UK Forum on Haemoglobin Disorders

WM QRS West Midlands Quality Review Service

Services for Children with Sickle Cell Disease or Thalassaemia
At
Leeds Teaching Hospitals NHS Trust

Quality Review Visit Report
Visit date: January 28th 2011

Report finalised: 22nd July 2011

CONTENTS

INTRODUCTION

This report presents the findings of the peer review visit to services for children with sickle cell disease or thalassaemia at Leeds Teaching Hospital NHS Trust, which took place on January 28th 2011. 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 Leeds Teaching Hospital NHS 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 parent 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 LEEDS TEACHING HOSPITAL NHS TRUST

Service (as at January 2011)	Patient numbers sickle cell disease	Patient numbers thalassaemia	Patients on long term red cell transfusions
Leeds General Infirmary	57 [46 SS]	6	8
Bradford	19	45	
Halifax/Huddersfield Hull Airedale	<5 <5 0	<5 0 6	<5 6

Substantial changes relating to the children's sickle cell and thalassaemia services had taken place over the months preceding the review visit. As part of a larger move of children's services from the St James' Hospital site to Leeds General Infirmary site [LGI] during 2010, the red cell services moved in October 2010, and at that time there was a change in medical leadership, and a lead nurse [0.2 WTE for this service] was appointed.

Accident and Emergency

Children requiring urgent care did not usually present to A&E, but to the Day Care area in routine hours, or to the designated in-patient ward out of hours. A copy of the clinical guidelines was in place in A&E. Initial analgesia for children with sickle cell pain crisis was usually with oral morphine, and the aim was for a first dose to be administered within 30 minutes of arrival. No audit had been undertaken to confirm this was happening in practice.

Outpatients Clinics

Children with sickle cell disease of thalassaemia attended a 'benign-haematology' clinic held weekly, run by the lead paediatric haematologist, the deputy, and an additional paediatric haematologist, the nurse specialist and one or two trainee doctors. Before clinics started there was a half hour professional meeting to discuss the children attending and to share any concerns or issues to be considered. The meeting was also attended by the designated social worker and a pharmacist. Out-patient nursing staff took blood samples on site and there was a facility for near-patient full blood count testing. The clinic facilities, and the adjacent Day Care area, were of a high standard. There was a separate teenage waiting area. No out of hours clinic appointments were available.

There were plans to increase out-patient provision, with a weekly MDT and additional clinic session at which annual reviews could be undertaken. MDT discussions and case review took place three monthly. Transcranial Doppler stroke risk assessment scans were offered at a different time of the week but it was hoped these could soon be offered within the regular clinic.

Day Care

The Day Care area was adjacent to the clinic area, and managed by the same nursing staff. It was open 8 am to 6 pm on weekdays. Children could attend for transfusion after school and if need be transfer to the in-patient ward to finish. Same-day pre-transfusion sampling was offered to save double trips. Nurses established venous access for children who had central line devices, and most other children. Paediatric doctors were called for those with difficult access but this was not reported to lead to undue delays. Weekend transfusions could be accommodated on the in-patient ward but this was apparently not requested often, and was mostly only being done for specific reason, for example, during exam times for an older child. The Day Care area was also used for children with acute problems presenting during the day, by-passing Accident and Emergency. There was sufficient space for this not to lead to problems in accommodating booked children booked also.

IN-PATIENT FACILITIES

Children requiring in-patient care, and those presenting acutely out of hours, were managed on one of two ward areas depending on their age, with a cut off of 13 years. There was a weekly ward cover system by one of six consultants in rotation, including paediatric oncologists as well as haematologists. In-patients were usually also seen by one of the paediatric haematologists and by the nurse specialist. If children needed red cell exchange transfusion for sickle cell complications, this was undertaken on site by a team out-reaching from the St James' site, during working hours. If a child needed an exchange transfusion out of regular hours, it was undertaken manually. For out of hours concerns around an in-patient, if the duty consultant was an oncologist, they sometimes needed to 'ring around' the haematologists if they needed clinical advice. The team had been considering the possibility of establishing a second 'benign haematology' out of hours rota between the four paediatric haematologists.

The in patient facilities were spacious, and bays include televisions and play stations for all children. The ward included a well set-up school room and a pleasant play area with three nursery nurses and one play specialist. There were good facilities for parents who wished to remain in hospital with their children, including chair beds in all bays, a shower room, sitting room and kitchen area.

COMMUNITY SERVICES

The community red cell nursing team, which also covered adult services, consisted of two WTE posts, one filled by two job-sharing nurses. They worked closely with the antenatal screening co-ordinators to deliver antenatal counselling and they shared partner testing between the teams. The community team were responsible for receiving results of newborn screening for haemoglobin disorders, and for undertaking home visits to advise families of results. Audit data against screening standards, provided after the visit, did not indicate compliance in terms of time to referral to clinic, or age of babies at the first clinic visit [see Appendix B, Quality requirements P3-5, p21].

The community nurses attended the first clinic appointment for newly identified babies and followed this up with a further visit or phone call. They had also been doing home visits to the families of all carrier babies identified by the screening programme. They received copies of discharge letters after children had needed in-patient care, and offered follow-up contact to the families. The team felt that the interface between the acute and community services worked well, and they also worked closely with the designated social worker. Attempts to offer support groups had not been successful as uptake was poor. The community team were expecting soon to come under the management of

Leeds Healthcare Trust. They were unaware of a service level agreement. They were not permitted to visit outside the Leeds area but could offer telephone advice to families living outside Leeds, or see them if they attended the community centre.

User Feedback

Only one parent came to meet the visiting team. Her experience of services had not been typical due to the nature of her child's condition, so it was not possible to get insight into the likely service experience of other users. It was unfortunate that the review team was unable to meet any parents of sickle children as they form the majority of the patients and it would have been useful to hear their experiences, especially relating to acute and out of hours care.

Linked Hospitals

The largest linked hospital was Bradford Royal Infirmary, which has a smaller sickle cell caseload than LGI, but a considerably larger thalassaemia practice. The previous specialist team lead had been offering outreach clinics there, but this had ceased some time before. Within the preceding two years, a new paediatrician had been appointed. Members of the review team talked to this paediatrician and a non-clinical manager by telephone. The service in Bradford appeared to run largely autonomously. Children were reviewed on a three to four monthly basis by the paediatrician working with a specialist nurse and nurse counsellor. The team in Bradford noted that specialist annual reviews were not currently being offered by the Leeds specialist team to Bradford children. Urgent clinical advice could be accessed from the centre, and the ready availability of telephone advice was appreciated. Children with sickle cell disease were offered TCD scans at LGI, but at the time of the visit, no outreach service was planned. Funding arrangements for the TCD service for Bradford children appeared to be uncertain.

Because of the large paediatric thalassaemia case load in Bradford, in excess of the number of children being managed at LGI, and because network links with Leeds appeared not to be robust, the visiting team subsequently decided that there should be an additional informal visit to Bradford to assess in more detail how services were functioning. Any conclusions from that visit will be included in a supplementary report.

Other linked hospitals were Calderdale Royal Hospital, covering Huddersfield and Halifax, Mid-Yorkshire Hospital serving Dewsbury, Pontefract and Pinderfields, Harrogate Hospital, York District Hospital, Hull Royal Infirmary and Scarborough District Hospitals. Small numbers of children were managed at these hospitals and they mostly attended LGI for annual review. Although there was a

functional paediatric oncology network covering these hospitals, the Leeds team acknowledged that the network for non-malignant conditions including haemoglobin disorders had lapsed and that meetings covering management of these disorders were not in place.

Commissioning Arrangements

The commissioner who met with the review team described the location of specialist and local haemoglobinopathy services in the region for which they were responsible. She had a good understanding of the regional prevalence of haemoglobinopathy and how it differed based on local populations. Haemoglobinopathy had been reviewed as part of their commissioning process and no areas of significant risk had been identified and therefore a more detailed piece of work had not been undertaken.

Review Visit Findings

Achievements

1 A quite new clinical team, supported by some longer serving members, had taken over this service at a time when it was also being re-located from one hospital site to another, and they had managed this transition well. A lot had been achieved in a short period of time, and there was a sense of enthusiasm and vision about what the team was working towards. The hospital team worked well with a strong and experienced community nursing team.

2 The new facilities were of a very high standard with good clinical adjacencies and an experienced and involved, flexible nursing team.

3 Attention to the needs of older children and teenagers as well as younger children was evident at all levels, with separate waiting areas in outpatients and separate ward facilities, including the flexibility to admit teenagers with red cell disorders to the Teenage Cancer Trust unit. Facilities for parents were also excellent.

Immediate Risks

No immediate risks were identified.

CONCERNS

1 There were no audits against key clinical standards, except for TCD screening uptake.

2 The functional links with Bradford Royal Infirmary as the major linked hospital with a large thalassaemia practice appeared to have broken down. The team at the specialist centre did not have knowledge of care standards there. Early consideration about how to ensure appropriate care for Bradford children was needed, including a discussion about whether the Leeds team was best placed to offer specialist support there.

3 There was no practical, written guidance for undertaking manual exchange red cell transfusion, although this is a procedure which might need to be performed out of hours by inexperienced doctors.

FURTHER CONSIDERATION

1 Some of the protocols and guidelines needed to be further developed into operational documents. At present, many were descriptive and useful educationally, but did not offer practically useful direction for front line staff delivering care, especially out of hours. Management support for this work, as well as to offer some data handling input and support the process of entering children onto the National Haemoglobinopathy Registry, may be beneficial.

2 The network in general is in the early stages of development, and some links which were previously in place appeared to have lapsed. Establishing these, or re-establishing them, will need managerial and commissioning support. Improved communication between the teams at the centre and the linked hospitals will also be needed if the network is to become fully functional. If it is decided that the specialist team at Leeds is to undertake annual reviews on all the children in the network, clinician's job plans are likely to need review.

3 Establishment of a trans-cranial Doppler stroke risk assessment screening programme at LGI in summer 2010 was an important development. However, with a relatively small number of sickle cell children in the network, these scans should be undertaken by no more than two operators, to allow each to undertake sufficient procedures to assure competence.

4 Funding arrangements for TCD screening of children with sickle cell disease in linked hospitals, particularly Bradford, need review to ensure this service continues to be offered.

5 Neither the acute nor community team could present data at the time of the visit to confirm whether the service was compliant with the screening standards concerning a) the time to informing the family of an affected newborn, and b) referral to and attendance at the clinic for the first time. Such data should be regularly reviewed with a view to improvement.

6 The community team visited the families of all children identified by the newborn screening programme as carriers for a haemoglobin disorder. This is not usually offered in other services and reviewers questioned its value. Consideration might be given to prioritising the tasks of the team so that they can to identify more time to work to the newborn screening standard timelines, attend clinic, see children when they are in-patients, and undertake more support work with children at home.

7 As out of hours cover could be by a paediatric oncologist, with no specialist training in the management of children with haemoglobin disorders, consideration should be given to how cover could be more appropriately provided, for example by establishing a second, benign haematology rota, or perhaps requesting cover from the adult haematology services working with the paediatric specialists.

8 The lead nurse had recently been appointed, and was contributing well to the service, but had not had training specifically relating to the management of haemoglobin disorders. Reviewers considered that this training was required within a short time frame.

9 A leaflet describing services for users, including key contacts and clinic and ward facilities, would be useful for families new to the service as well as those who had transferred over from the previous service at St James' Hospital. Suitable information was included in the patient held record for children with thalassaemia, which was seen in draft form.

10 Psychology support for the service was available, but not in the systematic way currently considered as good practice. There was no named psychologist for the service, and neuro-cognitive assessments were not available. For other support services, specialists with an interest in haemoglobin disorders were named for a few, for example cardiology, but not for all, for example, orthopaedics. Referral to a named specialist with an interest allows development of expertise in that consultant team for the specific problems children with haemoglobin disorders encounter.

11 Review of guidelines for starting routine penicillin should be considered. It is usual practice to start routine twice daily penicillin, as prophylaxis against pneumococcal infection in babies with sickle cell disease identified through the newborn screening programme, at the first

clinic visit without waiting for confirmation of the diagnosis. Waiting until a subsequent visit can engender delays and makes starting by the age of three months more difficult.

12 Some documents and guidelines were not dated, and had no planned review date.

GOOD PRACTICE

1 Children who need urgent assessment and treatment access the Day Care unit, or the in-patient ward out of hours. This allowed early access to the specialist team and bypassed delays in a busy A&E Department.

2 Aspects of the Day Care unit were judged to be strong, including extended hours of opening, the facility for on site blood sampling and near patient full blood count analysis, and pharmacy presence to advise on medication.

3 The weekly pre-clinic multi-disciplinary discussion about all children who are expected to attend allowed sharing of concerns, and clarification of each child's needs, across the team.

4 Several documents in regular use were outstanding including a flow chart for the management of acute presentations, patient information booklet about exchange transfusion, protocol for anti-thrombotic treatment, presentation of the analgesic ladder, and transfusion prescription chart.

5 The Regional newborn screening laboratory held tight control of babies identified through the screening programme, and checked that they have started accessing clinical services to ensure ongoing care.

APPENDIX 1: MEMBERSHIP OF THE REVIEW TEAM

Jon Currington	Senior Strategy & Planning Manager	East Midlands Specialised Commissioning Group
Marie Donohue	Consultant Haematologist	Nottingham University Hospitals NHS Trust
Sarah Lawson	Consultant Paediatric Haematologist	Birmingham Children's Hospital NHS Foundation Trust
Elaine Miller	Coordinator	UK Thalassaemia Society
Ravinder Raj	Information Officer	Sickle Cell & Thalassaemia Support Project
Katherine Stevenson	Haemoglobinopathy Specialist Nurse	Central Manchester University Hospitals NHS Foundation Trust
Anne Yardumian	Clinical Lead for Peer Review Programme	North Middlesex University Hospital NHS Trust

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	*But some standard resources were in use, for example the booklet given by the community team to families of babies identified to have sickle cell disease cover all of part 1. There was an excellent family information leaflet on red cell exchange.* *No evidence of age-appropriate information.* *A local service description leaflet was not yet available.* *Very little health promotional material was seen, although much is generally available; locally produced versions are not necessary.*

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	
3	Information should be available on transition to adult care. This information should cover all aspects of the transition (QR43).	N	
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	*A draft for patients with thalassaemia was in preparation and is comprehensive but some aspects of it were in need of updating, for example desferrioxamine was the only iron chelator included.*
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	*Facilities were all of a very high standard.*
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	*And comprehensive CME documentation presented.*
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.	N	*Experienced senior consultant acted as deputy. No CME documentation available.*
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.	N	*Although lead nurse was now in place, no evidence of formal training in haemoglobin disorders was apparent.*
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	*CPD records for the community nurses were not seen, although in discussion it was clear they frequently attend national update meetings.*
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	*But for many services no named contact was given – see further consideration point 10.*
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	*Included in Trust-wide guidance.*
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	*Social work support from a designated team member was strong. Psychology was under-provided.*
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 is in place but in practice the acute and community interface worked well.*
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	*Venepuncture competencies were seen, transfusion competencies were not seen but the visiting team was assured they were in place. No records of individual nurse training / competency were available.*
20	All staff involved in the care of children and young people with haemoglobinopathies should undertake regular child protection training.	N	*CP training record was only seen for lead paediatric haematologist.*
CLINICAL and REFERRAL GUIDELINES			
21	Guidelines should be in use covering a How to establish and confirm diagnosis b Parent and sibling testing	Y	*These were comprehensive and clear.*

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 sick 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	N	*Most guidelines were in place but most were descriptive rather than operational and no practical guidance was seen for a key procedure – manual red cell exchange transfusion – which can be needed out of hours.*

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	*Here too some guidance, especially in relation to management of children with thalassaemia, appeared descriptive rather than operational. However, local team users are reported to find the guidelines clear in a practical way.*

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).	N	*Some proformas were seen, but were not complete in covering aspects of this QR, and did not appear to be in regular use.*
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	*In place for children and families with sickle cell disease, not thalassaemia.*

Ref	Quality Requirement	Met?	Comment
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	
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.	N	*In place for thalassaemia but not for sickle cell, and more detail about pre-transfusion checks were considered necessary.*
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	*The guideline should specifically refer to the hospital transfusion guideline which is available on the intranet.*
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	*But lacked indications for splenectomy.*
33	Clinical guidelines should be in use covering: a Indications for exchange transfusion b Arrangements for carrying out an exchange transfusion.	N	*A list of indications was in place but no guidance for manual exchange, or details of how to refer to NBS team at St James' to undertake in routine hours.*
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	*No policy seen.*
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	*Patient held record for thalassaemia were not yet in use, and none seen for sickle cell children. Parent copies of clinic letters were inconsistent, from review of case notes.*
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	N	*No policy seen or evidenced from discussions.*
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	
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 out of hours clinic appointments offered, but phlebotomy and transfusion could be accessed out of routine hours.*

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	*But needed to be developed into operational guidance.*
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	*Although sparse at the time of the visit, and needed to be developed and made more operational.*
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.	N	*Although community team had undertaken some questionnaire evaluation of their part of the service.*
45	The SHT should run a programme of training and awareness of the management of patients with haemoglobinopathy for its main referring LHTs.	N	*And this needed to be addressed in the near future.*
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)	N	*As for 45.*
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	*Although no minutes of meetings or other written evidence was seen, the team gave assurance that this was in place.*

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	*The team acknowledged the need for these audits to take place in the near future. Data for TCD coverage was seen.*
51	Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry.	N	*No children yet entered onto the National Haemoglobinopathy Registry.*

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.	Y	

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	*This level of discussion had not taken place.*

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	*Data not provided.*
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	*[the first part of this standard overlaps with P3, and has been ignored]* *Data sent after the visit gave timelines for babies between 2005 and March 2010. Standard not met over recent years.*
P5	Timely confirmation of diagnosis for infants with a positive screening result**	N	*Data not seen.*
S1i	Failsafe to ensure ongoing care	Y	*The review team was assured that the screening laboratory lead has responsibility for this and that it was regularly monitored.*
S1ii	Up-to-date registers maintained of babies (cases) for which units are responsible	Y	*A database of newborn babies had been recently established at LG. This was not seen but the team was assured it was in place.*

** 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.