

INTRODUCTION

Sickle cell anaemia is a hereditary condition affecting the haemoglobin contained within red blood cells. It predominantly affects people of African or Afro-Caribbean origin, but can also affect people of Mediterranean, Middle Eastern and Asian origin.

When an acute sickle cell crisis occurs, the red blood cells change shape from the usual bi-concave discs to an irregular or crescent shape. These cells become unable to carry oxygen effectively, and begin to clump together. This leads to reduced blood flow in the capillaries causing tissue hypoxia and a marked reduction in the life of the cells involved.

HISTORY

A previous history of sickle cell anaemia and sickle cell crisis will be present in most cases, with the patient almost always being aware of their condition.

The crisis may follow as a result of an infection, during pregnancy, or after the patient has been anaesthetised.

These painful crises can result in damage to the patient's lungs, kidneys, liver, bones and other organs and tissues. The recurrent nature of these acute episodes is the most disabling feature of sickle cell anaemia, and many chronic problems can result, including leg ulcers, blindness and stroke. Acute coronary chest syndrome (ACS)^{1,2} is the leading cause of death amongst sickle cell patients.

Signs & symptoms:

- severe pain, most commonly in the joints of the arms and legs, but also in the back and abdomen
- difficulty in breathing
- high temperature, reduced oxygen (O₂) saturation, cough and chest pain may indicate ACS
- swelling of the joints
- pallor
- tiredness/weakness
- dehydration.

MANAGEMENT^{3,4}

- the patient will often be able to guide their care from the usual practice for them and they may have an individualised treatment plan available.

Follow **medical emergencies guideline**. In addition:

- administer high concentration O₂ (**refer to oxygen protocol for administration and information**) via a non-re-breathing mask, using the stoma in laryngectomy and other neck breathing patients. High concentration O₂ should be administered routinely, whatever the oxygen saturation, except in patients with chronic obstructive pulmonary disease (COPD) (**refer to COPD guideline**), as this helps to counter tissue hypoxia and reduce cell clumping
- check 12-lead ECG. This may be the only indication of the presence of ACS (**refer to ACS guideline**)
- patients with a sickle cell crisis will not have acute fluid loss, but may present with dehydration resulting in reduced fluid in both the vascular and tissue compartments, if they have been ill for an extended period of time. Often this has taken time to develop and will take time to correct. Gradual rehydration over many hours rather than minutes is indicated. Consider obtaining IV access.
- all sickle cell patients should be offered pain relief⁵ (**refer to pain management guidelines**), and this should initially be through administration of **Entonox** (**refer to Entonox protocol for administration and dosage**) (**NOTE:** Entonox should not be used for extended periods for sickle cell patients). Consider use of IV analgesia (**refer to pain management guidelines**)
- position the patient so as to minimise pain
- patients should not walk to the ambulance, as this will exacerbate the effects of hypoxia in the tissues
- unless there is a life-threatening condition present, patients in sickle cell crisis should be transferred to the specialist centre where they are normally treated.

Key Points – sickle cell crisis

- Sickle cell anaemia is a hereditary condition affecting the haemoglobin contained within red blood cells; the cells are irregular in shape and become unable to carry oxygen effectively
- Sickle cell crises can result in damage to the lungs, kidneys, liver, bones and other organs and tissues
- Sickle cell crises can be painful and patients should be offered pain relief
- Administer high concentration oxygen therapy
- Acute coronary chest syndrome (ACS) is a leading cause of death amongst sickle cell patients, therefore, check 12-lead ECG.

REFERENCES

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- ³ Gurkamal V, Harold B, Donohoe R. A Snapshot clinical audit examining the management of Sickle Cell Crisis: London Ambulance Service NHS Trust: Available from: http://www.londonambulance.nhs.uk/aboutus/trustboard/media/Trust%20Board_28Sept04.pdf, 2004.
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METHODOLOGY

Refer to methodology section.