

Sickle Cell in children

Training for schools

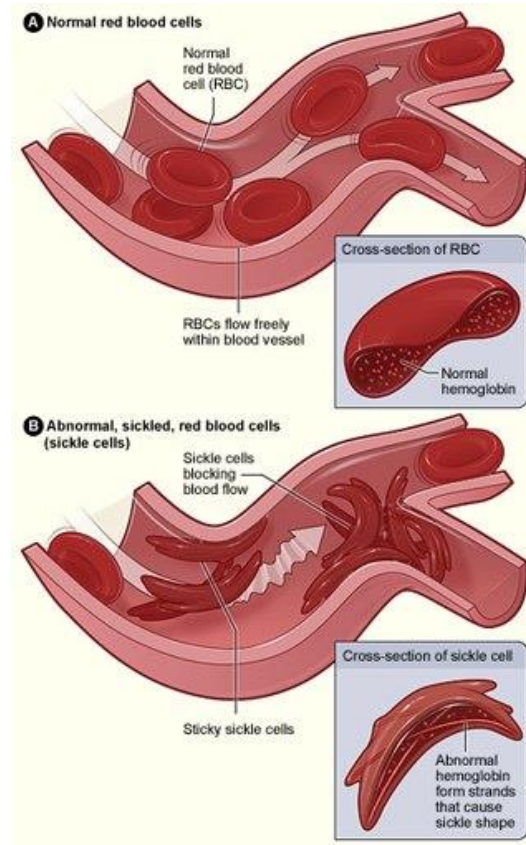
Introduction

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- Any questions

What is Sickle Cell?

- Sickle Cell is a life - long inherited blood disorder that means the red blood cells become sickle shaped and are unable to pass through the blood vessels effectively. This causes pain.
- Sickle cell can cause what's known as a "crisis" this means the child can develop pain anywhere in their body. This often needs hospital treatment consisting of pain relief and sometimes a blood transfusion.
- A crisis can also occur in the penis. This is called 'priapism' and is a non-sexual erection of the penis. Prompt treatment is needed for this to avoid any damage to the functioning of the penis.
- There are two types of Sickle Cell HbSS and HbSC, both can present as similar although it's considered that HbSC is a milder form of Sickle Cell

Red blood cells



How is Sickle Cell treated?

- Blood transfusions or blood exchanges
- Patients are advised to take penicillin, folic acid and vitamin D.
- Sickle patients which have HbSS are offered a medication called hydroxycarbamide. This reduces production of some red blood cells in the bone marrow so the bone marrow can make more protective red and white cells instead. The new cells are less sticky and fragile and help blood flow without painful blockages. It can also reduce pain. A Community Nurse or Nurse from our team may visit the patient at school to take blood if the child is taking this medication. This is to reduce the amount of time they are absent from school.
- In some rare cases patients will be offered a bone marrow transplant although this isn't always possible due to lack of matching donors.
- Children with HbSS are monitored yearly with a TCD. This is a transcranial doppler and is an ultrasound of the head which measures blood flow around the brain. This is needed as children with Sickle Cell can experience silent strokes.

What is Dactylitis?

Hand Foot syndrome (Dactylitis)

- * Dactylitis is severe pain that affects the bones of the hands, the feet, or both.
- * It's often the first symptom of sickle cell disease in babies.
- * Dactylitis is caused by blocked blood circulation.
- * Symptoms include extreme pain and tenderness, usually with swelling.



Precautions

- Children with Sickle Cell must be encouraged to drink and must always have access to water. Dehydration can cause a sickle crisis
- Extreme temperatures can also cause a crisis, for example children should be allowed to wear extra layers when cold and when in class kept away from draughts.
- Hot weather can also trigger a crisis however it is important for the child to be included in play times with peers. Teacher discretion is advised.
- After swimming the child should dry quickly to prevent a crisis and the pool facilitator could be approached to increase the temperature of the water when your school visits
- Unlimited access to the toilet – Sickle Cell can cause the kidneys to concentrate urine which means children make a large amount of dilute urine. The dilute urine fills the bladder quickly.
- Sickle Cell can cause silent strokes in children, if you notice a child is declining in academic ability or is unable to carry out tasks they could do competently previously please inform parents who can then contact us
- After PE if the child is wet or cold they should change immediately into dry clothes
- Some children with sickle cell may experience fatigue
- Having Sickle Cell may affect the child's school attendance if they are unwell or needing to attend hospital appointments

A Community Nurse or Nurse from our team may visit to take blood from a pupil

What to do in an emergency

- If the child is experiencing pain they could be administered pain relief at school. This should be provided in packaging labelled with the child's details and with permission from parent/carer.
- If the pain is not relieved by the paracetamol and/or ibuprofen call parents/carers for advice as they may need to come to hospital

What to include in a care plan

- Contact details of parents and emergency contacts
- GP
- Name of members of staff in charge of health conditions
- Medication, dose, time date etc
- What to do in an emergency
- Hospital staff contact details

Useful Resources

- <https://www.sicklecellsociety.org/> is a very useful source of information
- My friend Jen book series – Amazon



Our contact details

- Clinical Nurse Specialist's
Bristol Children's Hospital
01173428721
- Hayley Wiles – Benign Haematology Family Support
Worker
Bristol Children's Hospital
01173420658



University Hospitals
Bristol and Weston
NHS Foundation Trust

Any questions?

We are
supportive
respectful
innovative
collaborative.
We are UHBW.

