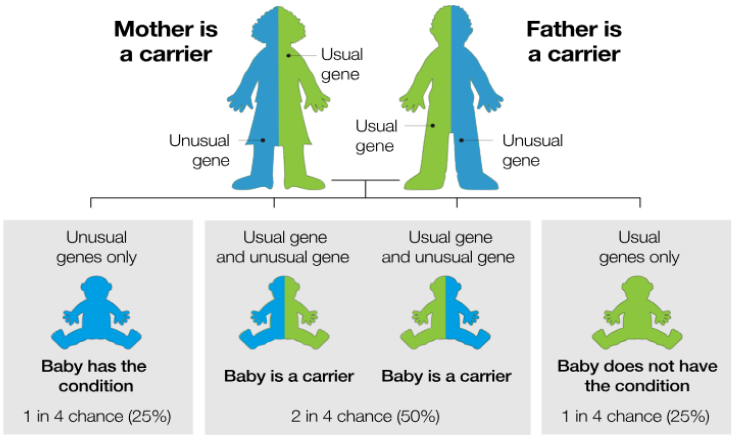
Whittington Health NHS Trust

What is Sickle Cell Disease

Patient information factsheet

- Sickle cell disease is an inherited blood condition.
- It is now the most common genetic condition in England, affecting about 15,000 – 18,000 people, mainly of African or Caribbean origin.
- To be born with sickle cell disease, a child would have to inherit a copy of the sickle cell gene from both their parents. If both parents are carriers of the sickle cell gene, the chance that their child will have SCD is one in four (25%).



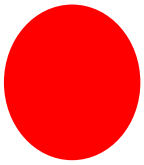
- There are various types of sickle cell disorders:
    - Sickle cell anaemia (Hb SS)
    - Haemoglobin SC disease (Hb SC)
    - Sickle beta thalassaemia (Hb S beta thalassaemia)
    - Haemoglobin SD disease (Hb SD)
- All sickle cell disease types can cause painful crises, but sickle cell anaemia HbSS is the most common. The disorder affects the red blood cells that contain a special protein called haemoglobin (Hb for short). The function of haemoglobin is to carry oxygen from the lungs to all parts of the body. When sickle haemoglobin gives up its oxygen to the tissues, it sticks together to form ‘long rods’ inside the red blood cells, making them rigid (sickle/half-moon shape), hence the name sickle cell.
- Normal red blood cells can bend and flex easily, however sickled red blood cells cannot squeeze through small vessels as easily, and this can cause blockage of the small blood vessels virtually anywhere in the body, stopping the oxygen from getting through to where it is needed. This in turn can lead to severe pain and damage to organs.

Sickle shaped red blood cell

- rigid / sticky
- tend to stick together
- Will block a blood vessel

= **PAIN**

Normal red blood cell



- flexible / soft
- can squeeze through small blood vessels
- Do not block blood vessels

What causes the red blood cells to sickle?

It is known that certain things can contribute/trigger off increased sickling, for example:

- Dehydration (reduced water in the body)
- Infection
- Sudden changes in body temperature due to cold / hot conditions
- Physical exertion / activity
- Increased lack of oxygen
- Stress

How will this affect me?

PAIN:

- Painful crisis can occur at any time and can be mild, moderate, or severe. It can affect any part of the body and can last from a few hours to many days.
- It may be advisable to attend the hospital if your pain is not controlled with your usual painkillers, so you can be given stronger pain medication.

INFECTIONS

- All people with sickle cell, whatever the type, are at risk of developing severe infections. This can range from mild infections, such as a cold, to potentially life-threatening infections like meningitis. This is because your spleen, which helps your body fight infections, does not work as efficiently as it should do.
- Penicillin, when taken as a prophylactic antibiotic twice a day, and vaccinations help reduce the risk to many infections. Taking Penicillin will not treat pain, and forgetting a dose does not give rise to pain, but it is very important in the management of sickle cell disorders.
- It is a good idea to have a thermometer at home to take your temperature if you are unwell. If it is 38 degrees centigrade and above, you need to seek medical advice.
- If admitted to hospital, you might need a different antibiotic to treat the infection.

ANAEMIA

- The red cells containing the sickle haemoglobin do not survive in the blood circulation as long as normal haemoglobin. Over time this will cause anaemia, as the body cannot make enough haemoglobin. This will cause you to become pale and at times tired.
- You may also find that you become short of breath quite quickly when you exercise or walk a long distance. This does not usually cause you any problems, as the body adjusts itself to this. Sometimes you may have to allow your body to rest a bit more than usual to make sure that you do not trigger off a sickle crisis.

JAUNDICE

- Yellowing of the whites of the eyes is common and is caused by a fluid called bilirubin that is formed when red blood cells break down in the body. The liver clears the bilirubin from the body, but if there is a lot of bilirubin, the liver may not be able to clear all the bilirubin away and the yellowing of the eyes may appear.
- The condition is normal, and you may notice more yellow at times than others.
- Sometimes your urine may be dark, this is for the same reasons and is not a problem.

'CHEST SYNDROME'

- Pain in the chest or difficulties in breathing may be due to sickling in the chest/lungs or severe infection. This is one of the most serious complications of sickle cell and requires urgent medical attention.
- It is strongly advisable when unwell with pains or infection that you do regular deep breathing exercises to try and prevent 'chest syndrome 'developing.

ENLARGED SPLEEN

- The spleen is an organ that lies on the left side of the tummy under the rib cage. The spleen helps to filter organism and damaged blood cells from our body.
- Sometimes, this gets jammed / blocked with sickle blood, resulting in sudden enlargement.
- This is most common in children under five years of age and could be fatal.

What other problems may I have?

- **Delayed growth:** Some people be smaller or shorter than their peers, but they will catch up later. Puberty may also be delayed but this is of no concern. Where this does not resolve by itself, it will be investigated further, and a referral will be made to the endocrine team.
- **Frequency of urine:** The need to pass urine more frequently is caused by the kidneys not concentrating the urine fully. You should not reduce your fluid intake because of this, as dehydration will cause sickling to happen. Bedwetting at night is also not uncommon: if this is a concern, please inform the team in clinic.
- **Gallstones:** These are small stones that can develop in the gallbladder, which can sometimes cause upper abdominal (stomach) pains. The stones are formed from the same bilirubin pigmentation that makes the eyes go yellow. If they start to cause you problems, then you may need to have your gallbladder removed.

- **Priapism:** A hard painful erection happens when the red blood cells block the blood vessels of the penis. Going to the toilet to pass urine and emptying the bladder often helps, as well as a warm bath or shower and gently walking, to help improve blood circulation. Painkillers are normally required. If the painful erection lasts for more than one hour, you should be seen at the hospital as a delay may cause problems with impotency later in life. Do not use ice or cold water to relieve pain, as this will cause more pain due to the blood vessels narrowing, and further sickling could happen.

How do I manage my sickle cell?

- We know that certain things can trigger off sickling pain as discussed above. The main thing therefore is to remain healthy and avoid the known trigger factors as much as possible. You need to drink plenty of water, keep warm, rest when you are not feeling your best and eat healthily.
- Apart from this you can do the same as your friends. **However,** it is very important that if you are unwell with a high temperature or pain in your chest/ breathlessness, you seek medical advice as soon as possible.
- **Pain** is probably the worst thing about your sickle cell and as you get older you need to learn how to manage the pain yourself. The best policy is to avoid the known trigger factors to prevent sickling.
- As you get older you will be able to assess your own pain and the best way to manage it.
- The most important thing is to take your painkillers and to drink plenty of fluids to 'flush' the sickle cells through your system.
- Most pain crisis can be managed at home, which is the best place for you to be. The only time you should definitely come to the hospital for assessment is if you have pain in your chest, difficulty in breathing, a high temperature or severe diarrhoea or vomiting and cannot keep any fluids down.
- Pains in your arms, legs, back, stomachs are safe to be managed at home.

Pain medication (analgesia)

- **Mild to moderate pain**

Paracetamol and Ibuprofen, these can be given together and will relieve pain, reduce inflammation around the area and reduce fever.

- **Moderate to severe pain**

Paracetamol and Ibuprofen as above and add Dihydrocodeine (this is slightly stronger and will relieve pain only).

- **Severe pain**

Paracetamol, Ibuprofen and Dihydrocodeine as above
Oramorph is a very strong pain killer and often used in hospital.

Other measures to relieve pain

- It is well reported that alternative measures can help to reduce your pain. These will be different for everyone, but the most used ones are:
    - Warm bath / shower
    - Heat pads / hot water bottle to painful area.
    - Massage to the painful area.
    - Plenty of rest
    - Diversional therapy: for example, listening to your favourite music, watching TV, reading, playing…
- **Remember:** Plenty of fluids, rest and painkillers will relieve your pain, not just painkillers.

Hospital appointments

- It is important that you have regular check-ups at the hospital at least once a year.
- If you have been unwell you may need to go more often. This is to support you with your sickle cell as well as monitor you for any problems that you may have as you get older.
- This will usually involve a routine blood test, any vaccinations due and a routine eye tests every 1 – 2 years if indicated.

Treatments options in sickle cell

- There are different **disease modifying / curative options** such as medication (Hydroxycarbamide), blood transfusions, bone marrow transplants and gene therapy. If you would like to learn more about these please speak to the team looking after you.
- **Healthy Eating / Food and Nutrition:** Healthy eating has been proven to improve general health if you have sickle cell. A nutritional diet and plenty of water are known to reduce the risks of having a crisis. A healthy diet will also reduce the risks of other health problems later in life such as heart disease, obesity and high blood pressure. Current guidelines are:
    - Eat three meals a day with healthy snacks in between, if required.
    - Eat five portions of fruit and vegetables each day.
    - Reduce the amount of fat in your diet, particularly saturated fat.
    - Increase your carbohydrate e.g. bread, pasta, potatoes, rice, yam.
    - Cut down on sugary foods.
    - Physical activity to your ability
- **Fitness** Having sickle cell does not mean that you cannot exercise to keep healthy. Some people with sickle cell find that they can be quite physically active without any problems. However, some people may have some problems with pain following strenuous exercise, so you need to find out what is best for you. Older people with sickle cell have found that regular gentle exercise has improved their general wellbeing, and they feel better and stronger in themselves when they have taken up some form of exercise.

The exercise can be anything from walking to the shops instead of going by car or bus, to dance, yoga or playing in a team.
Sports to avoid however are contact sports, such as rugby as trauma/impact has been known to trigger a pain crisis. Swimming is a good form of exercise, but for some people with sickle cell it can trigger a crisis. If you have been swimming as a child and have not had any problems, then continue but always avoid very cold water and ensure that you always have a warm shower and dry off after getting out especially during the winter.

Contact at Whittington Hospital

Children's clinical nurse specialist: 020 7288 3017

Adult Sickle Cell Specialist Nurse: 020 7288 5035

Useful websites

Sickle Cell Society website https://www.sicklecellsociety.org/

FRANK https://www.talktofrank.com/

Patient advice and liaison service (PALS)

If you have a compliment, complaint or concern please contact our PALS team on 020 7288 5551 or whh-tr.PALS@nhs.net

If you need a large print, audio or translated copy of this leaflet please email whh-tr.patient-information@nhs.net. We will try our best to meet your needs.

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