Haematology Department

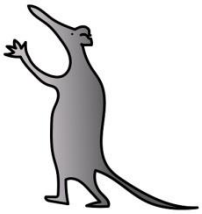
Living with Sickle Cell Disease – What is Sickle Cell Disease

Information for parents and carers

Introduction

This leaflet aims to provide you with general information about living with Sickle Cell Disease. If you are ever worried about your child please contact your Sickle Cell Team or take your child to the Accident and Emergency Department.

What is Sickle Cell Disease?



Sickle Cell Disease is a condition that affects the shape and function of red blood cells. It is inherited, which means that it is passed from a parent to their child.

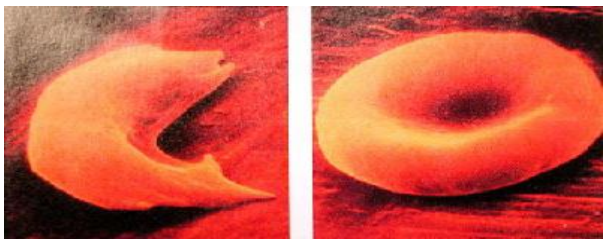
What is the function of a red blood cell?

Red blood cells transport the oxygen around the body. The red blood cells pick up oxygen from the lungs and move it around the body. When the red blood cells reach the tissues and muscles oxygen is released and waste gases such as carbon dioxide are collected and taken back to the lungs to be breathed out. The red blood cell then collects more oxygen from the lungs and goes around the body again.

Why are sickle red blood cells different?

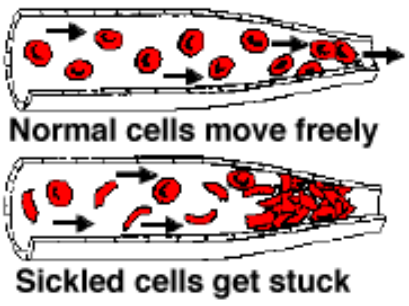
Usual red blood cells are round and flexible and contain normal haemoglobin (HbA). They can flow easily through blood vessels. The red blood cells of people with sickle cell disease contain haemoglobin called sickle haemoglobin (HbS). In sickle cell disease the red blood cell can change shape. This change in shape usually occurs when there is a lack of oxygen in the blood. This can be caused by reasons such as infection, dehydration, cold and trauma, known as trigger factors. Trigger factors are explained fully in the Trigger Factor leaflet.

Sickle red blood cell →



← Usual red blood cell

When the red blood cells become sickle shaped and rigid, it causes them not to flow freely through the veins and tissues. They get trapped and stop oxygen getting to where it is needed. This is known as a crisis or sickling.

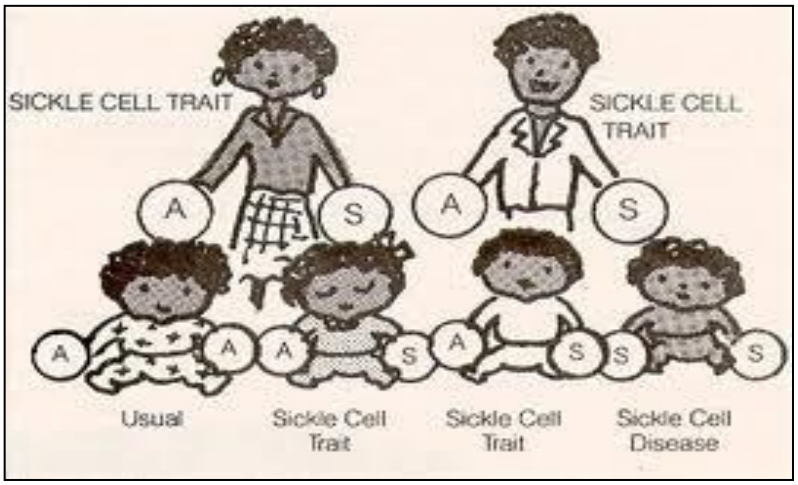


As blood flows throughout the body, a sickle cell crisis can occur anywhere, causing pain, anaemia (a drop in the haemoglobin) and damage to parts of the body affected by the crisis.

How did my child get sickle cell disease?

Sickle cell disease is a genetic or inherited condition, this means it can be passed down from parents to their children. We each carry two copies of the gene that makes up our haemoglobin. A gene is the code that makes up the body, for example the colour of your eyes or hair. We inherit one set of genes from our mother and one set from our father. People can have an unusual gene and it not cause any problems. These people are known as carriers or having a trait. It is when a person has two unusual genes that problems can occur.

This is the case in sickle cell disease. If two people who both carry the sickle cell gene (HbAS) have a baby and the baby inherits one HbS gene from their mum and one from their dad they will have sickle cell disease. Not every child from a couple that are sickle cell carriers will have sickle cell disease. There is a one in four chance every pregnancy that the child will have sickle cell disease or usual haemoglobin (HbA) and a one in two chance that the child will be a carrier.



The diagram above shows a mum and dad who do not have sickle cell disease but are both carriers.

Are there different types of sickle cell disease?

There are different genes that cause sickle cell disease. The most common gene is HbS but there are other genes that can interact with the HbS gene to give sickle cell disease. These genes are HbC, HbE, HbO, HbD and β Thalassaemia.

Why is it important to know about the sickle cell gene?

If you have a child with sickle cell disease then you and their other parent will both be carriers. To find out which unusual gene you carry, you will have to have a blood test.

If you change your partner it is important to know if they carry an unusual gene to see if there is a risk to any future children.

As sickle cell is inherited, you will have inherited the unusual gene from one of you parents. If you have brothers or sisters it is important that they get tested to see if they carry the unusual gene.

Can my child give their children sickle cell disease?

Your child has two unusual genes so will always pass one of these genes onto their child. If their partner does not have an unusual gene then all their children will be carriers.

If their partners have an unusual gene then there is a one in two chance (50:50) that their child will have sickle cell disease.

Further information

If you have any questions or want any further information please contact your Sickle Cell Team on 0151 252 5079.

Useful websites

www.alderhey.co.uk
www.sicklecellsociety.co.uk
www.sct.screening.nhs.uk

Alder Hey Children's NHS Foundation Trust is neither liable for the contents of any external internet site listed, nor does it endorse any commercial product or service mentioned or advised on any of the sites.

This leaflet only gives general information. You must always discuss the individual treatment of your child with the appropriate member of staff. Do not rely on this leaflet alone for information about your child's treatment.

This information can be made available in other languages and formats if requested.

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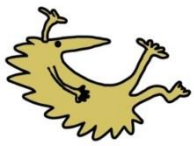

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