical follow-up to detect and manage complications associated with sickle cell disease.
- It avoids the need for repeat testing in the future.
- Extra care needs to be taken with the administration of anaesthetics.
- Thalassaemia trait may be confused with iron deficiency, leading to unnecessary iron treatment.

If two people who carry a variant haemoglobin or thalassaemia do become parents and there is a risk of a severe disorder in the fetus, special prenatal tests can be done to detect or rule this out.

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

Can Haemoglobin S or Thalassemia Be Prevented?

Haemoglobin S and/or thalassaemia cannot be prevented at this time, but a programme of health education, testing for the trait and genetic counselling can provide families with full medical information to help them have healthy children.

People who think they may have haemoglobin S or thalassaemia 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.
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On behalf of the Clinical Haematology Team at Christchurch Hospital

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