

I NEED TO KNOW ABOUT SICKLE CELL

Transfusion Fact Sheet Volume 4, Number 16
By Carolyn Wilson

What is sickle cell disease?

Sickle cell disease is a condition which causes the body to make abnormal haemoglobin. Haemoglobin is the protein in red blood cells responsible for carrying oxygen to all parts of the body (see our Fact Sheet *What Happens When I Need a Blood Transfusion?*, Vol 3, No 7,). This abnormal haemoglobin affects the shape of the red blood cells causing them to be crescent or sickle shaped.

What are the effects of sickle cells in the blood?

The sickle shape of red blood cells makes them inflexible and sticky. They can get stuck in small blood vessels blocking the flow of blood, depriving the tissues of oxygen and causing pain and organ damage. A severe attack of pain is known as a sickle cell crisis.

The abnormal red blood cells can get stuck in the spleen making people with this disease more prone to some infections. Sickle cells also don't last as long in the circulation as normal red blood cells. This leads to a lower haemoglobin level called 'sickle cell anaemia'. A patient may become pale, easily tired, short of breath and feel unwell as a result of reduced oxygen being carried around the body.

Can I catch sickle cell disease?

No. Sickle cell disease is a genetic disorder. People with sickle cell disease carry two genes (one from each parent) for the sickle haemoglobin. If a person has only one sickle gene, that person is a 'carrier' and has sickle cell trait.

How common is sickle cell disease?

This is an uncommon condition in Australia. It is most common among people of Asian, African, Caribbean, Mediterranean or Middle Eastern origin.

How is sickle cell diagnosed?

A special blood test can tell whether a person has sickle cell disease or is a healthy carrier.

What treatments are available?

People with sickle cell disease may need transfusions to increase their red cell counts. A medication called hydroxyurea



may also help improve haemoglobin levels. Giving oxygen and pain relief for some sickle cell crises is very important.

Can people with sickle cell donate blood?

A person with sickle cell disease is not eligible to donate blood.

BLOOD FACT

Sickle cell carriers only have one tenth the risk of dying from malaria infection.