UK Forum on Haemoglobin Disorders

WM QRS West Midlands Quality Review Service

Services for Children with Sickle Cell Disease or Thalassaemia At University College London Hospitals NHS Foundation Trust and Whittington Hospital NHS Trust

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Quality Review Visit Report
Visit date: March 31st 2010

Report finalised: August 14th 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 University College London Hospital NHS Foundation Trust and Whittington Hospital NHS Trust, which took place on March 31st 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 College London Hospital NHS Foundation Trust and Whittington Hospital NHS Trust for their hard work in preparing for the review and for their kindness and helpfulness during the course of the visit. Thanks too to the 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 COLLEGE LONDON HOSPITAL NHS FOUNDATION TRUST AND WHITTINGTON HOSPITAL NHS TRUST

Service (as at March 2010)	Patient Numbers Sickle cell disease	Patient Numbers thalassaemia	Patients on long term red cell transfusions
UCLH	34	19	12 thalassaemia
Whittington	125	25	17 [8 sickle cell, 9 thalassaemia]
Linked hospitals	not	yet	recorded.

INTRODUCTION

Over the last two years, the paediatric and haematology teams at University College London Hospital NHS Foundation Trust [UCLH] and Whittington Hospital NHS Trust [WH] have been working together

increasingly as one joint specialist Red Cell Disorder Unit, offering services at both sites, but with some shared senior medical staff, and shared protocols and management guidelines. The services are not jointly managed but team members indicated that the process of preparing jointly for the review visit had helped communication and understanding between managers of the services.

Some members of the review team [HA, EM, AY] first visited the UCLH site, and spent over an hour meeting staff, visiting outpatient, A&E, in-patient and day care facilities, and reviewing some case notes, before travelling to WH meeting the remaining members of the review team [JB, NW, PT] and the host team which included staff from both sites. The remainder of the day was spent at WH site, reviewing evidence and meeting teams from both sites.

The Royal Free Hospital NHS Trust [RFH] has also offered red cell services for this part of North Central Sector until recently, but no longer undertakes planned care for children with these conditions. The paediatrician who was previously responsible for those services on the RFH site now works for the majority of her sessions at the WH site. This is welcome support for the lead paediatrician.

Accident and Emergency

Children attending for urgent care at UCLH attend A&E, where they are 'fast-tracked' using a named emergency card system. In routine hours, there is a paediatric SHO and SpR for whom A&E calls are priority. There is also consultant paediatric presence in A&E until 9 pm on weekdays and 5 pm at weekends. A play specialist is present in A&E at times during the week. Protocols for the management of children with these conditions were being updated and awaiting 'sign off'. None were available electronically or in paper versions in the Department at the time of the visit.

At the Whittington site, families are asked to call the paediatric in-patient ward or clinical nurse specialist for advice if they think admission might be necessary. If they do come to the hospital they are asked to bypass A&E and go straight to the ward. If they present to A&E they are re-directed to the ward for care, unless seriously ill or needing resuscitation in which case they are stabilised in A&E first. The new paediatric guidelines for management of children presenting for urgent care were not on the intranet at the time of the visit, although they were accessible through a different paediatric drive. An older set of guidelines was available on the intranet which differed, in some aspects of recommended management, from current practice.

OUTPATIENT, DAY CASE AND IN-PATIENT FACILITIES

At the UCLH site, there is a weekly paediatric haematology clinic, in a new and spacious out-patient area. This is run jointly by the haematology and paediatric teams, and attended at intervals by additional specialists including endocrinologist, dentist, and a visiting paediatric haematologist undertaking trans-cranial Doppler assessments. Day care and in-patient care takes place on age-specific ward areas, divided into paediatric [under 12 years] and young persons [13-19]. These are on adjacent floors in the hospital's Tower Block and the facilities are of an extremely high standard, including a beautiful 'recreation room' and award-winning teaching services. There is an HDU bay on both wards, offering escalated care including CPAP. Almost all day case transfusions take place out of hours, at weekends. The expectation that planned transfusion should take place out of hours, so that children do not miss school, is commended. Bed availability is seldom a problem. Cannulation is undertaken by doctors, not ward nurses, for children attending for transfusion, except for the small number of children who have indwelling 'port' access. Children admitted or discharged within the previous week are discussed at a weekly MDT meeting. An electronic 'patient key record' is updated to include any important additional issues and medication changes. The team feel to a large extent this replaces the need for a patient-held record, although families do receive copies of clinic letters and discharge summaries.

At the WH site, there is a dedicated paediatric department which houses clinic, ward, and day care facilities. The medical records for the children with red cell disorders are held in lockable shelving in the clinic area, and are accessible to admitting teams at all times. The notes are marked with a red 'dot' to alert treating teams. Clinics starting at 4 pm are held on a quarterly basis for review of children with thalassaemia, led by the lead paediatrician and haematologist. Children with sickle cell disease attend a Monday morning clinic which is held weekly, led also by the lead paediatrician and a haematologist. There is a large interactive patient feedback screen in the clinic waiting area. Day case transfusions are offered until 9 pm, not at weekends, with most children attending after school. Nurses undertake cannulation for these children. The transfusions take place on a general day care area, which manages unscheduled as well as planned care, for example, care of children with acute exacerbation of asthma. This can sometimes hinder planning and the team would like to work towards separating these functions. Partly because of its dual function, the area has hospital beds rather than recliners, so it lacks the more relaxed atmosphere which planned day care / transfusion areas can achieve. Children presenting acutely usually bypass A&E [see above] and attend the ward directly. Their early assessment and care is recorded on a 'first hour' sheet. The ward facilities are

ample, with an HDU bay. The facilities are generally geared more to younger children than adolescents.

On both sites, children admitted urgently come under the care of the general admitting paediatric team, with input from the specialist paediatrician and haematology teams as needed. On the WH site, the paediatricians and haematology consultants undertake joint ward rounds.

Linked Hospitals

At both sites, children who receive most of their care from non-specialist teams at other hospitals are seen for assessment and management guidance, when such input is deemed necessary by the local hospital team. To date there has not been much emphasis on creating a formal network of linked hospitals sharing management protocols and having educational links. There has also been little systematic review and data gathering for children being managed by non-specialist teams. The teams have concentrated initially on creating the joint linked centre between UCLH and WH, and are hoping to move on to extending their network.

Review Visit Findings

Achievements

1 The two hospital paediatric and haematology teams have gone a very long way towards working as an integrated joint centre for the care of children and young people with red cell disorders. To a large extent teams on both sites are sharing practices and working to the same protocols.

2 The facilities at the UCLH site are outstanding, with separate highly age-appropriate provision for young people up to the age of 19.

3 Clinical teams across the two sites are committed to providing excellent services. The special energies and enthusiasm of the Whittington lead paediatrician and lead nurse are particularly evident.

4 Multi-disciplinary working between teams and across acute and community services is very strong, with both in-reach and out-reach work taking place.

5 Service user feedback is sought and taken seriously, evidenced in practice by a display board of user survey feedback on the ward at UCLH and an interactive screen for immediate comment in the out patient area at WH. The accessibility of the team, the use of the emergency card, and the speed of starting treatment after urgent presentation were all especially appreciated by the families who met the review team.

6 The materials and practice for managing transition to adult services are excellent, with an innovative workshop approach and positive feedback from young people on how this had worked.

7 Some of the patient information material was exceptionally good, including the home pain assessment and management 'action plan' for use in sickle cell crisis at home.

IMMEDIATE RISKS

No immediate risks were identified.

CONCERNS

1 At the time of the visit, there were no guidelines or protocols for the management of acutely presenting children available in the A&E Department in UCLH, either electronically or hard copy. A revised version was awaiting approval and uploading.

2 At the WH A&E, the guidelines accessible on the intranet were an old version, recommending approaches to management which are no longer current. The newer guidelines were less easily available via an i-drive which A&E staff could not access easily.

FURTHER CONSIDERATION

1 Some of the clinical guidelines, especially for thalassaemia, were considered not to be 'user friendly'. Some lacked detail at potentially important points [see appendix 2]. There are some issues of document control with not all guidelines having dates or review dates.

2 Patient information for families of children with thalassaemia was not easily available. Some old information leaflets, outdated in content, were still displayed around the hospital, for example, a sickle cell leaflet picked up on the in-patient ward at WH.

3 It was not quite clear the extent to which the service guidelines and protocols are applied and in regular use at the two sites. Some content appeared to apply in practice only to one site. For example, there is guidance in use at both UCLH and WH about nurse cannulation for children having transfusions whereas nurses did not undertake cannulation at the UCLH site.

4 The visiting team considered that children on monthly blood transfusions having to attend clinic for pre-assessment and blood sampling every month [UCLH site] may be excessive. It may be helpful to consider less frequent formal clinic attendance with pre-assessment and sample collection being offered in a different setting or out of hours on, perhaps, alternate months.

5 As clinical links exist with the Royal Free Hospital, it may be helpful to review with clinicians there the management of children and young people with red cell disorders, who attend the RFH A&E in an emergency. It was not clear to what extent the same protocols are in use at RFH, and how systematically the care of the children who present to A&E there is monitored.

6 Waiting time in the Monday morning sickle cell clinic at WH was reported by some of the local team and a user to be excessive on occasions. The team was aware of this and is addressing how it might be improved.

7 The current provision of scheduled and non-scheduled day care in the same clinical area [WH site] can result in problems for planning and booking cases, can lead to conflicts on staff time, and also makes it difficult to offer an appropriately relaxed environment for children attending for regular booked transfusion. If site options allowed, separating these two functions would be beneficial.

8 The visiting team felt that some consideration might usefully be given to clarifying some issues for children living on borough boundary areas, including clear understanding of referral patterns for affected newborn babies, and continued community specialist nurse support for those treated at UCLH or Whittington but living outside Camden and Islington.

9 There is an opportunity to engage with the local commissioning team as it appears, from the review team's meeting with a member of the North Central London Commissioning Agency, that this is the body responsible for commissioning services across both sites. Further discussion might lead to better reciprocal understanding and agreement about how engagement of specialist commissioning teams might best progressed.

10 The service arrangement described by the host teams as a 'network' is functioning, in practice, more as a two-site specialist centre. Links with other, non-specialist, hospital teams at present work on the level of seeing and advising on management of children referred in for specific difficulties or issues. Planning for more systematic partnership working with teams in these hospitals would allow a wider-functioning and more robust clinical network to be established, taking account of the needs of all the affected children across the geographical area.

GOOD PRACTICE

1 Up to date patient records are available at all times This has been achieved through the use of the electronic 'patient key record' at UCLH, and the storage of all medical records for the children with red cell disorders in the paediatric clinic area adjacent to the ward at WH.

2 Medical record folders are marked with a red sticker dot. Treating teams are alerted through the PAS that this is an 'emergency card holder' with a major red cell disorder.

3 Haematology consultants see patients in paediatric clinics from infancy. There are also joint ward rounds by paediatric and haematology consultants at WH. This helps teamwork and gives familiarity with the haematologist from the start which eases the process of transition to adult care.

4 The use of the emergency card on both sites facilitates rapid admission to the appropriate services and, at WH, direct ward access at all times. Together with the use of the 'first hour' sheet ensures focussed care very soon after arrival. Parents found the detailed 'check list' of symptoms very helpful in managing their children at home, and in guiding them when to seek medical attention.

5 The wall mounted guidelines on HDU [WH site] on when / how to refer out to PICU [CATS guidance] is a good example of information being available exactly when and where needed.

6 The operating procedures and service descriptions are clear and practically useful, for example giving detailed lists of specialist services with phone and fax numbers.

7 Play therapy provision is very good, with input at clinic, in the imaging department and the day unit as well as the ward.

8 There were many examples of good practice in services in the community. For example, the screening standards are well met and the counsellors aim to visit every primary school in Islington, which must enhance awareness as well as make their service easily visible and accessible where there are affected children. The community and outreach nurses visit homes and schools to take blood samples for hydroxycarbamide monitoring thus saving hospital visits for children and families. The provision of social work support, sometimes across borough boundaries, is strong.

APPENDIX 1: MEMBERSHIP OF THE REVIEW TEAM

Dr Paul Telfer	Senior Lecturer (Honorary Consultant) in Haematology	Barts & The London NHS Trust
Helen Appleby	Paediatric Sickle Cell Nurse Specialist	Guys & St. Thomas' NHS Foundation Trust
Elaine Miller	Co-ordinator	UK Thalassaemia Society
Neil Westerdale	Advanced Nurse Practitioner Haemoglobinopathies	Guy's & St. Thomas' NHS Foundation Trust
James Borland	Service Manager	Birmingham Children's 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	*Information for sickle cell families is excellent, including some outside-sourced, and some locally written, material. Noted are sheets about 'action plan at home' for sickle cell pain management, travel guidance, priapism information, hydroxyurea leaflet.* *Some information included for the review was outdated. Leaflets on the ward [WH] re sickle cell were also old and included guidance no longer applicable eg attending A&E for sickle cell pain crisis.* *A patient information sheet / card reminding those taking deferiprone of the importance of stopping the medication and having urgent full blood count if fever / symptoms of infection – in case of neutropenia / agranulocytosis - was not seen.*

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	*The clinic letters to GP contain some of relevant information but not all aspects were covered.*
3	Information should be available on transition to adult care. This information should cover all aspects of the transition (QR43).	N	*An excellent range of material was available for sickle cell disease, including self assessment sheet. Transition material for thalassaemia was not seen.*
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	*No patient held record was seen, except for 'emergency cards' at both sites, and transfusion record for long term transfused children at UCLH. These were filed in the patients' notes when the page was complete.*
5	The locally agreed patient-held record (QR4) should be in regular use within the LHT / SHT.	N	*See QR 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	*Facilities on UCLH site were outstanding, especially for young people to 19. At WH the environment is more appropriate for younger children.*
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.	N	*Y for WH* *WH lead nurse fully operational in role and CPD provided.* *For UCLH: the lead nurse has quite recently taken up post and at the time of the visit did not yet appear to work fully in all these roles, and had undertaken little specifically relevant CPD.*
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	*A full list of contact names and numbers was in place for both sites.*
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.	N	*Y for WH* *N for UCLH* *For HDU referral, this is covered in protocols. For CATS / PICU, this is wall mounted on HDU area of ward. A full audit was provided.* *For UCLH, a 'standalone' sheet was included in folder, referring to p10 of clinical guideline. This did not cover specific indications for admission to HD/PICU care.*
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	*Teaching and play provision were noted to be of a very high standard.*
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	*Antenatal and neonatal screening policy, and patient pathway were in place. No service level agreement was is in place. Co-operative working seems, in practice, to function well.*
19	A nurse with competency in cannulation, starting and supervising a transfusion should be available at all times at which children are attending for transfusion.	N	*Y for WH* *N for UCLH* *At UCLH nurses do not undertake cannulation, apart from 'port' access.*
20	All staff involved in the care of children and young people with haemoglobinopathies should undertake regular child protection training.	Y	*Details of training received for core team members.*

Ref	Quality Requirement	Met?	Comment
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	
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	But detail was lacking in parts eg for ACS in sickle cell it is indicated to check arterial gases, without guidance as to what steps to take depending on results. For biliary problems it is indicated to 'call King's' but no specific detail about contacts there. Some of the guidance was felt not to be user – friendly. Many documents lack dates or review dates.

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	*But some parts were not felt to be 'user friendly' eg iron chelation in form of review article reprint. Dates and review dates were again sometimes lacking.*

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	
26	Guidelines should be in use for referral of patients and their families to a clinical psychologist with experience in the care of patients with haemoglobinopathies (including the option for self-referral)	N	*Presented for sickle cell affected children, not thalassaemia.*

Ref	Quality Requirement	Met?	Comment
27	Guidelines for referral for consideration of bone marrow transplantation should be in use.	N	*Not seen in evidence for either condition.*
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	N	*In place for sickle cell but not seen for thalassaemia.*
29	Clinical guidelines should be in use covering review by a specialist nurse or doctor prior to transfusion to ensure that each transfusion is appropriate.	N	*As for 28.*
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 this was as a review article reprint, rather than practical guideline.* *Note: the team queried the use of Exjade in under 6 years olds – indications for change from desferrioxamine in these younger patients was not clear in the guidelines.*
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.	N	*Not available at time of visit in A&E Dept @ UCLH, and old set on intranet @ WH.*
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 initial visit protocol was seen.*

Ref	Quality Requirement	Met?	Comment
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	N	*No guidance covering children previously treated outside UK was seen.*
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 is in place, but examples of letters between the centre and referring hospitals for individual children show communication to be good.*
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 PHR is in use, but copies of clinic letters to families gives a record to refer to. The 'patient key record' at UCLH, and ready availability of case notes at WH, both help ensure up to date information is available to hospital staff, which meets this part of the intended aim of a patient held record.*
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	*This level of working with linked hospitals is not yet in place.*
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	*A quarterly evening clinic for thalassaemia patients review is in place at WH. There is no out of hours clinic provision for sickle cell at WH, and none at UCLH for either condition. Out of hours transfusion is available at both sites. Phlebotomy is available on the day unit until 9 p.m at WH. No out of hours phlebotomy availability at UCLH.*

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	*There is a written policy and the lead paediatrician gave a verbal description of practice. Section 3.6 includes a final point 'paediatrics exempt from UCLH DNA policy' – this is puzzling as it appears to be a paediatric DNA policy.*
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	*And this was viewed to be an excellent document, guiding good practice.*
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	*This was evidenced by a board on the ward at UCLH with results of most recent annual user feedback audit and, at WH, by conversations with users and prominent interactive feedback screen in OPD.*
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)	N	
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 were available.*

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.	Y	*Results of recently undertaken audits of all main aspects, for both sites, and both sets of conditions, were available.* *Note: DNA rates were given but this does not fully address the content of this sub-section for each condition ['effectiveness of action to contact families who DNA'] and given the high reported DNA rates this might be useful.* *No patients had died, but full policies are in place for managing this eventuality, at both sites.*
51	Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry.	N	*No patients yet entered onto NHR. The centres are working on a joined internal database and have consent from many families for NHR entry.*

Commissioners of Services

Ref	Quality requirement	Met?	Comment
52	Each Specialist Commissioning Group[1] should have agreed the location of services for its population: a Specialist Haemoglobinopathy Team/s for children and young people b Local Haemoglobinopathy Team/s for children and young people c The expected referral patterns to each SHT and LHT. d The type of patients (sickle cell and / or thalassaemia) who will be treated by each team.	N	*But the commissioner who attended the visit was aware of these services, and there is an opportunity for further discussion to improve reciprocal understanding.*

Ref	Quality requirement	Met?	Comment
53	Each Specialist Commissioning Group should have: a Compared the staffing, support services and facilities of each SHT and LHT located within its area (QR52) with the levels expected in QRs 7 to19. b Agreed a plan for the development of SHTs and LHTs located within its area. c Monitored achievement of the agreed plan at least annually. The agreed development plan should ensure that QRs 7 to 19 are met within 2 to 5 years.	N	

Additional Requirement – Screening Services

Ref	Quality requirement	Met?	Comment
P3	Timely communication of positive screening results (sickle cell disorder) – including a review of parental results	Y	*Detailed figures were not available at the visit but based on data from 12 babies over 4 years subsequently sent to review team lead, the standard is met.*
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) and seen by 3 months of age	Y	*[first part of this standard ignored, duplicates P3 with different target] As for P4.Only 1 of the 12 babies in the audit was not seen by 3 month of age, and this was as appointment not attended.1st attendance was by 4 months.*
P5	Timely confirmation of diagnosis for infants with a positive screening result**	Y	*In the case records checked, though full audit was not seen.*
S1i	Failsafe to ensure ongoing care	Y	*Process described verbally by hospital and community team.*
S1ii	Up-to-date registers maintained of babies (cases) for which units are responsible	Y	*This community service has had a robust linked antenatal and neonatal testing result database [CLANS] operating for over 10 years. This applies to Camden and Islington babies, not to all babies across a wider, yet-to-be-defined, geographical network.*

** Specified conditions: Hb-SS, Hb-SC, $HbSD^{Punjab}$, Hb-SβThalassaemia (β^{+}, β^{0}, δβ, Lepore), $Hb\text{-}SO^{Arab}$, Hb-S/HPFH