Sickle Cell Anaemia Policy

'Please believe me when I say I am in pain. The pain is like hammering into bone'

What is Sickle Cell Anaemia?

It is not infectious. Sickle cell disorders are inherited blood conditions. They affect thousands of people in this country.

There are 3 main types of sickle cell disorder:

- sickle cell anaemia
- haemoglobin sickle cell/haemoglobin C disease - SC
- sickle beta thalassaemia

All of them cause attacks of pain called **crises**, but sickle cell anaemia is the commonest and often the most severe type. In sickle cell anaemia the red blood cells tend to change from their normal round shape to a half-moon or sickle shape. This can cause a blockage in the blood flow of small blood vessels virtually anywhere in the body. These "sickle" red blood cells do not live as long as the normal 120 days and this results in a chronic state of anaemia.

More than 9 out of every 10 hospital admissions for sickle cell disorders are for painful crises. If not properly treated in certain cases, sickle cell crisis can cause premature death.

What causes Sickle Cell disorders?

Children can inherit sickle cell disorders if both of their parents carry the sickle cell trait. The parents themselves are usually perfectly healthy. The trait is found mainly in people from Africa, the Caribbean, the Middle East, Asia and the Mediterranean or their descendants.

Triggers to Sickle Cell Crises

Factors that can trigger the red blood cells to 'Sickle':

- a lowered level of oxygen or fluid in the tissues
- sudden changes of temperature
- infections
- strenuous exertion

Some people with sickle cell may have few problems, while others are seriously incapacitated. Others may find they have good and bad patches.

Crises

A **crisis** can affect different parts of the body.

The most common type is long bone pain. Children can experience painful swelling of the hands and feet. Young adults can suffer from pain all through the backbone accompanied by massive muscular spasm. Hips and shoulders may also be affected.

There are 2 types of pain that may indicate the possibility of more dangerous, possibly fatal, conditions:

- rib pain anywhere around the chest often precedes silting up of the lungs.
- abdominal pain often precedes liver malfunctions

The length of a crisis also varies – it can last for as little as several hours to as long as several weeks. Most crisis last 3 – 5 days, with pain throughout the day.

The treatments for a crisis depend on its severity. They can range from home treatment with mild pain killers such as paracetamol to hospital administration of powerful drugs.

The most difficult aspect of the disorder is dealing with the pain during a crisis. It generally comes without warning and sometimes with no apparent cause. The lack of control over when the pain is going to occur is very upsetting. The pain itself is sometimes so violent that some sufferers feel they cannot survive it.

Sports

Many children with sickle cell disease are keen on sporting activities but should not be pushed beyond their limitations as they are likely to start sickling. A fundamental problem is that affected children may not be able to balance their enthusiasm for sports against the limitations of their illness. These children may be quite adept at sporting activities, but may find that participation is accompanied by the pain of sickling crisis. The emotional and psychological implications are significant as feelings of frustrations may often overwhelm these children.

Children affected with this illness will inevitably recognise their own abilities. For example a boy taking part in a football match may want to sit down before half time and substitution should be made if he indicates that he is tired. Although teachers may be reluctant to rule out participation in any one sport, cross country running is very taxing physically for children with sickle cell disease and children may be admitted to hospital in pain at the end of these races. **These long distance runs should be avoided**.

Swimming is an enjoyable sport for many children but precautions must be taken that children are dried well once out of the pool. Afro-Caribbean children often have the type of hair that will not dry in a few minutes and this must be taken on board by teachers. Children with sickle cell disease should be encouraged to wear swimming caps and it is advisable to get the children out of the water several minutes before the end of the swimming lesson, as this would provide the extra time needed for children to dry off properly. If there is a warm area available the child should be allowed to wait in this area for class mates before making the journey home or back to school. **Extra time for drying off and warming up is important for reducing the risk of sickling after a swim.**

Gym exercises are a common feature of many schools' sporting activities. Some children are more prone than others to have joint sickling and this problem may become more pronounced with certain types of gym exercises. If children complain of pain with certain exercises it is advisable that these exercises are pursued more gently or changed to a different type of exercise or discontinued altogether.

School Journeys

School journeys are an exciting time for children however, for those with sickle cell disease extra preparation and planning may be required.

It is not necessary to exclude the child from any trip away with the school but the impact on health should be considered when the trips are planned.

The choice of location is of course determined by desirability, costs, relevance to the curriculum, but where medical facilities are poor or inaccessible the choice of location may be unsuitable for a child with sickle cell disease.

Medical Emergencies

Not all sickling crisis are medical emergencies. Children do usually have some warning of the onset of a crisis as there is a painful signal. If the child is able to cope, is uncomfortable but not distressed, is able to drink fluids and is communicable, there is time for the parent or guardian to collect the child from school.

The sudden onset of acute pain accompanied by much distress would indicate a major crisis. Another firm indicator of a major crisis is the appearance of shock or any such collapse. **In these situations a paramedic ambulance should be called immediately.**

Key Notes for Teachers

1. Sickle cell disease in general does not affect mental intelligence.
2. Children who have experienced strokes may have impaired mental function.
3. Delay in learning is usually attributed to the disruption caused by frequent hospital admissions.
4. Where children have missed significant time from school, due to ill health, educational support could be considered.
5. Children admitted to hospital may not be well enough to be discharged home but may feel able to undertake light school work whilst in hospital.
6. It is recommended that all children with sickle cell disease have routine eye testing from the age of 10 years.
7. Where children have difficulty reading, visual problems should initially be excluded prior to the consideration of other assessments.
8. As there is no cure for sickle cell disease more emphasis should be placed on the prevention of sickling crisis.
9. Most children are able to participate fully in the school's curriculum and can take part in sporting activities.
10. Activities that pre-dispose sickling crisis should be limited or discouraged eg. hiking, cross country, sprinting.
11. Sudden changes in temperature could cause sickling crises.
12. Pain brought on by activities usually ceases once these activities are stopped.
13. As a child with sickle cell anaemia should always drink plenty of fluids – remember he/she may need to go to the toilet more frequently.

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It is the intention to review this policy annually.