

Sickle Cell Crisis - update

INTRODUCTION

Sickle cell disease 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.

The red cells of patients with sickle cell disease are prone to assuming a permanently sickled shape when exposed to a variety of factors including hypoxia, cold or dehydration. These cells are prone to mechanical damage, hence the haemolytic anaemia in this group of patients, and to occluding the microvasculature leading to tissue hypoxia and pain or end organ damage.

HISTORY

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

A crisis may follow as a result of an infection, during pregnancy, following surgery or a variety of other causes including mental stress.

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 disease, and many chronic problems can result, including leg ulcers, blindness and stroke. Acute Chest Syndrome^{a,12}

^aAcute Chest Syndrome (also known as chest crisis)

This is a common and potentially life threatening complication of painful crises, and is often precipitated by a chest infection.

The patient becomes breathless, hypoxic and tachypnoeic / tachycardic over a short period of time. Chest pain is often present, and the hypoxia responds poorly to inhaled oxygen. Crackles are often present in the lung bases and will ascend rapidly to involve the whole lung fields in severe cases. Radiological changes follow late and patients may be critically ill with near normal radiology.

If a chest crisis is suspected, treatment should be initiated with inhaled oxygen and intravenous fluids. In hospital, intravenous antibiotics and urgent exchange transfusion will be instituted after discussion with the haematology team. Intensive care and mechanical ventilation may be required in some cases.

Pulmonary embolus is an important differential diagnosis.

is the leading cause of death amongst sickle cell patients.

Signs & symptoms (any of these may apply):

- severe pain, most commonly in the long bones and/or joints of the arms and legs, but also in the back and abdomen
- high temperature,
- difficulty in breathing, reduced oxygen (O₂) saturation, cough and chest pain may indicate Acute Chest Syndrome
- pallor
- tiredness/weakness.
- dehydration
- headache
- priapism.

MANAGEMENT³

- the patient will often be able to guide their care and may have an individualised treatment plan available; if this is the case this plan should be followed.

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

- **Oxygen** - helps to counter tissue hypoxia and reduce cell clumping. Administer supplemental oxygen to all patients including those with chronic sickle lung disease:
 - **children** - administer high levels of supplemental oxygen
 - **adults** - administer 2-6 litres of supplemental oxygen per minute via nasal cannulae or 5-10 litres of supplemental oxygen per minute via a simple face mask until a reliable SpO₂ measurement is available then adjust oxygen flow to aim for target saturation within the range of 94-98% or what is known to be normal for the individual patient.
 - administer supplemental oxygen via the stoma in laryngectomee and other neck breathing patients.
- Check 12-lead ECG in patients with chest pain to exclude obvious cardiac causes. (**refer to acute coronary syndrome guideline**).

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- 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 relief⁴ (**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 the use of opiate analgesia, preferably orally or subcutaneously rather than intravenously. The dose should be guided by the patient's hand-held record if available, otherwise refer to pain management guidelines.
- 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 disease is a hereditary condition affecting the haemoglobin contained within red blood cells; the cells are irregular in shape and occlude the microvasculature leading to tissue ischaemia.
- Sickle cell crises can result in damage to the lungs, kidneys, liver, bones and other organs and tissues.
- Sickle cell crises can be very painful and patients should be offered pain relief.
- Administer supplemental oxygen to all patients including those with chronic sickle lung disease.
- Acute Chest Syndrome is a leading cause of death amongst sickle cell patients and is characterised by hypoxia and tachypnoea.

REFERENCES

1. Yale SH, Nagib N, Guthrie T. Acute chest syndrome in sickle cell disease. Crucial considerations in adolescents and adults. *Postgraduate Medicine* 2000;107(1):215-8, 221-2.
2. Vichinsky EP, Neumayr LD, Earles AN, Williams R, Lennette ET, Dean D, et al. Causes and Outcomes of the Acute Chest Syndrome in Sickle Cell Disease. *N Engl J Med* 2000;342(25):1855-1865.
3. Standing Medical Advisory Committee. Report of a working party of the Standing Medical Advisory Committee on Sickle Cell, Thalassaemia and Other Haemoglobinopathies. London: HMSO, 1994.
4. Maxwell K, Streetly A, Bevan D. Experiences of hospital care and treatment seeking for pain from sickle cell disease: qualitative study. *BMJ* 1999;318(7198):1585-1590.

METHODOLOGY

Refer to methodology section:
http://www2.warwick.ac.uk/fac/med/research/hsri/emergencycare/prehospitalcare/jrcalcstakeholderwebsite/a-z/specific/sickle_cell/.