North East & Yorkshire Haemoglobinopathy Coordinating Centre

Sickle Cell Disorder for Adults

Information for patients, families and carers

Haematology - Haemoglobinopathy Coordinating Centre

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Sickle Cell Disorder (SCD) is an hereditary disorder of the red blood cells (RBC) which contain abnormal haemoglobin known as sickle haemoglobin (HbS). People are born with SCD, it is not contagious and is inherited from both parents each having passed on the gene for SCD.

There are different types of genotypes and these are determined by the types of genotypes, these are determined by the type of gene that is inherited.

Haemoglobin SS	HbSS	• Inheriting 2 HbS genes • One from each parent
Haemoglobin SC	HbSC	• One HbS gene and one HbC gene • One sickle gene from one parent and one abnormal HbC from the other parent
Haemoglobin (HbS) (beta) Thalassaemia Sickle Sß $^{+}$	HbS $ß^{+}$ thal	• One HbS gene from one parent and one gene for ß+ thalassaemia from the other parent
Haemoglobin Sß ^{0} (Beta-zero) thalassaemia	.HbS $ß^{0}$ thal	• One HbS gene from one parent and one Hb beta - zero thal gene from the other parent

Haemoglobin (HbS) with herediatry persistence of fetal haemoglobin	HbS/HPFH	• One HbS gene from one parent and high fetal haemoglobin levels (HbF) from the other parent. This is different to someone with HbSS and HPFH.

Sickle cell disorder may be suspected clinically if a patient from an at risk ethnic group presents with clinical signs that may suggest a painful crisis or acute complications of sickle cell. It can effect anyone, although it predominately affects people from African and Caribbean backgrounds. However Ethnicity is not always a marker of who may be at risk and a large majority of diagnosis are made as a result of the neonatal bloodspot screening programme in England.

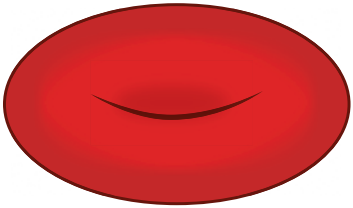
Diagnosis

Even if you have a diagnosis already, the hospital team will always take a blood test to confirm diagnosis is correct and the type of genotype. This will be requested at your first initial presentation to the hospital either in our red cell clinic or as an inpatient, however the GP can also conduct these especially if you wish for a partner to get tested too.

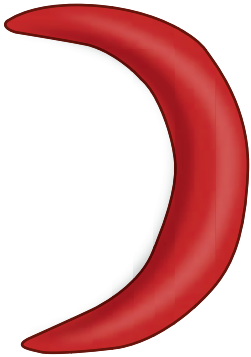
What is sickle cell?

- Haemoglobin is the substance that gives blood its red colour
- It is a protein which carries oxygen around the body.
- Normal red blood cells are disc shaped, like a donut without a hole.
- Sickle haemoglobin HbS is not like normal Hb it forms stiff rods within the red cell, changing it to a crescent or sickle shaped.

- They are not flexible and can stick in small vessels causing a blockage that slows or stops the flow of blood and when this happens Oxygen can't reach nearby tissues and these cause painful episodes which are known as a sickle cell crisis.
- The main symptoms of sickle cell disorder are episodes of pain.



Normal cell



Sickle cell

An acute painful sickle cell crisis is a medical emergency and may require immediate hospital admission if:

- **High temperature of 38 degrees and above on two separate occasions- fever**
- **Pain unresolved by paracetamol, ibuprofen and codeine**
- **Respiratory symptoms- breathless, unable to breathe**
- **Priapism- a persistent and painful erection of the penis**
- **Neurological symptoms – stroke**
- **Dehydration associated with diarrhoea and vomiting**

Treatment for you whilst you are in hospital may include stronger analgesia such as opioids i.e Oxycodone or Morphine to manage your pain, intravenous or oral antibiotics, intravenous fluids if unable to or

cannot manage oral and oxygen therapy if oxygen saturation levels below 95% on air.

You may need a blood transfusion or red cell exchange if clinically decided by the medical team on review which would be discussed with you beforehand.

What can trigger a crisis?

- Heat or cold- be aware of changes in temperature and try and take this into account
- Physical or psychological stress- listen to your body with exercise and only workout to a comfortable level. If you are struggling with your mental health please talk to us and we can refer you to our psychology service.
- Infection
- Hypoxia
- Drugs
- Dehydration- try to keep a bottle of water on you when out and about
- Alcohol
- Tobacco products
- Pregnancy- please let the team know if you are pregnant so we can provide you with extra support and complete a birth management plan.
- Fatigue- this could also be a sign of anaemia or vitamin deficiency, if tiredness lasts for long duration please consult the red cell team or GP.
- Hypoxemia- low blood oxygen

Symptoms and complications of sickle cell

- Pain
- Breathlessness
- Acute chest syndrome
- Chronic anaemia- feeling weak and fatigued
- Priapism- persistent, painful erection of the penis
- Increased risk of infections
- Organ damage- over time you may experience damage to your organs such as liver, kidney, lungs, heart and spleen.
- Tissue damage
- Eye problems- Retinopathy
- Dactylitis- swelling of hands and feet
- Leg ulcers- wounds or sores
- Shortened life expectancy

For more information please see the keeping well with sickle cell disease leaflet.

Managing sickle cell

- Antibiotics - used to help prevent infections, you may be asked if you would like a supply of prophylactic antibiotics which is commonly Penicillin (or erythromycin if allergic to penicillin) this is a supply to be taken everyday and is used as a precaution to prevent, rather than treat an infection. If you do not want to take antibiotics every day, you may be prescribed some to take as and when needed. However, you would still need to come in for a review.
- Immunisations- it is important these are kept up date, your GP should have a record of your vaccination history
- Pain killers- mild to moderate painful episodes may be managed with over-the-counter pain relief such as paracetamol and ibuprofen (unless you have a history of stomach ulcers) regularly until the episode is over. For severe pain it may be necessary for you to go to the hospital for a review and/or admission as stronger pain killers may be needed.
- Blood transfusions or red cell exchanges
- Hydroxycarbamide – Is a daily, oral medication given to people with sickle cell disorder to reduce short- and long-term complications of the condition. There is further guidance on this in our Hydroxycarbamide patient information leaflet or you can contact the clinical nurse specialist team for further information and support.

Living with a chronic health condition can be difficult at times and can affect you physically and psychologically, as your healthcare provider we can provide support and guidance with this which can include support letters for employment or referral to our services such as psychology and or our benefits advisor.

There is further information on this in the main haemoglobinopathy patient information leaflet.

A guide for employers and employees on work, employment and sickle cell disorder (SCD) can be found at:

http://sicklecellwork.dmu.ac.uk

One possible cure for the disorder is bone marrow transplant but it is only available for a limited number of sickle cell patients who have a suitable donor. Gene therapy is another potential cure for sickle cell disorder, it is estimated that around 50 patients, suitable for a stem cell transplant but without a matched donor, will receive gene therapy each year.

Use this space to make notes or record any questions you may have

Sickle cell trait

Sickle cell trait is inherited when one HbS gene is inherited from one parent, it will not develop into sickle cell disorder. It is usually found in 1 in 4 West Africans and 1 in 10 Afro- Caribbeans however it is also found in people who originate from the Mediterranean, Asia and the middle east.

The red blood cells (RBC's) in sickle cell trait are mostly normal with a round donut shape with some sickle shaped cells appearing in certain circumstances.

Most people with the trait are healthy and usually have no symptoms. You may experience pain at high altitudes (generally above 10,000 feet) including long-haul flying in unpressurised planes and mountain climbing. If undertaking any of these activities it is important to notify the company as you may need oxygen. Extreme exercise may also precipitate problems, it is advised that if you are a professional athlete your training programme should take this into consideration.

It is also important to inform the dentist or doctor before any treatment is given as a precaution especially as it is reported that anaesthetics can cause problems.

Things to consider

The trait is not an illness, however there are some considerations that need to be taken into account:

- You do not have symptoms from sickle cell trait, so it is a good idea for you and your partner to have blood tests so your sickle cell status is confirmed. This can help you plan for the future when starting a family. This can be conducted your GP.
- If you and your partner both have the trait, there is a 25% chance that any child conceived may have sickle cell disorder and a 50% chance that they will have the trait.
- If your partner does not have the trait, any children you have, have a 50% chance of having the sickle cell trait however will not have sickle cell disorder.

For more information and guidance you can find this at the Sickle Cell Society which provides support, advice and information to people with SCD and their families. Telephone number: **020 8961 7795** website: **www.sicklecellsociety.org**

If you have any further questions please speak to your specialist nurse or doctor for further information.

Please use the QR code or link below to take you to the Patient Information Leaflet (PIL) section of the North East and Yorkshire Haemoglobinopathy Co-ordinating Centre Website

Patient Information - NEYHCC

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