

North West London Haemoglobinopathy Clinical Network

Guidelines for the transfer of adult sickle cell disease patients to the intensive care unit / high dependency unit

1. Background

The National Confidential Enquiry into Patient Outcome and Death (NCEPOD) in 2008 analysed all haemoglobinopathy deaths in the UK over a two year period. 40 adult deaths occurred in sickle cell disease patients. The majority of deaths occurred in hospital. The main causes of death were stroke, multi-organ failure, acute chest syndrome, and pulmonary embolism.

This guideline focuses on identifying patients early that may benefit from escalation of care.

2. Sickle emergencies requiring urgent admission:

- Agonising pain requiring opiate analgesia
- Increasing pallor, breathlessness or exhaustion
- Marked fever (> 38⁰C)
- Tachycardia, tachypnoea, or hypotension
- Chest pains, signs of consolidation or desaturation
- Abdominal pain or distension
- Diarrhoea and vomiting
- Any abnormal neurological signs including headache, fitting, drowsiness
- Priapism > 4hrs

3. Indications for Escalation of care:

- Acute respiratory failure- as in acute chest syndrome where the oxygen levels are not maintained with supportive measures on the ward, or the patient is becoming exhausted from respiratory effort.
REMEMBER acute chest syndrome is associated with a significant morbidity and mortality. The mortality rate in adults is 4% increasing to 20% once ventilated. Prompt diagnosis and management, with early involvement of Haem Registrar/Consultant, is essential. It is often multifactorial in origin with infection, sludging/ sickling or thrombosis of pulmonary arteries and fat embolism giving a similar clinical picture.

Further guidance on patient care pathway is provided in Table 1, Appendix 1.

- Multi- organ failure
 - Planned post elective surgery
 - Pulmonary emboli with haemodynamic instability
 - Septic shock with haemodynamic instability
 - Renal failure requiring haemodialysis eg anuria, hyperkalaemia
 - Critically ill, unstable patients who require treatment and monitoring not available outside of an ICU
 - Acute stroke with altered mental status or coma
- Patients with a stroke need to be managed by a neurosurgical unit with access to automated exchange (Charing Cross Hospital via the hyper acute stroke unit). Discuss with the red cell attending/ on call consultant at Hammersmith Hospital.**

All cases should have a multidisciplinary approach. All cases for consideration of ICU care should be discussed directly with the consultant intensivist. If accepted to ICU the haematology team will review the patient daily and liaise with ICU staff to ensure multidisciplinary care. The haematology team are contactable at all times for advice and should be contacted if the patient's condition deteriorates.

3. Transfer to a Tertiary Centre

Central Middlesex Hospital and Hammersmith Hospital are the North West London Haemoglobinopathy Network's two tertiary acute care centres for adults. They provide a 24/7 service for advice on all sickle related queries. Automated red cell exchanges are available weekdays 9-5 at Central Middlesex and 24/7 at Hammersmith Hospital. Any patients that are being considered for red cell exchange can be discussed with the tertiary centres at any time. Please contact the red cell Spr or red cell attending/ on call consultant at either centre.

Once the decision has been made for exchange transfusion, this should be completed as quickly as possible at either the local or specialist centre depending on individual circumstances.

For those patients who are due elective surgery and require exchange transfusion, please refer to Dr Gavin Cho at Central Middlesex or Dr Mark Layton at Hammersmith Hospital, with the proposed date of surgery. A date for surgery should be made well in advance of the referral to allow sufficient time to prepare patients adequately.

Appendix 1

(Read in conjunction with the Network acute chest syndrome guidelines)

PaO ₂	All patients	Incentive spirometry (as per pain protocol)
	8.0 - 9.5 kPa	CPAP
	< 8.0 kPa	Discuss exchange transfusion with registrar/consultant and CPAP
	< 7.5 kPa	Exchange transfusion and CPAP
	< 7.5 kPa (on increasing insp O ₂ (60% or CPAP)	Intubate, IPPV-ventilation & exchange transfusion
PaCO ₂	> 6.7kPa or rising and patient tiring	Consider intubate, IPPV-ventilation & exchange transfusion

Table 1: Escalation of care guidance for sickle cell adult patients with deteriorating respiratory condition