UK Forum on
Haemoglobin
Disorders

WM QRS West Midlands
Quality Review Service

Services for Children with Sickle Cell Disease or Thalassaemia
At
University Hospitals of Leicester NHS Trust, Leicester Children's Hospital

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Quality Review Visit Report
Visit date: June 30th 2010

Report finalised: January 27th, 2011

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INTRODUCTION

This report presents the findings of the peer review visit to services for children with sickle cell disease or thalassaemia at University Hospitals of Leicester NHS Trust, Leicester Children's Hospital, which took place on June 30th 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 University Hospitals of Leicester NHS Trust, Leicester Children's 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 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 UNIVERSITY HOSPITALS OF LEICESTER NHS TRUST, LEICESTER CHILDREN'S HOSPITAL [LCH]

Service (as at 2010)	Patient numbers sickle cell disease	Patient numbers thalassaemia	Patients on long term red cell transfusions
Leicester Children's Hospital	~50	~10	
Northampton	~20		
Kettering	~40		

BACKGROUND

The clinical team providing services for children with sickle cell disease and thalassaemia at Leicester Children's Hospital [LCH] had seen some recent staff changes. After a period when it was unfilled, the paediatric haematology lead consultant post had been held since October 2008 by a long term locum with a special interest in red cell disorders. A data manager for the East Midlands red cell network was appointed in December 2009 and a part time clinical nurse specialist in February 2010. These posts were funded for the first 2 years by the specialist commissioning team. The recent appointment of a paediatric neuro-psychologist [1.5 sessions per week for the children's red cell

network] was another important addition to the team. The service worked closely with adult haematology and general paediatric services, and was well supported by the scientific technical team who had undertaken routine trans-cranial Doppler scans for stroke risk assessment since 2009.

There was a six to eight weekly MDT, which included core team members plus a pharmacist, members of the vascular team, data manager and psychologist. A four-monthly network meeting was hosted by the East Midlands Specialist Commissioning Group [EMSCG]. This co-ordinated outreach support service for the main linked hospitals at Kettering and Northampton and joint working with the other main centre in the region at Nottingham.

Many of the staff and service users who met the review team noted improvements in services since the new team had been in place, working with the support of energetic local management. The positive contribution of the East Midlands Specialist Commissioning Group in developing the team and providing special focus on these services across the network was acknowledged.

Accident and Emergency

Children usually accessed services for urgent assessment and management by presenting to the Children's Assessment Unit, (CAU), which is open 24 hours a day, 7 days a week. Should they present to A&E, they were triaged urgently and managed by paediatric SHO and Registrar grade doctors who were available in A&E at all times. First analgesia was usually intranasal diamorphine or morphine by injection. Oral morphine was used less often. An audit of time to analgesia for children presenting in pain in general gave average times of approximately 15 minutes. All children were managed according to the general pain guideline and no individualised protocols were necessary. Written guidelines were in place in the A&E as well as on the hospital 'intranet'.

The 16 bedded CAU assesses / admitted four to five children presenting with acute sickle cell problems each month. Paediatric SHO and Registrar grade staff were available at all times. Most of the nursing staff were able to take blood samples and insert IV cannulate. Acute pain relief was mostly with oral morphine, or parenteral morphine. Children could remain here for up to six hours, until a bed was available for admission, or until they were fit for discharge.

Outpatient Clinics

Two out-patient clinics were run each week. Children with sickle cell disease or thalassaemia usually attended on a Monday afternoon and the hospital nurse specialist worked with the lead paediatric haematologist in this clinic. The clinic was cancelled if the paediatric haematology lead was away. A

psychologist working in this clinic session was planned. There were also plans for trans-cranial Doppler stroke risk assessment scans to be undertaken at the same clinic visit. Currently these scans are undertaken in the same out-patient area but on a different day from the main clinic. An additional transition clinic is held every six to eight weeks, run by the paediatric and adult haematology consultants, and the paediatric and adult clinical nurse specialists. Haematology trainees have some input into the clinic, but no paediatric haematology or other paediatric trainees take part. The 'did not attend' rate is reported to be low.

Day Care

Planned transfusions were offered on the children's Oncology Day Unit, adjacent to Ward 27, the designated ward for admission of children with red cell disorders. The unit was open 8am to 6pm on weekdays, but there was some flexibility as to finish time. The unit also opened as needed to allow for weekend transfusions. Some of the Day Unit nurses were being trained to cannulate. At the time of the visit the CNS usually inserted an IV cannula before transfusion. If she was unavailable this was undertaken by paediatric medical staff but time from arrival to starting transfusions were reported to be short.

In-Patient Facilities

A recent pathway change had led to all in-patient children being admitted to a designated ward, ward 27, whenever possible and beds were usually available to allow for this. It was unusual for there to be more than one in-patient with red cell disorders at any one time. Children were managed by a paediatrician of the week, with input from the specialist team including the haematology consultant if the paediatric haematologist was away, and haematology trainee medical staff. Clinical guidelines in hard copy were available on the ward, as well as on the 'intranet'. Play input for in-patient children was felt to be good, with general or normalising play being offered as well as specific distraction or focus on procedure difficulties, but perhaps not sufficiently available. Good 'teaching boards' were noted in the clinic and the ward areas.

There was access to HDU and PICU as needed. If exchange transfusion was needed for children with sickle cell disease, this was usually undertaken manually by critical care staff on the HDU.

COMMUNITY SERVICES

The community team was based at a local Health Centre, and consisted of two nurses, and two A&C staff. One nursing post was unfilled at the time of the visit with an appointee due to start two months later. They undertook antenatal counselling for women identified to carry a haemoglobin disorder, and partner testing. Counselling for couples found to be at risk is undertaken by the adult haematology consultant with the clinical genetics team. The community nurses aimed to undertake home visits to families of affected newborn identified through the screening programme, and liaise with the GP and health visitor, before the first hospital clinic visit. They tried to attend that first visit, but did not usually attend other outpatient clinics on a regular basis as they found it difficult to identify the time to do so. The community nurse attended the two monthly MDT. The team also offered drop-in testing, undertook school visits and developed individual care plans with parents. The visiting team gained the impression that closer working between the hospital specialist and community teams would be beneficial, and that some more continued support in the community for affected children and families would be appreciated. It was reported that the community services and UHL have different IT systems, so sharing information between them can be a problem.

Organisation for sickle cell anaemia research [OSCAR] is a national charity which has had a strong local presence, and representatives came to meet the review team. They had provided a good deal of practical support and help to families affected by sickle cell disease, and continued to undertake awareness raising and educational activities in the community in a range of settings. They were funded entirely by public donation. Their activities, which had included hosting a well attended monthly support group, had been limited recently as they were no longer able to use their previous office base, and they had no premises from which to offer their services. This was felt to be regrettable as the organisation was providing some very useful input for families. Further discussion was felt to be needed between the voluntary and community and hospital services, as to how best they can work together in the interests of their clients.

LINKED HOSPITALS

Red cell services at Northampton and Kettering Hospitals were planned to link in with LCH when the network became more fully established. The Nottingham centre, also in the East Midlands, linked with Derby and Lincoln. At the time of the visit Northampton was supported by outreach visits from the team at Birmingham Children's Hospital, and children there could access TCD in Birmingham. Kettering to date had no specialist support on an outreach or in reach basis. However, plans were in

place, through the East Midlands Specialist Commissioning Team Network, to offer outreach clinic services, ideally with a vascular scientist and psychologist as well as medical and nursing team members, who would visit these two linked sites on a regular basis. The Network group was currently developing and agreeing shared clinical guidelines, and formalising communication links.

COMMISSIONING ARRANGEMENTS

The East Midlands Specialist Commissioning Group [EMSCG] working together with a senior nurse manager for service improvement and local service managers, has been highly effective in bringing focus on services for children and young people with red cell disorders in the region, hosting network meetings, and providing pump prime funding for key posts. This was initially in response to concerns expressed by local clinicians approximately two years previously at the failure to offer some basic services, such as trans-cranial Doppler stroke risk assessments for children with sickle cell disease. The Group continued to work with providers on network development, and clear monitored plans with timelines were in place. Some staff, for example the data manager and psychologist, worked across the East Midlands network, and regular meetings are intended to facilitate sharing of clinical expertise, and allow for case review and educational discussions as well as addressing operational issues. The EMSCG website hosted web-based resources for the network but a separate dedicated website was planned. The community services did not come under the Network Group at the time of the visit but this was under review.

REVIEW VISIT FINDINGS

ACHIEVEMENTS

1 This team was offering a very good service, and there was a real sense of momentum for change. Coming from a difficult staffing situation two years earlier, and having appointed some new and enthusiastic staff, the team was pulling the services together well with clinical services focussed in one area of the hospital, and with availability of extended hours services. Practice was supported by clear clinical guidelines which were of a very high standard. Users appreciated the services in general, and commented on recent improvements. They commented particularly on the helpful role of the hospital clinical nurse specialist, and they felt that the team in general knew them well.

2 There was evidence of good teamwork between the paediatric haematologist, the adult haematology team and the general paediatricians.

3 Engagement and support of specialist commissioners and local senior management had strikingly helped pull services forward – by 'pump prime' funding some key posts, and by taking a lead in network development. While this was still in early stages, clear and closely monitored plans were in place, and the work should solidify to give robust and excellent services.

IMMEDIATE RISKS

No immediate risks were identified.

CONCERNS

1 Not all of the recommended audits against key clinical standards for children with sickle cell disease appear to have been undertaken, for example, the numbers who have received pneumococcal immunisation or who have had annual review assessments.

2 The recently appointed hospital clinical nurse specialist post and the data manager post for the network were key to continued service provision and it was felt to be important that the team worked towards securing continued funding for these.

3 Some short term planned absence of the CNS was going to need cover, to allow continuity of service, and the fact that she was available only two days a week caused some problems for service users who are unsure how best to make contact when she is not on site.

4 Working with linked hospitals was not yet sufficiently established to ensure that all children and young people with red cell disorders within the network were receiving appropriate care from this specialist team, including annual review.

FURTHER CONSIDERATION

1 The community and hospital teams might usefully work more closely together. Although a service level agreement was in place, in practice to a large extent they appeared to work as two separate teams.

2 User experience feedback should continue to be sought, and shared between community and hospital teams. There was currently no support group to facilitate the gathering of feedback as well as providing opportunities for learning and family support. A branch of the charity OSCAR had been active locally and had hosted a well attended support group, but this had stopped since they had no base to work from. The clinical services might usefully engage with OSCAR to see how best user support can be offered. There was a sense that the major improvement in hospital services over the last couple of years had not yet been paralleled by a change in community service provision.

3 Some aspects of service could be streamlined to make them more convenient for users. For example, families whose children have regular transfusions sometimes had to go to four different contact points for blood sampling, transfusion, and collection of different sorts of supplies for home chelation – including their health centres and local pharmacies. The outreach paediatric nursing team might be able to visit these children at home to undertake pre-transfusion sampling, which the families would welcome, and perhaps the hospital team could prescribe supplies to be collected from the day unit at the time of transfusion.

4 The plans for children to have trans-cranial Doppler studies undertaken at the same time as their regular clinic visits, and to have the newly appointed psychologist in clinic, would enhance the service considerably. Offering the same joint clinics on an outreach basis for the major linked hospitals would be excellent. Establishing the planned outreach services, especially for Kettering, and for Northampton which is currently covered by outreach from Birmingham Children's Hospital, would allow this network to start functioning fully.

5 There were plans in place for the lead nurse to undertake some specialty training. This should take place as soon as practicable.

6 The service was largely delivered by the lead consultant and nurse specialist. Given the inevitability that out of hours and emergency services are mostly delivered by trainee doctors, involving them in the day to day services would be useful to ensure they have an appropriate understanding of the conditions. Training of junior doctors in the management of these conditions will also be important for future staffing of services here and elsewhere.

7 Some more joint working between the centre at Leicester and that at Nottingham may be mutually beneficial for example, more educational meetings and discussion of complex cases, especially as neither has very large patient numbers.

8 The day unit which the children with sickle cell and thalassaemia attend was signed 'Children's Oncology Day Unit'. A less specific unit name which includes both services maybe helpful, although user families who met the visiting team were not troubled by this.

GOOD PRACTICE

1 Recently established direct access to the Children's Assessment Unit at all times gave excellent access to care.

2 The team was able to start planned transfusions for children attending the Day Unit with minimal delay.

3 Play support was appropriately strong and was provided at two levels of service. Play leaders notes formed part of the written medical records, which reflected the fact that they work as part of the core team. The planned level of psychology support was also appropriate to the needs of these children.

4 Much of the patient information was excellent for example, the sheet detailing what to do after returning home from A&E or the Assessment Unit, the home pain score and management sheet, the TCD leaflet for parents, the service description leaflet and the information relating to the time of transition to adult services.

5 The Trust-wide pain assessment tool in general use reflected the fact that good pain management was focus for all services.

6 The patient transfusion integrated care pathway provided an excellent record of transfusions, and the inclusion of management of transfusion reactions on the same chart was ideal.

7 The training and assessment materials for venepuncture and cannulation were excellent.

8 Free availability of fruit and drinks in the clinical areas for children and families were noted.

APPENDIX 1: MEMBERSHIP OF THE REVIEW TEAM

Dr Asa'ah Nkohkwo	Chief Executive	Sickle Cell Society UK
Marie-Claire Ngeumshe	User Representative	
Dr Jenny Welch	Consultant Paediatric Haematologist	Sheffield Children's NHS Foundation Trust
Louise George	Haematology Nurse Specialist	Sheffield Children's NHS Foundation Trust
Jo Sutcliffe	Service Manager	Kings College Hospital NHS Foundation Trust
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	

Ref	Quality Requirement	Met?	Comment
2	Written information for the patient's primary health care team should be available covering at least: a All aspects of QR1 b The need for regular prescriptions, penicillin and analgesia (SC)	Y	*And the first clinic letter was very full and gave comprehensive information for primary care colleagues.*
3	Information should be available on transition to adult care. This information should cover all aspects of the transition (QR43).	Y	*Plentiful information was seen although the extent to which it was used in practice was not clear.*
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	*Not yet, although planned, for families of children with sickle cell disease. UK Thalassaemia Society record was in use for those with thalassaemia.*
5	The locally agreed patient-held record (QR4) should be in regular use within the LHT / SHT.	N	*As for 4.*
6	Services should be provided in a child friendly environment, including toys and books / magazines for children and young people of all ages.	Y	
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	*Although some further specialist training was needed, and planned to be undertaken as soon as possible.*
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	*Psychology service support was not yet optimal but would soon be in place as a psychologist had been appointed.*
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	
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.	Y	*A protocol outlining community service input was drafted, and needed to be agreed and finalised.*
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	*And training and competency assessment materials were outstanding.*
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	Y	
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	

Ref	Quality Requirement	Met?	Comment
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	*These guidelines were full and very good.*
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	*And the proformas were clearly in use in the medical records seen.*

Ref	Quality Requirement	Met?	Comment
26	Guidelines should be in use for referral of patients and their families to a clinical psychologist with experience in the care of patients with haemoglobinopathies (including the option for self-referral)	Y	
27	Guidelines for referral for consideration of bone marrow transplantation should be in use.	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.	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	*But there appeared to be two guidelines, and it was not clear whether one was for children with thalassaemia and one for other children. Amalgamation to a single guideline or clarity as to which guideline applies to which children would be useful.*
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)	N	*No protocol was seen. Communication appeared to be good in general, although as the network arrangement with central or outreach review was not yet established, [d] could not yet be assessed. No PHR for sickle cell yet in use.*
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 protocol was seen. Clinic letters copied to patients provide some family held record. Full network functioning was yet to start.*
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	Y	*Although no formal patient held record, copy clinic letters to parents appeared to be consistent, and fulfilled the same end.*
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	*As for 37*
40	An operational policy should be in use covering: a Teaching children, young people and their patients how to set up an administer subcutaneous desferrioxamine infusions b Encouraging children to participate in setting up and administering their own infusion c Regular assessment and updating administration techniques d Recording of assessments of administration techniques	Y	*But was dated 2004; this should be checked and updated or re-dated, if no changes needed.*
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	*Although out of hours phlebotomy and transfusions were now offered, no extended clinic hours.*
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	*No written protocol was seen but evidence of process in practice was understood from discussion with the team, and from medical records.*

Ref	Quality Requirement	Met?	Comment
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	*Materials were plentiful but the young people who met the team [aged 14 and 17 years] had not had the opportunity to use them.*
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	*Some user feedback had been sought by the community team, but the issues raised had not been shared with the hospital team.*
45	The SHT should run a programme of training and awareness of the management of patients with haemoglobinopathy for its main referring LHTs.	N	
47	The SHT should meet at least annually with its referring LHT teams to: a Identify any changes needed to network-wide policies, procedures and guidelines b Review results of audits undertaken c Review any critical incidents including those involving liaison between teams d Consider the content of future training and awareness programmes (QR45)	Y	
49	The SHT should meet at least annually with representatives of the neonatal screening programme to review progress, identify issues of mutual concern and agree action.	Y	

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	*Audit data were not available for rates of immunisation against pneumococcus, proportion of children having had annual review assessments. TCD audit was in progress.* *Audit data for children with thalassaemia was full, since 2008.*
51	Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry.	Y	*This was now in progress.*

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	*Local specialty commissioners were engaged to an unusual degree in planning and monitoring services and establishing network activities.*

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.	Y	

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	*Audit data to Dec 09 gave 100% compliance*
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]* *As for P3*
P5	Timely confirmation of diagnosis for infants with a positive screening result**	Y	
S1i	Failsafe to ensure ongoing care	Y	
S1ii	Up-to-date registers maintained of babies (cases) for which units are responsible	N	*Although a database was in use, the last baby entered onto it had date of birth August 09, so evidence was against it being systematically kept up to date.*

** 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.