Sickle cell disease in adulthood

Information for patients

Introduction

This leaflet explains more about sickle cell disease (SCD) in adults and provides an overview of the services offered by Whittington Health. If you would like further information, please speak to a member of our team.

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 in childhood 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 problem related to sickle cell disease, you can ask for your outpatient appointment to be brought forward.

If you are more unwell beyond your usual sickle cell crisis, you should attend the Emergency Department (A&E).

If you attend an Emergency Department (A&E) in another hospital, please inform us so we can liaise with the doctors looking after you in that hospital.

What is an emergency?

The following symptoms require immediate medical attention:

- Fever: 38°C or above
- Feeling very unwell
- Pain that cannot be controlled with painkillers at home
- New weakness, particularly if felt more on one side than the other
- If you think you are much more anaemic than usual
- Shortness of breath with or without cough

- Tummy problems – severe pain, diarrhoea, vomiting
- Priapism (see Priapism information leaflet)

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 at 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 us 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 recommendation 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 you have a busy life 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 haemoglobin 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.

Contact details

Specialist nurses

Matty Asante-Owusu (community matron, adults): 07920 711210
Edith Aimiuwu (nurse specialist, children): 07799 347161
Emma Prescott (nurse specialist, thalassaemia): 020 7288 5225

Haematology consultants

Dr Bernard Davis
Dr Farrukh Shah
Dr Ali Rismani

Paediatric consultants

Dr Andrew Robins
Dr Sara Hamilton

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Where can I get more information?

The Sickle Cell Society

54 Station Rd, London NW10 4UA
Tel: 020 8861 7795
Website: www.sicklecellsociety.org

The UK Thalassaemia Society

19 The Broadway,
London N14 6PH
Tel: 020 8882 0011
Website: www.ukts.org

Red Cells R Us

Sickle Cell & Thalassaemia Centre
17a Hornsey Street
London N7 8GG
020 3316 8853/8854
Twitter: @Red Cells R Us
Facebook: Red Cells R Us
Google+: Red Cells R Us (in community section)

NHS Sickle Cell and Thalassaemia Screening Programme

https://www.gov.uk/topic/population-screening-programmes/sickle-cell-thalassaemia

Patient advice and liaison service (PALS)

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

If you need a large print, audio or translated copy of this leaflet please contact us on 020 7288 3081. We will try our best to meet your needs.

Healthwatch England

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