Sickle Cell Disorders

Nigerian Version

A Guide to School Policy

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Health and Safety in Schools

This booklet has been produced based on research examining the experiences of young people with sickle cell disease in schools. An important part of school inclusiveness is recognising the importance of offering care to young people with long-standing illness, particularly since a major part of childhood is spent in attending school. The Nigerian School Health Policy of November 2006 calls for adequate resources and programmes that promote the physical, mental, safety and social well being of all categories of school children generally

What is Sickle Cell Disorder (SCD)?

Sickle cell disorder (SCD) is a collective name for a series of serious inherited chronic conditions that can affect all systems of the body. It is one of the most common genetic conditions in the world and affects around 1 in 50 of all babies born in Nigeria[1]. These sickle cell disorders are associated with episodes of severe pain called sickle cell painful crises. People with sickle cell disorder have a type of haemoglobin (called haemoglobin S (HbS) or sickle haemoglobin) which differs from normal adult haemoglobin (haemoglobin A or HbA). This can cause red blood cells to change shape and become blocked in the blood vessels, causing acute pain. Many systems of the body can be affected meaning that different key organs can be damaged and many different symptoms can occur in many different parts of the body. The main types of sickle cell disorder are sickle cell anaemia, haemoglobin SC disease and sickle beta-thalassaemia.

[1] *World Health Organization* (*2006*) *Sickle Cell Anaemia*. WHO Fifty-Ninth World Assembly A59/9, Geneva

How can the symptoms of sickle cell disorders (SCD) be prevented?

Certain factors have been identified as more likely to precipitate a painful sickle cell crisis. These include infections, cold and/or damp conditions, pollution, dehydration, strenuous exertion, stress, sudden changes in temperature, alcohol, caffeine, and smoking. Advice to people living with a sickle cell disorder on preventing crises includes taking all precautions against malaria, keeping warm, eating healthily, taking moderate exercise, taking plenty of fluids, seeking medical advice if they have a fever, feel unwell, extremely tired and in pain. Avoid smoking and alcohol, keep up to date with medications (including anti-malarial prophylaxis) and vaccinations, and try to reduce stress.

Preventive measures to support people with SCD

Since those with SCD are ill-suited to hard manual work, it becomes doubly important for them to receive a good education, proper medical and social care and sound careers advice.

***School absences*:** If schools/colleges do not have strong supportive frameworks on sickle cell disorder to reduce school absences, then studies have suggested that a pupil with SCD could miss weeks of schooling a year, most often in short absences of 2-3 days at a time. Most pupils with SCD do not feel supported by schools in catching up these absences. Such students who have high rates of school absenteeism are faced with risks of poor school performances. A minority may have very high levels of absences. It is important such pupils are not mislabeled as truant. Parents, other students and the wider community should be encouraged to view going to school as something that is normal for a young person with SCD. Education is an investment in the future of young people with SCD.

Good Practice: SCHOOL ABSENCES

One school has a policy of regular twilight catch-up sessions after school. The teachers take it in turns to staff the learning centre so that pupils who have missed a lesson for any reason can be helped to catch up in the presence of teachers. This not only helps the young person with sickle cell disorder catch up, but it does so without drawing attention to them as different from other pupils.

Good Practice: PEER SUPPORT PROGRAMMES

Another good policy is implementing a peer tutoring programme where learners, especially young people with SCD who have missed lessons, learn from their peer groups in school and after classes. Some schools in Nigeria have started the Student Tutoring, Mentoring and Counseling Programmes, where the individual concerns of children in the school are consciously addressed to enable learners to adjust, attend and improve performances in school. Children with SCD are likely to catch up on their missed lessons from their friendly peers. Remember that young people with SCD are well most of the time so they can be the ones who offer peer support to other young people who may miss school for other reasons than for sickle cell-related illness. This teaches the important lesson that young people with SCD can be givers of care and support as well as receivers of care and support.

***Water*:** Young people with SCD need to be well hydrated to reduce the likelihood of becoming ill. Have a ready supply of fresh drinking water available. Allow them to drink water in class. In some schools, each class has a plastic container, with a serving cup and each pupil is allowed to carry his/her drinking cup into the class. The eating and drinking policy in a school should be such that children with SCD are allowed to carry water into the classroom when in need. Ensure any water fountains are working and kept in the highest state of cleanliness so young people with SCD are not put off using them and risk of infection is kept to a minimum.

***Using the Toilet*:** People with SCD cannot concentrate urine as readily as others. They produce large quantities of dilute urine and need to go to the toilet more often. Do not restrict toilet breaks.

Good Practice: USING THE TOILET

One school has instituted a system of issuing the young person with a card stating that the young person has the right to excuse themselves during lesson in order to go to the toilet.

Tiredness: The person with SCD may experience severe anaemia. This may mean they feel tired, lethargic and unable to concentrate. They may feel tired to the point where they feel they need to sleep. It is important that teachers do not mistake this serious medical symptom of SCD for laziness. Climbing several flights of stairs several times per day to get to and from the classroom is physically demanding for some young people with SCD.

Leg Ulcers: Some young people with SCD may experience painful and recurrent leg ulcers, often around the ankle, that are slow to heal. Walking can be painful and make ulcers worse. Schools should ensure the physical environment is suitably adapted to support young people with leg ulcers.

Good Practice: SCHOOL BUILDINGS

The initial build and design of schools should include consultation with all relevant stakeholders but including learners with SCD and their families. The ideal design should consider all of the following:

- Access: A situation that prioritizes building high-rise school structures will NOT be beneficial to young learners with SCD who may be tired from anaemia or have ongoing pain from leg ulcers.
- Space: A dedicated space, separate from classrooms, should be available so that young people with SCD can take time out to rest when unwell, perhaps enabling them to return to classes later. The room could also be where their medicines and painkillers are kept securely and given to them when needed.
- Hygiene and infection control: school building materials and interior surfaces should be chosen with young people with lowered immunity, such as students with SCD, in mind, and should consider ease of cleaning and maintenance
- Toilet areas are in clusters so that it is a short distance to a toilet for a young person with SCD

***Physical Exercise*:** Avoid hard, physical exercise involving strenuous exertion that could precipitate a sickle cell crisis. Encourage moderate exercise. Listen to the young person who will come to know their own safe limits of physical activity. For SCD do not refuse requests if a young person asks to be excused or stop activity because of tiredness or pain. For children with SCD, cold or wet weather, or exposure of the skin to cooling wind may all be a trigger to episodes of illness. It is important to listen to the child and parent, and follow advice from their medical specialists.

Good Practice: MALARIA
Young people with SCD are especially vulnerable to deaths from malaria. One school has arranged for all classroom windows/entrances to be covered with mosquito nets. For all schools, consider brands of soaps impregnated with DEET. All children with SCD should be offered free anti-malarial prophylaxis. Encourage school staff, pupils and parents to hold an environmental clean up before the rainy seasons to reduce places for mosquitoes to breed within the immediate area around the school. Such anti-malaria activities helps the young person with SCD but also helps the other children who are also vulnerable, to varying degrees, of catching and becoming seriously ill with malaria.

Infection: Young people with SCD may have a damaged or missing spleen (the organ that helps to fight infections) and therefore make them vulnerable to infections. Enable safe storage and dispensing of any antibiotic drugs prescribed for the young person with sickle cell disorder.

Good Practice: TEMPERATURE
Young people with SCD may become ill if they are too hot or too cold. When it is cool allow warmer clothing to be worn in class. They should not be made to go outside during breaks when it is raining, cold or windy. Staff supervising breaks, such as assistants or dinner staff, should be instructed not to enforce this. Members of the School Based Management Committee should work with parents to agree school uniform or a policy to enable young people with SCD to wear suitable clothes at appropriate times of the year. Furthermore, in extreme weathers, hot or cold, school closing and opening times should be reviewed according to the seasons. It is important to avoid classroom congestion/overcrowding as some schools are overpopulated, and so facilities are overstretched. Overcrowding can cause the classroom environment to be stuffy, hot, and can lead to poor oxygen intake and further jeopardize the health of the young person with SCD. A person with SCD should be kept away from air conditioners as the cold air they produce can trigger a sickle cell crisis.

Individual Health Care Plans: All children with SCD should have individual health care plans, which should be reviewed yearly. As SCD has numerous possible complications affecting many systems of the body, it is important, where possible, to include a specialist sickle cell health worker in drawing up this plan. Other resources might include local Sickle Cell Clubs.

Good Practice: Individual Health Care Plans
All young people with a medical condition at school should have an individual health care plan. A school health worker with good knowledge of SCD and other chronic illnesses should be developed. A school health plan should be developed by the School Health Committee whose membership is clearly spelt out in the School Health Policy. Individual health care plans should be drawn up in consultation with the parents and the young person. The plans are individual but should cover: preventive measures to keep the child well at school; arrangements for giving pain medication to the child; what constitutes an emergency and what to do; key contacts, especially the consultant (the key health professional caring for the child); and a list of all school staff (not just teachers) who have attended a professional update on SCD. Ideally the plan is reviewed each year, and checked to ensure all staff the young person is likely to meet during their academic year. have received an update.

Medical Issues and Medical Emergencies for Sickle Cell Disorders (SCD)

Acute chest syndrome: Signs include chest pain, coughing, difficulty breathing, and fever. It can appear to be similar to flu like symptoms. However, it is important to see a consultant ASAP.

Aplastic crisis: This is when the bone marrow temporarily slows its production of red blood cells, usually due to infection with a virus called ParvovirusB19. This results in a severe drop in the red cell count and severe anaemia. Signs include extreme paleness and fatigue, and rapid pulse.

Fever: Children with sickle cell disorder are at increased risk for certain bacterial infections. A fever of 101° Fahrenheit (38° Celsius) or higher, could signal an infection. Children with sickle cell disorder and fever should be seen without delay by a consultant (paediatric haematologist).

Hand-foot syndrome (also called dactylitis): Painful swelling of the hands and feet, plus fever. It is most likely to occur in children under five. It is important nursery and pre-school staff are aware of this to avoid false accusations of non-accidental injury.

Painful crises: These may occur in any part of the body and may be brought on by cold or heat, infection or dehydration. The pain may last a few hours, up to 2 weeks, or even longer, and may be so severe that a child needs to be hospitalized. It is important to listen to the young person who will come to know whether the pain is mild/moderate and will pass (where schools can promote school inclusion by permitting rest and re-integration into school later that day) or whether they need to go to hospital.

Splenic sequestration crisis: The spleen becomes enlarged by trapping the sickle shaped red blood cells. This leads to fewer cells in the general circulation. Early signs include paleness, weakness, an enlarged spleen, and pain in the abdomen. It is important that nursery and pre-school staff are aware of this life-threatening event, as it is more likely in younger children.

Strokes: The higher risk is in children aged 2-10 years. Apply the FAST approach:

Facial weakness: can the person smile, or has their mouth or eye drooped?

Arm: can the young person raise both their arms above shoulder height?

Speech problems: can the person speak clearly and understand what you say?

Time: to get emergency medical help.

It can be difficult to differentiate the symptoms of stroke from those of a sickle crisis, where pain can result in restriction of movement. Children with SCD need a Transcranial Doppler Scan yearly from age 2 to assess risk of stroke.

Silent Strokes: Changes in a young person's behaviour or concentration or a sudden deterioration in the quality of their school work could be due to several reasons. In up to a fifth of young people with sickle cell disorders, small areas of brain damage may occur resulting from impaired blood supply. It is important for teachers to discuss with parents any changes to behaviour or educational achievement that might indicate such damage is occurring. It is important to help parents link up with local hospitals to investigate if such changed behaviour is owing to a silent stroke.

Priapism*:* An unwanted painful erection of the penis, unrelated to thoughts about sex. Urgent medical help should be sought if it lasts more than two hours.

How does someone get sickle cell disorder (SCD)?

Sickle cell disorders are inherited, that is passed on through the family. They are **not** infectious diseases and **cannot** be caught like coughs or colds. Sickle cell carriers are sometimes referred to as having sickle cell trait. Carriers have a normal and an affected gene. In the case of sickle cell carriers their red blood cells contain both normal haemoglobin (adult haemoglobin, haemoglobin A) and sickle haemoglobin (haemoglobin S). Carriers are usually perfectly healthy themselves, and may not know they have sickle cell trait unless they have a blood test. If someone is a carrier it cannot turn into a sickle cell disorder. For example, if both partners are sickle cell carriers (haemoglobin AS), then **in each pregnancy** there is a one in four chance that they could have a child with sickle-cell anaemia (haemoglobin SS, a type of sickle cell disorder); a one in four chance of a child with normal haemoglobin (haemoglobin AA), and a one in two chance of a child who is a sickle cell carrier (AS).

Good Practice: Sickle Cell in the Curriculum
One means of creating a positive school ethos is to make the curriculum relevant to the pupils. The pattern of genetic inheritance for sickle cell could be made central to the genetics part of the science curriculum. The UK Sickle Cell Society has produced guidance on how sickle cell can be incorporated into various key stages of a school curriculum **http://www.sicklecellsociety.org**

Pain: SCD is an unpredictable condition, variable over time and between different people. This creates uncertainty for the young person. The painful crises can come on quite suddenly. Pain can make a person grumpy, unresponsive and uncooperative. The pain of a sickle cell crisis can be mild, moderate or severe. Since pain is such a common experience for people with SCD it is vital that the school develops a policy for supporting children when in pain. The care plan needs to be worked out individually for each child, with input from teacher, school nurse, sickle cell nurse specialist, child and parents. It is very important that the policy includes instructions about giving painkillers (including who is responsible for administration, which pain killers and how to decide which one to give).

Medication: A key part of the Individual Health Care Plan should include arrangements for giving medication, and agreed procedures for assessing the severity of the pain. The key is to listen to the young person. Where pain is mild or moderate a key aim should be to keep the young person in school, by combining pain medication with an opportunity for rest and time out in a safe environment so that they can return to lessons later in the day. A blanket policy on not administering drugs or on having a young person collected as an outcome of administering any medication will in effect be an exclusionary policy for the young person with a sickle cell disorder.

Teacher Awareness: All staff should be made aware of sickle cell. Some schools cover this using part of training updates for teachers. Staff need to know what to do if the child has a painful crisis, how to recognize signs and symptoms of a stroke in young people with SCD, and learn to listen to the child if the child says they are feeling unwell. Ensure there are robust systems for relaying this information when the child has a supply teacher, when they change class or when they change school. Ensure the availability of a safe area for a pupil with SCD to recover and take time out from activities. They may be able to return to study later in the day.

Challenging Discrimination**:** Ensure that SCD is discussed as part of Personal, Health and Social Education and make sure that other pupils are challenged on any discriminatory views.

A Framework for an Individual Health Care Plan for Someone with Sickle Cell Disorder

<table>
<tr><td>Name:

Date of Birth:

School:

Current Class/Group:

Condition 1: Sickle Cell Anaemia (HbSS)
Condition 2:
Condition 3:

Date of Plan:

Review Each Year:</td><td> 
Photograph</td></tr>
<tr><td>PARENT/GUARDIAN/CARER CONTACTS

Contact Name:
Relationship:
Contact number:

Contact Name:
Relationship:
Contact number:</td><td>CONTACT NUMBERS

Emergency Contact Name:
Emergency Contact number:

Hospital (Haematologist) Name:
Hospital (Haematologist)Number:

Specialist Nurse Name:
Specialist Nurse Number:</td></tr>
<tr><td>KEYWORKER RESPONSIBLE IN SCHOOL:

Name:
Building/Department:
Contact Number:</td><td>FAMILY DOCTOR

Contact Name:
Contact number:</td></tr>
<tr><td colspan="2">Date of Health and Safety Risk Assessment Carried out by School:</td></tr>
<tr><td colspan="2">PREVENTION:

Key worker to ensure that each member of school staff is aware of importance of following preventive measures….

Unrestricted access to drinking water during class time
Unrestricted access to use of the toilet, including during class time
Taking all necessary precautions against malaria
Not forcing to undertake exercise if they say they are tired or in pain</td></tr>
</table>

MEDICATION

Name of medication:

Reason for medication:

Dosage:

Time of medication:

Special Considerations:

Medication will be stored:

Arrangement for Delivery to School:

Written Records Arrangement:

PAIN MANAGEMENT

The aim is to strike a balance between responding appropriately to medical emergencies and maintaining an inclusive school environment where a pupil with sickle cell disorder is not constantly sent home for episodes of minor pain. Ask the specialist sickle cell nurse or hospital consultant if there is a pain scale suitable for use in getting the young person with sickle cell disorder to say how severe the pain they are in. There are scales in which a young person is shown drawings of a series of cartoon faces ranging from happy (no pain) to sad and crying (most pain). Such a scale could be included in the individual health care plan. The following scale is for illustrative purposes only and any scale used should have the approval of the young person's hospital consultant (haematologist).

0	2	4	6	8	10
			✓		
I am not in any pain	I am in a little pain but don't need my medication	I feel if I have my medication I can be in class	I feel I need to have time out but may feel better later	I feel I need to go home	I feel I need to go to hospital

OTHER PARTICULAR NEEDS/ISSUES

This section can contain information specific to the young person's individual condition (for example, information about silent strokes, leg ulcers, priapism, headaches, seizures or other possible complications of sickle cell disorder).

Stakeholders in drawing up IHCP

Name of Person:	Signature:	Date:
Guardian/Carer:	Signature:	Date:
School Nurse:	Signature:	Date:
Sickle Cell Specialist Nurse:	Signature:	Date:
Teacher:	Signature:	Date:

School staff Who Have Received Sickle Cell Awareness Session:

Name:	Date:

Space to include examples of good practice developed by the school:

Water

[Name] is allowed to take his water bottle into assembly. He sits at the end of the row in order to be able to access this discreetly.

Exercise

Use other aspects of the sports activity to promote inclusion e.g. score-keeping, timing with stopwatch

For Sickle Cell Disorder Blood Transfusions

Work with hospital to schedule regular blood transfusions so that [name] is most energized at beginning of period of exams.

Further Information

RESEARCH

For a link to the research evidence underpinning the production of this information, please visit:
http://www.sicklecelleducation.com

This site includes resources originally developed for teachers in the UK, including:

My Pupil has Sickle Cell Disease [Leaflet]
My Friend has Sickle Cell Disease [Leaflet]
What to Do if You Suspect your Pupil is having a Sickle Cell Crisis [Poster]
Sickle Cell and Stroke [Leaflet]

SICKLE CELL VOLUNTARY GROUPS

SCORE Sickle Cell Cohort Research
http://www.score-international.org/

The UK Sickle Cell Society
http://www.sicklecellsociety.org

Sickle Cell Young Stroke Survivors (Cardan, Nigeria)
http://www.scyss.org

Sickle Cell Foundation, Nigeria
http://www.sicklecellfoundation.com/

OTHER RESOURCES

UK National Health Service Sickle Cell and Thalassaemia Screening Programme
http://www.sct.screening.nhs.uk
For a downloadable copy of *A Parent's Guide to Care and Management of Your Child with Sickle Cell Disease*

SCOOTER Open Education Resources for Sickle Cell and Thalassaemia
http://www.sicklecellanaemia.org/
For free sickle cell images and other open educational resources

A downloadable copy of this leaflet is available at:

www.sicklecelleducation.com
www.sicklecellanaemia.org
www.score-international.org

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