Guidelines for the Immediate Management of Paediatric Patients with Sickle Cell Disease (SCD) and Acute Neurological Symptoms

Document Information			
Version:	2	**Date:**	Sept 2014
Authors (incl. job title):	Sue Height Consultant Paediatric Haematologist Prof David Rees Honorary Consultant Paediatric Haematologist		
Responsible committee	**Child Health Clinical Governance & Risk Committee**		
Review date:	Sept 2016		
Target audience:	Paediatric haematology, paediatric medical and nursing staff		
Stakeholders/ committees involved in guideline development :	Paediatric haematology, paediatric medical and nursing staff		

For Child Health Clinical Guidelines Groups' use only	
Assessed by:	**Child Health Clinical Guidelines Group**
Assessment date:	09/09/2014
Approved by :	**Child Health Governance and Risk Committee**
Approval Date	16/09/2014
Author notified:	20/09/2014

For Evidence Based Practice Committee use only	
Ratified by:	
Date ratified:	
Reference No.:	
Date when guideline comes into effect:	

Document History

Document replaces: 1 Sept 2012

Consultation distribution (before ratification)

Sent to	Version	Date	Actions taken as a result
Professor Rees	2	Sept 2014	Agreed

Reviews and updates (including CGG comments)

Date	New version no.	Summary of Changes	Author of change/s
Sept 2014	2	Minor changes – addition of cardiolipin and b2 microglobulin antibodies	Sue Height
		Including neck vessels in neuro-imaging and TCDs	"
		Need to contact on call anaesthetist for GA for MRI	"

Dissemination schedule (after ratification)

Target audience(s)	Method	Person responsible
Paediatric Medical, Haematology and Nursing staff	On Kings Guidelines System	Guidelines process

Guidelines for the Immediate Management of Paediatric Patients with Sickle Cell Disease (SCD) and Acute Neurological Symptoms

Abstract Page

Background

This guideline is intended to assist medical and nursing staff in the immediate assessment and management of patients with sickle cell disease and acute neurological symptoms under the care of the paediatric haematologists at King's College Hospital.

Contents of guideline (quick reference guide to determine if guideline is useful to reader)

Guidelines for the Immediate Management of Paediatric Patients with Sickle Cell Disease (SCD) and Acute Neurological Symptoms

Definition/Background

This guideline is intended to assist medical and nursing staff in the immediate assessment and management of patients with sickle cell disease and acute neurological symptoms under the care of the paediatric haematologists at King's College Hospital.

Cerebrovascular Accident (CVA) - neurological event lasting >24hrs +/- new areas of abnormality on scan
Transient Ischaemic Attack (TIA) - focal event lasting <24hrs with no abnormality on scan

- The peak age for CVA is 2-7 years, and occurs in 8% of children with SCD.
- The anterior cerebral circulation is most commonly involved, leading to weakness, paraesthesiae, speech or behavioural abnormalities.
- Visual symptoms can occur in isolation and should prompt an urgent assessment

Pre – Requisites for Practitioner
This is intended for paediatric, haematology medical and nursing staff

Indications
Child with Sickle Cell Anaemia presenting with neurological symptoms who may have had a CVA or TIA

Contra-Indications
No Contraindication

Equipment required
No particular equipment is required

Presentation

The usual presentation is a child who is well, and has either weakness not associated with pain, or a history of a TIA – in some cases the history from the parents may be the only indication of a problem.
If either diagnosis is suspected there is a need for urgent management – admit, investigate and treat pending results of investigations.

Less commonly, patients are clinically sick in association with a neurological event ***which may include stroke,*** consider the following alternative/additional diagnoses and investigate and treat accordingly:

- Meningitis or encephalitis, fit or febrile convulsion, syncope, SAH, vaso-occlusion of calvarium, cerebral malaria, trauma, fat embolism, hypoglycaemia, drugs, abscess, or tumour.

Immediate Investigation and Management

- Admit to HDU
- BM stix, iv access and send urgent blood tests:
    - FBC, reticulocytes and film
    - Blood group (ABO, RhD and Kell) & antibody screen and urgent cross-match (request sickle negative blood).
    - Urgent exchange transfusion – contact PICU – protocol will be individualised for patients taking into account starting Hb and clinical condition.
    - DAX and CRP
    - INR/APTR/Fibrinogen and D-Dimers,
    - Haemoglobin analysis for HbS%, HbF% if patient not known to King's
    - Consider malaria screen, auto- and ds DNA antibodies, cardiolipin and beta2 microglobulin antibodies.
- Supportive treatment - iv fluids to maintain blood glucose, O_2, treat fever aggressively
- Neurological observations, BP and O_2 saturations
- Assess and secure airway (discuss with PICU urgently if reduced consciousness)
- Urgent CNS imaging (discuss with neuroradiology), usually urgent CT scan to exclude bleed or tumour first, and then arrange MRI/MRA of head and including neck vessels (sickle protocol). This usually requires GA or sedation for children <7years, and should not be performed until the child is clinically stable and the blood transfusion completed.
- If there is likely to be a delay in arranging the exchange, and the Hb is <8 g/dl, then an urgent top-up transfusion to Hb 10g/dl should be undertaken.
- Perform exchange transfusion as soon as blood is available - discuss with PICU
- If the CT scan shows an intracranial bleed, request an urgent neurosurgical opinion and intervention as necessary. Exchange transfusion will still be necessary but may be performed before, during or after the surgery depending on the urgency and timing of surgery.
- Steroids, aspirin, mannitol or hyperventilation are not routinely indicated for the management of ischaemic CVA in SCD
- Request paediatric neurology review as in-patient

Additional investigations:

- Urinalysis
- Viral serology - HSV, VZV, CMV, Parvovirus, Hepatitis A, B and C.
- Blood, urine and throat swab for cultures and ASO titre if febrile
- Consider LP after imaging if meningitis suspected
- Transcranial Doppler (TCDs) and assessment of extracranial vessels: discuss with vascular lab (Ext 33711)
- Urine & serum drug screen if altered mental status with no explanation

Subsequent investigations to be arranged

- MRI/MRA including neck vessels and perfusion-weighted images (discuss with Dr Jarosz or Dr Sibtain – consultant neuro-radiologists). Children <7~8 years may require GA and the ward paediatric staff will need to contact the on-call anaesthetist.
- TCDs including extracranial ICAs if not performed already
- ECG
- Sleep Study – contact Children's Community Nursing Team or discuss with respiratory team if remaining as an inpatient with possible OSA on ward.
- Trans-thoracic cardiac echo (discuss with Paediatric Cardiology Team at Evelina and download referral from ECH cardiology website to fax across). Further investigations may be needed to exclude a Patent Foramen Ovale – bubble studies or trans-oesophageal echo (under GA) – discuss with paediatric cardiology team at Evelina.

Further Management:

- Arrange for regular transfusions on Philip Isaacs' Ward (Ext 34200)
- Referral to Dr Keith Pohl, Consultant Paediatric Neurologist c/o Paediatric Haematology Secretary, KCH.
- Physiotherapy, Speech & Language Therapy Occupational Therapy referrals for assessment and treatment.
- Neuropsychometric Assessment – referral to Clinical Psychologist with Sickle Cell Team.

Referrals from other hospitals – arrange transfer to HDU directly with plan for exchange transfusion. Discuss with paediatric haematology consultant first.

Reference: Transfusion Guidelines for Neonates and Older Children British Committee for Standards in Haematology transfusion Task Force *2004 British Journal of Haematology* ***124;*** *433-453*