UK Forum on
Haemoglobin
Disorders

WM QRS West Midlands
Quality Review Service

Services for Children with Sickle Cell Disease or Thalassaemia
at
Royal London Hospital
[Barts and the London NHS Trust]

Quality Review Visit Report
Visit date: May 6th 2010

Report finalised: September 7th 2010

CONTENTS

INTRODUCTION

This report presents the findings of the peer review visit to services for children with sickle cell disease or thalassaemia at Barts and the London NHS Trust, Royal London Hospital [RLH] site, which took place on May 6th, 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 the Royal London Hospital 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 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 BARTS AND THE LONDON HOSPITAL NHS TRUST

Service (as at May 2010)	Patient numbers sickle cell disease	Sickle cell on long term red cell transfusions	Sickle cell on hydroxyurea	Patient numbers Thalassaemia Maj	Int
Royal London Hospital	340	38	10	11	8
Newham University Hospital	337	6	22	6	16
Whipps Cross Hospital	136			6	<5
Queen's Hospital [B, H & R NHS Trust]	227	7		7	-<5
Basildon [SW Essex]	64				
Essex - other	23	<5			
Totals	1127			59	

[small number of 'other haemoglobin disorders' omitted – but included in total figures]

BACKGROUND

The specialist team at RLH manage a large number of children and adults with sickle cell disease and thalassaemia on site, and support management of a very large number of affected children across a wide established network – the 'East London Haemoglobinopathy Network'. The network covers residents of City and Hackney, Newham, Tower Hamlets, and all of Essex. The children's service is led by two haematologists, who also run the adult red cell service, and a paediatrician, with junior medical staff and two on-site clinical nurse specialists. There is strong support from other team members including a dietician, teacher, and psychologist with the dietician attending the main weekly clinic, and the teacher and psychologist attending the weekly MDT. There is an active research programme and many major publications have come from the Department. It is anticipated that the recent appointment of a haemoglobinopathy service administrator will improve data quality and management across the network.

Services are currently provided in disparate sites around the old hospital buildings, pending the transfer in 18 months into the large new hospital building.

ACCIDENT AND EMERGENCY

There is a busy children's A&E Department and many children needing urgent care for sickle cell pain crisis or other acute problems present here. Triage is very prompt and a standardised proforma is used for 'clerking', which contains within it a copy of the analgesia flowchart. First analgesia is intra-nasal diamorphine to age 14 years, then usually transbuccal fentanyl. No individual patient care plans are held as all but a few receive this standard analgesia. Patients managed off-protocol are included in a 'protocol exception' list. Time to first analgesia averages 15 minutes which is well within the recommended period. The department's clinical management guidelines are available in hard copy and are easily accessed on the intranet. The pain management pathway for children with sickle cell is posted in a clinical area highly visible to A&E staff, as well as accessible via the intranet. The parents of children with sickle cell disease using the service reported no concerns with delay in receiving analgesia.

Outpatient Clinics

Weekly clinics, predominantly for children with red cell disorders, are held on a Wednesday morning in a children's outpatient department, run by the three consultant medical staff, trainee doctors and the CNS's. The dietician comes to this clinic and a dentist is available to see children on alternate weeks. Phlebotomy is available within the clinic and there is a play area with play therapist present. In addition, a clinic on a Thursday afternoon is run by the haematologists in which children needing longer appointments, such as first visit or annual review, are seen. TCD is offered in this session for children coming for annual review from out of the local area. A Friday afternoon routine TCD session for local children is available twice a month. TCD's are undertaken by the two haematology consultants. An outreach TCD service is offered at Newham Community Centre. Joint clinics with Paediatric Endocrinologist take place every three months, and with a Hepatologist approximately every six months. Interpreters are based in the outpatient clinic and are usually available when needed.

Although it is understood that significant investment in the fabric of the buildings is unlikely, given the planned move to the new hospital towers in under two years, the clinic rooms particularly are in a poor state of repair.

Day Care

Day case transfusions are offered on two different day care areas. Buxton Ward is a 7 am to 7 pm five-day ward. Ocean Ward is open Monday morning through to Saturday morning. Approximately 16 transfusions are undertaken per week for this patient group on Buxton. Four sessions take place on Ocean in the evening and are offered to older children. Provision of more out of hours bookings would be used if available. Cannulation is undertaken by one of the two nurse specialists for children on both wards. Other ward staff are not trained to do this and, in her absence, children wait for cannulation by a doctor.

In-Patient Facilities

In-patient care is on a designated ward, Ann Riches Ward. There are 4 to 5 admissions for sickle cell presentations each week, and it is unusual for bed pressures to require admission to a different ward. Staff are familiar with the management protocols, which are available in hard copy and on the intranet. There is a separate recreation room for adolescents, and a schoolroom run by a very involved and proactive teacher. Urgent exchange transfusions are undertaken on a high dependency

unit, and children with single organ failure can also be managed. Those requiring formal paediatric intensive care are transferred out, usually to Great Ormond Street Hospital, St Mary's Hospital (Imperial College Healthcare NHS Trust) or Guy's Hospital.

COMMUNITY SERVICES

Community nurses based at Newham Sickle Cell Community Centre and Hackney Sickle Cell and Thalassaemia Centre support children and families locally. A newly appointed nurse who will be leading the genetic counselling service in Tower Hamlets is expected to offer services to children there. Only a nurse from the Hackney Centre is regularly able to attend the weekly MDT at present. Staffing levels, and the model of service offered at the three Centres, differ. The modus operandi of the specialist team has evolved to reflect this. The lead nurse at the Hackney centre outreaches into parts of Essex [Barking Havering and Redbridge] where there is currently no community provision (see below).

LINKED HOSPITALS

Many of the hospitals linked with RLH are caring for very large numbers of children and there is considerable local expertise in the management of most clinical problems. Support needed from the specialist team therefore varies. All the local teams refer children with complicated issues and decisions about the management of iron overload are usually made by the RLH team.

Representatives of the large local hospitals, Newham, Homerton and Whipps Cross, were not able to attend the visit. Consultants from Queen's Hospital and Basildon Hospital did meet the review team, however, thus further detail about services is included only for these parts of Essex. These are important area for focus, however, with rapidly growing patient numbers outstripping current service provision.

The paediatric haematologist at Queen's Hospital manages almost 600 patients with red cell disorders, of whom about a half are children. This large service is currently under-resourced, with inadequate support for the lead consultant. There is only 0.2 wte nurse specialist time, no dedicated psychology input, and no specialist community staff. Consultant medical cover is available for times of leave. A business case for additional staff is currently being assessed. In the meantime, input from the senior Hackney Centre community nurse, working with at-risk couples and visiting families at home to give the diagnosis of an affected newborn, is gratefully acknowledged. Despite the sparse team, almost all care for children with sickle cell and thalassaemia is offered on site. TCD's are now locally undertaken and children seen by the RLH team, and the scan repeated, only if the local scan is

abnormal. Annual reviews take place at Queens, except for the children with thalassaemia for whom this is undertaken at RLH. Referral in for specialist management is usually restricted to those needing high dependency care and surgery, although telephone advice is sought - and readily available - for other queries. The RLH protocol for thalassaemia care is used. A local protocol is used for sickle cell management but this has been approved by the specialist team.

In SW Essex, patients are managed at Basildon Hospital by a consultant paediatrician with a primary interest in endocrinology, but with experience in haemoglobin disorders during her training. There is no named cover for the service at times she is away. Five years ago, there were about 5 affected children attending, now there are approximately 60, with about 20 new l cases being seen in the last year (11 newborn and 9 moving into the area). There is no day care provision for transfusions, and pressure on in-patient beds can lead to cancellation of booked dates. Two part time advanced nurse practitioners provide important support in the community.

Both consultants value the support from the specialist team, network meetings are useful and there have been helpful teaching seminars offered at both sites by the lead haematologist from RLH.

COMMISSIONING ARRANGEMENTS

Representatives from Children's Community Services at City and Hackney PCT, Public Health Services at Tower Hamlets and Disabilities Commissioning at City and Hackney came to meet members of the review team. The public health team work with the provider team particularly around the newborn screening programme, and are familiar with the range of services. Commissioning for acute service was not in the remit of any of the teams represented, and is considered to be part of the general paediatric commissioning arrangement. For the community services, however, there are clear arrangements and for City and Hackney [Newham and Tower Hamlets teams not available for discussion] performance targets are in place, and activity data are supplied by the Hackney Centre. It was agreed that discussion between the commissioners of the each community service, the community team themselves and the hospital specialist team would be very useful to ensure that the activities undertaken in the community best served the needs of affected children and families.

REVIEW VISIT FINDINGS

ACHIEVEMENTS

1 This is a strong service, with highly developed network links, offering care to a very large number of patients. The strength of medical and nursing leadership in developing the services, and the energy and commitment of all the care team members, were much in evidence. The recent appointment of a network administrator further supports the team's activity. Users and families value the care they receive very highly. In addition, there is considerable research activity and the team have published a number of key papers relating to the subjects of this review in respected peer reviewed journals.

2 Written protocols and guidelines are excellent, clear and simple, containing key information in an easily accessible form. Especially good examples included: the network guideline for referral of acutely ill children, the transition 'structured interview', and timelines charts for interventions at different ages. Some patient information was also found to be noteworthy, including the 'self-help' manual for children managing pain, silent stroke, and dietary information for those with sickle cell disease.

3 Input from support team members is excellent, in particular, the involvement of the teacher, who attends the MDT and makes school visits, and the dietician, who attends the clinic regularly. The multi-specialty shared clinics are comprehensive and the alternate weekly input from a dentist is very good.

4 The timeliness of communicating the diagnosis of sickle cell disease or thalassaemia to the families, and of children being seen first in clinic by 3 months of age is exemplary and exceeds the 'developmental' standard required.

IMMEDIATE RISKS

No immediate risks were identified.

CONCERNS

1 Some of the recommended audits, against key clinical standards for children with sickle cell disease, have not yet been undertaken.

2 Staffing provision, including medical, hospital specialist and community nursing, in parts of Essex is insufficient for the numbers of patients. Services for this large patient population are stretched to worrying extent. There is a business case [Queen's Hospital] awaiting approval for further staff. Reviewers hoped that this will be successful and that appointments to the necessary extra posts can be made in the near future.

FURTHER CONSIDERATION

1 In this network, serving a very large number of children and families, the roles and responsibilities of the specialist team based centrally at RLH and that of the experienced 'local' clinical teams, some of which each manage a larger caseload than specialist teams elsewhere, might need to be revisited and possibly re-drawn. This is particularly relevant because of the likely future growth in case numbers in some of the network areas. 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 an intermediate level provider, perhaps offering an annual review service for children from smaller units geographically close to them. The specialist team might focus on providing comprehensive care to their own local families and otherwise see children from any part of the network who have complex or difficult clinical issues. Discussions with commissioning teams on the location of services of different levels, as well as plans for developing and monitoring services, would be needed to support this change.

2 Information for parents and families of children with thalassaemia was felt to be sparse, and it did not cover all recommended issues.

3 The services are currently housed in old hospital buildings, and it is understood that significant investment to improve them is going to be unlikely given the move into the large new hospital premises expected within two years. The out patient clinic rooms in use, on the first floor of Fielden House, are dilapidated and could be renovated to a level suitable for use over the intervening period.

4 Children attending clinic may see several professionals during one visit, for example a consultant, a specialist nurse and a dentist. They may also receive TCD screening as well as an annual clinical review. It was not clear that these separate clinical contacts are appropriately recorded and coded.

5 There is currently no 'out of hours' provision of phlebotomy or out patient clinics. This should be considered in order to reduce children's time out of school for planned care.

6 Cover for absence of the specialist nurses is not available. In particular in the absence of the CNS who places venous cannulae before red cell transfusions; children have to wait for a doctor to undertake this procedure.

7 The medical and nursing team members from the Essex linked hospitals who met the review team indicated that they did not always receive all relevant information about their shared-care patients. This could include blood results, TCD results, and outcomes of MDT discussions at neuroradiology meetings.

8 There is an opportunity further to engage commissioners of the community services for discussion. With three different models across the three PCT's, some evaluation of what works well could be undertaken and shared so that appropriate focus can be given to priority activities. Likewise, the acute service providers, functioning as the hub of an excellent and growing clinical network, might work with commissioners to shape services for the future.

GOOD PRACTICE

1 The A&E clerking / admission proforma for children presenting with sickle cell pain contains a copy of the analgesia pathway so that errors, and variation between individual clinicians, is minimised.

2 The active move from parenteral to intranasal/transbuccal and subsequently oral analgesia avoids unnecessary cannulation and helps lead to later independent self care.

3 All the nurses on Buxton day care ward are trained and competent to undertake exchange blood transfusions.

4 Children on regular transfusion can attend the clinic area at any time to collect their pre-transfusion sample request forms, prepared for them by the CNS, and have blood samples collected.

5 Nurse led drop-in and monitoring clinics are very appropriate, led by the experienced nurse specialists on the team, and help reduce clinic waiting times.

6 There are prompt efforts to track children who do not attend for outpatient appointments, with a home visit frequently occurring after a single missed visit providing a robust failsafe.

7 There is a clear focus on separating children's services from adult services; for example, a separate dedicated children's diagnostic imaging department is available.

8 Leaflets and posters relating to sickle cell and thalassaemia are prominently displayed in the clinical areas, making this a highly 'visible' service, and helping to make service users feel included.

APPENDIX 1: MEMBERSHIP OF THE REVIEW TEAM

Dr Mark Layton	Consultant Haematologist	Imperial College Healthcare NHS Trust
Dr Olu Wilkey	Consultant Paediatrician	North Middlesex University Hospital NHS Trust
Helen Appleby	Clinical Nurse Specialist, Paediatric Haemoglobinopathies	Guy's & St Thomas' NHS Foundation Trust
Teresa Warr	Head of Service Development	South Central Specialised Services Commissioning Group
Elaine Miller	User Representative	UK Thalassaemia Society
Ravinder Raj	User Representative	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.	N	*Some excellent material for sickle cell families – pain self-help manual, 'silent stroke' leaflet for teachers.* *But information for families with thalassaemia was sparse, and no information was found regarding when to seek emergency advice, nor about iron overload problems.*

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	*But some of the necessary information was seen in clinic letters to GP's.*
3	Information should be available on transition to adult care. This information should cover all aspects of the transition (QR43).	Y	*Excellent material was presented.*
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.	N	
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 clinic rooms in outpatients were in a poor state of repair.*
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	*A list of expected competencies for nurses at various 'bands' was given, but no documentary evidence of individual competency assessments was provided.*
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	*Several areas of Essex [especially Barking, Havering and Redbridge] currently lack this even though, in some cases, patient numbers are high.*
14	There should be agreed cover arrangements for the outreach nurse specialist / counsellor.	Y	*In place for local boroughs, but as for 13.*

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 the level of input from some of these support team members is outstanding.*
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	*No SLA between the acute and community services is in place. An inter-PCT agreement about screening services was seen.*
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.	N	*In place for much of the time but cover for the CNS for these duties, when she is on leave, is not in place.*
20	All staff involved in the care of children and young people with haemoglobinopathies should undertake regular child protection training.	Y	*Training records for key staff were submitted and checked after the visit*
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	
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	

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	*The guideline for thalassaemia was seen, and the guidance for regular clinic visits for sickle cell affected children includes all aspects of this QR for annual review.*

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)	N	*No written guidance was in place although in practice referral can be at the request of patient / family or by member of professional team, and the psychologist attends the weekly MDT at which need for referral is considered.*
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	*All transfusion guidelines were excellent.*
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.	Y	
33	Clinical guidelines should be in use covering: a Indications for exchange transfusion b Arrangements for carrying out an exchange transfusion.	Y	
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	Y	
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	*Network guidelines regarding consultation for acutely unwell children are innovative and good.* *Copying letters to parents is inconsistent [from case notes], and linked hospitals indicated they do not always get full communication about shared children.*
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	*As self assessment.*
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	*A brief leaflet describing the use of desferrioxamine was presented, but no material regarding teaching of children or assessment of competency.*
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	*Available for transfusion but not for phlebotomy or clinic appointments.*

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	*Very strong – 'structured interview' approach is excellent.*
44	The team should have in place mechanisms for receiving feedback from patients and carers about the treatment and care they receive. a Mechanisms for involving patients and carers in decisions about the organisation of the services. b Mechanisms for encouraging the development of local support groups.	Y	
45	The SHT should run a programme of training and awareness of the management of patients with haemoglobinopathy for its main referring LHTs.	Y	*Self-assessed as 'no', but in practice the linked hospitals reported useful teaching visits by lead consultant.*
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	*Minutes of meetings seen.*
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	*Minutes of meetings seen.*

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	*Audits indicating proportion of patients taking penicillin, having pneumococcal immunisation, annual reviews within last 12 months, were not presented.* *Thalassaemia audits have been undertaken*
51	Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry.	N	*But the locally held database is excellent and the recently appointed service administrator is planning to establish link with 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	

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	Y	
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	*[the first part of this standard overlaps with P3, and has been ignored]* *Figures are excellent – 99% referred by 8 weeks and 99% seen by 12 weeks.*
P5	Timely confirmation of diagnosis for infants with a positive screening result**	Y	
S1i	Failsafe to ensure ongoing care	Y	*No specific policy for failure to attend first clinic but DNA policy and team clearly describe actions including escalating to social services if failing to attend*
S1ii	Up-to-date registers maintained of babies (cases) for which units are responsible	Y	*No babies are lost to follow up.*

** 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.