Sickle cell disease in adulthood

North Central London Haemoglobinopathy Network jointly with Whittington Health, Royal Free London and Luton and Dunstable NHS Foundation Trust

Joint Red Cell Unit
If you need a large print, audio or translated copy of the document, please contact us on 020 3447 9638. We will try our best to meet your needs.

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Introduction
This booklet explains more about sickle cell disease (SCD) in adults and provides an overview of the services offered by University College London Hospitals NHS Foundation Trust (UCLH). If you would like further information, please ask for our leaflet ‘Guide to services for people with red cell and iron disorders’.

What is SCD?
SCD is a disorder of the body’s red blood cells that is genetically inherited and lifelong. It is caused by an abnormality in haemoglobin (the part of red blood cells that carries oxygen round the body), which makes the red cells become inflexible and sticky. This in turn makes other blood cells as well as the vessels they travel in sticky. Blood flow becomes interrupted and an inflammatory cascade occurs — a process where one inflammatory reaction causes another. This process may cause pain and also problems in the organs where the sickling is happening. It also affects the immune system.

In SCD, there are problems that happen suddenly and are often painful — these are known as sickle cell crises. The long-term effects of crises and background sickling (that may have no symptoms) are known as chronic complications. The approach we take is to limit and prevent both of these from happening as much as possible.

What kind of problems can happen in SCD?
- Infection
- Gallstones
- Joint damage/destruction
- Loss of spleen function (making patients prone to infections)
- Damage to kidneys
- Leg ulcers
- Visual impairment
- Neurological damage which may lead to stroke
- Erectile dysfunction (priapism).
The signs and symptoms of crisis may include:

• Fatigue (feeling tired or weak)
• Pain
• Jaundice (yellowing of the white of the eyes)
• Paleness of skin, or inside the mouth or eyes
• Shortness of breath
• Dizziness
• Headaches
• Symptoms of infection, such as fever.

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

Treatment

You may be able to manage a simple painful sickle cell crisis at home by taking painkillers, resting, drinking fluids and keeping warm. However, if the pain becomes worse you may need stronger painkillers in hospital. We can help to control the pain in hospital, and also 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 feel 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 starting hydroxyurea or a long-term blood transfusion programme.

Some people have had a bone marrow transplant that has cured their sickle cell disease.
What to do in an emergency

If you have a non-urgent medical problem or a medical problem unrelated to your sickle cell disease, for example a rash or a twisted ankle, you can contact your GP or go to your local Emergency Department (A&E). If necessary, they can discuss your problem with us.

If you have a non-urgent red cell/iron problem, you can ask for your outpatient appointment to be brought forward. It may also be appropriate to have a review in day care.

If you are unwell, you should call the haematology advice line (office hours). If it is out of hours, please call the adult haematology advice line or paediatric helpline — contact details are at the end of this booklet. The haematology advice line is manned by an administrative staff member who will contact your clinical team. The adult haematology advice line and paediatric helpline are manned by a senior haematology ward nurse who will contact the on-call team.

You may be advised to attend the rapid access service in haematology day care, go to your GP or Emergency Department at UCLH, or to bring forward your clinic appointment. Sometimes the nurse will speak to one of the doctors to get further advice.

The day care rapid access service is only open for assessments between 09:00 and 16:00, Monday to Friday, after previous discussion with the CNS or medical team. Outside of these times, you will need to go to the Emergency Department (A&E).

Even if you are going to the Emergency Department (A&E), it is always best to call ahead so that we can try to streamline your assessment as much as possible.

If you attend an Emergency Department (A&E) in another hospital, it may take up to a few days to transfer you to UCLH. Although we are always happy to support other teams, we would not be able to coordinate your care at another hospital. For this reason, we advise that you come to the Emergency Department (A&E) at UCLH if you think the delay in getting there is not going to be dangerous.
**Note:** If you have an infectious illness, such as cough, cold, diarrohoea, vomiting or chicken pox, **do not** come to clinic or haematology supportive care unit. You should go to the Emergency Department (A&E) and we will see you there. These seemingly minor illnesses can be very serious for some haematology patients with very poor immune systems, for example those recovering from chemotherapy or stem cell transplants.

**What is an emergency?**
The following symptoms require immediate medical attention:

- Fever: 38°C or above
- Feeling very unwell
- Pain that can not be controlled with painkillers
- New weakness, particularly if felt more on one side than the other
- If you think you are much more anaemic than usual.

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

**How to prevent SCD**
As SCD is an inherited condition, each carrier (trait) parent may pass on an affected gene to their child. If both parents are carriers, the chance that their child will have SCD is one in four. If one person has the disease and the other is a carrier, the risk is greater — one in two. Therefore it is important that your partner is tested for SCD if you are planning a pregnancy.

If you would like to discuss this further, please speak to the red cell team or go to your local sickle cell counselling service.
If you or your partner becomes pregnant, it is extremely important that testing is done as early as possible. Please contact us immediately if this happens and we will organise further counselling and assessment.

**What can I do to stay well?**

- Maintain a good fluid intake
- Eat healthily
- 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 that you attend your clinic appointments so that we can review your health and monitor you for any complications.

Attending your clinic appointments is a requirement stipulated in the National Standards produced by the Sickle Cell Society together with the Department of Health. These appointments are every six to 12 months in healthy people but may be more frequent if you have health problems.

If you do not have an appointment, please ask your GP to refer you.

If your appointment is inconvenient, please **change it**. We understand that many of you have busy lives with many commitments and we will do our best to schedule an appointment that is convenient for you.

For further information on how to stay well, please see our leaflet “Medications, vaccinations and travel in sickle cell disease”.

National Haemoglobinopathy Registry (NHR)
The NHR is a new database of patients with red cell disorders (mainly sickle cell and thalassaemia) living in the UK. This database collects data that is required by the Department of Health from haemoglobinopathy centres. The central aim of the registry is to improve patient care.

Your red cell team will speak to you about this in detail and offer you an information leaflet.

Sexual health
Mortimer Market Centre offers STI (STD) testing, HIV tests and a wide range of sexual health services.

You need to book an appointment to attend the clinic, with the exception of walk-in HIV testing clinics. You can phone the clinic for advice if you have an urgent problem or to find out the opening times.

Mortimer Market Centre
Address: Mortimer Market Centre
         (off Tottenham Court Road)
         London WC1E 6JB

Appointments telephone: 020 3317 5100
Website: www.camdenproviderservices.nhs.uk/clinic/mortimer-market-centre

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 applicable.

If there are any changes to your medication, please let the red cell team know.
Stop smoking clinic
UCLH has joined forces with Smokefree Camden to establish two clinics a week. Accredited advisers from the National Centre for Smoking Cessation and Training (NCSCT) offer behavioural support and a range of nicotine replacement therapy patches. If you are interested, please ask your consultant to refer you.

Where can I get more information?

The Sickle Cell Society
Tel: 020 8861 7795
Website: www.sicklecellsociety.org

NHS Sickle Cell and Thalassaemia Screening Programme
Website: www.gov.uk/guidance/sickle-cell-and-thalassaemia-screening-programme-overview

The UK Thalassaemia Society
Tel: 020 8882 0011
Fax: 020 8882 8618
Email: office@ukts.org
Website: www.ukts.org

UCL Hospitals cannot accept responsibility for information provided by external organisations.
Contact details

Haematology advice line (office hours, adults and children):
020 3447 7359

Adult haematology advice line (out of hours):
07852 220 900

Paediatric helpline (out of hours):
  • nurse in charge 07961 081 645
  • ward T11S 020 3456 7890 ext. 71103 or 71143

Apheresis:
020 3447 1803

Address: The Joint Red Cell Unit
        Department of Haematology
        3rd Fl West, 250 Euston Rd
        London NW1 2PG

Website: www.uclh.nhs.uk/JRCU

Haematology consultants:
Professor John Porter
Dr Sara Trompeter
Dr Perla Eleftheriou
Dr Bernard Davis
Dr Farrukh Shah

Specialist nurses:
Bernadette Hylton (adults)
Nancy Huntley (apheresis)
Catherine Mkandawire (children)
Space for notes and questions