

## South Thames Children's Sickle Cell and Thalassaemia Network - Guidelines for the shared-care of children with inherited anaemias

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<b>Responsible committee or Director:</b>	Child Health		
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<b>Stakeholders/ committees involved in guideline development :</b>	Paediatricians, paediatric haematologists, haematologists		

**For Clinical Guidelines Groups' use only**


## **South Thames Children's Sickle Cell and Thalassaemia Network - Guidelines for the shared-care of children with inherited anaemias**

This guideline covers the shared-care of children with sickle cell disease, thalassaemia and other inherited anaemias between specialist (SHT) and local (LHT) haemoglobinopathy teams. It is aimed at all doctors and nurses involved in the care of these patients, both at King's College Hospital (KCH), the Evelina Children's Hospital (ECH) and in the hospitals who refer patients to KCH or ECH for shared-care (South Thames Children's Sickle Cell & Thalassaemia Network). Other members of the multi-disciplinary paediatric and haematology teams may find it helpful. It applies to patients aged 18 years and under who have not yet been transferred to the care of the adult services.

### **Background**

Children with sickle cell disease and thalassaemia are increasingly born in and moving to areas with a low prevalence of the condition. These children receive shared care between their local hospital and a specialist haemoglobinopathy clinic. It is very important that the child is known to the local haemoglobinopathy team, in that acute admissions and regular review will occur locally. The specialist haemoglobinopathy clinic provides annual reviews, specialist MDT clinics, transcranial Doppler scanning, telephone advice and inpatient care for patients who require or might require HDU/PICU. In our network, the specialist services are provided by King's College Hospital and the Evelina Children's Hospital, who work closely together. Most of the local haemoglobinopathy teams are in Southeast London, Kent, Surrey or Sussex, although children are also referred from all over the UK. Most of the local hospitals have low numbers of patients, although there are three with more than 200 patients each: Lewisham University Hospital, Queen Elizabeth Hospital Woolwich and Mayday University Hospital Croydon.

### **South Thames Children's Sickle Cell & Thalassaemia Network**

#### *Local Haemoglobinopathy Teams*

Dr Elizabeth Sleight  
Consultant Neonatologist  
Lewisham University Hospital  
Lewisham, London

Dr Paula Robertson  
Consultant Paediatrician  
Queen Elizabeth Hospital  
Woolwich

Dr Grant Marais  
Consultant Paediatrician  
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Dr Riaz Ahmed  
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Paediatric Department  
Medway Maritime Hospital  
Gillingham, Kent

### *Specialist Haemoglobinopathy Teams*

Dr David Rees, Dr Sue Height, Dr Moira Dick  
Department of Paediatric Haematology  
King's College Hospital NHS Foundation Trust  
London

Dr Baba Inusa, Dr Jayanthi Alamelu  
Department of Paediatrics / Haematology and Haemophilia  
Evelina Children's Hospital  
Guy's and St Thomas' NHS Foundation Trust  
London

### **Communication between Local and Specialist Haemoglobinopathy Teams**

- Letters will be written following every clinic attendance.
- All correspondence from the specialist haemoglobinopathy team will be copied to the local team.
- All correspondence from the local haemoglobinopathy team will be copied to the specialist team.
- All correspondence will be copied to the GP.
- Children with particular complications or needs will have a care plan which will be agreed by both hospitals.
- All shared-care patients will be offered a hand-held patient record, which will be filled in by both local and specialist teams.
- Regular network education and review meetings will be organised by the SHTs.

### **First Hospital Visit**

- The parents will be informed of the diagnosis according to local arrangements, which may involve specialist nurse counsellors, health visitors, GPs and paediatricians.
- The patient should be seen in hospital by three months of age at the local hospital, to which the child would be taken when acutely unwell.
- At the first hospital visit the diagnosis should be discussed with the parents and tests performed to confirm the diagnosis.
- Penicillin V prophylaxis should be started at or before the first visit.
- The LHTs and SHTs will be contacted monthly by the Neonatal Screening Laboratory to ensure that all affected babies have been seen in clinic.

### **Follow-up Appointments and Annual Review**

- The baby and family will normally be offered 3 monthly appointments until 2 years of age, and then 6-12 monthly appointments. These will be split between the local and specialist haemoglobinopathy teams.
- A suggested regimen is given below, although this may vary depending on local facilities, and the needs and wants of the baby and family.
  - Annual review blood tests should approximately alternate between SHT and LHTs.
  - TCDs and annual reviews will typically be at the SHT although for some hospitals with large numbers of patients, alternative arrangements may be made.

- Lewisham: weekly visiting TCD service from KCH with annual reviews by Dr Sleight and referral to SHT as necessary.
- Mayday Hospital Croydon: monthly joint clinic with Dr Marais, Dr Rees/Height and TCD service from KCH.
- Queen Elizabeth Hospital, Woolwich: monthly joint clinic with Dr Robertson, Rees/Height and TCD service from KCH.
- Other joint review clinics may be established throughout the network depending on patient numbers local needs.

	3 months	6 months	9 months	12 months	15 months	18 months	21 months	24 months	30 months	36 months	42 months	48 months
location	LH	LH	LH	SH	LH	LH	LH	SH	LH	SH	LH	SH
Blood tests	*			*			*			*		
TCD scan <sup>+</sup>				*				*		*		*
Annual Review				*				*		*		*
Other								23vPS				

LH=local hospital, SH=specialist hospital, 23vPS=23-valent pneumococcal polysaccharide vaccine (Pneumovax). <sup>+</sup> SCD patients only.

### Shared-care and Inpatients

- In general patients will be admitted to their local hospital initially.
- The specialist haemoglobinopathy teams will be happy to discuss the management of the patients at an early stage if appropriate.
- KCH or ECH should be contacted at an early stage if the patient is likely to require HDU or PICU support, including patients with neurological problems and acute chest syndrome.
- Contact details:
  - KCH
    - 9am-5pm
      - paediatric haematology registrar
        - 020 3299 9000 bleep 268
      - Paediatric haematology secretary
        - 020 3299 3773 or 0203 299 3242
    - 5pm -9am
      - On-call haematology registrar
        - 020 3299 9000 – ask switchboard to bleep
  - ECH
    - 9am-5pm
      - Paediatric Sickle Cell Team
        - 02071889432 / 02071887774
    - On-call paediatric registrar
      - 020 7188 7188 Bleep 0339

## **Hydroxyurea**

- Hydroxyurea will usually be started following discussions with both the local and specialist haemoglobinopathy teams, and review at either KCH or ECH.
- Hydroxyurea will usually be prescribed and monitored by the local hospital
- Patients taking hydroxyurea may be reviewed more frequently by the specialist haemoglobinopathy team, typically every six months.

## **Children receiving regular, long-term blood transfusions**

- This includes children with thalassaemia major, and those with SCD and cerebrovascular disease
- Regular transfusions will occur in the local hospital.
- Reviews will typically occur in at the specialist haemoglobinopathy centre every 3-6 months.
- Iron chelation will usually be started and monitored by the specialist haemoglobinopathy team, in conjunction with the local team.
- Audiometry and ophthalmology reviews will occur locally

## **Specialist Haemoglobinopathy Network Clinics**

- The following specialist, multidisciplinary network clinics occur, and children can be referred directly to these by either the local or specialist haemoglobinopathy teams.
- These are usually run by a specialist with input from by Dr Dick, Dr Height, Dr Inusa or Dr Rees.
  - Neurology clinic
    - Specialist consultant: Dr Keith Pohl
    - Location: KCH and ECH
    - Referrals: haemoglobinopathy and neurological problems
  - Renal clinic
    - Specialist consultant: Dr Caroline Booth
    - Location: ECH
    - Referrals: haemoglobinopathy with persistent or progressive proteinuria, hypertension or renal impairment
  - Neuro-revascularisation clinic
    - Specialist consultants: Mr Sanj Bassi, Mr Christos Toliias, Dr Keith Pohl
    - Location: KCH
    - Referrals: progressive or very severe cerebral vasculopathy despite optimal transfusion therapy
  - Bladder clinic
    - Specialist consultant: Dr Anne Wright
    - Location: ECH
    - Referrals: nocturnal enuresis or day-time enuresis
  - Hepatology clinic
    - Specialist consultant: Dr Dino Hadzic
    - Location: KCH
    - Referrals: hepatitis, sickle hepatopathy
  - Orthopaedic clinic
    - Specialist consultant: Mr Om Lahoti

- Location: KCH
  - Referrals: avascular necrosis, osteomyelitis
- Ear Nose and Throat
  - Specialist consultant Mr Ian Hore
  - Location: ECH
  - Referrals: sleep-disordered breathing, nose bleeds
- Cardiology
  - Specialty Contact: Dr John Simpson
  - Location: ECH
  - Referrals: cardiac murmurs, possible pulmonary hypertension

Dr David Rees  
May 2010