UK Forum on
Haemoglobin
Disorders

WM QRS West Midlands
Quality Review Service

Services for Children with Sickle Cell Disease or Thalassaemia
at
North West London Hospitals NHS Trust

Quality Review Visit Report
Visit date: April 30th 2010

Report finalised: December 20th 2010

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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 North West London Hospitals NHS Trust, Central Middlesex Hospital [CMH] site which took place on April 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.

Although this is a split site Trust, with acute and planned paediatric services taking place on both Northwick Park Hospital [NPH] site and CMH site, the great majority of the activity relating to this service is at present focussed on the CMH site, and most of the comments relate to the documentation in place, and the facilities, there. Where there is relevant activity at NPH, for example planned transfusions for a small number of children with thalassaemia, some information given verbally to the review team at the visit is also included in the body of the report.

ACKNOWLEDGEMENTS

I would like to thank the staff of North West London Hospitals 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 NORTH WEST LONDON HOSPITALS NHS TRUST

Service (as at April 2010)	Patient numbers sickle cell disease	Sickle patients on long term red cell transfusions	Patient numbers thalassaemia
Central Middlesex Hospital site	221	6	<5
Northwick Park Hospital site			<5

The sickle cell and thalassaemia services at North West London NHS Hospitals Trust are long established, particularly at the CMH site – where the great majority of the activity still takes place. The paediatric services are highly integrated with the adult service, and both are well supported by community services from the Brent Sickle Cell and Thalassaemia Centre and the skills of a full time clinical psychologist. There is currently no acute service clinical nurse specialist but a post in the Brent Centre recently vacated by a retirement is being re-shaped to undertake some of the acute clinical nurse specialist role.

Accident and Emergency

There is a small but busy paediatric A&E, at which children with sickle cell disease are triaged as 'priority'. They are then assessed in and out of hours by a team of nurses who also work on the in-patient and day care area. Most of the regularly attending children are well known to this nursing team. First analgesia may be oral or parenteral morphine as needed, taking account of the medication already given and the pain score. PCA's are used subsequently if preferred. Times to first analgesia are audited and reported to be within the target 30 minutes and usually less. Children with sickle cell disease seldom attend the Northwick Park A&E. If they do they are managed according to the same guidelines and if they need admission are moved over to the Central Middlesex in-patient ward.

Outpatient Clinics

Out-patient rooms are adjacent to the other clinical areas. There is a weekly red cell clinic, run by the service's lead paediatrician and lead haematologist, a doctor in training, together with the psychologist and a community CNS. During the course of the clinic, children and families may choose to see a number of these specialists separately to discuss different issues, during the course of the clinic. The last clinic appointment is 4.30 pm, and although the clinic frequently 'over-runs', there is no facility for after-hours appointments. Blood samples are taken within the clinic area. Regularly transfused patients are reviewed in clinic approximately 3 monthly. At the end of the clinic, the professionals meet to discuss the children and families they have seen, to share any concerns and agree management plans. Trans-cranial Doppler scans for stroke risk assessment are offered on Saturday mornings in the Radiology Department.

Day Care

There is a 4 station assessment / observation bay opposite the nurses' station in the in-patient area, which is immediately adjacent to the children's A&E. This is spacious with 4 hospital beds and chairs. The staff did not report any difficulty in accommodating booked children for treatment here. The unit is open 9am to 5pm Monday to Friday. Evening transfusion is also offered and, at the Northwick Park site, weekend transfusions are available for the thalassaemia patients who attend there. Pre-assessment of children for transfusion, and cannulation, is undertaken by paediatric doctors. The clinical areas are all close together and so doctors are easily available and no delays in starting transfusion were reported.

In-Patient Facilities

There are 6 in-patient beds, used for all paediatric admissions. There is good play support. Hospital teaching was not felt to be necessary as length of stay is short. There is a pleasant play room for younger children, and a specific relaxation area for older children and teenagers. Most transfer to adult services from 16 but, if transition has not taken place, it is possible for them to be admitted to the paediatric ward up to the age of 18.

Community Services

Community services are provided mainly by a team of 5 nurses based at the Brent Sickle Cell and Thalassaemia Centre [BSCTC], located in the hospital site at Central Middlesex and by two other community specialist nurses, one employed by NHS Ealing and Harrow and the other by Hounslow. The duties of the Brent team include managing the newborn screening programme results across the 14 PCT's of the old North West London Region, and providing continuing support for children in Brent, Harrow, Kensington & Chelsea, Hillingdon, East, North and West Hertfordshire. They also provide clinical supervision and support for the carrier counselling health visitors and clinical supervision for the community specialist nurses in the sector. A specialist midwife is responsible for antenatal counselling and specialist maternity services.

The professional services manager at the BSCTC is the named lead haemoglobinopathy nurse for the sector. All the specialist nurses and counselling health visitors undertaking carrier counselling have a quarterly meeting to discuss clinical practice, develop shared protocols and guidelines, audits and educational issues. The meetings also include sickle cell and thalassaemia specialist nurses from

Hammersmith & Fulham and Hounslow, and those from St Mary's, Hammersmith and Queen Charlotte Hospitals.

There are monthly support group meetings running in Willesden and in Ealing.

LINKED HOSPITALS

The hospitals with which this team links are to some extent historical. There are some out of sector links to – Bedford, Barnet, Welwyn Garden City and Ipswich. In-sector linked hospitals include West Middlesex. Some of these hospitals also share care of children with other specialist teams. There are other local hospitals, for example Ealing, which do not link with CMH. There is very little joint working between CMH and the specialist team at St Mary's Hospital [Imperial].

No local hospital team members attended the review visit but during brief telephone conversations with paediatric consultants at West Middlesex and Bedford Hospitals it was understood that they continued to manage their affected children predominantly locally, that they referred children systematically for TCD screening, and otherwise referred children about whom there were specific concerns. For other children annual reviews are being undertaken by the local teams without input from the specialist team. Central Middlesex protocols are in use in Bedford. At West Middlesex there is a briefer local set of protocols but the recently updated CMH guidelines are being considered for use there also. Neither local paediatrician had attended educational or management meetings with the specialist team although the Bedford consultant indicated that he had been invited to join meetings held at CMH. Both reported that if they needed advice about an individual patient, the CMH lead paediatrician and haematologist were readily available and always very helpful.

COMMISSIONING ARRANGEMENTS

Commissioners from Brent and Harrow PCT's came to meet a member of the review team. The services are commissioned as part of acute services for maternity and children. Commissioners knew about the services in general terms. They felt they had a good reputation, and they had no concerns about them. They felt they had become more involved with providers in the period leading up to this visit. The role of the 'managed network' for services was not clear to the review team, apart from hosting meetings to discuss protocols and guidelines. The way in which different hospitals in the sector link with the teams at CMH / St Mary's Imperial appears to be largely ad hoc, and some link with both.

PLANNED RECONFIGURATION OF SERVICES

At the time of the visit consultation was in progress about some significant changes to the configuration of paediatric services between NPH and CMH. Although this has no impact on services at the time of the review, the potential changes were highlighted before and during the visit, as they will impact on the sickle cell and thalassaemia services if agreed and implemented. Described in a document 'Better services for local children – a public consultation for Brent and Harrow', the main changes proposed are that, as from Autumn 2010, there will be no overnight in-patient paediatric care at the CMH site, so that any children needing admission will be transferred to NPH in-patient facilities. It is estimated that only approximately 150 transfers per year will be needed, although the red cell team indicated that one or two children with sickle cell disease needed admission each week, which suggests that they constitute between one and two thirds of those needing transfer for in-patient care. 24 hour, 7 day a week A&E facilities would remain at CMH and a 12 hour per day consultant-led paediatric assessment service would also remain. If this change goes ahead, it is likely that some of the planned care for children with red cell disorders, such as booked transfusions, would transfer to CMH from NPH.

Some of the CMH staff and user families expressed concern about the possible impact of this change on their services. While in practice the great majority of the care needed would still be offered at CMH, if families know that admission is only possible at NPH they might preferentially take their children there for assessment if acutely unwell, or may choose instead [and this was felt to be more probable by some] to access St Mary's Hospital; for those living in the south of the catchment area, this is easy to reach and offers a comprehensive specialist service.

REVIEW VISIT FINDINGS

ACHIEVEMENTS

1 This well established service, now with the benefit of some relatively new, committed and enthusiastic team members, continues to offer very good clinical care. The close working between paediatric and adult service staff, and between hospital and community teams is noteworthy. All staff are working essentially as part of an integrated service. This integration of teams from the start of the child's clinical journey means that the process of transition in mid-to-late teens is much less challenging than is often the case.

2 The immediate physical co-location of children's A&E, day care and in-patient facilities, and the integrated nursing team which staffs all three, works really well for patients, who greatly value the familiarity of experienced clinical staff when they attend, and the prompt and expert care they receive.

3 Clinical psychology input into the service is unusually strong, and is offered systematically and regularly to all children and families from the start instead of just 'as needed' / as referred by the medical or nursing teams. While being in line with the recommendations of the national care standards, this is quite unusual in practice.

4 The clarity and quality of many of the written protocols and guidelines were judged by the review team to be outstanding. They were available in full on the intranet and in hard copy in the clinical areas.

IMMEDIATE RISKS

No immediate risks were identified.

CONCERNS

1 The extent to which the links with the named hospitals referring into this specialist service was actually functioning as a systematic network was not clear. The current arrangements appear to work more as an 'ad hoc' clinical support service for the outlying hospitals for particular cases. This is highly valued by the local teams but relies on those teams to identify and refer any children with problems and some may be going unidentified. Work towards

sharing protocols is in hand, and the availability of TCD screening for children from any referring unit is excellent. Annual reviews for most of the children in outlying units are undertaken, if at all, by the local team without specialist input which is not in-keeping with the guidance in the national care standards. This is of concern in smaller units where staff have less experience in caring for children with red cell disorders.

2 Data about the children using the service is entered onto the EHR [European Haemoglobinopathy Register] but this is not readily available to the clinical team, who were unable at the visit even to access it for demonstration. Making use of entered data for routine clinical audits, or to inform clinical practice in any way, must therefore be difficult. Further, the EHR now captures data only for patients attending CMH and a couple of other sector hospitals, so is functioning as a local database but not one which is currently available or useful for the clinical staff running the service.

FURTHER CONSIDERATION

1 Links with the local haemoglobinopathy teams, in terms of training and network development, are evolving but need some more work, as above. Links with the specialist team at St Mary's Hospital, Imperial might also usefully be re-considered, in terms of protocol sharing [in both directions], audit, teaching and training and for joint consideration of difficult clinical issues. The roles of the CMH and St Mary's teams across the NWL network might also usefully be discussed.

2 It may be useful and interesting to undertake some user feedback survey for this particular patient group. General mechanisms for receiving and using patient opinion were described, but none specifically for this service.

3 The families who met members of the review team reported that they would have valued more continued input and support from the community team in the early weeks following the diagnosis of their child, and continued nurse input to schools as the children grow through the system would also be much appreciated.

4 The sickle cell patient held record is very new, and so it was not possible to see how useful / how much used it might prove in practice. In the meantime, ensuring that the family have up to date information about their child relies on their receiving copies of clinics letters. This appears to be inconsistent in practice. Study of the medical records made available to the team showed several instances where this had apparently not happened.

5 With a large practice such as this, encompassing many older children, consideration might be given to providing out of hours clinic facilities to minimise time lost from school. Medical staff are willing to undertake this work but some additional support staff would be required.

6 Access to urgent paediatric urology care, for management of fulminant priapism, can be a problem with many delays being reported. Also, access to a specialist paediatric neurologist to guide management decisions for children with stroke is currently not in place. These are London wide issues and links across a range of providers will need to be put in place. The programme clinical lead will pull together such issues as the London visits continue, and try to progress them subsequently.

7 The beginnings of direct engagement between providers of this service and commissioners, around this visit and around the possible service reconfiguration, is an opportunity which can hopefully be built on and lead to further constructive dialogue.

GOOD PRACTICE

1 The multi-professional meeting at the end of each clinic allows for exchange of important information and leads to appropriate ongoing management decisions.

2 The overall facilities are good, with excellent co-location, and the focus on the adolescent users, even in a small ward area, is laudable.

3 The patient information available was felt to be very comprehensive, and the fact that much of it is available through the Brent Sickle Cell and Thalassaemia Centre website is really useful.

4 The use of a 'trolley' containing necessary documentation relating to the service, and patient information material, for use across the clinical areas makes access for all staff easy and visible. The laminated 'contact sheet' gives specific phone numbers / addresses for all the key support teams and saves searching through lengthy documents for this information.

5 Some of the individual service documents were unusual and innovative and deserve special mention, including standard letter templates, an 'out of area' transfer sheet, 'specialist nurses paediatric contact' check list with actions and discussions detailed by age of patient.

6 The transitional material in general is strong with flexibility around age at which transition actually occurs. A good letter for 16 to 18 year olds to carry when they had not yet transferred to adult care indicates that they should be admitted to a paediatric bed.

7 A checklist of routine monitoring tests to be taken on regularly transfused children, by month of year, is a simple and robust system to ensure all tests are taken as recommended. This is laminated and displayed at the nurses' station outside the day care area.

8 The availability of TCD scans on a Saturday morning minimises unnecessary school time lost for this routine test.

APPENDIX 1: MEMBERSHIP OF THE REVIEW TEAM

Dr Marie Donohue	Consultant Haematologist,	Nottingham University Hospitals, Nottingham.
Neill Westerdale	Advanced Nurse Practitioner Haemoglobinopathies	Guy's & St. Thomas' NHS Foundation Trust
Jo Sutcliffe	Service Manager	Kings College Hospital NHS Foundation Trust
Ravinder Raj	User Representative	
Oliver Mmounda	User Representative	
Dr Anne Yardumian	Clinical Lead for Peer Review Programme	

APPENDIX 2: COMPLIANCE WITH QUALITY REQUIREMENTS

Specialist Haemoglobinopathy Teams

Ref	Quality Requirement	Met?	Comment
INFORMATION AND SUPPORT FOR PATIENTS AND THEIR CARERS			
1	Written information should be offered to patients and their families covering at least: 1 A simple explanation and description of the condition, how it might affect the individual, possible complications and treatment. For sickle cell disease this information should include: a Problems, symptoms and signs for which emergency advice should be sought b How to manage pain at home, including how to avoid pain, and non-pharmacological interventions c Importance of adequate fluid intake d Use of antipyretics for fever e Importance of regular antibiotics and full immunisation f How to feel the spleen and its significance. For thalassaemia this information should include: a Problems, symptoms and signs for which emergency advice should be sought b The importance of maintaining good haemoglobin levels by transfusion c Potential problems of iron load and how it can be managed. 2 Details of the services available locally including: a Clinic times and how to change an appointment b Key contact name and number c Alternative contact if key contact away d Who to contact for advice out of hours e How to use emergency services f Ward usually admitted to and its visiting times g Community services and their contact numbers h Details of support groups available. 3 Health promotional material including a Inheritance and implications for other family members b The importance of a good diet and regular exercise c Implications for travel d Age appropriate information on avoiding smoking and excess alcohol consumption e Age appropriate information on contraception and sexual health f Where to go for further information, including useful websites and national voluntary organisations.	Y	

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	
3	Information should be available on transition to adult care. This information should cover all aspects of the transition (QR43).	Y	
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	
5	The locally agreed patient-held record (QR4) should be in regular use within the LHT / SHT.	N	*Although patient held record was presented, there was no evidence of its use in practice at the time of the visit. The user families who met the review team were unaware of it.*
6	Services should be provided in a child friendly environment, including toys and books / magazines for children and young people of all ages.	Y	*And the focus for older children / adolescents in a relatively small in-patient area was commendable.*
STAFFING and SUPPORT SERVICES			
9	The SHT should have a nominated lead paediatrician / paediatric haematologist consultant with an interest in the care of patients with haemoglobinopathies who should have responsibility for guidelines, protocols, training, audit relating to haemoglobinopathies and overall responsibility for liaison with referring LHTs. The lead consultant should undertake CME activity of relevance to care of children and young people with haemoglobinopathies.	Y	
10	The lead consultant (QR9) should have a named deputy who will provide cover for absences. The named deputy in the SHT should undertake CME activity of relevance to care of children and young people with haemoglobinopathies.	Y	
12	The SHT should have a lead haemoglobinopathy nurse with responsibility, with the lead consultant (QR9), for guidelines, protocols, training, audit relating to haemoglobinopathies and liaison with referring LHTs. The lead nurse should have specific training in the care of patients with haemoglobinopathies.	Y	
13	The SHT should have a nurse specialist or counsellor who provides outreach support for patients in the community. This nurse specialist / counsellor should have specific training in the care of patients with haemoglobinopathies.	Y	
14	There should be agreed cover arrangements for the outreach nurse specialist / counsellor.	Y	

Ref	Quality Requirement	Met?	Comment
15	Access to the following staff and services should be available: a MRI and CT scanning b Transcranial Doppler ultrasonography (SC) c Hospital dental services d Genetics services e Bone marrow transplantation services f Contraception and sexual health services g Consultant cardiologist h Consultant endocrinologist i Consultant hepatologist j Consultant neurologist k Consultant ophthalmologist l Consultant orthopaedic surgeon m Consultant obstetrician n Child and adolescent mental health services	Y	*As per 'contact list' laminated and attached to information trolley.*
16	The following services should be available a Paediatric high dependency care b Paediatric intensive care There should be agreed criteria for admission to each level of care.	Y	
17	The following support services should be available: a Interpreters b Social work c Play specialist d Hospital teacher (in-patient care only) e Child psychologist f Dietician	Y	*And psychology level of input is very high.*
18	A service agreement for support from community services should be in place. This service agreement should cover, at least: a Guidelines for involvement of community paediatric services in the care of patients with haemoglobinopathies b Role of community services c Exchange of information between hospital and community services and vice versa d Arrangements for liaison with schools.	N	*A service agreement was presented but it did not cover liaison with schools, or exchange of information between teams although, in practice, communication between services is good.*
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	*Although not nurse led, the ready availability of doctors undertaking these duties and working across a small clinical area meets this QR.*
20	All staff involved in the care of children and young people with haemoglobinopathies should undertake regular child protection training.	Y	*Training records by level and date were seen for core team members.*
CLINICAL and REFERRAL GUIDELINES			
21	Guidelines should be in use covering a How to establish and confirm diagnosis b Parent and sibling testing	Y	

Ref	Quality Requirement	Met?	Comment
22	Clinical guidelines should be in use covering: a Recommended immunisations b Immunisations, other prophylaxis and travel advice prior to travel abroad. c Penicillin prophylaxis while awaiting clarification of diagnosis (sickle cell disease only)	Y	*Lacking only a recommended alternative to penicillin prophylaxis for children who are allergic to it.*
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	*Urology urgent referral was delayed in practice.(See 'further considerations' section of main report).*

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	*And these were assessed to be very good guidelines.*

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	*Only management of post-splenectomy complications including thrombocytosis, was not included.*

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	*Although no specific referral guidelines were seen, the ready availability of the psychologist in clinics means that families can access him when they wish to.*
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	*These are very strong guidelines.*
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	*This was missing only guidance as to where the transfusions will take place, and specific limit on how many cannulation attempts an individual clinician should make before handing over to another.*
31	Clinical guidelines for chelation therapy and monitoring iron load should be in use including: a Indications for starting chelation b Choice of regime c Dosage and dosage adjustment d Clinical assessment of tissue damage e Monitoring of serum ferritin f Use of non-invasive estimation of organ-specific iron loading heart and liver by T2*/R2 g Management of side effects of chelators.	Y	
32	Clinical guidelines for the management of thalassaemia intermedia should be in use including: a Indications for transfusion b Monitoring iron loading c Indications for splenectomy.	Y	
33	Clinical guidelines should be in use covering: a Indications for exchange transfusion b Arrangements for carrying out an exchange transfusion.	Y	*Yes clear guidelines in place including for automated red cell exchange in young children.*
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	*Although part b] not met patient held record not in systematic use at the time of the visit.*
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	*Essential elements are covered in the general new outpatients guideline for both conditions.*
37	A protocol should be in use covering arrangements for care between SHT and LHT for ongoing care. This protocol should ensure that, to facilitate effective shared care: a All patients have an up to date patient held record and details of their care plan. b The LHT and the patient's GP have received details of the patient's care plan.	N	*But regular use of the patient held record now being introduced, and systematic copying of clinic letters to parents, should allow practice to meet this requirement in the near future*
38	A protocol should be in use covering: a Updating patient-held records b Offering patients a permanent record of consultations at which changes to their care plan are discussed. c Recording changes of key contact d Giving further information (QR1) as patients' and families' needs change	N	*No protocol seen, but in practice should soon be met, as for 37.*
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 in place. No specific agreements with LHT's.*
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	
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	*Nearly met as phlebotomy and transfusion can be accessed out of hours but no out of hour clinic facility in place at present.*

Ref	Quality Requirement	Met?	Comment
42	A protocol should be in use covering: a Follow up of children who do not attend b Communication and follow up of children who move to another area	Y	
43	A protocol should be in use covering transition to adult care. This should ensure: a Age guidelines for timing of the transfer. b Involvement of the young person in the decision about transfer. c Involvement of primary health care, social care and adult services in planning the transfer. d Allocation of a named coordinator for the transfer of care. e A preparation period and education programme relating to transfer to adult care. f Communication of clinical information to the adult services. g Arrangements for monitoring during the time immediately after transfer to adult care.	Y	*Very good material presented.*
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	*Support groups in place, and general mechanisms for receiving feedback from patients but none specific to this 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	*No educational or business meetings in place for other hospitals in network*
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	*Lead nurse described regular strategy and case based meetings.*

Ref	Quality requirement	Met?	Comment
DATA COLLECTION and AUDIT			
50	The LHT / SHT should have audited compliance with key standards including: **Patients with sickle cell disease:** a Proportion of patients taking regular penicillin b Proportion of patients fully immunised against pneumococcus c Proportion of patients (HbSS and HbSβ^0) who have had Transcranial Doppler ultrasonography undertaken within the last year d Proportion of patients who have had their annual multi-disciplinary review within the last year e Effectiveness of action to contact families who have not attended for follow up appointments. f Review of the care of any patients who have died. **Patients with thalassaemia:** a Proportion of patients on chelation therapy b Proportion of patients who have had their annual multi-disciplinary review within the last year c Adequacy of recording of: ▪ Pre-transfusion Hb levels ▪ Regular monitoring of iron level ▪ Complications of iron overload ▪ Height / weight progression ▪ Spleen size ▪ Support for home chelation programme (QR40) d Effectiveness of action to contact families who have not attended for follow up appointments. e Review of the care of any patients who have died.	N	*Audits reported for penicillin / antibiotic prophylaxis, and immunisations, but not for TCD coverage or annual reviews completed.* *Note: Figures for TCD coverage were collated and sent after the date of the visit.* *An audit covering a very small number of thalassaemia patients was presented, although more are being treated in the Trust [6 across the two sites]*
51	Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry.	N	*Data are entered onto the EHR, but this services a different function and clinical staff cannot access or make use of it easily.*

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	*Audit undertaken, 60% families seen 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	*Part a ignored [duplication of P3 but with different % required]* *Part b – 60 % referred by 8/52* *Part c – 40% seen by 13/52*
P5	Timely confirmation of diagnosis for infants with a positive screening result**	Y	

S1i	Failsafe to ensure ongoing care	Y	*Although a comment made by clinicians, that the only denominator available [for audit purposes eg TCD coverage] is the number of children who actually attend clinic, sheds some doubt on this.*
S1ii	Up-to-date registers maintained of babies (cases) for which units are responsible	Y	*Entered onto EHR, though as clinical team cannot easily access data, the review team questioned the practical usefulness of this.*

** 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.