UK Forum on
Haemoglobin
Disorders

WM QRS West Midlands
Quality Review Service

Services for Children with Sickle Cell Disease or Thalassaemia at King's College Hospital NHS Foundation Trust

---

Quality Review Visit Report
Visit date: May 20th 2010

Report finalised: December 5th 2010

CONTENTS

INTRODUCTION

This report presents the findings of the peer review visit to services for children with sickle cell disease or thalassaemia at King's College Hospital NHS Foundation Trust which took place on May 20th, 2010. The purpose of the visit was to review compliance with the '*Quality Requirements for Health Services Caring for Children and Young People with Haemoglobinopathies', 2009*. The visit was organised by the West Midlands Quality Review Service.

ACKNOWLEDGEMENTS

We would like to thank the staff of King's College Hospital NHS Foundation Trust [KCH] for their hard work in preparing for the review and for their kindness and helpfulness during the course of the visit. Thanks too to the parents who took time to come and meet the review team. Thanks are also due to the visiting team (Appendix 1) and their employing organisations for the time and expertise they contributed to this review.

SICKLE CELL AND THALASSAEMIA SERVICES AT KINGS COLLEGE HOSPITAL

Service (as at 2010)	Patient numbers sickle cell disease	Patient numbers thalassaemia	Patients on long term red cell transfusions	Patients on hydroxy carbamide
KCH	478 [326 SS, 101 SC]	~ 20, 4 thal major, +1 post BMT	25	30
Mayday Hospital	214 [138 SS, 51 SC]	<5	8	
QE Hospital Woolwich	201 [141 SS, 43 SC]		<5	
Lewisham Hospital	244	<5	7	
Dartford, Medway, Brighton	20	<5		

Services for red cell disorders in children are very well established on this site, where one of the earliest dedicated clinics in the UK was started. The service is run by two paediatric haematologists, one with an academic contract [4 PA's for red cell services], the second with 4 PAs devoted to the red cell service and responsibility also for paediatric haematological malignancies. There is also a community paediatrician [3 PA's for this service] with very long experience who comes into KCH to do clinics, and see in-patients. The 'Variety Children Hospital' within KCH has 70 beds plus 36 cots in the neonatal unit. There are 8 PICU beds and an 8 bedded High Dependency Unit. KCH is also part of 'Kings Health Partners', a collaboration intended to work

across clinical services, research and education, with Guys and St Thomas' NHS Foundation Trust [GSTT], South London and Maudsley NHS Foundation Trust, and King's College London.

For red cell services, KCH and GSTT each provides a service for large numbers of locally resident children with some subspecialties particularly developed on one or other site. Children may be referred to specialty clinics on either site.

This area has particular challenges in terms of the larger network, whereby local hospitals with services run by general paediatricians are linked to the centre so that members of the specialist team can support the local service, see complex children and, at least for the smaller local hospitals, systematically see all patients for annual review. The prevalence of the condition in south east London, as in east London, is so high that many of the linked local hospitals themselves have a very large caseload, with over 200 children in many cases. There are over 1,000 affected children included in the area covered by this network. Some previously low prevalence areas have seen a very rapid increase in local patient numbers over the last five years, for example an increase from zero to 50 children in Medway, and from three or four to 100 in Sidcup. When families move, they sometimes choose to continue to receive care from the out-of-area hospital they were previously attending. The network at present is complex, and more of a 'web' than a hub-and-spoke arrangement. The KCH team has made particular efforts to support its large local hospitals, providing regular outreach visits to three of them. Arrangements with some more distant hospitals, including Dartford, Kingston, and others in Kent and Sussex, are not yet formalised. The lead paediatric haematologist would like to work toward a system whereby KCH provided systematic annual review for all children in local hospitals with less than 50 children receiving care for these conditions. The team is actively engaged in research, and there is an established teaching programme for staff within and beyond the hospital.

Accident and Emergency

Children needing acute assessment and management are asked to go to the paediatric A&E. There is a two bedded 'clinical decision' area where children can remain if they need more than four hours care but not formal admission. Children do not have a 'fast triage' system through reception. There are fewer presentations for pain than previously. The A&E team said that some children present having taken no analgesia at home, and may respond to simple analgesia. Approximately 50% children attending A&E require opiate analgesia and all of these are admitted. Children are managed with a generic pain protocol.

There were 377 in-patient spells for sickle cell related problems in the last year. A few children attend regularly and a system is in place for these to be notified to the specialist team so that discussion about improved management can take place. The review team met only one family so it was not possible to get a sense of how well A&E management works from the users' perspective.

IN-PATIENT FACILITIES

Admission is to a 14 bedded in-patient ward. This is a good facility with many cubicles, staffed by nurses who are familiar with acute sickle cell problems. A pain sister helps with in-patient management. The children are admitted under the paediatrician of the week. They are seen by the paediatric haematologists, sometimes the community paediatrician, and the sickle cell clinical nurse specialist, usually on a daily basis. Children needing urgent red cell exchange are moved to a high dependency bed. There is an enthusiastic new play leader, managing a team of five staff in total. There is a well-stocked teaching room used by in-patients and those awaiting transfusion on the Day Unit, with an excellent team of teachers who seemed sensitive and knowledgeable about the needs of haemoglobinopathy patients. The school room caters for children from 5 to 17 years and the teachers liaise closely with schools, tailoring their input to the specific needs of the child. Home tuition after discharge is available for children living in Lambeth.

HIGH DEPENDENCY UNIT (HDU) / PICU

This is an excellent 'state of the art' facility. There are close working links between this team, led by an enthusiastic consultant, and paediatric haematology. All nurses on the HDU can undertake red cell exchange transfusions. An electronic system for recording observations and clinical interventions is being rolled-out for children on this unit.

OUT-PATIENT CLINICS

A large weekly morning clinic is run, usually by both the paediatric haematologists, the community paediatrician, a Specialist Registrar, and the acute and community clinical nurse specialists. The psychologist attends most weeks. The psychologist, currently on maternity leave (but with cover arranged) works four days a week on this service and aims to undertake neurocognitive assessment on the children who have cerebrovascular problems annually, although this sometimes proves difficult in practice. Trans-cranial Doppler [TCD's] scans to assess stroke risk are undertaken by an experienced vascular sonographer in the same clinic. Around 30 children attend each week. There is currently no specific adolescent clinic at which medical handover between the paediatric haematologists and adult haematology lead takes place but the paediatric and adult CNSs meet children together when they can from the age of about 13 years. A 'Darzi' post has recently been approved for a doctor to focus on transition arrangements for a variety of long term conditions, including red cell disorders. The paediatric neurologist from Guys and St Thomas' comes to KCH to do a joint clinic every two months.

Annual review for the transfused children or those with complex needs is in place. There is a calendar indicating which child should be reviewed each month, and the date the review visit took place is recorded. The process is supported by an on-line 'prompt' for electronic requesting of investigations to be undertaken. The

visiting team did not get confirmation that annual reviews were consistently undertaken on other children although there is a policy in place guiding what should be covered in the annual review visit for all children.

DAY CARE

Children having booked transfusions are managed on a separate Day Unit, adjacent to the in-patient ward. This is open during usual working hours and has recently started opening on Saturday mornings for children with red cell disorders. It is staffed by a nursing team which also staff the outpatient area. A paediatric doctor is based on the unit in the mornings, after which cover is by the ward team. Most of the nurses on the day unit can cannulate and set up transfusions. Children usually come in the morning for pre-transfusion samples, and then go to the schoolroom for a couple of hours before starting the transfusion. Samples can also be taken on the day unit prior to transfusion, or in out-patients phlebotomy (which is currently open 9 to 5 but might soon be extended to 8 to 8). Details of all the regularly transfused children are held on the electronic patient record, and updated after a monthly MDT at which each of these children [and those receiving hydroxycarbamide] are discussed. The MDT is organised by the Paediatric Sickle CNS and attended by the Day Unit staff, paediatric haematologist[s], sometimes the community paediatrician and junior doctors, the community nurse and the paediatric psychologist.

COMMUNITY SERVICES

The large community team covers Lambeth, Southwark and Lewisham and also works with the acute teams at Guy's and St Thomas', and Lewisham Hospitals. One team member particularly covers the paediatric caseload, as well as antenatal screening and managing the results of the newborn screening programme for Southwark and Lambeth which covers most of the KCH local catchment. Two further clinical nurse specialists (CNSs) work with children from the age of 12, and support adult services. The hospital service works with families from further afield, for example, Kent, but, apart from Bexley, there is no specialist community nursing service in Kent. The paediatric nurse works in the KCH clinics fortnightly, and the adult nurse joins her on alternate weeks. The adult nurse meets and talks with older children to consider transition to the adult service, using an assessment questionnaire. Medical referral to the adult service takes place when a young person is ready for transition. The teams also undertake school liaison, school visits if needed and work with the SENCO for children with specific learning difficulties. The nurse team does not currently have an active role in the support group, although they attend by invitation for teaching or discussion. Previously, there were some workshop-type educational days for young people run by the paediatric community nurse, who also set up a 'buddying' system for older children. The hospital team feel that the community specialist service works well, and every effort is made to ensure effective liaison between acute and community services. The community service is soon to be merged managerially with GSTT.

LINKED HOSPITALS

The main linked hospitals, each managing in excess of 200 children with sickle cell disease and very small numbers with thalassaemia, are Mayday Hospital in Croydon, Queen Elizabeth Hospital in Woolwich and Lewisham Hospital. A monthly outreach clinic by one or other of the paediatric haematologists takes place at Croydon and Woolwich with discussion and annual [or more frequent] review of severely affected children, or those with complex needs. Trans-cranial Doppler screening takes place in parallel, undertaken by a member of the KCH team using portable equipment. Lewisham Hospital was offered the same service but to date has opted just to have the TCD outreach service rather than the level of clinical support provided to Croydon and Woolwich. All three hospitals were been visited separately and more detail about their services is available in those reports.

Link with other network hospitals are less well defined (see Introduction). Children are usually referred to KCH when the local paediatrician is concerned about some aspect of their clinical progress or care. Children are usually referred to KCH for TCD's. Many of the hospitals in Kent, Surrey and Sussex appear to link with KCH and Guy's and St Thomas'. Some also link with St George's.

COMMISSIONING ARRANGEMENTS

The assistant director for acute commissioning for the Lambeth, Southwark and Lewisham Alliance, which will shortly join Bexley, Bromley and Greenwich to make the South East London commissioning team, met the review team. He and his team have London-wide responsibility for acute service activity at KCH, but would not cover activity at more distantly located hospitals. They have not had specific discussion about staffing and service arrangements for children with red cell disorders, but do have a bi-monthly quality review meeting which includes some senior KCH members as well as PCT members, addressing performance against targets, and discussing quality assurance process within the Trust. He felt he would be aware of any concerns about this service, had they existed. He indicated that the report of this visit, as for other external reviews, would be taken for discussion at their quality review meeting.

REVIEW VISIT FINDINGS

ACHIEVEMENTS

1 This is a very good service, providing high quality care for local children who come to KCH for all their care. There are well-organised arrangements for providing appropriate levels of specialist support to services at the largest linked hospitals, as recommended in the national clinical care standards. The specialist team outreach to two of the large local hospitals is ideal in terms of quality of service and convenience for children and families, as well as providing useful clinical teaching for the local teams. The role of the hospital based CNS is noteworthy. She now has a prescribing qualification, and her work provides a strong focus around which the team functions. The community team, with input from an experienced community paediatrician, play a positive part in service provision. Some of the support team are excellent, notably the HDU / PICU team, teachers, and play specialists, and team working in general is strong.

2 Appointment to the new 'Darzi' post focussing on transition from paediatric to adult services for children with a variety of long term conditions is innovative, and is likely further to develop the nurse-led process for children using this service.

3 The service seems to have a high profile in the Trust. It has the interest and support of the senior management team, and local management is energetic, involved and valued by the clinical team.

CONCERNS

1 Child protection training records are kept, but indicate that some of the staff involved in this service do not have either the required level of training, or there is no record of their level and date of training.

FURTHER CONSIDERATION

1 Discussion with commissioners is needed about the model and location of services to meet the needs of the population. Plans for developing and monitoring services will then need to be agreed. The KCH specialist team is already working very effectively, with outreach clinical and TCD clinics, in support of two of its large linked hospitals. As local expertise in those units is strong, it is probably not necessary for the specialist team to see every child every year. There are opportunities across this very large network, working with partners at GSTT, to divide up responsibilities for supporting some of the other linked hospitals. Discussion between the two specialist providers might include planning a co-ordinated approach, so that each linked hospital works mainly with one or other specialist team, at

least for affected newborn. This would enable a more systematic referral pattern to be established. The feasibility of providing outreach services in local hospitals, even if only 6 to 12 monthly could then be assessed. This might be helpful in achieving the aim of each child managed by the smaller linked hospitals having specialist input.

If many hospitals in the network have patient numbers to justify outreach, senior medical and specialist sonographer staffing levels in the KCH team needed to staff these services will need to be considered. Alternatively, it may be appropriate for one or more of the larger linked hospital teams to act as intermediate level providers, perhaps offering an annual review service for children from smaller units geographically closer to them than to the centre. The specialist team might focus on providing comprehensive care to their own local families and seeing children from any part of the network who have complex or difficult clinical issues.

The University Hospital Lewisham [UHL] team has opted to have outreach TCD services only without clinical outreach. In order to ensure equity of care across the linked hospitals, the KCH team may wish to have further discussion with UHL managers and clinicians.

2 A & E guidelines include review of adequacy of analgesia at 1 hour. Most other guidelines suggest review within 30 minutes. An audit of time from arrival in A&E to time to first analgesia, and time to adequate analgesia, might be helpful to provide assurance that analgesia for children attending in acute pain is as prompt and effective as patients and the team would wish.

3 Clinical guidelines are mostly excellent. Not all are available on the intranet. Some were lacking in parts, for example there are no thalassaemia intermedia guidelines, none relating to referral for bone marrow transplantation, and there is no mention of malarial prophylaxis in the clinic guidance.

4 Although the information for patients and family presented is wide-ranging and good, very little was seen on display in the clinical areas for children and families to take and read. It may be helpful to review availability of information in the places which families use.

5 The facilities were in temporary disarray because of building works. Some details could be addressed quickly, including improving the play equipment on the ward and in the clinic rooms.

6 Children sometimes travel between KCH and Evelina Children's Hospital at GSTT for clinical input by the most appropriate subspecialty team. They also travel for routine investigations, for example echocardiogram. Travel could be reduced if this service was provided on a sessional basis at KCH.

GOOD PRACTICE

1 When children are acutely ill in hospital they are reviewed frequently, usually daily, by the consultant paediatrician, a paediatric haematologist and the hospital based CNS.

2 Children attending A&E routinely have a finger prick 'Haemaccue' haemoglobin measurement if they are not going to have a formal blood count.

3 The guideline for exchange transfusion is clear, available on the wards, and all HDU nurses can undertake this procedure.

4 Many of the clinical guidelines are strong, including for the management of chest syndrome, the shared care guideline including a list of key contacts for all network professionals, and the cannulation competency document.

5 Two 'clinical decision' beds are available on A&E so that a child can be observed for longer than four hours but do not have to be formally admitted. The practice of flagging frequent A & E attenders to the specialist team is commendable.

6 Performance against the neonatal screening standards for 2009 was excellent, with all babies referred and seen in clinic by the target ages.

7 Use of a mouth swab for DNA confirmation of the diagnosis at first clinic avoids the need for venepuncture at this time.

8 The focus on haemoglobin SC disease as a separate condition is interesting. This is the subject of some of the team's current research. The lead clinician's suggestion that this should be taken into account in further clinical guidance, as not all the requirements for children with SS necessarily apply equally to those with SC, is noted.

9 Young people are 'buddied' with others of similar age as they may use this support more readily than attending a formal support group.

10 There is a systematic case-based monthly MDT for transfused or complex children, attended by acute and community team members, with updating of the electronic record after each meeting.

11 Much of the patient and family information is excellent, including the clinic leaflet, the information pack for carers, the care plan for management in schools, the handbook for school nurses, and ethnically appropriate dietary information.

12 The new electronic clinical observation recording and management system in use on HDU was felt to be a useful and robust way of monitoring acutely sick children.

13 The entry of 420 children on the National Haemoglobinopathy Registry to date is outstanding.

APPENDIX 1: MEMBERSHIP OF THE REVIEW TEAM

Dr Farrukh Shah	Consultant Haematologist	Department of Haematology
Helen Appleby	Clinical Nurse Specialist, Paediatric Haemoglobinopathies	Guy's & St Thomas' NHS Foundation Trust
Karen Madgwick	Transfusion Practitioner	North Middlesex University Hospital NHS Trust
Sally Riley	Deputy General Manager, Women & Children	Whittington Hospital NHS Trust
Elaine Miller	Co-ordinator	UK Thalassaemia Society
Ravinder Raj	Information Officer	Sickle Cell & Thalassaemia Support Project, Wolverhampton
Dr Anne Yardumian	Clinical Lead for Peer Review Programme	

APPENDIX 2: COMPLIANCE WITH QUALITY REQUIREMENTS

Specialist Haemoglobinopathy Teams

Ref	Quality Requirement	Met?	Comment
INFORMATION AND SUPPORT FOR PATIENTS AND THEIR CARERS			
1	Written information should be offered to patients and their families covering at least: 1 A simple explanation and description of the condition, how it might affect the individual, possible complications and treatment. For sickle cell disease this information should include: a Problems, symptoms and signs for which emergency advice should be sought b How to manage pain at home, including how to avoid pain, and non-pharmacological interventions c Importance of adequate fluid intake d Use of antipyretics for fever e Importance of regular antibiotics and full immunisation f How to feel the spleen and its significance. For thalassaemia this information should include: a Problems, symptoms and signs for which emergency advice should be sought b The importance of maintaining good haemoglobin levels by transfusion c Potential problems of iron load and how it can be managed. 2 Details of the services available locally including: a Clinic times and how to change an appointment b Key contact name and number c Alternative contact if key contact away d Who to contact for advice out of hours e How to use emergency services f Ward usually admitted to and its visiting times g Community services and their contact numbers h Details of support groups available. 3 Health promotional material including a Inheritance and implications for other family members b The importance of a good diet and regular exercise c Implications for travel d Age appropriate information on avoiding smoking and excess alcohol consumption e Age appropriate information on contraception and sexual health f Where to go for further information, including useful websites and national voluntary organisations.	Y	*Almost all is available and good. Some travel guidance especially for thalassaemia Is not yet available.*

Ref	Quality Requirement	Met?	Comment
2	Written information for the patient's primary health care team should be available covering at least: a All aspects of QR1 b The need for regular prescriptions, penicillin and analgesia (SC)	Y	
3	Information should be available on transition to adult care. This information should cover all aspects of the transition (QR43).	Y	
4	The SHT and its linked LHTs should have agreed a patient-held record for recording at least: a Information about the patient's condition b Current management plan c Regular medication d Named contact for queries and advice e Alternative contact for times when key contact is away.	Y	*Although it was not certain to what degree this was in regular use. It is indicated that not all parents want to use and some of the medical team do not see its value.*
5	The locally agreed patient-held record (QR4) should be in regular use within the LHT / SHT.	N	
6	Services should be provided in a child friendly environment, including toys and books / magazines for children and young people of all ages.	Y	*Although the temporary disruption caused by building works is inevitable and short term; some aspects of provision might be improved in the meantime, for example replacement of old or damaged toys in play room on ward and in clinic.*
STAFFING and SUPPORT SERVICES			
9	The SHT should have a nominated lead paediatrician / paediatric haematologist consultant with an interest in the care of patients with haemoglobinopathies who should have responsibility for guidelines, protocols, training, audit relating to haemoglobinopathies and overall responsibility for liaison with referring LHTs. The lead consultant should undertake CME activity of relevance to care of children and young people with haemoglobinopathies.	Y	
10	The lead consultant (QR9) should have a named deputy who will provide cover for absences. The named deputy in the SHT should undertake CME activity of relevance to care of children and young people with haemoglobinopathies.	Y	
12	The SHT should have a lead haemoglobinopathy nurse with responsibility, with the lead consultant (QR9), for guidelines, protocols, training, audit relating to haemoglobinopathies and liaison with referring LHTs. The lead nurse should have specific training in the care of patients with haemoglobinopathies.	Y	
13	The SHT should have a nurse specialist or counsellor who provides outreach support for patients in the community. This nurse specialist / counsellor should have specific training in the care of patients with haemoglobinopathies.	Y	
14	There should be agreed cover arrangements for the outreach nurse specialist / counsellor.	Y	

Ref	Quality Requirement	Met?	Comment
15	Access to the following staff and services should be available: a MRI and CT scanning b Transcranial Doppler ultrasonography (SC) c Hospital dental services d Genetics services e Bone marrow transplantation services f Contraception and sexual health services g Consultant cardiologist h Consultant endocrinologist i Consultant hepatologist j Consultant neurologist k Consultant ophthalmologist l Consultant orthopaedic surgeon m Consultant obstetrician n Child and adolescent mental health services	Y	
16	The following services should be available a Paediatric high dependency care b Paediatric intensive care There should be agreed criteria for admission to each level of care.	Y	
17	The following support services should be available: a Interpreters b Social work c Play specialist d Hospital teacher (in-patient care only) e Child psychologist f Dietician	Y	*And input by play leaders and teaching staff is enthusiastic and strong. Unfortunately the team was not able to meet the psychologist at this visit [on maternity leave].*
18	A service agreement for support from community services should be in place. This service agreement should cover, at least: a Guidelines for involvement of community paediatric services in the care of patients with haemoglobinopathies b Role of community services c Exchange of information between hospital and community services and vice versa d Arrangements for liaison with schools.	N	*Only 'appendix 3' from a document which had no author or date was offered.*
19	A nurse with competency in cannulation, starting and supervising a transfusion should be available at all times at which children are attending for transfusion.	Y	
20	All staff involved in the care of children and young people with haemoglobinopathies should undertake regular child protection training.	N	*Records were available but indicated that some staff had no record of training; others were trained to a level below that which would be appropriate for their degree of involvement with paediatric practice.*
CLINICAL and REFERRAL GUIDELINES			
21	Guidelines should be in use covering a How to establish and confirm diagnosis b Parent and sibling testing	Y	

Ref	Quality Requirement	Met?	Comment
22	Clinical guidelines should be in use covering: a Recommended immunisations b Immunisations, other prophylaxis and travel advice prior to travel abroad. c Penicillin prophylaxis while awaiting clarification of diagnosis (sickle cell disease only)	Y	*Guidance about the importance of malaria prophylaxis for children travelling to W Africa was omitted, although included in the patient information material. The need for, and details, about penicillin prophylaxis was also not included here, although it was covered in the OPD guidelines. It should also be included in this section about infectious disease prevention.*
23	Clinical guidelines should be in use covering possible acute presentations including, at least: **For patients with sickle cell disease:** a Fever and infection including major sepsis b Acute pain c Acute anaemia d Stroke and other acute ischaemic events e Acute chest syndrome f Acute splenic sequestration g Abdominal pain / jaundice h Priapism i Changes in vision, including urgent referral for an ophthalmologic opinion j Indications for 'top-up' and for exchange transfusion and practical protocol for undertaking exchange transfusion **For patients with thalassaemia:** a Fever and infection including major sepsis b Unexpected cardiac, hepatic, endocrine decompensation. These guidelines should ensure that patients are transferred to the paediatric ward for assessment as quickly as possible	Y	*Only guidance as to management of acute changes in vision was omitted*

Ref	Quality Requirement	Met?	Comment
24	Clinical guidelines should be in use covering routine out-patient monitoring and management between formal annual progress review visits, including: **All patients:** a General assessment of well-being including school attendance b Any difficulties adhering to treatment or other difficulties with care c Monitoring growth and development d Checking for palpable spleen **Patients with sickle cell disease:** a Discussion to ensure analgesics available for home use understood and effective b Informal assessment of cognitive function, learning and behavioural difficulties, including referral if needed c Checking for history of priapism in boys d Checking for and management of nocturnal enuresis e Checking all necessary immunisations up to date f Penicillin therapy and alternative therapy for children who are allergic to penicillin g Monitoring of oxygen saturation by pulse oximeter h demonstrating to parents / carers how to check for splenic enlargement i Monitoring of Hb levels, renal function tests, liver function tests j Indications for early referral to specialist haemoglobinopathy team (LHT only) **Patients with thalassaemia and regularly transfused sickle cell:** a Checking adherence specifically to chelation therapy and any difficulties b Monitoring Hb levels, iron levels, liver function and other biochemistry c Indications for early referral to specialist haemoglobinopathy team	Y	

Ref	Quality Requirement	Met?	Comment
25	Clinical guidelines should be in use covering **annual specialist review** visits including, at least: **All patients:** a Regularity of school attendance and reasons for absence b Any difficulties adhering to treatment or other difficulties with care c Monitoring growth and development, including review of centile charts d Health promotion and healthy lifestyle advice and support e Contraception and sexual health in relevant age group f Travel advice g Review Hb levels, renal and liver assessment, other biochemistry h Hepatitis B vaccination i Monitoring of hepatitis B antibodies and action to be taken should levels fall j Monitoring for hepatitis C in transfused patients k Discussion and preparation of child for any planned surgery l Indications for consideration of splenectomy m Preparations for splenectomy including recommended immunisations n Treatment of complications of splenectomy, including persistent thrombocytosis. **Patients with sickle cell disease:** a As QR 24 plus b Monitoring and screening for neurological complications, including transcranial Doppler ultrasonography c Indications for imaging to assess the extent of cerebrovascular disease d Indications for overnight oxygen saturation monitoring (sleep study) e Indications for echocardiography including possibility of pulmonary hypertension **Patients with thalassaemia:** a As QRs 31, and 32 where applicable [thalassaemia intermedia] plus b Review annual red cell consumption c Review adequacy and appropriateness of iron chelation regimen d Consideration of options for helping children, young people and their families to adhere to chelation therapy e Audiometry and ophthalmology check for those on desferrioxamine, from age 10 f Cardiological assessment (from age 10) g Testing for endocrine abnormalities (from age 10) h Bone mineral density assessment (from age 10).	Y	*Included in out-patient guidance but in practice the team indicated this was only systematic for regularly transfused, or otherwise complicated, children.*

Ref	Quality Requirement	Met?	Comment
26	Guidelines should be in use for referral of patients and their families to a clinical psychologist with experience in the care of patients with haemoglobinopathies (including the option for self-referral)	Y	
27	Guidelines for referral for consideration of bone marrow transplantation should be in use.	N	
28	Clinical guidelines should be in use covering: a Indications for regular transfusions b Investigations and vaccinations prior to first transfusion c Monitoring of haemoglobin levels	Y	*Indications for transfusion and investigations before first transfusion are included in guidance 'blood transfusion in children with haemoglobinopathies'.*
29	Clinical guidelines should be in use covering review by a specialist nurse or doctor prior to transfusion to ensure that each transfusion is appropriate.	Y	
30	A Transfusion Policy should be in use covering: a Area/s where transfusions will usually be given b Procedures for checking blood c Staff allowed to undertake cannulation d Recommended number of cannulation attempts e Maximum rate and volume of transfusion according to size of child f Monitoring the transfusions g Management of transfusion reactions h Arrangements for medical cover during transfusion i Arrangements for ensuring that a nurse with appropriate competences is in attendance throughout the transfusion.	Y	
31	Clinical guidelines for chelation therapy and monitoring iron load should be in use including: a Indications for starting chelation b Choice of regime c Dosage and dosage adjustment d Clinical assessment of tissue damage e Monitoring of serum ferritin f Use of non-invasive estimation of organ-specific iron loading heart and liver by T2*/R2 g Management of side effects of chelators.	Y	
32	Clinical guidelines for the management of thalassaemia intermedia should be in use including: a Indications for transfusion b Monitoring iron loading c Indications for splenectomy.	N	
33	Clinical guidelines should be in use covering: a Indications for exchange transfusion b Arrangements for carrying out an exchange transfusion.	Y	*Clear guidelines and all HDU nurses trained to undertake this procedure.*
SERVICE ORGANISATION and LIAISON WITH OTHER SERVICES			
34	Clinical guidelines for acute and out-patient monitoring and management (QR23 and QR24) should be available and in use in appropriate areas including A&E, clinic and ward areas.	Y	*Hard copy in place but not all guidelines on the intranet.*

Ref	Quality Requirement	Met?	Comment
35	A protocol should be in use covering the initial clinic visit for patients with haemoglobinopathies covering, at least: a Giving each patient information relevant to their condition (QR1) b Giving each patient their patient-held record (QR4) c Allocation of a named contact for queries and advice to each patient. d Discussion of arrangements for future treatment and care e Sending the GP information relevant to their patient's condition (QR2)	Y	
36	A protocol should be in use covering the initial clinic visit for patients previously treated outside the UK, including: a Full medical history and examination b Investigations c Referral to other specialist services (QR15) d All aspects of QR28	N	*No specific detail about such children was included.*
37	A protocol should be in use covering arrangements for care between SHT and LHT for ongoing care. This protocol should ensure that, to facilitate effective shared care: a All patients have an up to date patient held record and details of their care plan. b The LHT and the patient's GP have received details of the patient's care plan.	Y	*The document 'guidelines for the care of inherited anaemias at Kings College Hospital, Guy's and St Thomas' sickle cell network' will largely cover this requirement.*
38	A protocol should be in use covering: a Updating patient-held records b Offering patients a permanent record of consultations at which changes to their care plan are discussed. c Recording changes of key contact d Giving further information (QR1) as patients' and families' needs change	N	*Not many patients use patient held record, and letters to GP's are not systematically copied to parents / carers.*
39	The SHT and its linked LHTs should have agreed a policy on the communication of clinical information, management plans and important decisions regarding treatment between clinical teams. This protocol should cover information from specialist clinic visits as well as contacts with SHT / LHTs. The protocol should be specific about frequency / indications for communication with the patient's GP	Y	
40	An operational policy should be in use covering: a Teaching children, young people and their patients how to set up an administer subcutaneous desferrioxamine infusions b Encouraging children to participate in setting up and administering their own infusion c Regular assessment and updating administration techniques d Recording of assessments of administration techniques	Y	
41	Patients and families should have choice of attending for blood tests, clinic appointments and blood transfusions 'out of hours' to minimise disruption to normal life	N	*Out of hours transfusion is now available. Clinic and phlebotomy out of hours services are being considered but not yet in place*

Ref	Quality Requirement	Met?	Comment
42	A protocol should be in use covering: a Follow up of children who do not attend b Communication and follow up of children who move to another area	Y	
43	A protocol should be in use covering transition to adult care. This should ensure: a Age guidelines for timing of the transfer. b Involvement of the young person in the decision about transfer. c Involvement of primary health care, social care and adult services in planning the transfer. d Allocation of a named coordinator for the transfer of care. e A preparation period and education programme relating to transfer to adult care. f Communication of clinical information to the adult services. g Arrangements for monitoring during the time immediately after transfer to adult care.	Y	
44	The team should have in place: a Mechanisms for receiving feedback from patients and carers about the treatment and care they receive. b Mechanisms for involving patients and carers in decisions about the organisation of the services. c Mechanisms for encouraging the development of local support groups.	Y	*User feedback in general, for example on the in-patient ward, is sought and a full out-patient satisfaction survey has recently been undertaken.*
45	The SHT should run a programme of training and awareness of the management of patients with haemoglobinopathy for its main referring LHTs.	Y	
47	The SHT should meet at least annually with its referring LHT teams to: a Identify any changes needed to network-wide policies, procedures and guidelines b Review results of audits undertaken c Review any critical incidents including those involving liaison between teams d Consider the content of future training and awareness programmes (QR45)	Y	
49	The SHT should meet at least annually with representatives of the neonatal screening programme to review progress, identify issues of mutual concern and agree action.	Y	*The lead paediatric haematologist is also the Director of the Neonatal Screening laboratory, and systems for ensuring that babies reach clinical services are in place. However a broader meeting to include community nursing team, to address process issues and any other concerns, may prove additionally useful.*

Ref	Quality Requirement	Met?	Comment
DATA COLLECTION and AUDIT			
50	The LHT / SHT should have audited compliance with key standards including: **Patients with sickle cell disease:** a Proportion of patients taking regular penicillin b Proportion of patients fully immunised against pneumococcus c Proportion of patients (HbSS and HbSβ^0) who have had Transcranial Doppler ultrasonography undertaken within the last year d Proportion of patients who have had their annual multi-disciplinary review within the last year e Effectiveness of action to contact families who have not attended for follow up appointments. f Review of the care of any patients who have died. **Patients with thalassaemia:** a Proportion of patients on chelation therapy b Proportion of patients who have had their annual multi-disciplinary review within the last year c Adequacy of recording of: ▪ Pre-transfusion Hb levels ▪ Regular monitoring of iron level ▪ Complications of iron overload ▪ Height / weight progression ▪ Spleen size ▪ Support for home chelation programme (QR40) d Effectiveness of action to contact families who have not attended for follow up appointments. e Review of the care of any patients who have died.	Y	*All the recommended audits have been undertaken within the recent past plus additional, useful clinical audits.*
51	Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry.	Y	*A very large number of children are already entered onto the NHR.*

Commissioners of Services

Ref	Quality requirement	Met?	Comment
52	Each Specialist Commissioning Group[1] should have agreed the location of services for its population: a Specialist Haemoglobinopathy Team/s for children and young people b Local Haemoglobinopathy Team/s for children and young people c The expected referral patterns to each SHT and LHT. d The type of patients (sickle cell and / or thalassaemia) who will be treated by each team.	N	*But the acute commissioning team are aware of the services and would be made aware of any concerns about them and plan to include consideration of this report at their two monthly quality assurance meetings.*

Ref	Quality requirement	Met?	Comment
53	Each Specialist Commissioning Group should have: a Compared the staffing, support services and facilities of each SHT and LHT located within its area (QR52) with the levels expected in QRs 7 to19. b Agreed a plan for the development of SHTs and LHTs located within its area. c Monitored achievement of the agreed plan at least annually. The agreed development plan should ensure that QRs 7 to 19 are met within 2 to 5 years.	N	

Additional Requirement – Screening Services

Ref	Quality requirement	Met?	Comment
P3	Timely communication of positive screening results (sickle cell disorder) – including a review of parental results	N	*But in most cases the delay was outside the control of the community nursing team.*
P4	Effective follow-up of infants with positive screening results (sickle cell disorder) – all babies to be registered with a local clinic/centre (or clinic working as part of clinical network)	Y	*[Ignore part a which is a repetition of P3 but with different standard target]* *Last year this standard was completely met.*
P5	Timely confirmation of diagnosis for infants with a positive screening result**	Y	
S1i	Failsafe to ensure ongoing care	Y	
S1ii	Up-to-date registers maintained of babies (cases) for which units are responsible	Y	

** Specified conditions: Hb-SS, Hb-SC, $HbSD^{Punjab}$, Hb-SβThalassaemia (β^{+}, β^{0}, δβ, Lepore), $Hb\text{-}SO^{Arab}$, Hb-S/HPFH

NOTE. Different QR's are not comparable in terms of their importance, or likely impact on the quality or outcomes of the service, and a figure summarising the number of QR's met cannot be used to assess the overall standard of the services provided or to compare this Trust's services with others.