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For more information

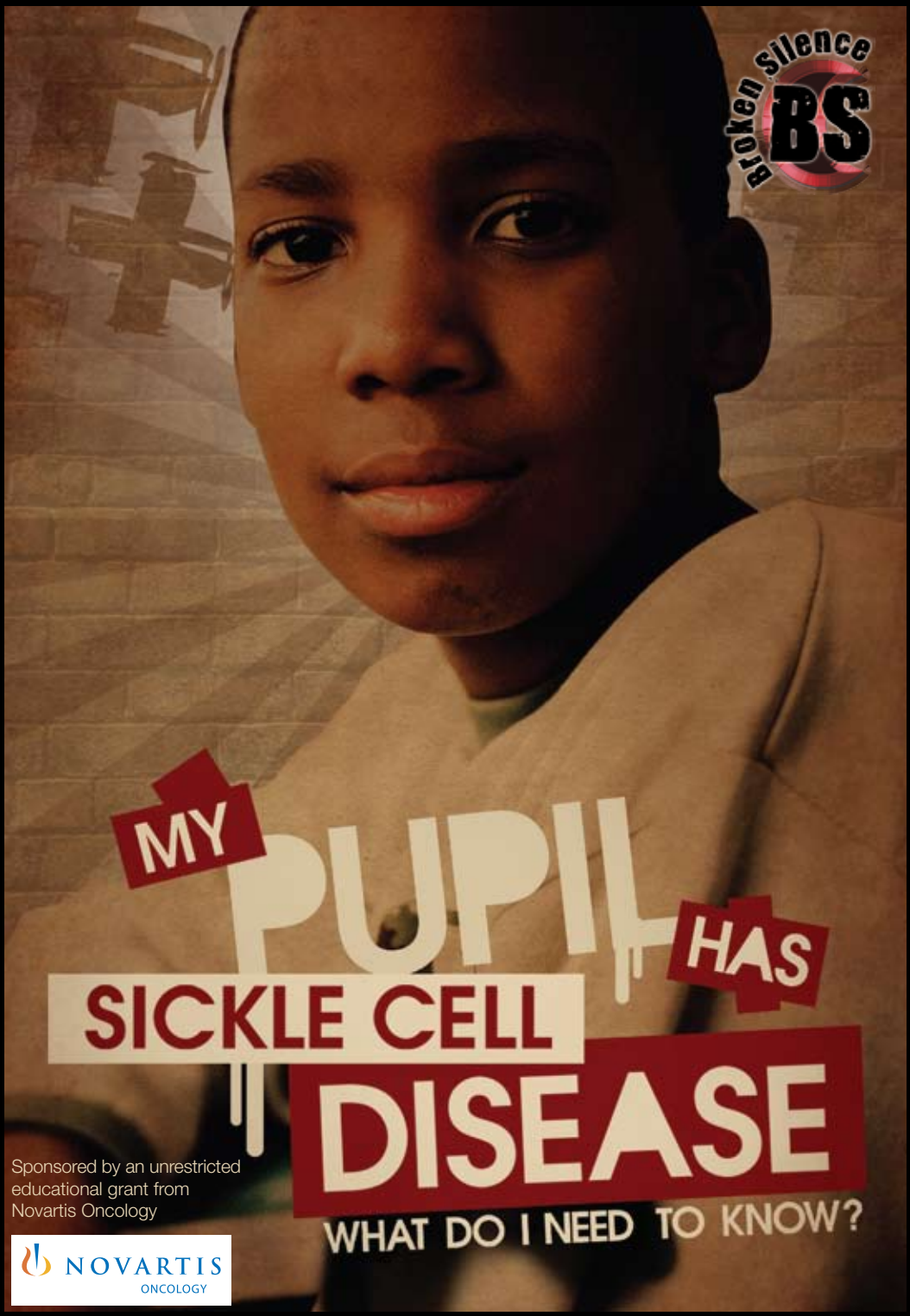
For more information on sickle cell disease or Broken Silence visit www.brokensilence.org or contact Broken Silence via e-mail – brokensilence@brokensilence.org

Useful websites

1. http://www.brokensilence.org/
2. http://www.nhs.uk/Conditions/Sickle-cell-anaemia/Pages
3. www.sickle-thal.nwlh.nhs.uk
4. www.theironfiles.co.uk

References

1. http://www.brokensilence.org/
2. NHS Direct Online Health Encyclopaedia. Available at: http://www.nhsdirect.nhs.uk/articles/article.aspx?articleId=361&sectionId=983 Accessed: 07 July 2008
3. http://www.nhs.uk/Conditions/Sickle-cell-anaemia/Pages
4. http://www.scyss.org/awareness.html
5. Simon Martin Dyson, Hala Abuateya, Karl Atkin, Lorraine Culley, Sue Elizabeth Dyson and Dave Rowley (2010) Reported school experiences of young people living with sickle cell disorder in England *British Educational Research Journal* [Research funded by the Economic and Social Research Council, Grant RES-000-23-1486]



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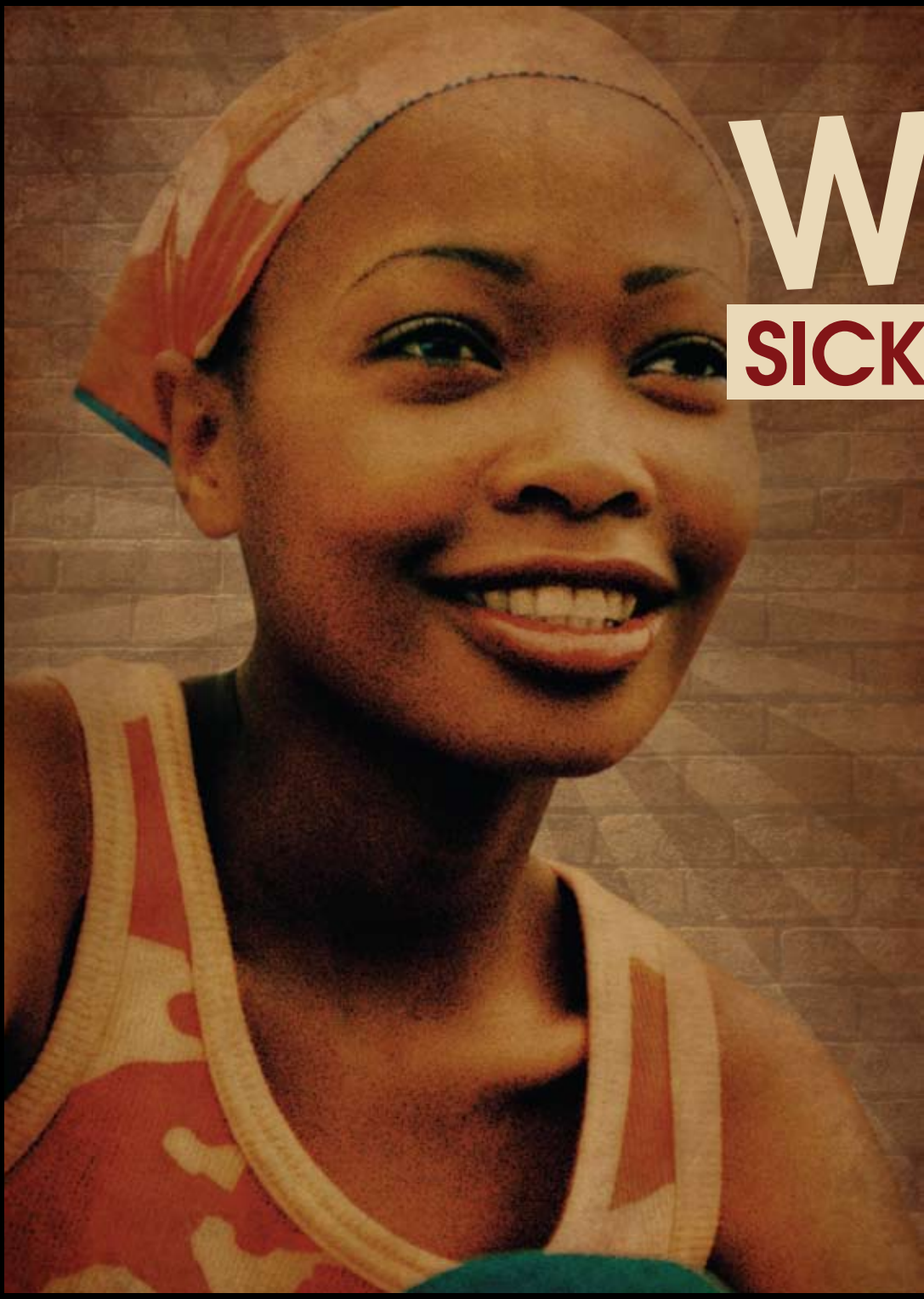
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SICK

WHAT IS SICKLE CELL DISEASE?

Sickle cell disease (or SCD) which includes sickle cell anaemia is an inherited blood disorder which potentially affects every part of the body and is caused by the presence of two altered genes. It is estimated that there are 15,000 adults and children living with SCD in Britain at present and the numbers are increasing. The most severe form of the disease affects 1 in 50 people of West/Central African descent and 1 in 300-400 of people of African-Caribbean descent.[1]

For people without SCD, red blood cells in the body are flexible, and therefore easily able to perform the vital function of transporting oxygen from the lungs to the rest of the body.[2]

For people with SCD, the haemoglobin in their red blood cells is faulty, causing the shape and texture of the red blood cells to change, forming 'sickle' or C-like shapes. This makes it far more difficult for them to move through small blood vessels. The cells also do not survive as long and are not as plentiful as red blood cells in non-sickle cell sufferers.[2]

The sickle cells can get stuck in small blood vessels and stack up, causing blockages and starving organs and tissue of oxygen-carrying blood: this is a sickle cell crisis. Physical symptoms of a sickle cell crisis can vary, from periods of severe pain, to major complications such as stroke, damage to the liver, kidneys, lungs, heart and spleen, or even death.[2]

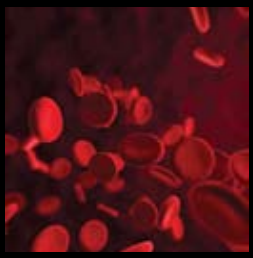
For many SCD sufferers another problem is that because their red blood cells contain sickle haemoglobin, the cells do not last as long as those of someone without the disease, leading to a chronic state of anaemia.[2] This may mean the pupil with SCD becomes tired, finds it difficult to concentrate, and this may be mistaken for being lazy or inattentive.

People with SCD can suffer from the disease to varying degrees. Some have mild forms of SCD, whilst for others it can be far more severe, leading to the need for regular blood transfusions.[2]

Sickle cell disease sufferers can lead reasonably normal and long lives, if the disease is managed by timely recognition and appropriate treatment.[2]

WHAT IS THE PUPIL GOING THROUGH?

It is important to acknowledge that the symptoms of SCD can vary from person to person, as can their severity. Every child is different but the common symptoms are outlined below:

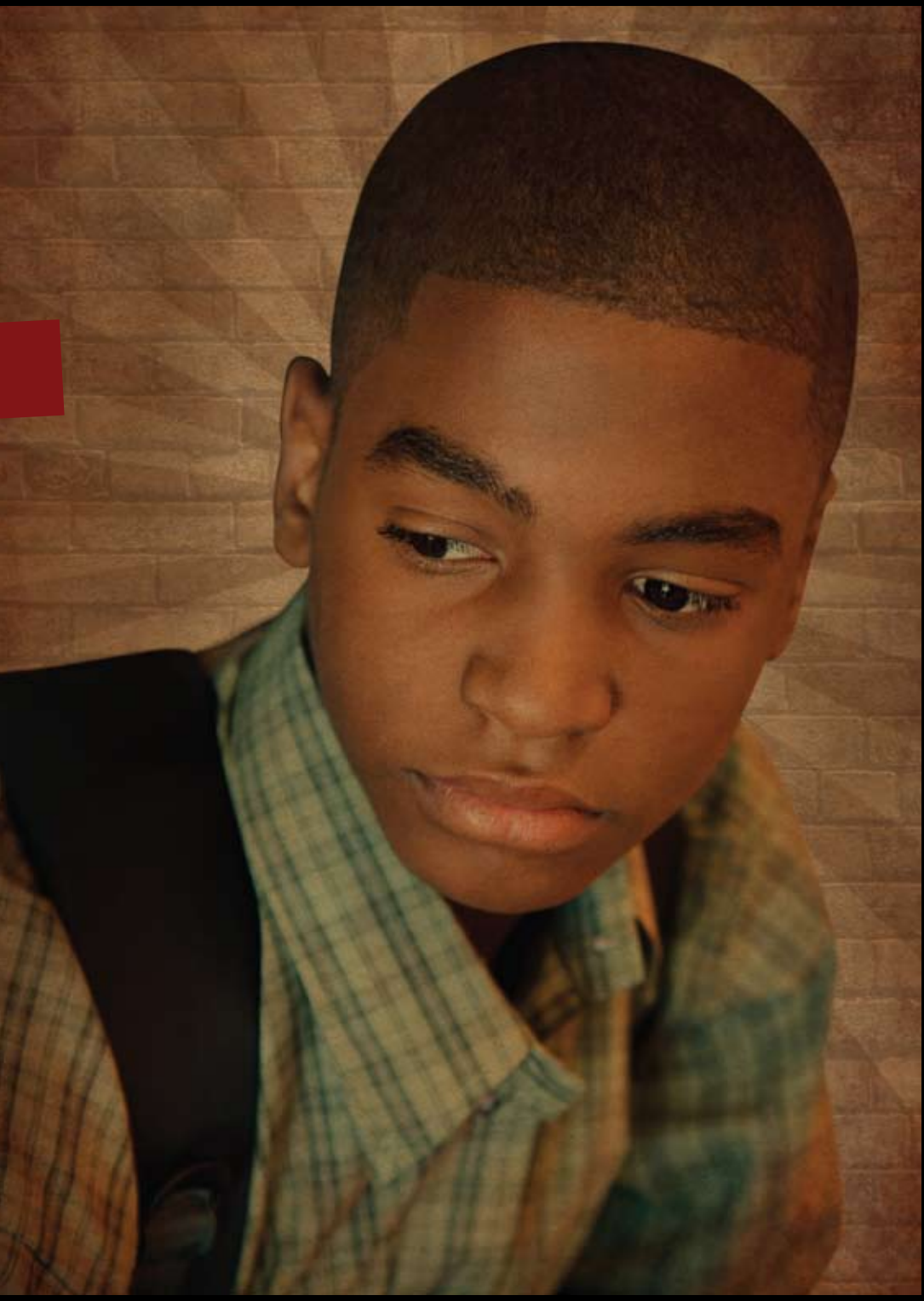


Normal blood cells

Blood cells of a person with SCD

- Anaemia – As your pupil's body is not getting all the oxygen it needs, they will often feel tired, weak and out of breath
- Jaundice – Caused by the liver not working properly due to the strain of having to remove broken down sickled cells, one sign of this might be yellow eyes
- Swelling of the hands and feet
- Frequent infections – The sickle cells can also damage the spleen – an organ that helps the body to develop immunity and fight infections. Failure to develop good immunity will increase their risk of getting an infection. From babyhood people with sickle cell disease are given daily antibiotics to reduce the risk of infections[3]

Schools can help by making arrangements for pupils with SCD to receive their antibiotic medicine and painkillers when required.



SICKLE CELL CRISES

Throughout their lives, sickle cell sufferers can expect to experience sickle cell crises. The severity and frequency of sickle cell crises varies from person to person and can be different each time. Some people experience one or two per year and will be able to control the symptoms using over-the-counter (OTC) or prescribed painkillers, whereas others may require hospital treatment.

Symptoms of a crisis to look out for:

- Gradually worsening pain in the bones and joints
- Severe pain in the abdomen with rigidity (inflexibility) of the muscular wall
- Fever
- Difficulty breathing and a stabbing chest pain on breathing
- Weakness on one side of the body and, if the brain is affected, seizures are possible
- Pain in the upper abdomen from the liver and spleen
- Persistent and painful erections (priapism) in boys/men[2]

A sickle cell crisis can often occur without warning, or due to a number of reasons. Precautions can be taken to reduce the risk and severity of a sickle cell crisis:[2]

- Drinking plenty of water, and remaining hydrated especially in the summer
- Avoiding heavy or strenuous exercise
- Eating healthily
- Avoiding extreme temperatures, particularly the cold and wind and also the heat in summer
- Avoiding stress

What to do if you suspect a pupil is having a sickle cell crisis

If a child is experiencing a mild pain crisis, you can provide them with plenty of fluids and allow them to lie down and rest or do whatever makes them feel comfortable; as long as you have had prior agreement from their parents. If necessary, they can be given a mild painkiller and the parents should be informed immediately. Treating mild pain quickly and effectively can prevent it from becoming severe.

If the child is complaining of severe pain or is experiencing the symptoms of a dangerous complication, such as chest pain or problems breathing, they may need hospital treatment. Hospital treatment can involve pain relief, blood transfusions, supplemental oxygen and antibiotics.

S tay with pupil but alert other members of staff

I f they ask for their painkillers, provide them

C all parents immediately, if unavailable, call hospital doctor

K eep other pupils calm

L isten. Let the pupil remain in whatever position they feel comfortable. They will tell you if they need their parents or the hospital called

E valuate child for signs of stroke, if you suspect a stroke, dial 999

Risk of stroke

Of all people with SCD, children have the highest stroke risk. The risk is greatest between ages of 2 and 16. Children with SCD are 300 times more likely than normal children to have a stroke. Of the children who have had a stroke and survived, 70% will have a second stroke and it usually occurs within 36 months of the first stroke.[4] A stroke should be treated as a medical emergency and it is important to act fast. There is generally a three-hour window of time for treatment.[4]

The first signs that someone has had a stroke are very sudden. Symptoms include:

- Numbness, weakness or paralysis on one side of the body (signs of this may be a drooping arm, leg or lower eyelid, or a dribbling mouth)
- Slurred speech or difficulty finding words or understanding speech
- Sudden blurred vision or loss of sight
- Confusion or unsteadiness
- A severe headache

Use the Face Arm Speech Test (FAST)

Three simple checks can help you recognise whether someone has had a stroke

These are:

Facial weakness: Can the person smile? Has their mouth or an eye drooped?

Arm weakness: Can the person raise both arms?

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

Why is it important to be aware of the needs of a pupil with sickle cell disease?

New research published in the British Educational Research Journal entitled, 'Reported school experiences of young people living with sickle cell disease in England' highlights that the lack of awareness of sickle cell disease in some schools is having a serious negative impact on the education of children living with the condition.[5]

A survey of 569 young people with sickle cell disease in England has found such pupils miss considerable periods of time from school, typically in short periods of two or three days. One in eight has school absences equating to government-defined 'persistent absence' and students with sickle cell report that they are not helped to catch up after these school absences.[5]

A lack of appreciation of the measures needed to prevent exacerbation of sickle cell symptoms was also apparent in the research:[5]

- Half the children reported not being allowed to use the toilet when needed and not being allowed water in class
- A third reported being made to take unsuitable exercise and being called lazy when tired
- Children surveyed perceived both the physical environment (temperature, school furniture) and the social environment (being upset by teachers or other pupils) as triggers to episodes of their illness

'Teachers may know that a child has sickle cell but they might not appreciate the full range of factors that can influence the child. Current frameworks do not appear to be supporting the inclusion of children with sickle cell disease in schools, therefore I am proud to support the Broken Silence initiative in its aims to educate and raise awareness of sickle cell both amongst young people and their teachers.'
Lead researcher, Simon Dyson

KEY CONSIDERATIONS IF YOUR PUPIL HAS SICKLE CELL DISEASE

It is important to understand that sickle cell is a complex and painful disease that the child will have to live with for the rest of their life.

Sickle cell disease is an inherited blood disorder and is NOT contagious.

Key considerations:

HYDRATION It is important that children with sickle cell disease drink plenty of water in order to remain hydrated. Furthermore, since their kidneys are unable to concentrate urine they need to be permitted to use the toilet more frequently

MONITOR EXERCISE Children with sickle cell may be unable to participate in strenuous physical exercise lessons because their cells have difficulty carrying oxygen. Particular attention should be paid when exercising outside during the winter as cold weather may trigger a crisis

MUST STAY WARM It might be necessary for children with sickle cell to wear warm clothing (even their coat) at all times. Their body temperatures are different to somebody without sickle cell

UNDERSTANDING The anaemia associated with sickle cell can cause tiredness and apparent lack of concentration; teachers are encouraged to be understanding in these circumstances

ABSENCES Children with sickle cell may miss days at school due to their illness; teachers are encouraged to allocate time to assist the child to catch up with their work as and where necessary

DISCRETION Do not force the child to disclose their sickle cell, provide avenues in order for them to approach you for help anonymously

WORK TOGETHER Work closely with the child's parents in order to effectively communicate regarding progress and areas for improvement

UNIQUE Remember that sickle cell disease is different in each person. Not everybody suffers from sickle cell in the same way. What works for one child might not be the same as the next