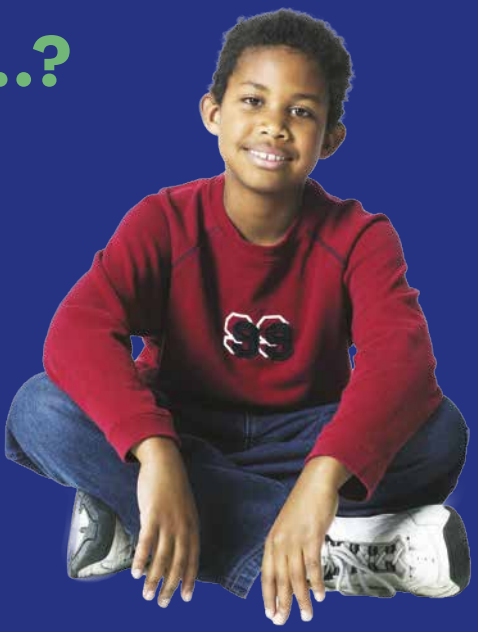
SICKLE CELL SOCIETY

Did You Know?

About sickle cell 11—16

Did You Know...?



Contents

Did You Know...? has been put together for you to understand more about sickle cell. Lots of young people ages 11 to 16 have given their ideas and thoughts on what should be included in this guide. It is important to know about sickle cell, what happens, how to cope and where to get help.

We hope Sickle Cell Did you Know? will help you to learn more about your condition and encourage you to ask questions.

What is Sickle Cell?

Sickle Cell Disorders are a group of inherited blood disorders. You may have heard different terms to describe a Sickle Cell Disorder:

- **Haemoglobin Disorders**
- **Sickle Cell Disorder**
- **Haemoglobinbopathies**

Where does the name sickle cell come from?

The sickle shaped red blood cell is named after a farmer's tool called a sickle that is used to cut grass and hay.

Genes: determine the inherited characteristics parents pass on to their children: height, features, hair colour or eye colour

Haemoglobin: If you have a Sickle Cell Disorder it is your haemoglobin that is affected.

Q: What is haemoglobin?

A: Haemoglobin is made up of two parts. One part called Haem is made up of iron. The other part is called Globin, this is protein that joins to the Haem, it contains alpha and beta chains.

Q: What does this mean?

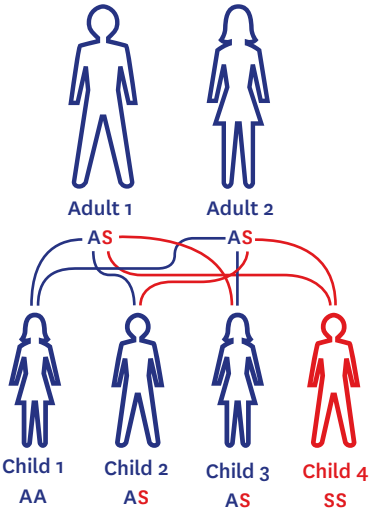
A: If you have sickle cell the beta chain is affected.

Hb is short for haemoglobin

Haemoglobin is a protein in the red blood cells. The haemoglobin's task is to carry oxygen to all the tiny cells of the body that make up organs. All cells of the body need oxygen. Haemoglobin gives blood its red colour when it is carrying oxygen from the lungs. There are lots of different haemoglobin types found in people around the world. Everybody inherits one haemoglobin gene from their mum and one gene from their dad. Haemoglobin A is the most common usual haemoglobin type. Some people have unusual haemoglobin such as sickle haemoglobin C or any other haemoglobin.

Sickle Cell Trait

Sickle Cell Trait means that a person has inherited one unusual haemoglobin S gene and one usual haemoglobin A gene from their parents AS. A person who carries the trait will not experience a crisis in normal circumstances



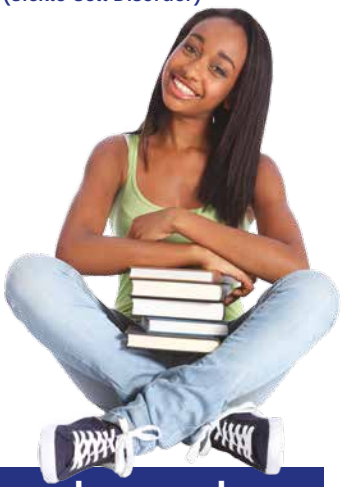
AS = Sickle Cell Trait - No disease
AA = Normal Adult Hemoglobin
SS = Sickle Cell Anemia
(Sickle Cell Disorder)

Red Blood Cells

Red blood cells are made in the bone marrow in the centre of long bones and the chest bone (sternum), skull bone (cranium) and pelvis

Normal red blood cells usually live for 120 days and are then cleaned up and destroyed by the spleen. The spleen is the size of a fist, it helps fight infection and sits alongside the stomach.

Normal red blood cells are soft and tiny and are able to squeeze through tiny blood vessels, they look like a soft doughnut.



Q: So what is the difference in someone with a sickle cell disorder?

A: The red blood cells change shape and are not soft and stick a little bit more to the blood vessels.

If the sickle shaped red blood cell cannot travel through the tiny blood vessels very easily oxygen cannot get to the cells, tissues and organs of the body that need it. When the cells of the body don't receive enough oxygen they can cause complications.

Pain (Crisis)

Not enough oxygen in the cells of your body can cause a pain, which is known as crisis. There are some things you are probably aware of but may not realise that can cause this shortage.

Too much Exercise: Shortage of oxygen during exercise can cause the red blood cells to sickle.

Stress: Everyday stress from things like exams can cause red blood cells to sickle.

Infections: Infections can cause red blood cells to sickle.

Cold Weather: Blood vessels become narrow in the cold making it hard for cells to flow, which can lead to a crisis.

Not Drinking Enough: When you don't drink enough water the blood vessels become thick and sticky and the red blood cells cannot travel through the blood vessels very easily.

Be active but know your limits

'Stress is anything that makes you feel worried, angry or unhappy' Jasmin aged 14

Pain (Crisis)

Pain could be your most common problem and something you know lots about. Everybody experiences pain differently. You probably have your own ways of coping and things you do when you feel pain in different parts of your body.

Painkillers are a common way to relieve pain but... there are other things you can try too.

'Sickle pain is different to other types of pain'
Alisha aged 13

Have you tried?

- Warm bath
- Massage
- Relaxing
- Drinking plenty

Take Control

If you think about your pain you will feel it more. Taking your mind off the pain can help. You could try:

- Reading
- Listening to music
- Hot water bottle
- Eating food you like
- Phoning a friend
- Watching TV
- Sleeping
- Think of a place that means a lot to you
- Playing computer games

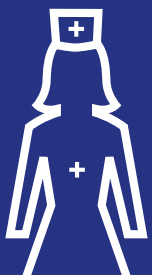
Manage your pain

Thoughts can change the way you feel pain. Try relaxing and thinking of somewhere you would like to visit on holiday maybe a beach with the sun beating down.

Pain Killers

There are different types of pain killers. There are some painkillers you take at home like paracetamol, ibuprofen and codeine. If you have a crisis and go into hospital you may have painkillers intravenously*, these are usually morphine or pethidine.

Write down any questions you have.
It is OK to ask your doctor or nurses questions



*Intravenous means given directly into the veins

Some Complications

When you have a crisis your doctor or nurse may use words that you do not understand. This section explains some complications.

Anaemia

Q: When I was in hospital my friend in the next bed had anaemia, what is it?

A: Anaemia is when there is not enough red blood cells or haemoglobin. It will make you feel tired. It is very common for people with a sickle cell disorder.

Stroke

A stroke is caused by sickling of the red blood cells in the blood vessels of the brain. Doctors and nurses watch for this very closely.

Priapism

Priapism is a painful erection of the penis. Don't be afraid to tell someone if you have this or if you have been passing urine a lot more often or it is painful when you pass urine. It could indicate an infection that caused the priapism.

Chest Syndrome

Sometimes a crisis occurs in the lungs and blood builds up that can cause breathing problems.

Jaundice

Some of you may have had jaundice. You will notice sometimes the whites of the eyes turn a yellow colour. Jaundice occurs when red blood cells come to the end of their life. Sickle red blood cells have a shorter life and produce lots of bile when they are cleaned up, this causes the jaundice.

Bedwetting

Bedwetting is known as enuresis. Some of you may experience this frequently even in your teenage years. With sickle cell disorders the urine is very dilute. Don't worry about this, it does pass. There is a lot of help available. Don't stop drinking plenty.

Managing Sickle Cell Disorders

Usually when you are admitted to hospital you will have different treatments. It is normal to have a drip, antibiotics, painkillers and vitamins.

This section of 'Sickle Cell Did You Know...?' explains some of the treatments

Blood Transfusions

Sometimes because of a bad crisis the doctor may decide to give you a blood transfusion. A blood transfusion may be necessary for a number of different reasons: Worsening of anaemia, Reduce sickle haemoglobin, Prevent frequent strokes. Having regular blood transfusions can mean after a while the body builds up iron. This is known as iron overload. The body has no natural way of getting rid of the iron.

Q: So what is the problem? We need iron don't we?

A: Yes, we all need iron, but too much can damage the body and lead to serious problems, such as iron overload.

Q: What can be done about this extra iron? It needs to be gotten rid of if it damages the body doesn't it?

A: Yes. There is a drug called Desferrioxamine*, it is an iron chelator, it removes the extra iron from the body. Exjade (Deferasirox) is another treatment, taken daily. It is an oral iron chelator developed for treating iron overload.

Q: But having the pump for eight hours a day sounds really uncomfortable!

* Desferrioxamine is also called desferal and iron chelation therapy Desferal can be given in two ways:At the same time as the blood is given through the drip. Through a pump. A needle is placed under the skin and an electronic pump gives the desferrioxamine continuously. This is usually for eight hours a day a few days a week. The desferrioxamine is given in these ways as it works best given continuously, not just one injection.

A: It can be uncomfortable and noisy. Many young people are worried about having desferrioxamine through a pump. This is understandable. But remember it does help.

Hydroxyurea

Hydroxyurea is a drug you may have heard about or take yourself. It is sometimes given to young people who experience severe disorders. It can help reduce the number of crisis and how bad they are, it is taken in the form of a tablet.

Q: This sounds great, why am I not taking it?

A: It is usually only given to people whose Sickle Cell is quite bad.

Bone Marrow Transplant

Bone marrow transplant involves having a donor. A donor is a volunteer that gives some of their bone marrow to someone who needs it. Bone marrow is a fluid in the middle of the bone that makes all your blood cells which are red blood cells, white blood cells and platelets. The marrow is taken from the donor and given to the person who needs it in a drip.

It is OK to be worried. Talking to someone can help. Ask questions if you are not sure about

Staying Healthy

Everybody needs to stay healthy. There are lots of things people can do to help them stay healthy. If you have sickle cell staying healthy is even more important. Being healthy can make you feel fit and well and give you more energy. You are also less likely to get infections. There are lots of different things you can do to help yourself stay healthy.

- Eat well
- Exercise – remember to drink more when exercising
- Rest and relax
- Drink lots (don't drink fizzy drinks too often)
- Keep warm in cold weather- you can be fashionable and dress warmly
- Keep cool in hot weather – remember to drink more

Try relaxing

lie down, close your eyes, tense all the muscles in your body (not where you have pain) starting with your head, screw your face up and tense all your body your arms and hands, your legs down to your toes. Hold your body like this for a few seconds and then relax. Practice this when you feel stressed (remember, do not tense your muscles where you feel pain.

Try to eat fruit and vegetables everyday, they contain lots of vitamins and minerals that will help keep you healthy

Below are some medicines you may take every day to help stay healthy

Penicillin

Penicillin is an antibiotic that fights infections caused by bacteria. As infections can cause a crisis most of you will take penicillin every day, this is called prophylaxis.

Prophylaxis means guarding against disease

Folic Acid

Most of you will take folic acid every day. Folic acid is a vitamin that helps treat and prevent anaemia. It is needed because your body uses up folic acid when it is making new red blood cells.

Fruit and vegetables contain folic acid

School

- School can be stressful whatever age you are. You may have different feelings about school.
- You may miss school when you are ill and this could worry you.
- Sometimes it can be hard to catch up with work.
- You could feel left out from your friends.
- Your teachers and friends may not understand what a Sickle Cell Disorder is or how it can affect you.
- You may not always be able to join in with sports.

There are some things you can do to help your teachers and friends learn about Sickle Cell Disorder

- You could give them a booklet explaining about sickle cell
- You could ask your nurse counsellor to talk to the school
- You could let them know about the Sickle Cell Society web site – www.sicklecellsociety.org

To help you when you are away from school:

- You can ask the school to send work to your home
- You can ask the support of your friends when you are away from school
- You could ask friends to collect any homework you may be missing

Join a local young persons' Sickle Cell support group

Think positive, feel positive

Quick Quiz

Here is the quick quiz some of you wanted. Answer true or false to the question below and see how you do

Q1 Hb is short for Haemoglobin? TRUE/FALSE

Q2 Haemoglobin is a protein found in red blood cells? TRUE/FALSE

Q3 Everybody inherits two haemoglobin genes, one from mum and one from dad? TRUE/FALSE

Q4 In sickle cell disorders shortage of oxygen can turn red blood cells into a sickle shape? TRUE/FALSE

Q5 It is good to be active? TRUE/FALSE

Q7 It is helpful to ask questions? TRUE/FALSE

Visit our website for answers **www.sicklecellsociety.org**

Thank you to all the young people who took part in advising how this booklet should be produced.

Remember if there are any questions you have or something you are not sure about just ask!

'I think we should know what is going on in our bodies'
Sarah aged 15

'It is good to know how we can stay healthy'
Olu aged 14

'I want to understand what the doctors are saying'
Steffi aged 12

'I want to take control'
Elijah aged 15

'If we know about sickle cell we can explain it to others that don't'
Dominic aged 13

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SICKLE CELL SOCIETY

Information, Counselling Caring for those with Sickle Cell Disorders and their families

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www.sicklecellsociety.org

Brent

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Quick Quiz answers: All the answers are true!