UK Forum on
Haemoglobin
Disorders

WM QRS West Midlands
Quality Review Service

Services for Children with Sickle Cell Disease or Thalassaemia
at
Imperial College Healthcare NHS Trust

Quality Review Visit Report
Visit date: March 12th 2010

Report finalised: December 15th 2010

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INTRODUCTION

This report presents the findings of the peer review visit to services for patients with sickle cell disease or thalassaemia at Imperial College Healthcare NHS Trust which took place on March 12th 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 Imperial College Healthcare 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 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 IMPERIAL COLLEGE HEALTHCARE NHS TRUST

Service (as at 2010)	Patient numbers sickle cell disease	Patient numbers thalassaemia	Other haemoglobin disorders
Imperial College Healthcare NHS Trust	173 [131 SS, 32 SC, 11 Sβ thal] [18]	116 *, including 61 post BMT [25]	15
Luton and Dunstable	43 [<5]	13	
Milton Keynes	61		
Wycombe	11	7 [7]	
Ealing	120 [5]	8 [8]	

Numbers on long term transfusions are shown in brackets.

* includes children with Hb H disease.

BACKGROUND

Imperial College Healthcare NHS Trust [ICHT] is a very large organisation, encompassing St Mary's Hospital, Hammersmith Hospital and Queen Charlotte's and Chelsea Hospitals together with some smaller specialty hospitals; with Imperial College Medical School and its academic and research campus, it forms an overall Academic Health Science Centre. Haemoglobin disorders are managed within the Paediatric Haematology and Stem Cell Transplantation programme, managed by a Clinical Lead consultant plus three other consultants, one the Professor of Paediatric Haematology. A fifth consultant is shortly to be appointed who will have particular responsibility for developing adolescent services, and network arrangements, for children and young people with red cell disorders. There is a senior haemoglobinopathy CNS. A second CNS post (0.7 wte for haemoglobinopathy and 0.3 wte for haemophilia), is currently advertised. There is a strong team of additional nurses and allied health professionals. There are four paediatric haematology doctors in training at SpR level and two at SHO level. A 'data officer' has recently been appointed.

Network arrangements with ICHT are well established for hospitals in the Thames Valley, including Luton and Dunstable, Milton Keynes, Wycombe and Watford. Consultants from the first three attended and met the review team to discuss the arrangements in place for them [see below]. For more local DGH's, Hillingdon and West Middlesex Hospitals, the arrangements are less clear cut. Central Middlesex / Northwick Park Hospitals [North West London Hospitals Trust] operate separately to a large extent and view themselves as a separate specialist service. Ealing Hospital, however, has close working arrangements with ICHT and the consultant paediatrician from Ealing also met the review team [see below].

There is a very active allogeneic bone marrow transplant [BMT] service, undertaking more transplants for red cell disorders than for malignant conditions. This service is highly rated by its referring units and much evidence relating to its activities, audits, and excellent outcomes was offered to the review team. BMT falls outside the remit of this review, however, and so further details are not included here.

ACCIDENT AND EMERGENCY

Children and families requiring non-elective assessment and care are invited to access the short stay ward and some do come through this route, but others come to A&E. Out of hours, access is through A&E, where first analgesia for severe sickle cell pain may be oral or parenteral morphine. Intra-nasal morphine is in use for some children attending A&E, but not for sickle cell children at the time of the

visit. Although the review team did not see this at the visit, the host team in later correspondence assured us that a database, 'Symphony', is in use in the Department which records the analgesia given and time administered. There is a facility for staff to check what medication has been used and found helpful on previous attendances. An audit of time to first analgesia was not available at the time of the visit.

Day Case and In-patient Facilities

Children with haemoglobin disorders may be managed as in-patients on a specialist ward [Grand Union] shared with other haematology patients and Infectious Disease patients. There are four positive pressure rooms and two negative pressure rooms. The BMT children are also managed on this ward. Within the ward area there is a day care area with some sofa seating and some beds. PICU is immediately adjacent to Grand Union ward. Sometimes, less sick children are cared for on the general paediatric ward [Great Western]. There is a teaching room, offering a lively and imaginative programme. Apart from A&E, which is some floors down but accessible by lift, all facilities are closely adjacent and conveniently accessed. All are of a high standard and child-friendly.

Meeting with Medical and Nursing Staff from Network Hospitals

Consultant medical staff from Luton and Dunstable, Wycombe and Ealing Hospitals, and the nurse specialist as well as consultant paediatrician from Milton Keynes, met the review team as part of this visit. Each was able to give patient numbers which are included in the Table above.

Until quite a short time before the visit, the working relationship with ICHT and these hospital teams had been informal, with the local hospital teams having ready access to the specialist team at ICHT for discussion, clinic review and transfer as needed, but without systematic inclusion of all managed children. Mostly only the sickest from each local hospital, or those with specific needs [for example, for central venous access], had therefore been seen at ICHT at the time of the visit. All the linked hospital teams had referred children to ICHT for BMT, and / or PICU and been very happy with the service and ongoing communication with the centre.

Luton, which previously linked more with the Whittington Hospital / University College London Hospitals, has now more formally linked with ICHT, and uses clinical guidelines based on those from ICHT. An outreach clinic is now established at Luton, the ICHT Clinical Lead visiting there three monthly so that all children can have formal annual review. There are plans to purchase a portable

TCD machine so that children can have stroke risk assessment undertaken as part of the outreach review.

Milton Keynes and Wycombe teams similarly discuss with, or refer in to, ICHT any children who are causing clinical concern and the teams use ICHT-based guidelines for care. Technically these hospitals fall into the Oxford SHA but both teams strongly feel that the range of services, and the specialist nature of the experienced ICHT team, make this a more appropriate referral centre for their children than Oxford. ICHT has therefore increasingly become their clinical hub. Three monthly ICHT team outreach clinics for annual review, including TCD, are planned for Milton Keynes. Wycombe families generally travel to ICHT in for their reviews.

Transition was highlighted as being a problem, especially at Milton Keynes, where there are a large number of adults, possibly as many as 250. Reviewers were told that there is no adult haematologist there who has an identified interest in red cell disorders. The paediatric team continue to manage the 'children' up to the age of approximately 19, but are concerned that robust arrangements for transition to adult services are not currently in place.

Ealing Hospital has a large paediatric red cell clinic population, managed together with the adult haematologist. ICHT-based guidelines are used and specific children with clinical concerns are referred to ICHT. All 'high-risk' children are already seen at the ICHT. There is now a shared social worker, and Ealing children and families attend the ICHT support group. An outreach clinic to cover all children for annual review and TCD is planned. Transition to the ICHT team for adult care [previously Hammersmith Hospital but now split across that site and the St Mary's site] has been established for many years.

All the linked hospitals from which the team met representatives were very satisfied with the willing and timely support from the ICHT team. They were enthusiastic about formalising these clinical links and about the establishment of outreach services. This should ensure that recommended care is delivered to all affected children.

Screening pathways were reported to be robust. The community nursing lead could name the individuals responsible for informing families of affected newborn and ensuring the babies are seen at clinic and started on penicillin by the recommended time. There were some discrepancies between the accounts of the community and hospital teams about the proportion of babies who did not reach the paediatric clinic for their first visit by the recommended age of three months, and the reasons for this.

Review Visit Findings

Overall the visiting team considered the quality of the services provided at ICHT for children with red cell disorders and families was very high.

Achievements

1 The Paediatric Haematology team is clearly very strong, clinically and academically. There is good clinical leadership from the Professor and Clinical Lead and all members of the team are committed and enthusiastic about the services they provide. There is evidence of excellent multi-disciplinary team working, with a holistic approach to the care of children with red cell disorders. User feedback confirmed the very positive way in which the services are viewed and appreciated.

2 There are rapidly growing and strengthening links with other hospitals in the network, particularly those in the Thames Valley area. The clinical teams there greatly appreciate the ready support from, and excellent communication with, the specialist team at ICHT.

3 Two newly approved posts are expected to allow continued improvement in service delivery and recording. The planned fifth consultant will focus on developing facilities and services for older children and adolescents and also will enhance the formal network links with other hospitals. The recent appointment of a 'data officer' should allow more comprehensive recording and use of patient level data.

4 There is a well developed 'sleep study' service with excellent facilities for investigation of children with sickle cell disease.

Immediate Risks

No immediate risks were noted.

Concerns

1. Many of the recommended audits of performance against key clinical standards have not yet taken place.

2. Figures presented to reviewers by the community nurse representative from Brent Sickle Cell and Thalassaemia Centre suggested that only 50% of babies identified through the newborn screening programme were seen in Paediatric Haematology Clinic by three months of age,

compared with a minimum standard of 90%. Figures supplied later by ICHT for all new babies seen during 2009 showed that 73% of babies were seen within the target three months. According to the data supplied by ICHT there were delays between the date on the referral letter and the date the letter was received by the hospital. Both teams need to clarify the position and the reasons for any delay. The fact that the community team and the acute Trust have different data raises questions about the effectiveness of communication between them.

FURTHER CONSIDERATION

1. Some attention should be given to document control as many of the documents were undated or had no review date.

2. The adequacy of junior doctor induction and arrangements for accessing guidelines may benefit from review. A junior A&E doctor asked to call up the clinical guidelines for managing children presenting with acute sickle cell pain accessed an internet based guideline from Chelsea and Westminster rather than the correct, intranet-based guideline.

3. Transition patient information and guidance is not yet available. It is anticipated that this will be in place by the time the new adolescent clinic opens in April 2010, with input from the fifth consultant when appointed.

4. At the time of the visit no patients were entered onto the National Haemoglobinopathy Registry. It is expected that the new data manager post will help in this respect.

5. As in other areas of London there was uncertainty about commissioning arrangements for services for children with red cell disorders. The commissioners who met the review team expressed interest in having activity and outcome information reported to them, and further dialogue about these services.

6. The working relationship with some of the more local hospitals within the network, for example Central Middlesex and Northwick Park, was unclear, Clinical links were better established with the more distant hospitals in the Thames Valley area. The paediatric haematology team at Central Middlesex Hospital team have requested a separate review visit, and insights into the services across of North West London might be gained from this.

GOOD PRACTICE

1 Clinical guidelines overall are comprehensive and excellent.

2 There is a very good provision and imaginative use of play therapists across the range of services for children with red cell disorders.

3 There are plans to move into a larger day care area, but the current area was noted to be relaxed, comfortable and user-friendly.

4 There is strong advice and support to the outlying hospitals with which there are established network links

5 The visiting team was impressed by the 'sticker' alert used on maternity notes to notify midwives if a baby is at risk of having a major haemoglobin disorder, in order to prompt newborn testing.

6 The multi-disciplinary team overall is clearly committed to giving excellent services; the pharmacy and dietetic input was felt to be particularly strong

7 The team's involvement with, and promotion of, the user support group and use of feedback from users was evident.

APPENDIX 1: MEMBERSHIP OF THE REVIEW TEAM

Dr Sarah Lawson	Consultant Paediatric Haematologist	Birmingham Children's Hospital NHS Foundation Trust
Neil Westerdale	Advanced Nurse Practitioner Haemoglobinopathies	Guys & St. Thomas' NHS Foundation Trust
Linda Brooks	Haematology Nurse Specialist Paediatrics	Bradford Teaching Hospitals NHS Foundation Trust
Marie Claire Ngeumshe	User Representative	
Dr Anne Yardumian	Clinical Lead for Peer Review Programme	
Sarah Broomhead	Quality Manager	West Midlands Quality Review Service

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	*The patient information is good, lacking only age-appropriate information on avoiding smoking and excess alcohol consumption (3d). For (3e) general information was available but there was no specific consideration of children with red cell disorders.*

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	*Information to GP's does not appear to cover all the aspects detailed in QR1, but clinic letters do highlight need for regular prescriptions as in b.*
3	Information should be available on transition to adult care. This information should cover all aspects of the transition (QR43).	N	*There is some material already available [a letter introducing the young person to the transition clinic, and a 'handover' proforma] and work is in progress to development age–appropriate, condition-specific information.*
4	The SHT and its linked LHTs should have agreed a patient-held record for recording at least: a Information about the patient's condition b Current management plan c Regular medication d Named contact for queries and advice e Alternative contact for times when key contact is away.	Y	*The parents of younger children use the card insert designed for inclusion in the 'red book'. It is not clear if the large booklet is in regular use. The UK Thalassaemia hand held record was included in evidence, but the review team was unable to check the extent to which it was in routine use.*
5	The locally agreed patient-held record (QR4) should be in regular use within the LHT / SHT.	Y	*In place for children with sickle cell, but not so clear for those with thalassaemia*
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	*QR's 9 – 14 are all fully met with highly qualified, experienced and appropriate staff members.*
10	The lead consultant (QR9) should have a named deputy who will provide cover for absences. The named deputy in the SHT should undertake CME activity of relevance to care of children and young people with haemoglobinopathies.	Y	
12	The SHT should have a lead haemoglobinopathy nurse with responsibility, with the lead consultant (QR9), for guidelines, protocols, training, audit relating to haemoglobinopathies and liaison with referring LHTs. The lead nurse should have specific training in the care of patients with haemoglobinopathies.	Y	
13	The SHT should have a nurse specialist or counsellor who provides outreach support for patients in the community. This nurse specialist / counsellor should have specific training in the care of patients with haemoglobinopathies.	Y	
14	There should be agreed cover arrangements for the outreach nurse specialist / counsellor.	Y	

Ref	Quality Requirement	Met?	Comment
15	Access to the following staff and services should be available: a MRI and CT scanning b Transcranial Doppler ultrasonography (SC) c Hospital dental services d Genetics services e Bone marrow transplantation services f Contraception and sexual health services g Consultant cardiologist h Consultant endocrinologist i Consultant hepatologist j Consultant neurologist k Consultant ophthalmologist l Consultant orthopaedic surgeon m Consultant obstetrician n Child and adolescent mental health services	Y	*All specialists and services are available on the St Mary's Hospital site with the exception of T2* MRI. This is due to start on site within weeks of the visit.*
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	*There are close spatial and functional links between haematology and PICU. It might be helpful to include bleep numbers /contact details in protocols for emergency transfer of children*
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.	N	*The review team did not see a Service Level Agreement with the community service team.*
19	A nurse with competency in cannulation, starting and supervising a transfusion should be available at all times at which children are attending for transfusion.	Y	
20	All staff involved in the care of children and young people with haemoglobinopathies should undertake regular child protection training.	Y	*List of trained staff with level / dates of training was not made available at the visit, although records have now been received.*
CLINICAL and REFERRAL GUIDELINES			
21	Guidelines should be in use covering a How to establish and confirm diagnosis b Parent and sibling testing	Y	*Although it did not specifically mention sibling testing for thalassaemia*

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	*Overall these are excellent.* *The section relating to eye problems outlines usual referral for screening but not specifically the management of acute change in vision [i].* *The thalassaemia guidelines had no review date.*

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	*These are mostly good and comprehensive, but the omission of 'h' [listed under' l' in the self-assessment return] that is, 'demonstrating to parents / carers how to check for splenic enlargement' was felt to be important. It is covered in the new patient section of the guidelines, but should also be included here.*

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 team did not see these at the visit, but have subsequently seen full documentation which was in use at the time of the visit.*
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	

Ref	Quality Requirement	Met?	Comment
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	*Although the team could not find this information among the evidence at the visit, full guidelines covering the content of this QR, which were in place at the time of the visit, were subsequently seen.*
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	*A protocol for automated exchange needs to be developed.*
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	*But the need for document control – having a single dated version of guidelines in each clinical area – is noted; see also' further consideration' section of main report.*

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	*Material submitted after the visit, which the visiting team had not found, but which was in use at the time of the visit, confirmed compliance.*
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	*Not yet in place although it is anticipated that the fifth consultant post, shortly to be appointed to, will focus on developing these aspects of the network.*
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 protocol was available there was evidence of use in practice.*
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	*There was no agreed policy but in practice this is becoming established.*
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	*Although no formal policy in place, this is covered in practice by a checklist used by the CNS.*
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	*Offered for transfusion, and soon to start for adolescent clinics, not for blood testing.*
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	

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.	N	*There is a 'transition data' chart in use, but no written protocol covering all these requirements. The appointment of fifth consultant will progress this.*
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	*Evidence of plentiful activity and use of feedback.*
45	The SHT should run a programme of training and awareness of the management of patients with haemoglobinopathy for its main referring LHTs.	Y	*And local teams confirmed involvement.*
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	*With many of the linked hospitals and plans to put in place for others. [although audits were not systematically being undertaken – see QR 50 and main report].*
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 after the visit confirmed this takes 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	*There is an audit of immunised patients, but other aspects appear not to have been subjected to audit.* *Thalassaemia children are included in a transfusion audit, but other aspects not met.*
51	Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry.	N	*No children yet registered on the National Haemoglobinopathy Registry but this is shortly to be addressed by the newly appointed data manager.*

Commissioners of Services

Ref	Quality requirement	Met?	Comment
52	Each Specialist Commissioning Group 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	*No submission was received from commissioners and it was not possible to clarify arrangements with them in discussion during the visit.*
53	Each Specialist Commissioning Group should have: a Compared the staffing, support services and facilities of each SHT and LHT located within its area (QR52) with the levels expected in QRs 7 to19. b Agreed a plan for the development of SHTs and LHTs located within its area. c Monitored achievement of the agreed plan at least annually. The agreed development plan should ensure that QRs 7 to 19 are met within 2 to 5 years.	N	

Additional Requirement – Screening Services

Ref	Quality requirement	Met?	Comment
P3	Timely communication of positive screening results (sickle cell disorder) – including a review of parental results	N	*Information presented by community services representative indicated only 43% results to parents by 4 weeks.*
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	*[first part of this standard ignored, duplicates P3 with different target] Regarding the time to referral to Paediatric clinic, the community nurse lead indicated at the visit that 100% of babies were referred by 8 weeks. The hospital team disagreed, and subsequently provided data for all newborns seen in 2009 which indicated that there can be a significant delay between referral letter being typed and received at the hospital. 73% babies were seen by three months compared with a minimum standard of 90%.*
P5	Timely confirmation of diagnosis for infants with a positive screening result**	N	*It was not possible to assess this. No data were presented.*
S1i	Failsafe to ensure ongoing care	Y	*The community nurse talked through the process in detail and the reviewers were assured it was in place.*
S1ii	Up-to-date registers maintained of babies (cases) for which units are responsible	Y	*A list of all children seen at ICHT was presented. There is currently no complete list of all the children managed in hospitals across the whole network.*

** 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.