



SICKLE CELL DISEASE

What are sickle cells?

- Red blood cells carry oxygen through the body. Normal red blood cells are round and soft, and flow easily in the blood vessels. They are shaped like this: 
- Some people have red blood cells which are hard and pointy, and shaped like a sickle or a crescent: 
- Sickle shaped cells can stick together and block small blood vessels, if the person has an infection, becomes very cold, or is dehydrated. When the blood vessels are blocked, less oxygen reaches that part of the body. Blocked blood vessels can cause pain, infection and organ damage. This is most serious when the lungs and brain are affected.

Sickle cell trait and sickle cell disease

- The tendency to have sickle shaped cells is passed from parents to children, in the same way that physical appearance is passed on.
- People with both round and sickle shaped red blood cells have *sickle cell trait*. They are healthy.
- People with only sickle shaped red blood cells have *sickle cell disease*. They may have problems with their health.

When both parents have *sickle cell trait* their children may have:

- Normal round red blood cells, or
- Sickle cell trait (with round and sickle shaped red blood cells), or
- sickle cell disease (with sickle shaped red blood cells).

How is sickle cell disease diagnosed?

A simple blood test can tell if you have sickle cell disease or sickle cell trait.

What are the health problems associated with sickle cell disease?

The health problems associated with sickle cell disease include:

- Anaemia, where the person looks pale, feels weak, and may have a faster heart beat than usual
- Jaundice, where there is yellowing of the skin and whites of the eyes, and possibly abdominal pain
- Pain, usually in the hands, arms, legs, chest and stomach
- Chest pain, coughing, and difficulty breathing
- Frequent infections
- Poor appetite and failure to grow and thrive
- Painful erections of the penis which can occur even in young boys and last a long time
- Difficulties with vision
- Headaches, seizures (fits), weakness of the arms and legs, speech problems, a facial droop, and loss of consciousness, if oxygen flow to the brain is reduced.

How is sickle cell disease managed?

There is no cure for sickle cell disease, and treatment continues for life. People with sickle cell disease may need to take medicines every day, even if they are well. Treatment may include:

- Vaccinations against certain infections
- Antibiotics to prevent infection
- Folic acid to improve production of red blood cells.

Caring for a person with sickle cell disease

People with sickle cell disease feel better if they:

- avoid becoming too cold or too hot
- drink lots of water – aim for 8 glasses a day
- eat healthy food
- get lots of rest and lots of sleep
- have regular medical checkups.

Adults with sickle cell disease can improve their health if they:

- limit the amount of alcohol they drink
- quit smoking
- talk to their family doctor if planning a pregnancy.

When to go to the Emergency Department

People with sickle cell disease should be taken to the Emergency Department of the nearest hospital if they have:

- Fever (temperature of 38° Celsius or higher)
- Jaundice (yellowing of the skin and eyes)
- Sudden paleness (check the palms of the hands)
- Weakness or inability to move a part of the body (such as an arm or a leg)
- Difficulty walking
- Unusual behaviour or confusion
- Loss of consciousness or a seizure (fit)
- Diarrhoea or vomiting
- Chest pain or difficulty breathing
- Severe pain, including painful erections in males
- A sudden change in vision.

When you arrive at the hospital, tell the staff that the person has sickle cell disease.

Further information is available from