NHS SCOTLAND

SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

Paediatric Guidance

Acute Chest Syndrome

NOTE
This guidance is not intended to be interpreted or used as a standard of care. Standards of care are determined based on the full range of clinical information available for an individual case and may change as scientific knowledge, technology, and clinical practice evolve. Following the recommendations in this guidance does not guarantee a successful outcome in every situation, nor does it encompass all appropriate methods of care or exclude other acceptable approaches that may achieve the same results. Clinical judgement remains the responsibility of the healthcare professional(s) involved in the patient's care. Clinicians must also be mindful of the requirements of patient autonomy and patient-centred care, which may involve a patient opting (after discussion of the available treatment options) for a treatment which is not the treatment recommended in the guidance. Where treatment involves deviation from national guidance, or local guidance derived from them, this should be fully documented in the patient's records at the time the decision is made.

This document has been prepared by NHS Public Services Delivery Scotland (PSD Scotland) on behalf of the Scottish Paediatric & Adult Haemoglobinopathies network (SPAH). Accountable to Scottish Government, PSD Scotland works at the heart of the health service providing national strategic services to the rest of NHS Scotland and other public sector organisations to help them deliver their services more efficiently and effectively. SPAH is a collaboration of stakeholders involved in the care of patients with a haemoglobinopathy, who are supported by an NSS Programme Team to drive improvement across the care pathway.

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Stakeholders involved	Consultant Paediatric Haematologists Consultant Paediatricians Haematology Nurse Specialists Paediatric Physiotherapists
Review by Pharmacist	N/A
Methodology used	Evidence-based literature review and engagement with clinical experts within the field of Paediatric Haematology and Sickle Cell Disease
Rationale	The network was established to support and develop haemoglobinopathy services throughout Scotland in improving standards of clinical care for patients. The network supports the delivery of evidence based, patient centred care through the development and implementation of clinical guidelines, care pathways and information resources utilising a once for Scotland approach. Document first created in 2016. To aid healthcare professionals in NHS Scotland in clinical decision making on the management of patients with sickle cell disease presenting with acute chest syndrome
Scope	Haematology specialists and general paediatricians managing children with sickle cell disease
Approval process	The guideline was approved by the SPAH Paediatric Guidance and Protocol subgroup on 10 March 2026. This subgroup has the authority to develop, review and endorse guidelines. This has been agreed by the SPAH Steering Group and noted in its Terms of Reference. See appendix for list of Paediatric Guidance and Protocol Subgroup Members.

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Background

Acute chest syndrome in sickle cell disease is an acute illness characterised by fever and / or new respiratory symptoms or signs 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.

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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, mycoplasma and consider legionella antigen (urine)
- Capillary blood gases if SaO_2 less than 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 (less than 200, or greater than 50% fall from baseline), 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 acute 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 guidance can be referred to.
- Oxygenation. Maintenance of adequate oxygenation is essential. Options include face mask oxygen, nasal cannulae, optiflow, CPAP and ventilation. Ideally maintain SaO_2 greater than 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 Guidance also provides additional information.

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- Consider regular bronchodilators by nebuliser in patients with wheeze, demonstrable reversible airways obstruction or history of asthma but these 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 in the Exchange Transfusion guidance.

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 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.

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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 can 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 potassium as required.
- IV therapy can be stopped once the patient is stable and pain is controlled with documentation of adequate oral intake.

References

For more comprehensive guidance please refer to reference 1.

1. British Committee for Standards in Haematology Guideline on the Management of Acute Chest Syndrome in sickle cell disease. British Journal of Haematology 2015 169 492-505
2. Sickle Cell Disease in Childhood Standards and Recommendations for Clinical Care 3rd Edition (November 2019) www.sicklecellsociety.org/resource/paediatricstandardsrescource/ (accessed 16/06/26)
3. Acute Chest Syndrome in Sickle Cell Disease. Bhasin and Sarode. Transfusion Medicine Reviews 37 (2023)

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Appendix - Guidelines and Protocols subgroup membership

Name:		Job Title:	Organisation:
Katrina	Adams	Non-Malignant Haematology Nurse Specialist	NHS GG&C
Ruth	Allen	Consultant Paediatric Radiologist	NHS GG&C
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