Sickle cell disease in children

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, braille or translated copy of this booklet, please contact us on 020 3447 9638. We will do our best to meet your needs.

Contents
Introduction 3
What is SCD? 3
What kind of problems can happen in SCD? 4
Can SCD be prevented? 4
Treatment of a sickle cell crisis 5
What to do in an emergency 5
What is an emergency? 6
What can trigger a sickle cell crisis? 6
What can I do to help my child stay well? 6
National Haemoglobinopathy Registry (NHR) 7
As your child gets older... 8
Contact details 9
Where can I get more information? 10
Space for notes and questions 11
Introduction

This booklet explains more about sickle cell disease (SCD) in children and provides an overview of the services offered by University College London Hospitals NHS Foundation Trust (UCLH).

What is SCD?

SCD is a lifelong condition that affects the body’s red blood cells. It is the most common inherited genetic disease in the UK – every day a child is born with sickle cell disease.

As it is an inherited condition, it cannot be caught from others with SCD. Parents may have the disease themselves, or they may be carriers of the sickle cell gene or an abnormal haemoglobin. If both parents pass their sickling gene to their child, he/she will be born with SCD.

In sickle cell disease, the red blood cells can’t maintain their round shape as they squeeze through narrow blood vessels. They then become crescent or sickle-shaped, giving the name to the condition.

The red blood cells are also sticky, making the other blood cells and the vessels they travel in sticky too. The sticky cells can stop the blood from flowing, causing pain and potentially other problems in the organs where the sickling is happening. This is called a sickle cell crisis.

The good news is that there is a lot that can be done to prevent sickle cell crises. Many people with SCD lead normal lives – they socialise, study, work and have a family.
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 whites of the eyes)
- Shortness of breath
- Dizziness
- Headaches
- Symptoms of infection, such as fever.

Can SCD be prevented?

If your child has a sickle cell disease, it means that they have inherited two abnormal haemoglobin genes from you and your partner, one of which has to be a sickle gene. The other gene can be a sickle gene, a beta thalassaemia gene or another abnormal haemoglobin gene, such as haemoglobin C, haemoglobin D, haemoglobin O Arab.

Carriers of sickle cell have one sickle cell gene only and the other haemoglobin gene is normal. 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.
We will discuss this in more detail with your child when they enter their teenage years to make sure they understand the risk of their children inheriting SCD too.

**Treatment of a sickle cell crisis**

A simple painful crisis is when your child has pain but is otherwise well. You may be able to manage it at home by giving your child painkillers, fluids and keeping them warm. However, if the pain becomes worse your child 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 their condition.

Sometimes the crisis may not be particularly painful or, more importantly, your child may feel unwell from the start. This is not a simple painful crisis, and your child will need to be assessed to check if they need treatment or are developing any problems. If your child is unwell even if they have no pain, you should always contact us as you may need to come to hospital (contact details are at the end of this leaflet).

If your child has frequent or severe crises, the team looking after them will discuss the treatment options with you. They may include starting hydroxyurea or a long-term blood transfusion programme.

Some people have also had a bone marrow transplant that has cured their sickle cell disease.

Gene therapy is another treatment option currently used in clinical trials. This is when a normal gene is inserted into the patient’s genes to allow them to produce normal haemoglobin. We hope that this will offer a cure to more patients in the future.

**What to do in an emergency**

Please go to the Emergency Department (A&E) with your child’s ‘paediatric passport’ and a paediatric specialist will see you. They should discuss your problems with the haematologist on call.
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
• New weakness, particularly if felt more on one side than the other
• If you think your child is 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. Sometimes crises happen out of the blue.

What can I do to help my child stay well?
There are things you can do to help your child stay well and prevent sickle cell crises. It is important that:
• All medicines are taken as instructed.
• Your child drinks enough fluids and has a good balance of nutrition and activity.
• Infections are treated quickly.
• Their vaccinations are up to date.
• Your child is warm at all times. If you go swimming, make sure you dry them off quickly.
• You help your child to develop strategies to cope with life’s ups and downs – we know that stress is an important factor in sickle cell disease.
• You attend your child’s clinic appointments so that we can review their health and monitor them for any complications.
In the past we only offered hydroxyurea if a child had a lot of problems with their sickle cell disease. A more recent study (known as baby HUG) demonstrated good effects and tolerability of hydroxyurea in newborn babies. Therefore we recommend it if there are any problems, but also offer parents an option to start hydroxyurea as a preventative treatment before any problems occur.

Attending your child’s clinic appointments is a requirement stipulated in the National Standards produced by the Sickle Cell Society together with the Department of Health and Social Care. These appointments are every three months until your child is two years old, and then every six months. If there are any problems or complications, we will see your child more often.

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

For further information, please see our leaflet ‘Medications, vaccinations and travel in sickle cell disease’.

**National Haemoglobinopathy Registry (NHR)**

The NHR is a database of patients with red cell disorders (mainly sickle cell and thalassaemia) living in the UK. This database collects data to help facilitate an understanding of the type of problems faced and treatments offered to these patients.

Your red cell team will speak to you about this in detail and offer you an information leaflet. For more information, please visit www.nhr.nhs.uk
As your child gets older...

As your child gets older, we will expect them to take more responsibility for their health. This is to ensure that they are able to look after themselves when they leave home to live independently. It is as important for them to learn the skills needed for a day-to-day adult life, such as paying bills or working, as it is to be able to navigate their own health needs.

This process is called transitioning and we will help and support them throughout that time. Transitioning happens gradually over their teenage years so that by the time your child is 18, they will feel confident in:

- their knowledge about their condition
- keeping their clinic appointments and attending them on their own if they wish
- ordering their medicines
- organising their vaccinations.

As your child gets older, they may want to talk to the team looking after them in private. They may have questions they would like to ask but may feel uncomfortable to discuss them with you. This is normal and we will be happy to see them on their own.
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 T11 south 020 3456 7890 ext. 71103 or 71143

Apheresis:
020 3447 1803

Address: The Joint Red Cell Unit
Department of Haematology
3rd Floor 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 Emma Drasar
Dr Bernard Davis
Dr Farrukh Shah

Specialist nurses:
Bernadette Hylton (adults)
Nancy Huntley (apheresis)
Nina Gorman (children)
Where can I get more information?

**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

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

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Space for notes and questions