

GSTFT Clinical Practice Guideline

Management of Acute Chest Syndrome in Children with Sickle Cell Disease

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Background

- Pulmonary complications are the leading cause of morbidity and mortality in children with Sickle cell disease (SCD).
- The acute chest syndrome (ACS) is the most frequent reason for hospital admission in children, while in adults it is secondary to painful vaso-occlusive crises. In early childhood there is usually a history of infection, whereas in older children and adults it more typically occurs during or following a vaso-occlusive crisis or after a general anaesthetic.
- ACS is likely to be multifactorial in origin with infection, fat embolism (1), sickling and endothelial activation likely to be important.
- Children with SCD will often develop pneumonia. Broadly speaking, these patients are usually mildly unwell, do not have bilateral chest signs, do not have chest pain and are well oxygenated in air. It should be noted that any child with a sickle cell crisis may go on to develop ACS and as such all children in sickle cell crisis should be closely monitored for such a complication.
- There is very little in the way of randomised controlled trials in the treatment of acute chest syndrome in sickle cell disease as demonstrated by the Cochrane reviews on the subject (2-5). The recommendations here are based on often small or retrospective or non-randomised studies.

Definition of ACS

The appearance of a new pulmonary infiltrate on chest x-ray, accompanied by pleuritic chest pain, fever, tachypnoea and abnormal chest examination (6) .

Presentation:

- fever, tachycardia
- Pain (often pleuritic) in chest wall, abdomen, spine
- Respiratory distress, hypoxia, cough
- Sign of lung consolidation, often bilateral, generally starting at the bases, but may be unilateral and difficult to distinguish from pneumonia. Note symptoms and signs often precede x-ray changes.

Management

1. **Initial assessment – all patients in whom ACS is suspected should have the following performed ASAP within 30 minutes on arrival to A+E or if developing ACS during an admission for another complication.**
 - Routine clerking as per all sickle patients with particular vigilance aiming to identify those patients who are showing evidence of severe illness requiring urgent admission, transfusion and ventilatory support.
 - Alert the Haemoglobinopathy team within 4 hours
 - Intravenous access
 - Observations/ Investigations see below
2. Observations / Monitoring – hourly minimum
 - HR, BP, SaO₂, RR, Temp
 - Conscious level
 - Use Paediatric Early Warning System (PEWS) charts
 - pain score
 - Admit all patients who have a differential diagnosis of ACS
3. Blood tests
 - FBC and Reticulocytes, U+E, LFT, CRP, LDH,
 - Cross match 35ml/kg blood for exchange transfusion (discuss with transfusion laboratory)
 - Blood cultures, serum lactate levels
 - Throat swab/ NPAs for virology sputum culture, M. pneumoniae IgM, Parvo B19 IgM
 - Arterial blood gases – If saturations <94% OR clinically deteriorating and likely to need ventilation OR decreased conscious state
4. Radiology
 - CXR
 - CT head if neurological concerns
5. Oxygen
 - Oxygen replacement therapy should be used to keep SaO₂>99% in all patients regardless of patients baseline/steady-state oxygen saturations.

6. Intravenous fluids

- Give to all patients with ACS unless very well with sufficient oral intake. Starting rates- 0.9% saline + 5% Dextrose 2/3 rd maintenance / review.

0-1 years	80 ml/kg/24 hours
1-3 years	60 ml/kg/24 hours
4-6 years	50 ml/kg/24 hours
≥7 years	40 ml/kg/24 hours
- Rate of infusion may need to be modified based on fluid losses, fever and of course be aware of syndrome of inappropriate ADH secretion (**SIADH**) and reduce fluids to 2/3rd standard maintenance. Monitor plasma Sodium in all patients. Restrict fluids if hyponatraemia develops
- Close attention should be paid to monitoring fluid balance, in particular avoiding fluid overload which may precipitate or exacerbate respiratory failure.

7. Antibiotics

- It is considered standard practice to administer antibiotics with up to 14% of children under 2 having documented bacteraemia (7) and particularly as it may be difficult to differentiate between ACS and pneumonia.
- Cefuroxime 50mg/kg 8hrly and Clarithromycin should be given to all patients (IV Erythromycin if unable to take oral medication)- paed formulary for dosages
- If clinical deterioration occurs after 24-48 hours of first-line antibiotics change to Tazocin (Piperacillin/Tazobactam) and Gentamicin while continuing Clarithromycin.
- Adjust antibiotics as per microbiology advice once culture results available.

8. Blood transfusion

- Blood transfusion is the mainstay of treatment for a chest crisis.
- Aim for a post transfusion Hb of ≤10g/dl Hct≤0.30.
- In a severely anaemic patient this may be achieved with a simple top up, however, other patients will need an exchange blood transfusion. This should be performed as soon as possible, and may need to be repeated until desired parameters are reached and patient is stable or improving.
- See protocol for exchange and top up transfusion (Do URL link here)

9. Analgesia

- Adequate pain control is essential.
- Monitor for opiate toxicity.

10. Physiotherapy

- Incentive spirometry can prevent the development of ACS and is indicated in all children presenting with chest or back pain and SCD (reference). Incentive spirometers are kept with Clinical Nurse Specialist (Helen Appleby) in Haemoglobinopathies on level 6, Evelina Children's Hospital.
- Chest physiotherapy may be helpful in those with atelectasis/consolidation and a productive cough.

11. Dexamethasone

- 0.3mg/kg at 12 hourly intervals for a total of 4 doses. Shown by a prospective study to reduce hospital stay, prevent clinical deterioration, and may reduce the need for blood transfusion (8), though not all studies demonstrate this.

12. Bronchodilators

- Give a trial of regular nebulised salbutamol in patients with wheeze, history of airway hyper-reactivity, previous benefit from a trial of nebulisers or clinical deterioration. Mucus plugging is extremely common in ACS (over 80% have significant mucus plugs on bronchoscopy (9) though once again no RCT here either (4)) Consider mucus plugs as possibility in patients with wheeze who have no response to bronchodilators. Physio may be indicated in these cases

Complications

1. Deteriorating patients

Patients with ACS require close monitoring as clinical deterioration can be abrupt and severe. Studies indicate that up to 10% may need ventilatory support.

Signs of deterioration

- Decreasing level of consciousness
- Progressive hypoxia on air, Increasing tachypnoea >50 (1st year) >40 (>1yr)
- Increasing oxygen requirements to maintain (or failure to reach) SaO₂ ≥96%

- High blood lactate , Progressive pulmonary infiltrates on chest x-ray

Management of the deteriorating patient

- Discuss early on (as soon as have assessed the patient) with Consultant Paediatrician /Paediatric Haematologist and PICU team.
- **Now Discuss with Retrieval Unit.....**
- Consider the following options
 - Re-assess fluid balance in particular looking for evidence of fluid overload.
 - Assess for opiate toxicity.
 - Exchange transfusion – indicated in rapidly deteriorating patients with extensive chest infiltrates and hypoxia that does not correct with high-flow oxygen therapy. Requires central venous access and should be performed on PICU.
 - Mechanical ventilation
 - Indicated for rapidly deteriorating or tiring patients with severe hypoxia that does not correct with high-flow oxygen. All ventilated patients should undergo exchange transfusion.
 - Inhaled Nitric Oxide may be of benefit in ACS (no good evidence for this (2)). We would not go down this line as a matter of routine as it is “cosmetic”. Only if we cannot oxygenate and standard therapy fails. Early bronchoscopy is indicated in all sickle chests where possibility of mucus plugging is high (rapid changes/ new areas of atelectasis/plugging on CXR. If mucus plugs are present , intratracheal DNase with physiotherapy is used
 - Cerebral event
 - There is a strong correlation between ACS and neurological events. These may include seizures, silent infarcts, cerebral haemorrhage and reversible posterior leuco-encephalopathy syndrome. Care should be taken to control hypertension and to avoid a haematocrit >0.30 after transfusion. Consider urgent CT/MRI if new neurological symptoms or signs develop.

2. Chronic chest syndrome

A chronic restrictive lung deficit can follow repeated episodes of ACS and in its severest form can lead to pulmonary hypertension in later life. Whenever possible all patients should have pulmonary function testing carried out 6-8 weeks after recovery from ACS, looking in particular at flow/volume loops and gas transfer.

Discharge from hospital

Prior to discharge patients should:-

- Afebrile, off IV antibiotics for at least 24 hours
- Have normal/baseline oxygen saturations with no requirement for supplementary oxygen-except if pre-existing oxygen saturation is known to be sub-normal
- Have normal RR/HR/BP

Follow up should be arranged in the sickle cell clinic for 1 week post discharge. Pulmonary function testing should be carried out 6-8 weeks following recovery from each acute episode.

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