

Sickle Cell Trait

If you inherit a gene from one of your parents, called a sickle cell gene, you have a condition called sickle cell trait. If you inherit a sickle cell gene from both parents, you get a double dose of the sickle cell gene. This causes a condition called sickle cell disease (SCD).

What is sickle cell trait?

Sickle cell trait means having one gene for a condition called sickle cell disease (SCD). This in itself does not normally cause problems and sickle cell trait is not considered as a disease. It is extremely rare for it to cause problems or complications, which mainly occur under conditions of severe physical stress (explained below).

Sickle cell trait is important because your children can inherit the sickle cell gene. If both parents have sickle cell trait or sickle cell disease, their children could get a double dose of the sickle cell gene, which would give them the serious condition called SCD. This is relevant if you are pregnant or wanting to start a family.

Who gets sickle cell trait?

Anyone can have sickle cell trait but it is most common in people whose family origin is Black African, Black Caribbean or Black British. It also occurs in people who originate from the Middle East, India and Eastern Mediterranean areas. In other populations, sickle cell trait is unusual but can occur.

In England, about 240,000 people carry a sickle cell gene.

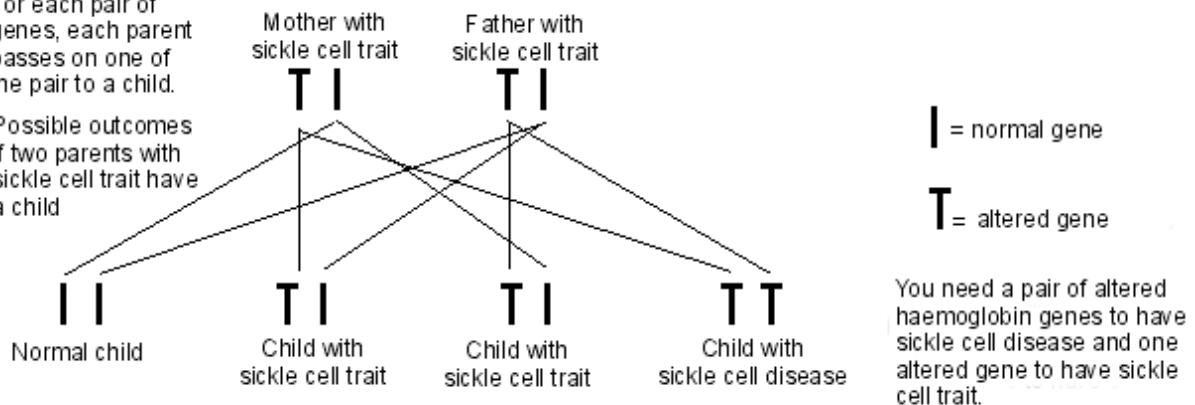
What causes sickle cell trait?

You inherit haemoglobin genes from both parents. One sickle cell gene gives you sickle cell trait; two sickle cell genes give you SCD.

Sickle cell inheritance

For each pair of genes, each parent passes on one of the pair to a child.

Possible outcomes if two parents with sickle cell trait have a child



This means that: if both parents have sickle cell trait, their children have: a 1 in 2 chance of having sickle cell trait, a 1 in 4 chance of having SCD, and a 1 in 4 chance of having no sickle genes (as shown in the diagram above).

If one parent has SCD and the other has sickle cell trait, their children have: a 1 in 2 chance of having sickle cell trait, and a 1 in 2 chance of having SCD.

If one parent has sickle cell trait and the other has no sickle genes, their children have: a 1 in 2 chance of having sickle cell trait, and a 1 in 2 chance of having normal haemoglobin genes.

Actually, the inheritance of SCD is slightly more complicated than that. This is because there are other haemoglobin genes which can interact with sickle cell trait. If you have one of these genes plus a sickle gene, you also get SCD (the combination behaves like two sickle cell genes). Examples of these interacting genes are HbC, beta thalassaemia, Hb Lepore and HbO Arab.

The result of these genes is that an important body chemical called haemoglobin is altered. This is what affects the red blood cells and makes them change into a sickle shape. With sickle cell trait, you have only small amounts of altered haemoglobin, so the blood cells are not much affected (unless conditions are very extreme). With sickle cell disease, there is more altered haemoglobin and the red blood cells easily change into sickle shape, which causes various problems. These are described in more detail in our separate leaflet called Sickle Cell Disease.

Sickle cell blood test

Sickle cell trait is diagnosed in the same way as SCD. It involves a blood test. The test may be offered to pregnant women and if necessary their partners. Babies may also need the test.

Sickle cell trait symptoms

It is extremely uncommon for people with sickle cell trait to have any symptoms. Complications of sickle cell trait are also uncommon. Most of the complications which do occur are due to extreme physical conditions, and so are usually preventable. The possible complications are:

Complications of severe low oxygen conditions or severe dehydration

These can occur in conditions where oxygen is extremely low – such as flight in an unpressurised aircraft, very high altitude or problems during a general anaesthetic. Also, if you become severely lacking in fluid in the body (dehydrated).

In these situations, sickle cells can form and can block small blood vessels. This may cause episodes of pain in bones, muscles or the spleen. If in the spleen, it is called splenic infarction and may cause pain in the tummy (abdomen) or chest. These problems may be severe enough to make you very unwell and to need treatment in hospital. They will usually clear up with treatment and when oxygen and hydration levels are back to normal.

To avoid possible complications, you should

- Avoid flying in unpressurised aircraft.
- Avoid visiting very high altitudes.
- Keep your fluid intake up, especially if you have a fever or are exercising.

Complications of severe physical exercise

Sometimes people can become seriously ill as a result of extreme physical training. This can particularly occur if it is a forced pace, under very hot conditions, if they are unused to the training, or if they do not drink enough fluid. Anyone can become ill under these conditions, but people with sickle cell trait are probably at higher risk.

In these conditions, people with sickle cell trait may get pain episodes or pain in the spleen (as explained above).

How can complications of severe exercise be prevented?

The following are a few recommendations which may be useful to prevent complications occurring:

- Tell your trainer, sports association, doctor and nurse that you have sickle cell trait, and make sure they are aware of the guidelines.
- Avoid extremes such as heat, very high altitude, exertion to the point of collapse, or sudden increase in exercise without proper training. You can still take part in sports if you take precautions. There are professional athletes who have sickle cell trait.
- When exercising, drink enough fluid. Have breaks for drinks, rest and cooling.
- Sports where you can pace yourself are safer than a forced pace.
- If you have symptoms such as pain or difficulty breathing, stop, report your symptoms, rest and rehydrate. Get medical advice if symptoms don't improve quickly.
- Build up your training gradually.
- If you also have asthma, make sure it is treated and well controlled.
- Extreme activities may not be suitable - you will need medical advice.
- As with any condition affecting the blood, it is healthier not to smoke.

Other complications

There are some other conditions which seem slightly more common in people with sickle cell trait. However, there is still uncertainty as to how far these conditions are linked to sickle cell trait. Possibly, you may be more prone (compared with people without the trait) to:

- Blood in the urine (haematuria) - this may be noticeable, or in a tiny quantity found only on urine tests. It may affect about 1 in 50 people with sickle cell trait. If you have blood in the urine, it should be investigated (tests done) to see if there is any particular cause. Treatment is needed in some cases.
- A form of kidney cancer. If you develop weight loss, loin pain, fever, and/or tummy pain, along with blood in the urine, always see a doctor straightaway. You should be referred urgently for investigation.
- Bladder or kidney infections (urinary tract infections).

- A blood clot in the leg or lung (deep vein thrombosis (DVT) or pulmonary embolus).

To help prevent these problems, advice is to avoid dehydration by drinking enough fluid. To prevent a DVT or a pulmonary embolus, keep mobile and move your legs when on a long journey or flight. See separate leaflets called Deep Vein Thrombosis, Preventing DVT When You Travel and Pulmonary Embolism for more details.

If you have sickle cell trait, you are more at risk of severe complications if you catch malaria. If you visit any area where there is a risk of malaria, it is absolutely essential that you take antimalarial tablets. You must also take precautions to avoid being bitten by mosquitoes.

Symptoms of sickle cell trait in children

Younger children are extremely unlikely to have any symptoms. They will usually be no more prone to becoming ill than children who do not have sickle cell trait. Older children and young people who do heavy physical exercise may have the same complications as adults (see above).