# Scottish Paediatric & Adult Haemoglobinopathy Network

## Paediatric Guideline - Acute Chest Syndrome

Acute sickle chest syndrome is an acute illness characterised by fever and / or respiratory symptoms accompanied by new infiltrates on chest x-ray.

It is likely to be multifactorial in origin. Infection can precipitate or complicate ACS, sickling within the pulmonary vasculature leads to infarction and sequestration and fat embolism may also contribute. The distinction between infection and sickling is difficult and management principles should be the same for the two conditions.

Commonly pain in the thorax, upper abdomen or spine leads to hypoventilation, which may be exacerbated by opiate analgesia reducing respiratory drive. Basal hypoventilation leads to regional hypoxia which triggers localised sickling with subsequent infarction and consolidation. Thus, a vicious circle is set up with sickling leading to progressive hypoxia and in turn to further sickling.

Acute chest syndrome is one of the major causes of death from sickle cell disease. A high index of suspicion is needed to detect and treat early. Patients should be treated aggressively irrespective of disease phenotype.

### Triggers

- Infection
- Painful vaso-occlusive crisis
- Post operative
- Opiate induced hypoventilation
- Pulmonary embolus
- Fluid overload

### Symptoms

- Pain (often pleuritic) in chest wall, upper abdomen and/or thoracic spine (T-shirt distribution) but may be absent in younger children
- Dyspnoea / breathlessness
- Wheeze and cough which may be a late symptom.

## Signs

- Fever
- Tachypnoea, tachycardia, increased work of breathing.
- Hypoxia – a useful predictor of severity and outcome
- Signs of lung consolidation, including wheeze and bronchial breathing

Physical signs often precede x-ray changes.

## Differential diagnosis

Sickle lung and pneumonia are often clinically and radiologically indistinguishable. However, consolidation in the upper and/or middle lobes, without basal changes, is suggestive of chest infection rather than sickle chest syndrome. Bilateral disease is most likely due to sickling, but atypical pneumonia should be considered. Pleuritic pain may also be due to spinal/rib/sternal infarction, or from subdiaphragmatic inflammation.

## Investigations

- Baseline FBC, UE and creatinine and LFT, CRP
- Chest x-ray – repeat if normal initially but ongoing clinical suspicion
- Blood, throat and sputum cultures
- Respiratory viral infection screen – nose and throat swabs in viral transport medium for respiratory viruses and mycoplasma.
- Capillary blood gases if $SaO_2$ < 94% - recommend pulse oximetry and CBG to monitor $CO_2$/acid base if required
- Baseline HbS and extended phenotype cross match (if being considered for transfusion)

Markers of severity include worsening hypoxia, increasing respiratory rate, falling platelet count (<200), falling Hb and multilobar changes on chest x-ray

## Management – General Measures

- Inform the Consultant on call for haematology at your regional referral centre if chest syndrome suspected. Discuss patients with PICU/HDU as early as possible
- Transfer of children in low prevalence paediatric units should be considered as soon as the condition is suspected. This may require transfer by the regional retrieval team dependent on the patient's clinical condition. The Inpatient Specialist Management of acutely unwell children with haemoglobinopathies guideline available on the SPAH Paediatric Guideline website page.
- Oxygenation. Maintenance of adequate oxygenation is essential. Options include face mask oxygen, nasal cannulae, optiflow, CPAP and ventilation. Ideally maintain $SaO_2$>96%.
- Pulse oximetry on air at least 4 hourly together with pulse, BP and respiratory rate. Consider continuous pulse oximetry if clinical concerns and CBG
- IV fluids: individualised and guided by the patient's fluid balance and cardiopulmonary status – (see below Fluid Management).

- Pain control: Ensure adequate pain control but with care to avoid opiate induced hypoventilation (see acute pain management guideline)
- Antibiotics: IV Piperacillin Tazobactam and oral azithromycin or clarithromycin (or as per local antibiotic policy).
- Physiotherapy: Incentive spirometry and other measures in conjunction with the physiotherapist. The Physiotherapy Guidelines are available on the SPAH Paediatric Guideline website page.
- Consider regular bronchodilators by nebuliser in patients with wheeze, demonstrable reversible airways obstruction or history of asthma but should not be used routinely
- Consider systemic steroids in severe ACS or acute asthma

## Management – Transfusion

The purpose of transfusion is to:

- enhance oxygen-carrying capacity and improve tissue oxygen delivery
- reduce HbS concentration to reduce sickling
- prevent progression to acute respiratory failure

Transfusion commonly results in impressive improvement within hours.

Simple transfusion is indicated for patients with:

- mild or moderate chest syndrome, particularly with hypoxia and/or falling Hb levels
- aim for a post transfusion Hb level of 100g/l

Exchange transfusions are used to:

- reduce the HbS concentration rapidly;
- avoid problems associated with increased fluid volume and viscosity

Exchange transfusion is indicated when there is evidence of:

- clinical and/or radiological deterioration despite simple transfusion
- worsening x-ray changes
- severe clinical features (see above)
- or if simple transfusion is not possible due to high baseline Hb

Automated RBC exchange transfusion may be performed if available or using a manual method - details of manual exchange procedure is available on the SPAH Paediatric Guideline website page.

## Fluid management

Dehydration occurs readily in children with sickle cell disease due to impairment of renal concentrating ability and may aggravate to sickling due to increased blood viscosity. Patients with acute chest syndrome also have respiratory compromise and may have cardiac compromise so fluid overload must also be avoided.

**Careful assessment of individual fluid status, administration of an appropriate hydration regimen and close monitoring of fluid balance is therefore imperative.**

The ill child should be assessed for the degree of dehydration by the history; the duration of the illness; by clinical examination; and (if known) weight loss. Hb and PCV (Hct) may be elevated as compared with the child's steady state values. These children normally have a low urea and so slight elevation is significant.

An IV line should be established whenever parenteral opiates have been given, or if the patient is not taking oral fluids well. In the less ill patient who is able to drink the required amount, hydration can be given orally. As an alternative consider a nasogastric tube in an alert patient.

- IV hydration should be commenced on admission at maintenance rates or appropriate to individual fluid status.
- A fluid chart should be started and kept carefully, both input and output.
- Fluid balance must be reviewed regularly (at least 12 hourly) to correct dehydration and avoid fluid overload.
- Check urea and electrolytes at least daily and add KCl as required.
- IV therapy can be stopped once the patient is stable and pain is controlled with documentation of adequate oral intake.

**For more comprehensive guidance please refer to the BSCH Guideline on the Management of Acute Chest Syndrome in sickle cell disease. British Journal of Haematology 2015 169 492-505**

*NOTE*
*This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined on the basis of all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.*