

Sickle Cell and Thalassaemia:



Education, Health & Care



A Guide to School Policy

Professor Simon Dyson

Version 2.0

June 2016



<http://creativecommons.org/licenses/by-sa/4.0/>

This work is licensed under Creative Commons. Except for the ERSC logo on the final page, it may be freely used and distributed provided original authorship is acknowledged. It may be adapted for re-use, provided the resultant work is shared back with the sickle cell and thalassaemia communities by offering the revised material to the Sickle Cell Open: Online Topics and Education Resources (SCOOTER) project at www.sicklecellanaemia.org.

Simon Dyson
Unit for the Social Study of Thalassaemia and Sickle Cell,
De Montfort University,
Leicester, United Kingdom
sdyson@dmu.ac.uk

We would like to thank the following for their critical appraisal of earlier drafts of this policy leaflet:

- Professor Elizabeth Anionwu, CBE, Emeritus Professor of Nursing, University of West London.
- Suzi Crawford, Specialist Nurse Counsellor, Soho Health Centre, Birmingham.
- Dr Paul Telfer, Senior Lecturer in Haematology at Queen Mary, University of London , and Honorary Consultant Haematologist at St Bartholomew's and The Royal London NHS Trusts.

This is an update of the previous version released on February 14th 2012. The original plus a number of translated versions can be found on the SCOOTER website:

<http://www.sicklecellanaemia.org/search-our-resources#social>

If you have any questions about the SCOOTER project, which was part of the 2009-2012 UK Open Educational Resource Programme you can contact Vivien Rolfe (project director) via Twitter [@vivienrolfe](https://twitter.com/vivienrolfe)

Education, Health and Care in Schools

This booklet has been produced based on research examining the experiences of young people with sickle cell disorder in schools in England. 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. Section 100 of the **2014 Children and Families Act** in England places a duty on the appropriate school authority (for example, board of governors or proprietor), who must make arrangements for supporting pupils at the school with medical conditions. Pupils with sickle cell disorder or thalassaemia come under this legislation and guidance.

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 2,000 of all babies born in England¹. 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. Despite its name sickle beta-thalassaemia is a sickle cell disorder and is distinct from beta-thalassaemia major described below.

What is Beta-Thalassaemia Major?

Beta-thalassaemia major is a serious inherited blood condition in which the red blood cells are nearly empty of haemoglobin, the key part of the blood that carries oxygen around the body. The first life-saving step of treatment involves having blood transfusions every 3-4 weeks for the rest of their lives. This extra blood introduces extra iron into the body that the body cannot get rid of easily. The second step of treatment involves drugs that get rid of the excess iron. Depending upon the individual's suitability for particular drugs some may take these orally, either by tablet or in a drink, whilst others may have to have injections that are delivered slowly over 10-12 hours, 5-7 days a week. Some young people with SCD, identified by screening as being at high risk of a stroke, may also be on regular blood transfusions and drugs or injections to reduce iron in the body. For both thalassaemia major and SCD attendance at regular hospital outpatient clinics, attendance for regular blood transfusions and attendance for treatment may all impact on school attendance.

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 keeping warm, eating healthily, taking moderate exercise, taking plenty of fluids, seeking medical advice if they have a fever, avoiding smoking and alcohol, keeping up to date with medications and vaccinations, and trying to live a stress free life.

¹ Streetly, A., Latinovic, R., and Henthorn, J. (2010) Positive Screening and carrier results for the England-wide universal newborn sickle cell screening programme by ethnicity and area for 2005-7, *Journal of Clinical Pathology*, 63: 626-629

Preventive measures to support people with SCD or beta-thalassaemia major

Since those with SCD or thalassaemia are ill-suited to hard manual work, it becomes doubly important for those with SCD or thalassaemia to receive a good education 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. A minority have absences at or beyond levels defined by government as persistently absent. It is important such pupils are not mislabeled by education welfare officers as truant and their parents pressured to account for themselves if such absences are the result of serious episodes of illness.

Good Practice: School Absences

One school has a policy of regular twilight catch-up sessions after school. This learning centre is staffed on a rota basis 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.

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. Do not restrict drinking water in class. Ensure 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. They produce large quantities of dilute urine and need to go to the toilet more often. Do not restrict toilet breaks for children.

Good Practice: Using the Toilet

One school has instituted a system of issuing the young person with a laminated 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. Young people with beta-thalassaemia major are likely to be tired towards the end of their 4 week cycle of transfusions. It is important that teachers do not mistake serious medical symptoms of SCD or beta-thalassaemia major 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. In some cases issuing a personal lift pass may be appropriate.

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 or beta-thalassaemia major 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. Obligatory sports and gym sessions out of doors in cold and wet weather is a potent stimulant to crisis for some children. It is important to listen to the child and parent, and follow advice from their specialist medical teams about this.

Good Practice: Swimming

Young people with SCD are advised not to become cold. This may happen when swimming in unheated pools or by delays in drying off afterwards. It is always important to listen to the views of parents and the young person about inclusion in activities such as swimming. One school made an arrangement with their local swimming pool to have the pool temperature increased when the class with the child with SCD was due to take part in a swimming session. This ensured inclusion of the young person in a valued school activity without making the young person feel separate from peers.

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

Temperature: Avoid activities that require outdoor work in cold or damp conditions; avoid under-heating of classrooms, especially mobile classrooms; maintain good ventilation of study areas. Allow coats to be worn in class, and permit the child with SCD to stay inside at break in cold or wet and windy weather.

Good Practice: Temperature

Young people with SCD may become ill if they are too hot or too cold. Schools should work with the young person to establish agreed warmer clothing for indoor use within school. They should not be made to go outside in cold or rainy or windy weather during breaks. Staff supervising breaks, such as assistants or dinner staff, should be instructed not to enforce this. One school was aware that its complement of rooms included mobile classrooms that readily became too hot or too cold. It used its timetabling system to ensure that classes with a child with sickle cell disorder were timetabled away from such unsuitable classrooms. This kept the young person with SCD safe, but did so without drawing attention to him as different from others.

Individual Education, Health and Care Plans: All children with SCD/ thalassaemia should have education, health and care plans, which should be reviewed yearly. As both SCD and thalassaemia have numerous possible complications affecting many systems of the body, it is important, where possible, to include a specialist sickle cell or thalassaemia nurse in drawing up this plan.

Good Practice: Individual Education, Health and Care Plans

All young people with a medical condition at school should have an individual education, health and care plan². One local area works with the local specialist sickle cell nurse counselor, as well as the school nurse, the parents and the young person themselves, to draw up these plans. The plans are individual but cover as a minimum: 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 who will be the key health professional caring for the child; and a list of school staff (including school secretaries, canteen staff, caretakers, not just teachers) who have attended a professional update on sickle cell/thalassaemia). The plan is reviewed each year and the list of staff who have received an update checked against those the young person is likely to meet during their academic year.

² Section 37 of the 2014 Children and Families Act

Medical Issues and Medical Emergencies for Sickle Cell Disorders

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 paleness, 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 by a consultant without delay.

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 or dehydration. The pain may last a few hours or 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 or 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 dial the emergency number for an ambulance.

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 have 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 are evident on a sensitive brain scan (MRI scan) resulting from impaired blood supply. It is important to liaise with the young person's medical consultant in order 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) or beta-thalassaemia major?

Sickle cell disorders and beta-thalassaemia major are inherited, that is passed on through the family. They are **not** infectious diseases and **cannot** be caught like coughs/colds. Sickle cell or thalassaemia carriers are sometimes referred to as having sickle cell/thalassaemia 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 trait unless they have a blood test. If someone is a carrier it cannot turn into sickle cell disorder or beta-thalassaemia major. For example, if both partners are sickle cell carriers (haemoglobin AS), then **in each and every 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 and Thalassaemia 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 both sickle cell and thalassaemia are an integral part of the genetics part of the GCSE science curriculum. The Sickle Cell Society has produced guidance on how sickle cell can be incorporated into various key stages of the national curriculum

<http://www.sicklecellociety.org> The UK Thalassaemia Society <http://www.ukts.org> has advice for science teachers on using thalassaemia for lessons on genetics including online DVDs.


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 and 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/thalassaemia. Some schools cover this using part of a teacher day. 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 or thalassaemia 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/thalassaemia 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 Education Health & Care Plan for Someone with Sickle Cell Disorder

<p>Name:</p> <p>Date of Birth:</p> <p>School:</p> <p>Current Class/Group:</p> <p>Condition 1: Sickle Cell Anaemia (HbSS)</p> <p>Condition 2:</p> <p>Condition 3:</p> <p>Date of Plan:</p> <p>Review Date:</p>	<div data-bbox="906 392 1289 761" style="text-align: center;">  </div> <p style="text-align: center;">Photograph</p>
<p>PARENT/GUARDIAN/CARER CONTACTS</p> <p>Contact Name: Relationship: Contact number:</p> <p>Contact Name: Relationship: Contact number:</p>	<p>CONTACT NUMBERS</p> <p>Emergency Contact Name: Emergency Contact number:</p> <p>Hospital Consultant Name: Hospital Consultant Number:</p> <p>Specialist Nurse Name: Specialist Nurse Number:</p>
<p>KEYWORKER RESPONSIBLE IN SCHOOL:</p> <p>Name: Building/Department: Contact Number:</p>	<p>GENERAL PRACTITIONER:</p> <p>Contact Name: Contact number:</p>
<p>Date of Health and Safety Risk Assessment Carried out by School:</p>	
<p>PREVENTION:</p> <p>Key worker to ensure that each member of school staff is aware of importance of following preventive measures....</p> <ul style="list-style-type: none"> • Unrestricted access to drinking water during class time • Unrestricted access to use of the toilet, including during class time • Keeping warm: permitting coat/hat/gloves indoors, not sending outside in break • Not forcing to undertake exercise if they say they are tired or in pain 	

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 education, health and 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.

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/Thalassaemia Specialist Nurse:	Signature:	Date:
Teacher:	Signature:	Date:

School staff Who Have Received Sickle Cell/Thalassaemia Awareness Session:

Name:	Date:
-------	-------

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.

Exams

Ensure plan is in place should they need additional time. Consider small, well heated room and supervised access to toilet during exams.

(For Beta-Thalassaemia Major or 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 for teachers 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]

If You Teach a Child with Thalassaemia [Leaflet]

UK SICKLE CELL VOLUNTARY GROUPS

The Sickle Cell Society

<http://www.sicklecellsociety.org>

Organization for Sickle Cell Anaemia Research

<http://www.oscarbirmingham.org.uk>

<http://www.oscarsandwell.org.uk>

<http://www.oscarbristol.org.uk>

Sickle Cell Young Stroke Survivors

<http://www.scyss.org>

UK THALASSAEMIA VOLUNTARY GROUPS

UK Thalassaemia Society

<http://www.ukts.org> (includes on-line DVDs)

OTHER RESOURCES

National Health Sickle Cell and Thalassaemia Service Screening Programme

<https://www.gov.uk/guidance/sickle-cell-and-thalassaemia-screening-programme-overview>

SCOOTER Open Education Resources for Sickle Cell and Thalassaemia

International Versions of the Guide to School Policy

BRAZIL (Portuguese) version

<http://www.sicklecellanaemia.org/open-education-resources/sickle-guide-for-schools-portuguese-translation>

Nigerian (English, Yoruba, Hausa, Igbo versions)

<http://www.sicklecellanaemia.org/open-education-resources/sickle-cell-guide-for-schools-nigerian-english-translation>

<http://www.sicklecellanaemia.org/open-education-resources/sickle-cell-guide-for-schools-yoruba-translation>

<http://www.sicklecellanaemia.org/open-education-resources/sickle-cell-guide-for-schools-nigerian-hausa-translation>

<http://www.sicklecellanaemia.org/open-education-resources/sickle-cell-guide-for-schools-igbo-translation>

INTERNATIONAL GOOD PRACTICE

BRAZIL

In the Brazilian state of Minas Gerais, CEHMOB - Minas Gerais Haemoglobinopathies Education and Support Center - a multi-disciplinary health education special project for sickle cell disease developed by NUPAD (the Newborn Screening Programme of the Medical School, Federal University of Minas Gerais) has produced an entire 30 hours distance learning course on sickle cell designed for people who deal with education. They have adapted and translated this Guide to School Policy which is part of the course. The distance learning course is part of the project *Knowing to Care: Sickle Cell Disease in School*. The project is part of the State Department of Education (SEE), more specifically, the Superintendence of Special Issues. The course is being expanded to cover the five Brazilian states with the highest incidence of the disease: Minas Gerais, Bahia, Rio de Janeiro, Pernambuco and Maranhão.

INDIA

In 2013 the Maharashtra State Board for Secondary and Higher Secondary Education provided for students with sickle cell anaemia to have an extra 20 minutes per each hour of a public exam, such as Grade 10 School Secondary Certificate and Grade 12 Higher School Secondary Certificate.

NIGERIA

Nigerian educators have adapted the guide to incorporate their peer-to-peer education initiative and refer to environmental clean-up campaigns to reduce mosquitoes and malaria. The guide has subsequently been translated into: **Yoruba. Hausa and Igbo**

A downloadable copy of this leaflet is available at:

www.sicklecelleducation.com

www.sicklecellanaemia.org

This leaflet was first produced in 2011 on the basis of research funded by the United Kingdom Economic and Social Research Council (RES-000-23-1486) a research grant held by Simon Dyson, Lorraine Culley and Sue Dyson (De Montfort University); Karl Atkin (University of York) and Jack Demaine (Loughborough University).



Version 2.0 Date: 19th June 2016