CRISES IN SICKLE CELL DISEASE

Adults with sickle cell disease most commonly experience painful crises. Acute chest syndrome, which results from a combination of pulmonary infarction, infection, fat emboli and red cell sequestration, occurs less commonly in adults than in children but is more severe and can be fatal. Most of this guideline is appropriate for management of all patients but specific advice on management of acute chest syndrome is also included

RECOGNITION AND ASSESSMENT

Symptoms and signs

- Severe pain (usually in extremities, back or abdomen)
- Dehydration
- Enlarged liver or spleen
- Bone pain
- Low grade fever (<38°C) even in absence of infection

Acute chest syndrome

- Pain in chest wall, upper abdomen and thoracic spine
- Fever
- Tachypnoea
- Tachycardia
- Auscultatory crackles and bronchial breathing consistent with bilateral basal consolidation (consolidation in upper or middle lobes is more suggestive of pneumonia)

Previous history

- Ask about:
- previous episodes and optimal analgesia
- family history
- infection
- extremes of temperature
- stress (physical/emotional)

Investigations

Presence of sickle cells in blood film does not correlate with clinical events

- FBC
- Differential WBC
- Reticulocytes
- Biochemical screen
- Group and save (new patients – obtain full red cell phenotype)
- Pulse oximetry
- Chest X-ray only if infection or acute chest syndrome (see below) suspected
- Painful bones need not normally be X-rayed

Acute chest syndrome

- Arterial blood gases (ABG) on air
- Chest X-ray
- Cultures of sputum and blood

IMMEDIATE TREATMENT

- Analgesia
- diamorphine 2.5 - 5 mg SC 4 hrly
- avoid pethidine
- diclofenac 50 mg orally 8 hrly if no contraindications
- Fluid replacement. May be given orally or via NG tube if venous access poor (central lines carry high complication rate)

Avoid using veins in ankles/feet for venous access; cannulation carries high risk of leg ulceration

- glucose (4%) and sodium chloride (0.18%) 1 L by IV infusion over 3 hr; then glucose (4%) and sodium chloride (0.18%) with potassium chloride 20 mmol/L by IV infusion every 6 hr
- monitor U&E for dilutional hyponatraemia

Always use commercially produced pre-mixed bags of infusion fluid and potassium chloride. NEVER add potassium chloride to infusion bags

- If SpO_2 <95%, give O_2
- Antibiotics only if evidence of infection present and after cultures taken
- In patients with rib/thoracic spine pain, incentive spirometry may reduce incidence of acute chest syndrome by encouraging maximal inspiration – discuss with physiotherapist (alternatively, encourage patient to take 10 maximal inspirations every hour)

Acute chest syndrome
As above plus:
Antibiotics

Assume penicillin allergy only if convincing history of either rash within 72 hr of dose or anaphylactic reaction. If any doubt about whether patient truly allergic to penicillin, seek advice from a microbiologist .	
Amoxicillin 500 mg IV 8 hrly **plus** clarithromycin 500 mg IV by infusion into large proximal vein 12 hrly	Clarithromycin 500 mg IV by infusion into large proximal vein 12 hrly **plus** gentamicin – see **Prescribing regimens and nomograms *in Trust Antibiotics Policy***
As soon as oral route available: amoxicillin 500 mg orally or via NG/PEG tube 8 hrly **plus** clarithromycin 500 mg orally or via NG/PEG tube 12 hrly	Clarithromycin 500 mg orally or via NG/PEG tube 12 hrly

- O_2 via face mask
- if PaO_2 remains low (<9 kPa) or progressively declining with evidence of bi-basal consolidation, discuss exchange transfusion with haematologist
- Nebulised salbutamol

Manual partial exchange transfusion

Perform exchange or transfusion therapy in patient with hypoxia and multi-lobe involvement

- Target Hb is 9-10 g/dL
- Crossmatch required amount of blood as packed cells
- Establish venous access if not already available
- If Hb <5 g/dL, transfuse packed cells until Hb 9-10 g/dL
- If Hb ≥5 g/dL, the following method is favoured for rapid partial exchange:

Step 1
- Bleed 1 unit (500 mL) of blood from patient, infuse 500 mL of sodium chloride 0.9%

Step 2
- Infuse 2-3 units of blood (target Hb 9-10g/dL)

Step 3
- If patient has large red blood cell mass or continues to deteriorate, repeat steps 1 and 2

SUBSEQUENT MANAGEMENT

- Assess efficacy of analgesia after 1 hr and adjust dose if necessary
- Painful crises usually last about one week
- Once pain controlled, reassess analgesic regimen daily and taper dosage gradually, changing to oral morphine as dosage is reduced (1 mg SC diamorphine = 3 mg oral morphine)
- If Hb falls below 5 g/dL, especially if reticulocyte count also decreased, crossmatch and transfuse two units

MONITORING TREATMENT

- Respiratory rate hrly after opioid started
- Pulse oximetry
- Fluid balance
- Hb (after each exchange) to ensure it does not exceed 10-11 g/dL and increase risk of hyperviscosity
- target HbS of 30-35% is useful in some patients but less valuable than clinical response
- Consider visual analogue scale to record pain intensity and response to analgesia

Acute chest syndrome
- ABG
- FBC and reticulocytes
- Pulse oximetry

DISCHARGE AND FOLLOW-UP

- Discharge home when pain controlled by oral medication
- Provide three to four days' supply of analgesia
- Do not prescribe parenteral opioids on TTO