Whittington Health NHS

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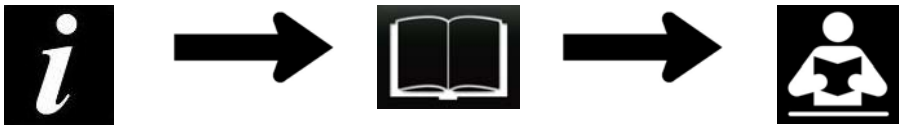
Sickle cell disease, the joint red cell unit

Whittington Health cannot accept responsibility for information provided by external organisations.

If you would like this leaflet in another format, for example large print or audio, or the services of a translator, please contact us on 020 7288 5437. We will do our best to meet your needs.

A patient's guide



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.... caring for you

Introduction

The purpose of this booklet is to provide adult patients who have sickle cell disease (SCD) with a brief guide of the disease. If you would like further information, please ask for our larger patient information leaflet which has details of our sickle cell service in greater depth.

What is sickle cell disease?

Sickle cell disease is an inherited condition of the body's red blood cells that causes lifelong anaemia. It is caused by an abnormality in haemoglobin – the part of red blood cells that carries oxygen round the body - that makes the cells become inflexible and sticky. This in turn makes other blood cells as well as the vessels they travel in sticky too. Blood flow becomes interrupted and there is inflammatory cascade - a process where one inflammatory reaction causes another in turn. This process may cause problems with pain and also problems in the organs where the sickling is happening. It also affects the immune system.

There are problems that happen suddenly (acute problems) such as painful crisis and chest crisis that are emergencies and require urgent medical attention. There are also the long-term effects of crises and background sickling (that may have no symptoms) that are known as chronic complications. The approach we take is to limit and prevent both of these from happening as much as is possible.

Contact details

The Archway Centre,
681-689 Holloway Road, London N19 5SE
Appointments Tel: 020 3317 5252
Website: www.archwaycentre.com

Consultants:
Dr Bernard Davis
Dr Farrukh Shah
Dr Ali Rismani

Nursing Staff:
Ms Matty Asante-Owusu
Ms Emma Prescott
Ms Sarita Kataria

Contact details:
Haematology secretaries:
020 7288 5437 /5144 /5035

Address:
Department of Haematology,
Whittington Health
Magdala Avenue
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Where can I get more information?

Sickle Cell and Thalassaemia Centre
17a Hornsey Street
London N7 8GG
Tel: 020 3316 8853/8854
www.islington.nhs.uk/Your-NHS-services/sickle-cell-and-thalassaemia.htm

The Sickle Cell Society
54 Station Road
London NW10 4UA
Tel: 020 8861 7795/8346
www.sicklecellsociety.org

Travel vaccinations
Please get all the routine vaccinations as recommended for your area of travel (e.g. Meningococcal ACWY and hepatitis A vaccine if travelling to area of increased risk of infection).

Insurance
Get travel insurance that incorporates repatriation costs. Declare your sickle cell disease or your insurance will be invalid. The Sickle Society may be able to provide you with a list of amenable insurers.

Fertility
We offer a range of services to couples presenting with fertility problems. These issues can be discussed in clinic following which we will refer the couple to a dedicated consultant (Miss Amma Kyei-Mensah). Miss Kyei-Mensah will initiate initial investigations and institute referrals as necessary.

Family planning
You may wish to discuss contraception with your GP, local family planning clinic or community sickle cell nurse. You should tell them you have sickle cell disease and any other associated medical problems if you have them. Do let us know of any changes to your medication.

Sexual health
The Archway Centre offers free and confidential a wide range of sexual health services including tests for HIV and other sexually transmitted diseases and contraception. You need to book an appointment to attend the clinic with the exception of walk-in HIV testing clinics where an appointment is not required. Patients can phone the clinic for advice if they have an urgent problem or to find out the opening times.

What kind of problems can happen in sickle cell disease?

- painful crisis – this is the commonest problem
- infection – sickle cell patients are prone to infection because of poor function of the spleen
- gallstones
- joint damage/destruction
- damage to kidneys
- leg ulcers
- visual impairment
- damage to the nervous system which may lead to stroke
- painful sustained erections which can lead to long-term problems with initiating or maintaining erections (Priapism)

Signs and symptoms of crisis may include:

- pain
- fatigue (feeling tired or weak)
- jaundice (yellowing of the white of the eyes)
- paleness of the mucous membranes
- shortness of breath
- dizziness
- headaches
- symptoms of infection, e.g. fever

If you develop any of the following symptoms: high fever, chest pain, shortness of breath, or the pain does not feel like the usual sickle pain, weakness (especially one sided), change in vision or extreme tiredness, we would advise patients to seek medical attention straight away.

Treatment
A simple painful sickle cell crisis may be managed by the patient at home by taking pain killers, resting, drinking fluids and keeping warm. However, if the pain becomes worse you may need stronger painkillers in hospital. In hospital we can help control the pain, treat the underlying cause (trigger) and any complications relating to your condition.

Sometimes the crisis may not be particularly painful or more importantly you may be medically unwell from the start. If you are unwell even if you have no pain, you should always contact us as you may need to come to hospital.

We may also suggest other ways of managing your condition with preventative treatments. We usually recommend this if the frequency of sickle cell crises increases or if an attack was particularly severe. Treatment options include commencing Hydroxyurea or starting a long term blood transfusion programme.

Where to go in an emergency
Please go to emergency department **without delay** if you have any of the following:

- pain that is not controlled with your usual pain killers
- fever (high temperature)
- difficulty in breathing
- problems with your vision
- new weakness particularly if on one side of the body
- if you think you are much more anaemic than usual
- feeling very unwell for any other reason even if it is unrelated to sickle cell

Air travel
Keep mobile (walk around at least every half hour), well hydrated, no alcohol. If you are well and in a pressurised cabin you will not normally need extra oxygen. There is a possible increased risk of splenic infarct (blood blockage in the spleen) from air travel, and this should be considered by your doctor if you have pain in the left upper part of your abdomen.

Surface travel
Drink plenty of fluids, keep mobile, avoid fatigue.

Antibiotics
Take your regular penicillin, if you are penicillin allergic, remember the alternative prescribed by your GP e.g. erythromycin. Ask you G.P. for a treatment course of antibiotics in case needed, however, if you are truly unwell you will need to go and see a doctor without delay. You should seek hospital treatment for dog bites and tick bites. Seek early treatment for other infections, remember that if you have sickle cell disease, your spleen does not work well and therefore you are more susceptible to infections. This risk is decreased with vaccinations and your penicillin but is not eliminated.

Malaria
People with sickle cell are at risk of malaria, even if previously resident in a malarial area. It is important to wear protective clothing, use insect repellent and mosquito nets and to take preventative anti-malarial medication. It may be advisable to start the medicine a little earlier than recommended to check that you are not having any side effects. Take curative anti-malarial medication with you if you are staying far from medical care. You should be aware of your G6PD status as some medications may cause problems in those with G6PD deficiency.

These may include strong painkillers, hydroxyurea (hydroxycarbamide) and iron chelation medication such as desferrioxamine (Desferal)and deferasirox (Exjade) for people with too much iron in their body as a result of transfusions. Other medications should be prescribed by the G.P.

Vaccinations
The following vaccinations are recommended and should be administered at your GP surgery. We ask you to tell us when you have had them to keep your hospital records up to date. We know that some of you may have missed the normal childhood vaccination schedule or may have been vaccinated abroad where the schedule may have been different. We ask that you discuss this with your GP so that they can bring you up to date with any vaccinations you may have missed.

- Pneumovax – once every five years
- Haemophilus influenzae type b (HiB) vaccine: a single dose should be given if not already received
- Conjugated Meningococcal C vaccine: a single dose should be given if not already received. Additionally, Meningococcal ACWY vaccine should be given if you are travelling to tropical countries
- Hepatitis B vaccination – three or four doses if never previously vaccinated. Booster doses every few years will be required
- Influenza vaccine every year

Travel
Travel can lead due to an increased risk of crises due to thrombosis, infection, fatigue, dehydration and climate change.

- If you have a non urgent medical problem e.g. a rash or twisted ankle then it is appropriate for you to go to your GP or local emergency department. They can always discuss your case with us.
- If you have a non urgent problem related to sickle cell, then you can ask for your outpatient appointment to be brought forward.
- Note: if you have an infectious illness, e.g. cough, cold, diarrhoea, vomiting, chicken pox etc. Please **do not** come to clinic, instead go to the emergency department and we will see you there.

What can trigger a sickle cell crisis?
Common triggers for a sickle cell crisis include: infection, stress, dehydration, cold/hot weather or sudden changes in temperature or nothing at all. Sometimes they happen out of the blue.

How to prevent sickle cell disease
As mentioned earlier, sickle cell disease is an inherited condition. This means that if two people who carry the sickle gene have a child, each carrier (trait) parent may donate an affected gene to their child. The probability of producing a child with sickle cell disease is one in four of every pregnancy if both parents are carriers. If one person has sickle cell disease and the other is a carrier the risk is greater at one in two.

If you want to discuss this further, please go to your local sickle cell counselling service or ask to speak with us. If you or your partner becomes pregnant it is extremely important that testing is done as early as possible. If this happens please tell us immediately and we will facilitate further counselling and assessment.

What can you do to stay well as a person with sickle cell disease?

- maintain a good fluid intake
- healthy eating
- exercise
- make sure infections are treated quickly
- maintain a good balance of nutrition and activity
- ensure your vaccinations are up to date
- avoid smoking
- take penicillin twice a day for life
- ensure you attend your clinic appointments so that we can review your health and monitor you for the development of complications from your condition.

- This is a requirement stipulated in the National Standards produced by the Sickle Society in conjunction with the Department of Health. These appointments are 6 -12 monthly in healthy people but may be more frequent in people who have health problems.

- If you do not have an appointment please get your GP to “choose and book” or write a short referral.
- If your appointment is inconvenient, please **change** it. Do not just not turn up. We understand that you have a busy life with many commitments and we will facilitate clinic appointments wherever possible.

Medications
People with sickle cell disease require certain medications.

Antibiotics: people with sickle cell disease are prone to infections because the spleen does not work well. All people with sickle cell disease should be on lifelong penicillin twice daily as this has been shown to reduce illness and death from particular infections.

The usual dose prescribed is 250mg twice daily, but 500mg twice daily may also be used. Amoxicillin (dose: 250mg twice daily) may also be used for the same purpose of infection prevention. If you are allergic to penicillin, then your GP will prescribe an alternative, usually clarithromycin (250mg twice daily) or erythromycin (250mg twice daily).

Opposite to popular belief, taking these antibiotics long-term does not make you less able to respond to antibiotic treatment when you are acutely ill.

Folic acid: we also recommend that people with sickle cell disease take folic acid tablets daily. Red blood cells last about three months in people without sickle cell but may last less than a month in some people with sickle cell disease because they are being broken down at a faster rate. This is what gives rise to the anaemia and jaundice (yellowing of the eyes.

Folic acid is a B vitamin that is required for the manufacture of DNA. People with sickle cell need to have adequate stores of folic acid in their body to be able to replace the red blood cells that are being rapidly destroyed. The usual dose of folic acid is 5mg once daily and your GP will normally prescribe this.

Other medications: medications that require hospital administration or monitoring will be prescribed in the haematology clinic.