Sickle Cell Disease - Acute Chest Syndrome - Clinical Guideline, management and referral pathway

Ref: CG-HAEM/2023/003

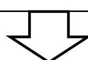
Presentation with respiratory signs and symptoms (including hypoxia +/- new CXR infiltration

Fever ++ Tachypnoea + Cough ++ Wheezing +/- Chest pain ++ Skeletal pain ++ Dyspnoea ++ Haemoptysis+

A high clinical suspicion is vital to early diagnosis

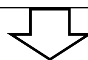
Acute Chest Syndrome can be a severe life-threatening condition

Think ahead of Exchange Transfusion and need for transfer to Nottingham City Hospital



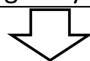
Investigations

CXR, FBC, UEs, LFTs Blood group and screen (or crossmatch)
Blood cultures Sputum cultures Serology/urine for atypical respiratory organisms
SpO2 ≤95% (on air) on pulse oximetry or a fall in SpO2 of 3% or more from a steady state should be confirmed with ABG promptly



Management

Fluids, Pain relief, Oxygen, Antibiotics +/- bronchodilators
Contact and inform on-call Consultant Haematologist for advice
Above investigations may need to be repeated in the deteriorating patient
Inform Blood Bank early on about possible blood transfusion requirement
Top ups/Manual exchanges are possible locally, discuss with Haematologist on call, If an automated exchange is required the patient should be transferred to NCH, this transfer should be arranged by the Derby/Burton Haematologist talking to the Nottingham non-malignant team



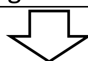
Falling pO2

Hb<70g/l or fall in of >Hb30g/d

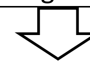
Top up transfusion

ITU outreach team review

Low threshold for Exchange Transfusion



pO2(air) <9.0kPA
Hb >70g/dl and/or rapid deterioration and/or indicators of severity (Fall in Hb >20g/dl, fall in platelets >200, falling GCS, increasing RR



pO2(air) <8.0kPA
pCO2 >6.0kPa

or Rapid deterioration

ITU review

Consideration of NIV/IV

Consider Exchange Transfusion

Transfer to Nottingham for Automated Exchange

Document Controls:

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Consultation with:	
Approved By:	CDCS Dec 2023
Review Date:	Dec 2026
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