Sickle Cell Crisis [276–279]

1. Introduction

- Sickle cell disease is a hereditary condition affecting the haemoglobin contained within red blood cells.
- 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.
- The signs and symptoms include (**any of those listed below 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
  - stroke
  - high temperature
  - difficulty in breathing, reduced oxygen (O_2) saturation, cough and chest pain may indicate Acute Chest Syndrome
  - pallor
  - tiredness/weakness
  - dehydration
  - headache
  - priapism.

2. Incidence

- There are different types of sickle cell disease found mainly in people of African or Afro-Caribbean origin, but can also affect people of Mediterranean, Middle Eastern and Asian origin. In the United Kingdom it is estimated that 15,000 adults and children suffer from sickle cell disease with 1 in every 2,000 babies born with the condition.

3. Severity and Outcome

- 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] is the leading cause of death amongst sickle cell patients.

4. Pathophysiology

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

5. Assessment and Management

For the assessment and management of patients with sickle cell crisis refer to Table 3.42 or Figure 3.4.

Methodology

For details of the methodology used in the development of this guideline refer to the guideline webpage.

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.**
- **Patients with sickle cell disease can be dangerously ill but in no pain (e.g. aplastic crisis, stroke, hepatic sequestration, PE, etc.).**
- **Acute Chest Syndrome is a leading cause of death amongst sickle cell patients and is characterised by hypoxia and tachypnoea.**

[a]**Acute 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 are likely to 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.