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Scottish Paediatric & Adult Haemoglobinopathy Network

Acute Chest Syndrome in Sickle Cell Disease

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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.

Acute chest syndrome is the leading cause of mortality in adults with HbSS. CONTACT HAEMATOLOGY REGISTRAR ON CALL AS SOON AS SUSPECTED

Acute Chest Syndrome is a spectrum of disease from a mild pneumonic illness to severe respiratory distress syndrome. It is characterised by fever and/or respiratory symptoms and new pulmonary infiltrates on CXR. It can evolve, often developing 24-48 hours after an acute painful crisis. The condition can develop rapidly and clinicians should have a high index of suspicion.

Key Features (patient may not have all of these)

- SaO2 Sats on air <94% or PaO2 <9kPa
- Fever, Cough, Chest pain
- Respiratory distress/hypoxemia
- New opacity on chest x-ray
- Worsening anaemia
- Bilirubin stained sputum
- **Antecedent painful crisis**

Symptoms and Signs

- Hypoxia
- Pain in chest wall, upper abdomen, and/or thoracic spine.
- Signs of lung consolidation; usually bilateral and, generally, starting at the bases (can progress in hours).
- Fever
- Tachypnoea
- Tachycardia
- Wheeze
- Cough may be a late symptom
- Physical signs often precede X-ray changes

Clinical features overlap with those of pneumonia. Consider also in differential diagnosis or as triggering factors

- Pulmonary Embolism
- Fluid overload
- Opiate narcosis and hypoventilation
- TRALI if recently transfused

Initial investigations

- Arterial blood gases on air (if SpO2<95%)
- Chest X-ray
- Group and save – please state patient has ‘Sickle cell disease’ on form (will also need rhesus typing if not previously done)
- Full blood count and reticulocytes
- Renal and liver function tests
- Ferritin and LDH (if significantly raised consider fat embolism syndrome).
- Blood cultures, sputum cultures, Respiratory viral screen, respiratory atypical serology (including Chlamydia, legionella, mycoplasma)
- Consider CTPA only if high clinical suspicion of pulmonary embolism

Initial Management

- Call Haematology Registrar/Consultant
- Monitor SaO2 (on air initially), pulse, respiratory rate
- Oxygen therapy to increase oxygen saturations >95%
- Analgesia as required
- IV fluids – with close monitoring of fluid balance
- IV Co-amoxiclav 1.2g tds plus clarithromycin 500mg bd (or ceftriaxone 2g daily and clarithromycin to cover atypical pneumonia), for 5-7 days (consult local antibiotic policy if penicillin allergic)
- Bronchodilators if there is evidence of wheeze or a history of asthma
- Blood transfusion can be lifesaving in the context of severe acute chest syndrome and is likely to be required in the majority of cases of acute chest syndrome though not all. This should be discussed with on call haematology consultant including out of hours.
- Patients requiring transfusion are likely to need managed in HDU/ITU setting

Additional supportive management

- Incentive spirometry – 10 deep breaths 1-2 hourly when awake
- Chest physiotherapy
- Consider one dose of furosemide 0.5-1mg/kg if signs of fluid overload
- Prophylactic Low Molecular Weight Heparin

Transfusion

Note: Sickle cell patients should have Rh and Kell matched sickle negative blood

- **Simple transfusion,** performed early may help prevent progression of a chest crisis, and may be indicated while arranging exchange. A maximum target Hb of 100-110g/dl (not higher) is recommended.
  - If the patient's pO2<9 kPa on air or the Hb is <60g/l, consider initial top up transfusion.

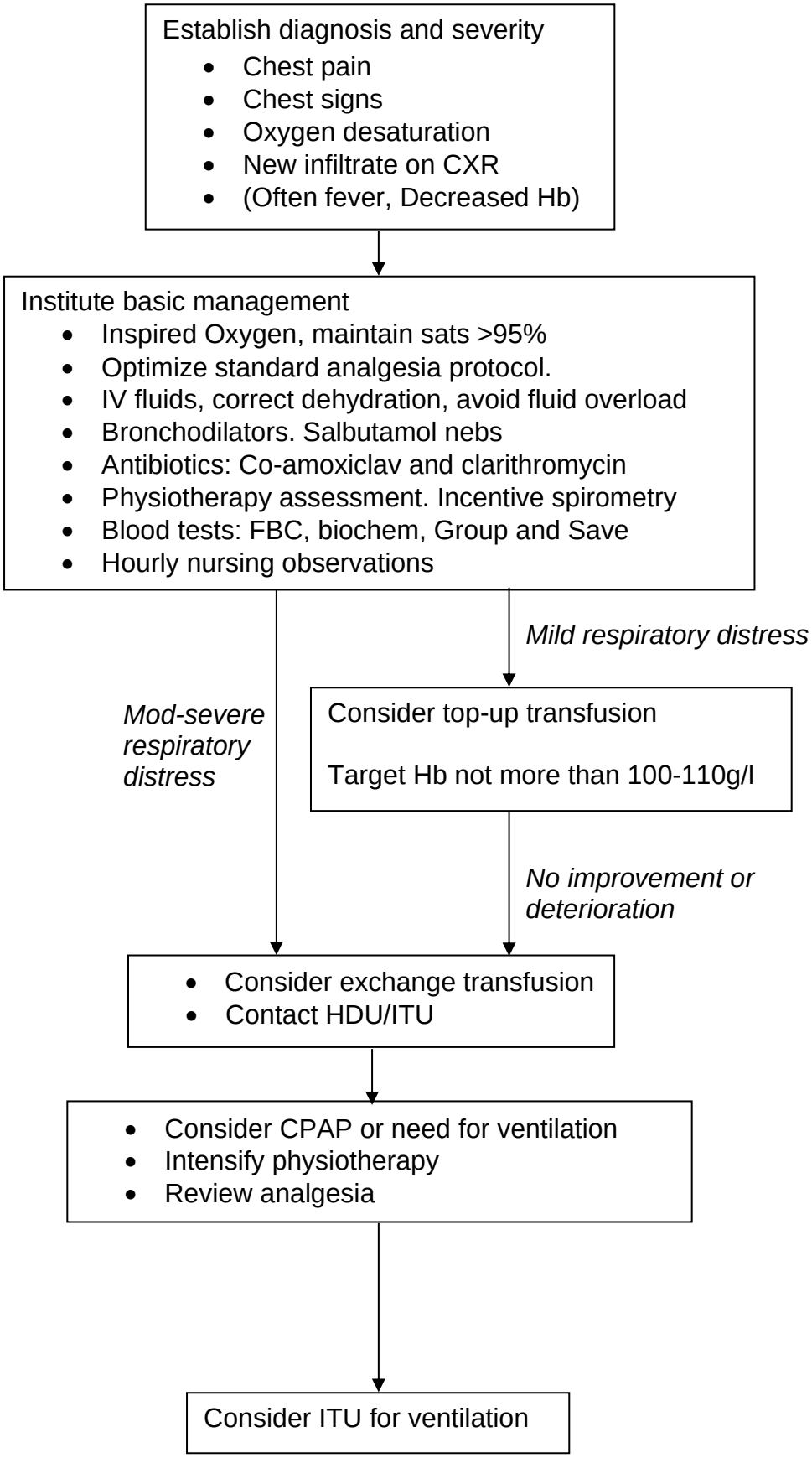
- **Exchange blood transfusion** can be done by the automated or manual method and is indicated in patients with severe respiratory symptoms, deterioration despite simple transfusion or who start with a high Hb concentration (>90g/l) (See "Red Cell Exchange transfusion in adults with Sickle Cell disease" guideline on SPAH website)
  - For either manual or automated exchange transfusion:
    - Cross match 8-10 units of blood depending on size.
    - Source blood warmer if manual
    - Ensure adequate venous access
    - Automated exchange will require insertion of Gamcath unless peripheral venous access is excellent.

Monitoring

- Ensure that vital signs are taken and documented hourly (more frequently if patients' condition is deteriorating)
- Continuous oxygen saturation monitoring (on air) and repeat ABGs if deterioration on oxygen therapy
- Maintain strict fluid balance
- CXR should be repeated in the deteriorating patient
- Daily FBC, reticulocytes, U and Es and LFTs until improving (compare patients baseline values)

Discharge Criteria – only in consultation with Haematologist

- Improved pulmonary symptoms and adequate oxygenation on room air
- Afebrile >24hours and negative cultures if applicable
- Stable/rising haemoglobin
- Tolerating adequate oral fluids and able to take medications orally
- Adequate pain relief on oral analgesia
- Hydroxycarbamide should be considered for prevention of recurrent ACS – decision should be discussed with/taken by Consultant Haematologist.



Taken from the British Society for Haematology "Guideline on the management of acute chest syndrome in sickle cell disease" British Journal Of Haematology, 2015, 169, 492-505