



Sickle Cell Disease in Children

A patient's guide

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Introduction

This booklet explains more about sickle cell disease (SCD) in children and provides an overview of the services offered by Whittington Health.

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.

Parents may have the disease themselves, or they may be carriers of the sickle cell gene causing 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.

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 this 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 where this can be controlled and also treat the underlying cause (trigger) and any complications relating to their condition.



Sometimes the crisis may not be particularly painful but your child may feel unwell from the start. Your child may need to be assessed to check if they need treatment or are developing any problems, you should always contact us as he/she may need to come to the 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 follow the instructions on the emergency card (ring clinical nurse specialist during working hours or I for ward and CAU out of hours.)

If brought by ambulances 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 you have at home
- Difficulty in breathing
- New weakness, particularly if felt more on one side than the other
- If you think your child is much more anaemic than usual.
- Sudden Enlargement of the spleen
- Painful erection lasting more than an hour.

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, physical exhaustion etc. Sometimes crises happen without a trigger.

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 new born babies. Therefore we now give parents an option to start hydroxyurea as a preventative treatment before the age of one if they have certain type of sickle cell disease before they experience any complication.

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 and in some cases once a year. 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 the treatments offered to these patients, and it is important to developing a good service.

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

Specialist nurses

Emma Prescott (nurse specialist, thalassaemia)	020 7288 5225
Matty Asante-Owusu (community matron, adults)	07920 711210
Edith Aimuwu/ (nurse specialist, children)	07799 347161
Olivia Kudom (nurse specialist, sickle cell adults)	07887 987931

Haematology consultants

Dr Bernard Davis
 Dr Farrukh Shah
 Dr Ali Rismani
 Dr Emma Drasar

Paediatric consultants

Dr Andrew Robins
 Dr Janine Younis

If you or your family have any other questions please do not hesitate to contact any of the above healthcare professionals at Whittington Health.

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



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.whitthealthPALS@nhs.net

If you need a large print, audio or translated copy of this leaflet please contact us on 020 7288 3182. We will try our best to meet your needs.

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