Wessex and Thames Valley Haemoglobinopathy Network

Adult Haemoglobinopathy Service

Acute chest syndrome in patients with sickle cell disease

Introduction
The Acute Chest Syndrome (ACS) is an unique pulmonary illness in patients with sickle cell disease (SCD) injury that may lead to acute respiratory distress syndrome and death. ACS is a leading cause of death in adults with sickle cell disease and is important as early recognition and aggressive treatment will reduce morbidity and mortality. The key to successful management of ACS is awareness, anticipation of its development, particularly in high-risk patients, and timely intervention with specific treatments such as oxygen and blood transfusion.

Recurrent episodes of ACS can lead to chronic lung disease.

Diagnosis
ACS may be defined as the combination of:
- Respiratory symptoms - cough, sputum production, dyspnoea, tachypnoea, wheezing, chest pain, hypoxia or fever > 38.0°C
- New pulmonary infiltrates on CXR

Risk factors for development of ACS include:
- Haemoglobin SS phenotype
- Low steady state HbF%
- Past history of ACS
- Infection
- Anaesthesia
- Pregnancy
- Rib infarction
- Sedation with opiate analgesia
- Recent / current painful crisis

Presentation
- ACS may be a presenting feature but equally may occur some days into an acute vaso-occlusive crisis.
- There can often be a rapid clinical course
- Pain is characterized by a ‘T-shirt’ distribution and its severity will usually cause splinting of the diaphragm, further impairing oxygenation resulting in progressive hypoxia. Coughing is a late symptom.
- Signs of lung consolidation (often bilateral), such as bronchial breathing, crackles or reduced breath sounds, accompanied by tachycardia and tachypnoea. If ACS is clinically suspected, a positive chest x-ray defines the disease, but a single negative chest x-ray cannot exclude it.

Investigations
- Chest x-ray
- Arterial blood gases: these should be done on room air even if the patient is on inspired oxygen
- FBC, retics, Hb HPLC (if genotype unknown)
Urea, creatinine and electrolytes, liver function tests, calcium, CRP
Group and save (if not already performed); extended phenotype if not known
Blood, sputum and urine cultures
Serum/Ur ine for atypical organism screen (Mycoplasma, Legionella)
Nasopharyngeal viral swabs

**Clinical Management**

All patients with SCD should be assessed for ACS at presentation to hospital and during admission, irrespective of the reason for admission.


Consider ACS if any of the following clinical features are present:

- Pain in chest wall, upper abdomen, and/or thoracic spine.
- Cough (may be a late symptom)
- Signs of lung consolidation; usually bilateral and generally starting at the bases.
- Fever > 38°C
- Tachypnoea
- Tachycardia
- Hypoxia (O₂ sats < 92% on room air or declining >2% from previously documented baseline saturations)

**Management – haematology referral**

- If the patient has presented to a non-haematology service (e.g. emergency department or medical take):
  - **OUH**: please alert the haematology registrar on bleep 1836 (during office hours) or via switchboard (after 5pm and at weekends).
  - **DGH**: During office hours contact Haematology SpR or Consultant on call; out of hours for emergency advice Registrar / Consultant may contact Consultant haematologist on call.
- Patients with suspected ACS at **OUH** are best managed on the clinical haematology ward. **DGH**: they should remain under the acute medical service until a member of the haematology team has formally agreed to take over.
- Where there is no scope for transfer to the haematology ward, management should be guided by haematology input.
- Decisions regarding transfusion MUST be taken by the Haematology team
- Contact Critical Care team according to local early warning score criteria

**General management**

- Monitor pO₂ on air, pulse, respiratory rate, BP
- Oxygen therapy to increase oxygen saturations >96%
sickle-cell-disease/S8-painful-crises-in-scd.pdf

- IV fluids. Maintain careful fluid balance, avoiding fluid overload
- Antibiotics: since ACS may have an underlying infective aetiology, and since its clinical manifestations may be difficult to separate from respiratory infection, empiric broad-spectrum antibiotic cover is advised. Referral to local antibiotic guidelines. Atypical organisms must also be covered.
- Bronchodilators may be of benefit if there is evidence of bronchospasm or wheeze or history of obstructive / reversible airways disease.
- Incentive Spirometry (refer to physiotherapy)
- Transfusion – see below

**Transfusion**

Early transfusion may be lifesaving and can result in improvement within hours. It aims are to enhance oxygen-carrying capacity, improve tissue oxygen delivery and reduce HbS concentration, thereby reducing sickling and preventing progression to acute respiratory failure.

**Top-up Transfusion**

Recent guidelines (2014) reiterate the importance of early top up in ameliorating the clinical course of incipient chest syndrome. Top-up is advised for stable patients with early features of ACS, particularly with falling Hb levels. Aim for post-transfusion Hb of no more than 100 g/l. Where the patient’s resting Hb is >90 g/l top-up transfusion is unlikely to be feasible.

**Exchange Blood Transfusion**

Indicated in the context of:
- Rapid or significant clinical deterioration
- Worsening CXR changes / multi-lobar involvement
- \( \text{O}_2 \) sats < 90%, suggesting \( \text{PaO}_2 < 70 \text{ mmHg} \)
- Baseline Hb > 90 g/l

For full exchange: Assess venous access; aim for Hb S < 25% and HCT not > 0.35 (see NSSG/Red cell/Exchange transfusion guidelines for further detail [link here](#))

**Critical Care Support**

Support should be requested from the Intensive Care Team for patients with oxygen saturations <90% despite maximal \( \text{O}_2 \) supplementation; for those with evidence of respiratory failure (\( \text{PaO}_2 < 60 \text{ mmHg} \) or \( \text{PCO}_2 > 50 \text{ mmHg} \)); and for those with evidence of a rapid decline in clinical status. For these patients, ventilatory support may be required.

**Primary Prevention of ACS**

- ACS should be anticipated in high-risk situations e.g. peri-operatively.
- Consideration of pre-emptive exchange transfusion
Secondary Prevention of ACS
- Recurrent ACS is associated with reduced survival.
- Hydroxycarbamide has been shown to reduce the frequency of ACS by 50%.
- Observational data suggests regular transfusions can also prevent ACS
- All patients suffering ACS should have outpatient review within one month of discharge to determine if hydroxycarbamide or a transfusion programme will be of benefit.

References


Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK © Sickle Cell Society 2018

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