Whittington Health NHS Trust

Sickle Cell Disease (SCD) in Children

A parent's guide

Introduction

This guide 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 (also called the 'sickling gene') causing abnormal haemoglobin (red blood cells). If both parents pass their sickling gene to their child, they will be born with SCD.
- In SCD, the red blood cells cannot keep their round shape as they squeeze through narrow blood vessels (arteries or veins). 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 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 (stones in the gallbladder)
- Joint damage/destruction
- Loss of spleen function (making patients prone to infections)
- Damage to kidneys
- Leg ulcers
- Visual impairment (loss/changes in eyesight)
- Neurological damage (to brain or nerves) which may lead to stroke
- Erectile dysfunction (for example priapism- long term erection)

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 SCD, it means that they have inherited two abnormal haemoglobin (red blood cells) 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 (drinks) and keeping them warm.
- However, if the pain becomes worse, your child may need stronger painkillers in hospital, where this can be controlled, and we can 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 they may need to come to the hospital (contact details are at the end of this leaflet).
- If your child has regular or severe crises, the team looking after them will discuss the treatment options with you. This may include starting Hydroxycarbamide or a long-term blood transfusion programme or treatment such as a bone marrow transplant.
- Some people have had a bone marrow transplant that has cured their sickle cell disease.
- Gene therapy is a potential option currently used in clinical trials. This is done by editing the faulty gene in a patient's bone marrow stem cells so that the body produces functioning haemoglobin (red blood cells), to reduce symptoms of SCD and thalassaemia.
  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 the Clinical Nurse Specialist during working hours on 07799 347 161. During out of hours, contact Ifor ward on 0207 288 5442/4989
- If brought by an ambulance, 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?

Get medical help straight away if your child experiences the following:

- 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 can 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.
  - Keep your child warm in cold temperatures.
  - Your child drinks enough fluids and has a good balance of nutrition and activity.
  - Infections are treated quickly.
  - Their vaccinations are up to date.
  - 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 SCD.
- 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 Hydroxycarbamide if a child had a lot of problems with their SCD.
- A more recent study (known as baby HUG) highlighted the good effects of hydroxycarbamide on newborn babies.
- We now give parents an option to start Hydroxycarbamide as a treatment, from 9 months old, if they have certain types of SCD, before they experience any complications.

Appointments

- You must attend your child's clinic appointments. You are required to do this by 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 you are unable to make your appointment, **please change it.** We understand that many people have busy lives, including children's school trips and exams, and we will do our best to give you an appointment that you are able to attend.

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 information that can help medical professionals understand the type of problems patients with these disorders face.
- It also stores information on all the treatments offered to these patients. This is important information to have because it helps us develop 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...

- We will expect them to take more responsibility for their health.
- We do this to make sure that they can look after themselves when they leave home and live independently.
- It is important that they learn both the skills needed for day-to-day adult life (paying bills or working) and be able to look after their own health.
- 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 discussing 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 711 210
Edith Aimiuwu/ Emma Savie-Disu (Nurse Specialist, children)	07799 347 161
Olivia Kudom/ Nicola Nwocha (Nurse Specialist, Sickle Cell adults)	07887 987 931

Haematology consultants

Dr Ryan Mullally
Dr Ali Rismani
Dr Emma Drasar

Paediatric consultants

Dr Janine Younis
Dr Arpana Soni

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.PALS@nhs.net

If you need a large print, audio or translated copy of this leaflet please email whh-tr.patient-information@nhs.net. We will try our best to meet your needs.

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Facebook.com/WhittingtonHealth

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