UK Forum on
Haemoglobin
Disorders

WM QRS West Midlands
Quality Review Service

Services for Children with Sickle Cell Disease or Thalassaemia
At
Guy's and St Thomas'
NHS Foundation Trust

Quality Review Visit Report
Visit date: May 13th 2010

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CONTENTS

INTRODUCTION

This report presents the findings of the peer review visit to services for children with sickle cell disease or thalassaemia at Guy's and St Thomas' NHS Foundation Trust which took place on May 13th 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 Guy's and St Thomas' NHS Foundation Trust 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 GUY'S AND ST THOMAS' NHS FOUNDATION TRUST HOSPITAL NHS TRUST

Service (as at May 2010)	Patient numbers sickle cell disease	Patients on long term red cell transfusions	Patient numbers thalassaemia
Evelina Children's Hospital	510 [75% SS]	35 sickle cell [12 for stroke]	9 intermedia
Lewisham	244	7	<5
Queen Elizabeth Hospital Woolwich*	~ 200	<5	<5
Queen Mary Sidcup	~ 100	<5	<5
Medway Hospital	~ 50	<5	<5
Darrent Valley Dartford	~ 60	<5	

[QEH Woolwich also links with the specialist team at King's College Hospital [KCH] – see 'Linked Hospital' section p 5]*

Evelina Children's Hospital [ECH] forms part of the large NHS Foundation Trust, Guy's and St Thomas' [GSTT]. All the acute children's services for the Trust are provided on the ECH site. Community services for Lambeth, Southwark and Lewisham are soon to be merged managerially with the GSTT

Trust. The Trust links with Kings College Hospital in King's Health Partners, functioning as an academic health sciences centre, and there are also some clinical links with the paediatric haematology service there.

Services for children with sickle cell disease and, to a lesser extent, thalassaemia which is not prevalent in the areas served, are very well established. They are clinically led by a named lead paediatrician and three hospital clinical nurse specialists, a named lead CNS, a nurse consultant trainee, and a nurse focussing, together with a part time psychologist, on transition-age children. The team is well supported by a range of professionals within the hospital and there are strong links with the community nursing team. The ECH team offers comprehensive care for around 500 children living locally and specialist input for children in a number of linked hospitals. Some of the linked hospitals care for a large number of children with sickle cell disease.

Accident and Emergency

Children needing urgent care present to the A&E Department which is small and busy. Guidelines are in place there, in hard copy, as part of a comprehensive 'Emergency Medicine' manual which is given to all junior doctors. Children arriving with sickle cell pain crisis are seen as soon as they arrive, and administration of first analgesia is usually within 15– 30 minutes. Intra-nasal diamorphine is usually offered first, depending on what the child has already been given. Service users commented that there can be delay in transfer from A&E to the ward because of bed availability. A folder kept in A&E, and updated by the lead nurse, holds individualised treatment protocols and a copy of the last clinic letter. Up-to-date clinical information is therefore available for staff when a child presents. There is also a weekends 'admissions box' where details of children attending or admitted are kept for the information of the acute and community teams.

Outpatient Clinics

Outpatient clinics are held in a spacious general clinic area with a large central waiting hall full of play equipment, including a large and obviously well used spiral slide! A clinic on Fridays for children with red cell disorders lasts most of the day. Trans-cranial Doppler studies take place in the morning. The clinic is run by the lead paediatrician and lead CNS, with two or three specialist registrars, often the transition CNS and the transition psychologist. The community nurse sometimes attends. About 20 children attend each week. A set of guidelines are available in hard copy and on the intranet, and some patient information leaflets are also held on the intranet and can be printed and given to families as needed. A transition clinic takes place monthly, in which the lead paediatrician and adult

haematologist who will take over clinical care, see the children together. The transition CNS and psychologist also work in this clinic, often the community nurse, and sometimes one of the adult psychologists also. Transition days, a mixture of entertainment and education and discussion, take place with children of different ages in school holidays. There is a session for the group preparing for independent living after leaving home. Shared clinics with nephrology take place approximately 2 monthly, and with neurology three to four times a year. There is a play specialist available in clinic. Phlebotomy is available 8.30 to 5, but children wishing to come later can use the adult facility which is open until 6 pm. A pharmacy is immediately adjacent to the out-patient area.

DAY CARE

Booked transfusions take place in a day ward, which is also used for children with a variety of other conditions. It manages about 15 children per day in six beds. It is open from 7.30 am – 8 pm, but most regularly transfused children attend on Saturdays, when the unit is opened mostly just for their treatment. A junior doctor is based on the unit, so there is minimal delay in the children being 'clerked' and the transfusion prescribed. At weekends, a covering doctor has to be called but delays are usually slight. Nursing staff otherwise manage the children, including inserting intravenous cannulae. Most of the team are competent to do this. A file is held on the ward including details of all the regularly transfused children, including the volumes of blood they usually receive, their chelation doses, and flow charts of their pathology results. These are also uploaded onto a spreadsheet. A play specialist is available on the unit just one day a week which staff felt was insufficient. Certainly if children are attending with difficulties and fears around procedures, more availability would be beneficial. The visiting team was interested to see disposable curtains in use on this unit, and also the in-patient ward, which are not available in most hospitals and which are a useful infection control measure.

IN-PATIENT FACILITIES

The in-patient ward, shared with other specialties, is bright and pleasant. Staff there are very familiar with the management of children with sickle cell disease, there being 50 to 60 admissions per year. Guidelines were in place on the ward. Children are admitted under the care of the admitting paediatrician, and the lead paediatrician for the service sees them about twice a week or as necessary, and the lead CNS usually visits daily. There is input from a nurse from the specialist pain team while the children are in hospital, and some children have continued follow-up from this service after discharge. There is no day room facility for older children. They can use an adolescent

area on the floor below when sufficiently mobile. There is a large teaching area, with different sections for use by children of varying ages and needs.

COMMUNITY SERVICES

This service is covered by a team of community specialist nurses based locally, who cover the populations of Lambeth, Southward and Lewisham. The nurse who particularly covered the children and families attending ECH has recently retired but her replacement has been appointed and will start in post shortly. The team often attend out-patient clinics to discuss children with the acute team. They visit the families of affected newborn to give the diagnosis and early education, and refer to the acute team for first clinic appointment. Approximately 18 newborn children were referred to ECH last year. The team are active in chasing up children who fail to attend for clinic appointments, and finding and referring on to the appropriate team any who move out of area, which is reported to be a frequent occurrence.

LINKED HOSPITALS

This network, together with its partner organisation, Kings College Hospital, in principle supports services for children with these conditions across a very wide area, extending throughout Kent and down to the Sussex Coast. In practice, there are clear links between the team at ECH and that in Queen Mary Hospital Sidcup, Medway, and Darrent Valley. Queen Elizabeth Hospital Woolwich has links with both ECH and King's College Hospitals, referring children for different clinical problems to each. Those needing cardiology, renal, neurology or urology input are usually referred to ECH. The KCH team now provide monthly outreach clinic support and a TCD service there, and acutely ill children are usually transferred to KCH. Some of these linked 'local hospitals' are managing very large caseloads, for example over 200 children in Woolwich [which was to be visited separately]. Links with Lewisham Hospital are now more towards King's College Hospital.

There are good communication arrangements between these units, through a 'SKILS' group which meets 3 to 4 times a year to discuss clinical issues and guidelines / protocols. A document outlining network pathways including for referral, is in discussion. The paediatricians from Medway and Sidcup who met the review team confirmed that these meetings were useful for them.

A member of the review team met the paediatric consultant from Medway, and from Sidcup, so more detail is included about these services at these two sites.

At Medway, approximately 50 children with sickle cell disease attend the hospital; a growth from none 3 to 4 years ago. There have been some new births but this sudden growth mostly reflects population movement. The paediatrician who cares for them was trained in an area where sickle cell disease was prevalent. Care is predominantly local with a joint clinic run by the paediatrician and haematologist. Telephone advice is readily available from ECH team, and transfer of acutely ill children takes place – for example a child needing exchange transfusion will usually be transferred. Children attend for TCD screening at the hospital they were attending before moving into the area; it is estimated that about 50% of children have so far been screened, although no local audit of this or other clinical standards has been undertaken. An outreach service, to include mobile TCD scanning, would be ideal for this service.

At Sidcup, about 100 children attend with sickle cell disease, and at Bromley, which is part of the same NHS Trust, about 10. Here too there has been a very rapid increase in numbers, with Sidcup treating only about four children five years ago. Fourteen affected newborn children were seen in the last year. The paediatrician who manages these children sees ECH as the specialist centre he refers to. In practice this only applies to those about whom he is worried. Those who are referred for one reason or another will usually have TCD's and full review at the centre, but he estimates this applies to less than half of his patients. He is concerned about this patchy coverage, and would like to work towards an outreach service. He has been told that the consultant who undertakes TCD scans on the ECH site is willing in principle to undertake this outreach work. A small number of children with thalassaemia major are treated at Sidcup, and they each attend a different paediatrician, which is a cause for some concern.

COMMISSIONING ARRANGEMENTS

A public health consultant from Lambeth met the review team lead, and later there was a meeting with a specialist commissioner from the London Specialised Commissioning Group and the lead commissioner for GSTT from the Lambeth, Southwark and Lewisham [LSL] Commissioning Alliance (now integrating into a new South East London acute commissioning unit). It was explained that there is no specialty commissioning for these services in London, at least at present, and the role of specialised commissioning was likely to be in helping to co-ordinate arrangements and advise PCT's about what should be commissioned, rather than undertaking any direct commissioning. The first pan-London Forum held in February 2010 was the start of the dialogue between commissioners and providers. The lead commissioner for acute services was aware of the services offered at ECH, but not the detail of staffing levels, activity, or how the broader network was developing. The community services are commissioned by Lambeth PCT for LSL, although they are soon to merge managerially

with Guy's and St Thomas'. There was discussion about the need for a broad approach to commissioning services, from the low volume very specialist work though to routine and community based care. The possibility of commissioning the whole service so that care pathways intended to reduce the need for hospital care did not result in financial disadvantage was also discussed.

REVIEW VISIT FINDINGS

ACHIEVEMENTS

1 This is an excellent service overall, with strong medical and nursing leadership in the acute service and strong support from psychology and play services. There is evidence of very good team-working within the hospital extending across to the community team who attend MDT's and ward rounds. There is appropriate support from members of the haematology team.

2 The hard work of the community nurse team is acknowledged. They are working in difficult circumstances with many mobile families and their support in general, and efforts to track children who do not attend appointments and / or move out of area, are noted.

3 Both the acute and community teams are highly valued by user families, who find them friendly and approachable and commented that they 'feel like family'.

4 The focus on the time of transfer from paediatric to adult services is clear. The Trust has appointed some staff members to lead on this, and there is now an outstanding transition service. The materials for use by older children, the regular meetings focussed on increasing knowledge and independence at different ages and stages, including at the time the older teenagers are leaving home, are exemplary.

5 Facilities are good in general, and especially in the out-patient department, teaching and play areas.

6 There is a strong research base, and an active teaching programme for professionals involved in care of children with sickle cell disease and thalassaemia across the UK.

7 The trans-cranial Doppler service was established here well before most other centres, and before stroke risk assessment using this technique became recommended practice. Supported by input from an experienced paediatric neurologist, this aspect of the management of children with sickle cell disease has been at the leading edge of practice.

CONCERNS

1 Data regarding compliance with the newborn screening programme standards were not complete, so it is not clear whether families are informed by the time their affected newborn is four weeks old, or how many infants are seen in clinic by three months of age. More detailed audit is needed for the team to assure themselves of compliance against these targets. [Note: between the time of the visit, and finalising the report, data has been provided for 18 babies born between January and September 2009; these are now included in Appendix 2].

2 If the specialist team is expected to take some responsibility for ensuring that all children in the network are accessing appropriate levels of care, the fact that it was reported that a small number of children with thalassaemia major are treated at Queen Mary's Hospital Sidcup, all under the care of different paediatricians, should be investigated further and different care arrangements considered for them.

3 There were no reported audits relating to children with thalassaemia.

4 There are apparently no clinical guidelines, or patient and family information, relating to thalassaemia intermedia although there are about nine children with this condition under the care of the specialist team. In networks such as this, where the number of children affected with sickle cell far exceeds those with thalassaemia, care must be taken that the latter receive adequate focus.

FURTHER CONSIDERATION

1 Some of the audits against key clinical standards were either old (Pneumovax II coverage, 2003), or incomplete (penicillin prescription and adherence to treatment, which formed part of a small audit of transfused patients numbering only 13 of the total treated). It will be important to undertake wider audit of these aspects of care.

2 Some of the premises are rather small and cramped, particularly children's A&E and the Day Care area.

3 Document control arrangements are not robust. Some of the written documents were not dated, or did not have a review date, and for some it was not clear if they were in draft form, or agreed and now in use.

4 The door of the Day Care ward has a sign indicating that it is the 'Haemophilia Centre', and it might be worth adding to this to include children using this service, or making a more general sign to cover all Day Care attendances.

5 In this network, serving a very large number of children and families, the roles and responsibilities of the specialist team based centrally at GSTT and that of the experienced 'local' clinical teams might need to be revisited and possibly re-drawn, bearing in mind the geographical areas of likely future growth in case numbers. Some of the 'local teams' manage a larger caseload than specialist teams elsewhere. It is not feasible, and probably not necessary, for the specialist team to review every child from across the whole network every year, and the expertise of the local teams at linked hospitals managing large caseloads must be acknowledged. As in other very high prevalence areas, it might be appropriate for one or more of the larger linked hospital teams to act as intermediate level provider, 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 otherwise see mostly those children from any part of the network who have complex or difficult clinical issues. For some highly specialist services, for example TCD screening, an outreach service would be ideal. There will need to be discussions with commissioning teams so that the location of services of different levels provided for their population, as well as plans for developing and monitoring services, can be agreed.

6 There are opportunities across this very large network, working with partners at King's College Hospital further, to develop a system of support for the many large linked hospitals. This would enhance the quality of care for all the children served. Outreach clinical and TCD services are already established by the KCH team at two of the busiest linked hospitals. Discussion between the key specialist providers might include planning a co-ordinated approach of linkage across the network, so that each linked hospital predominantly works with one specialist team, at least for affected newborn, and a more systematic pattern referral is established. It could then be decided if patient numbers in any one local hospital justify an outreach service there, even if only six – twelve monthly, or whether it is necessary for families to travel to the centre for their specialist reviews. - Some agreed geographical

boundaries for working in this way is likely to be needed if the aim of each of the children managed by the smaller linked hospitals having specialist input is to be met.

GOOD PRACTICE

1 Patient and family information relating to sickle cell disease is strong. The reviewers especially noted the enuresis leaflet, and one outlining the process of MRI scans.

2 There is a 'sickle cell care plan' describing fully all aspects of care for difficult or complex patients, for example those who cannot communicate easily.

3 The 'cannulation plan' held at the back of children's records, with a sticker to alert staff to it, is excellent and must help children gain confidence about the procedure, knowing that their preferences and concerns are clearly understood and taken into account.

4 There is a fortnightly MDT attended by the extended team including the community nurse.

5 There is close collaboration with the pain team in managing in-patients, with the facility for continued out-patient input for children with particular difficulties.

6 Some of the practice in A&E is noteworthy. In particular, there is a very comprehensive 'Emergency Medicine Guide' given to all junior doctors, a file is held in A&E which includes detailed individualised care plans and a copy of the last clinic letter ensuring up to date information is to hand whenever a child attends, and a box is in place where details of children attending / being admitted are placed for the attention of the hospital and community nurse specialists.

7 In the Day Care area, a file is held containing data relating to all the regularly transfused children, including their regular transfusion volumes and a log of pathology results. This is also uploaded onto a database.

8 Communication between teams across the network is enhanced by 'SKILS' meetings [Sickle kids in London South] three – four times per year, at which individual cases and shared guidelines and protocols can be discussed.

9 Finally, the review team noted the disposable curtains in the ward areas, which are novel, and likely to be valuable in assuring children and families of the Trust's commitment to infection control.

APPENDIX 1: MEMBERSHIP OF THE REVIEW TEAM

Dr Corinna McMahon	Consultant Haematologist	Our Lady's Children's Hospital, Dublin
Dr Gavin Cho	Consultant Haematologist	North West London Hospitals NHS Trust
Christine Williams	Service Director	Sickle Cell & Thalassaemia Centre, Hackney
Valerie Baker	Service Manager	University Hospitals of Leicester NHS Trust
Elaine Miller	User Representative	UK Thalassaemia Society
Rajpal Singh	User Representative	
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	*But information for families of sickle cell affected child did not include mention of checking the spleen. User parents however confirmed that this was taught. Some good health promotion materials. The out of hours contact guide was felt not to be clear, and no alternative language materials were presented.* *For children with thalassaemia and their families, information seen was very sparse, including only a rather old information sheet about desferrioxamine.*

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)	N	*Only regarding prevention of infection, seen in a generic letter to be sent to GP's after first visit by newly diagnosed infant.*
3	Information should be available on transition to adult care. This information should cover all aspects of the transition (QR43).	Y	*This is really strong, in written information and in practice.*
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	*At least some users do hold their records.*
5	The locally agreed patient-held record (QR4) should be in regular use within the LHT / SHT.	N	*Apparently now there are no more patient held records to give to families.*
6	Services should be provided in a child friendly environment, including toys and books / magazines for children and young people of all ages.	Y	*Outpatient and ward facilities are excellent, A&E and day care areas are somewhat small and cramped.*
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	*All are available in-house.*
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	*Guidance is in place and also based on frequent assessments of care needs and Paediatric Early Warning Signs.*
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 strong psychology and play input was evidenced, especially in regard to the transition process, with a dedicated team focussing on these years.*
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	*A draft, and out of date, document was seen which did not cover all aspects of this QR although the visiting team were informed that a meeting to address this was planned soon after the visit.*
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	*The team were clear that this was met in practice, although staff training records vs NSPA SPN14 were not seen.*
20	All staff involved in the care of children and young people with haemoglobinopathies should undertake regular child protection training.	Y	
CLINICAL and REFERRAL GUIDELINES			
21	Guidelines should be in use covering a How to establish and confirm diagnosis b Parent and sibling testing	N	*No specific guidelines were seen which covered this.*

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	*These were good, excepting no travel advice guidance; however, this was covered in information given to patients / families.*
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	*All good except no guidance on acute visual disturbance was included.*

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	

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	*Focus particularly on transition age rather than younger children.*
27	Guidelines for referral for consideration of bone marrow transplantation should be in use.	Y	
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	*Transfusion and chelation guidelines were all felt to be very good.*
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	*Although small numbers, some children are treated and some guidance is needed.*
33	Clinical guidelines should be in use covering: a Indications for exchange transfusion b Arrangements for carrying out an exchange transfusion.	Y	*And user friendly, easy to follow.*
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	

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	
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.	N	*But the document ' guidelines for the care of inherited anaemias at Kings College Hospital, Guy's and St Thomas' sickle cell network' once agreed 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	*No protocol in place, but copy letters to patients seen in practice. A pilot evaluation of the patient held record was seen, although the supply of these has now run out.*
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	N	*No written guidance seen.*
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	*No available out of hours clinic appointments, although regular transfusions are mostly at weekends and out of hours phlebotomy can be accessed.*

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	*Good written policy and community nurses confirm it is used in practice.*
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	*Excellent documentation and practice discussed. Users confirm it works well.*
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	*Active engagement in support group activity by professionals.*
45	The SHT should run a programme of training and awareness of the management of patients with haemoglobinopathy for its main referring LHTs.	Y	*Annual meeting in place.*
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	*SKILS meeting three – four times per year.*
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	*There are regular meetings with the community nursing team who manage the newborn screening results and screening standards included for discussion at these meetings.*

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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.	N	*Detailed audits seen regarding expectations / concerns of transition age children, coding, and regularly transfused children. Last Pneumovax audit 2003. A limited number of children had compliance with regular penicillin V included as part of an audit of pain management. TCD audit is up to date and detailed.* *No thalassaemia audits were presented.* *No review of patients who have died were included.*
51	Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry.	Y	*This is progressing well.*

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	

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	*71%, from figures gathered and sent by community team after the visit [for 18 babies born Jan – Dec 2009].*
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)	N	*But very close – 88% [figures as for P3].*
P5	Timely confirmation of diagnosis for infants with a positive screening result**	N	*44% [figures as for P3].*
S1i	Failsafe to ensure ongoing care	Y	*Policy seen and discussed.*
S1ii	Up-to-date registers maintained of babies (cases) for which units are responsible	N	*No register was seen.*

** 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.