Sickle Cell Disease – Acute Chest Syndrome (ACS)

ACS can be severe in all sickle genotypes
Call SORT for all cases, we will coordinate with local haematology

Recognition / Evaluation

Fever, chest pain, hypoxia < 94% on air, cough, wheeze, increased work of breathing, tachypnoea, crackles, bronchial breathing
Often 1-3 days after onset of painful veno-occlusive crisis
The chest examination may be normal!

Immediate Care

Give Oxygen aim SpO_2 >= 95%
Maintenance adequate hydration
Monitor and target fluid balance
(risk of pulmonary oedema)
Analgesia
Antimicrobials
(treat all empirically for pneumonia and cover atypicals)
Bronchodilators
(if acute bronchospasm or history of asthma)
Chest physiotherapy
Incentive Spirometry
*(start with 10 max inspirations every 2 hours during day and while awake at night **OR** consider positive pressure devices)*

Immediate Investigations

CXR *(?infiltrates, may be normal early)*
Consider ABG for PaO_2
(note SpO_2 may be normal in chronic anaemia)
FBC + blood film + reticulocytes
U&Es, LFTs, LDH, coag screen, CRP
(monitor sodium and beware hyponatraemia, systemic fat emboli, risk multi-organ failure)
Request Group & Save urgently
(alloimmunisation is common and expect delays)
Blood cultures
Atypical serology and urinary antigens
Sputum and nasopharyngeal aspirate for culture and PCR for respiratory viruses and influenza
ECG
Consider start Rx and request CTPA if suspicion PE
Hb electrophoresis – HbS levels?

Before Discharge

Penicillin V / Folic acid
Pneumococcal and Seasonal Vaccinations
Discussing start Hydroxycarbamide

Consider Differentials
(note these may all trigger ACS)

- Pneumonia
- Opiate narcosis
- Pulmonary oedema
- Pulmonary embolism
- TRALI
- Alveolar hypoventilation due to pain

Predictors of Severity

↓Sats , ↑RR
↓Hb ↓Platelets
Multi-lobar involvement on CXR

SEVERE DISEASE
(Call SORT Early)

Use NIV or HFNC Early
Consider Invasive Ventilation with:

Worsening hypoxaemia or
Severe dyspnoea or
Respiratory acidaemia (pH < 7.35)

Consider Simple Transfusion

Give 10ml/kg PRCs if Hb >10g/L below baseline or Hb <90 g/L
Target Hb: 90-110 g/L
If not anaemic consider moving straight to exchange transfusion

Consider Exchange Transfusion

Worsening hypoxaemia, rapidly progressive disease
Order 40ml/kg of PRCs
Targets:
Hb 100-110 g/L
HbS <30-40%
Aim HbS <30% if severe enough for invasive ventilation
Consider corticosteroids after exchange if not improving