UK Forum on
Haemoglobin
Disorders

WM QRS West Midlands
Quality Review Service

Services for Children with Sickle Cell Disease or Thalassaemia
At
Central Manchester University Hospital NHS Foundation Trust;
Royal Manchester Children's Hospital

Quality Review Visit Report
Visit date: June 23rd 2010

Report finalised: December 2nd 2010

CONTENTS

INTRODUCTION

This report presents the findings of the peer review visit to services for children with sickle cell disease or thalassaemia at Central Manchester University Hospitals NHS Foundation Trust, Royal Manchester Children's Hospital which took place on June 23rd 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 Central Manchester University Hospitals NHS Foundation Trust, Royal Manchester Children's Hospital [RMCH] 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 CENTRAL MANCHESTER UNIVERSITY HOSPITALS NHS FOUNDATION TRUST, ROYAL MANCHESTER CHILDREN'S HOSPITAL

Service (as at 2010)	Patient numbers sickle cell disease	Patient numbers thalassaemia	Sickle cell patients on long term transfusions	Other haemoglobin disorders
RMCH	162	34	12	29
Blackburn	~ 5	11 + <5 BTI	<5	
Wythenshawe	~ 10			

RMCH is situated in the large new complex of hospital buildings in central Manchester. It opened just a year before the visit. The buildings are visually very striking, and house a comprehensive range of clinical, research and academic activities. There is a large paediatric haematology department, which offers haemophilia, oncology and bone marrow transplant services. There are four paediatric haematologists, and seven [one post currently unfilled] specialist nurses are involved in the red cell services: one lead CNS for benign haematology, one whose focus is the children with sickle cell disease, one whose focus is the children with thalassaemia, and one who is the transfusion nurse lead. The paediatric haematologists work as two 'teams' of two, so that the if the clinical lead for haemoglobinopathy is on leave, his partner, the service deputy lead, covers. Services are essentially delivered by the consultant and specialist nursing staff. The service has grown rapidly, from approximately 50 children in 2004 to over 200 in 2010. There is a dedicated haematology unit, where the in-patient ward, out-patient clinic, consultant offices and bone marrow transplant unit are co-located. Children's medical records are stored on the ward for ease of access.

Accident and Emergency

Access for children needing urgent care is via the large, busy, and very well-equipped A&E Department. There are four paediatric trained A&E consultants and there is consultant presence in the department 9am – 7pm on weekdays and for six hours on Saturdays and Sundays. Children attending with sickle cell crisis or other complications are triaged urgently and managed according to the shared pain protocol. Time to administration of first analgesia was audited in March 2009 and is due for re-audit since the move into the new premises. A 12–bedded children's Clinical Decision Unit is adjacent to A&E and can be used for children needing longer than four hour care but not formal admission. One of the specialist nurses checks every morning to see if any children with red blood cell disorders have been admitted during the night so that they can be seen on the ward or followed up at home as needed.

Outpatient Clinics

There is a weekly haemoglobinopathy clinic, in which transcranial Doppler scans are undertaken. The clinic is run by (usually) two consultants, and sometimes a haematology Specialist Registrar. The 0.5 WTE psychologist and the specialist hospital and community nurses also work in the clinic. The lead clinician sees babies and families for their initial visit in the community centre rather than in the hospital clinic. This new baby 'clinic' runs 3 weekly. There is a weekly red cell multi disciplinary team meeting attended by team members as above and also a social worker. Transition clinics are

planned. At present the psychologist and one of the community nurses work with children at ages 11 and 14 introducing aspects of transition, with input from the adult specialist nurse at 16 years. Up to the age of 19 children can choose when to transfer to adult services although transition usually happens sooner.

Day Care

Day care is offered in a large, bright day unit, open from 8 am to 8 pm Monday to Friday, with a variety of beds and reclining chairs. There is some flexibility for transfusions running until after 8 pm. Cannulation is undertaken by the specialist nurses. Saturday transfusions can be arranged on the ward, but families do not often choose this and there is a wait for a paediatric doctor to come to the ward and cannulate at weekends. Exchange blood transfusion when needed is undertaken on the high dependency unit.

In-Patient Facilities

The in-patient ward 84, like all the facilities, is of a very high standard. All the bays and single rooms feel light and airy as they have windows either to the outside or to the large atrium at the centre of the hospital, which has a transparent roof. Nineteen beds are currently open but there is potential to open more. A fold down full size bed for a parent is adjacent to every bed, and there is a day room for parents. There is a pleasant play room and a school room, and teachers liaise closely with schools when children are admitted. An adolescent day room is being established. Most children needing admission come to this ward. Children are admitted to a general paediatric ward instead if there are no available beds but the specialist nurses input to their care on all wards they are admitted to. There are usually between one and four inpatients with sickle cell disease.

Community Services

There is a very well established Manchester Sickle Cell Centre [MSCC] across a road from the main hospitals. There are six specialist nurses, an overall Centre lead, and five others. Staffing has increased over the last two years. The 0.5 WTE psychologist is also based at the Centre and a benefits adviser comes to see clients one half day per week. Their work is primarily with Manchester families but they offer some input across Greater Manchester and, on a case by case basis, to newly identified infants across East Lancashire. For Manchester families, they aim to visit the family twice before the first clinic attendance, which is at the Centre rather than the hospital. The team have developed their own very good information packs for new parents. There is a well maintained data

base of babies going back to the start of newborn screening in 2004. One of the nurses has a special role in home care and transition services. It is planned to meet the mid-teen patients and their families in the near future to develop the transition process more fully. The community services are governed by a service specification drawn up by its commissioners, who work closely with the Centre team and there is a comprehensive quarterly performance management assessment undertaken using a 'continual improvement' tool.

Linked Hospitals

The clinical lead for the RMCH service has been working with smaller DGH's over very many years, holding regular joint clinics at Blackburn, which has the largest patient number outside of the RMCH catchment at (approximately 25 children) Wythenshawe and Leighton. Other linked hospitals are Rochdale, Bolton, and Tameside. The small number of children living in these areas attend RMCH for review and Doppler screening.

Consultant paediatricians from Blackburn and Wythenshawe met the review team. They reported that they were very happy with the support they receive from the RMCH team. It was always easy to reach a consultant for advice and if a patient needed transfer to the Centre there was usually no problem. Both said they would value having RMCH clinical guidelines available for their local use. The Blackburn paediatrician routinely copies the RMCH lead clinician into all letters, as a way of ensuring that he is happy with clinical decisions she is making.

There is a Regional Haemoglobinopathies Group, convened by the specialist commissioner for these services, which includes members of the RMCH team, Alder Hey Liverpool. The Blackburn paediatrician is the DGH member on this group.

Commissioning Arrangements

The specialist commissioning lead locally is involved with these services, and has recently set up the pan-Regional group described above. This has met twice so far, aiming to ensure appropriate access to investigation and management for all children and adults with red cell disorders in the NW Region and Merseyside. It is currently developing clinical guidelines to be used across the region. There is no benchmarking of staffing or resource, but activity is monitored against an agreed plan.

REVIEW VISIT FINDINGS

ACHIEVEMENTS

1 This long established service is delivered by some very experienced and long serving team members, and by some new members as the team has grown in line with its activity. It appears to be providing excellent services to children and families with haemoglobin disorders. There is close working between the acute and community teams, and some particular activities in both settings are singled out as 'good practice' below. The fact that the services appear to work so smoothly only one year after moving into new premises is noteworthy.

2 User feedback confirms that children and families have great confidence in the hospital and community professional teams, reporting that they are easily accessible and take every effort to make care as easy as possible for the users.

3 The facilities are all quite outstanding. The fact that they are largely co-located, with the community centre just a short walk away, helps towards the integration of services.

4 Support for the linked hospitals, in a well established network arrangement, is appreciated by the local teams.

5 Membership of the Board of Governors includes some young people which is likely to help maintain focus on what really matters to them as users of services.

IMMEDIATE RISKS

No immediate risks were identified.

CONCERNS

1 Some audits are available, but none against most of the key standards governing care of children with these conditions. Services appear to be excellent but these audits need to be undertaken to provide assurance that this is the case. This is a well resourced team, and it should be possible to undertake these clinical audits in the near future.

2 There are no guidelines for the management of children with thalassaemia who present with any acute complications.

FURTHER CONSIDERATION

1 While having a consultant and nurse-delivered service has obvious quality benefits, given how much out of hours assessment and treatment is inevitably delivered by junior medical staff, more involvement of trainees in the regular delivery of care should be considered. A less immediate consideration is the need for junior doctors to receive sufficient training to be the next generation of consultants in this growing sub-specialty. RMCH senior managers are already in discussion with the Deanery about a lack of paediatric general trainees.

2 Linked hospitals would value sharing of clinical guidelines across the network. This is work in progress through the pan-Regional group but in the meantime, some units have no written guidelines in place and others are using guidelines gleaned from different centres.

3 As some of the linked hospitals have very small numbers, consideration might be given to providing short written guidance about what presentations in children with sickle cell disease or thalassaemia should prompt admitting teams to contact the Centre for advice and / or transfer immediately.

4 User families commented on the amount of time missed from school by children who have regular transfusions. Further discussion about how best to provide weekend transfusions for the quite large number of service users would be beneficial. This will probably be included in the work soon to take place about transition and services for the older child.

GOOD PRACTICE

1 Much of the written information for user families is excellent, especially: the local packs for parents of affected newborn, the local comprehensive service description which includes a range of contact details for access to useful medical and non-medical services, and the parents' teaching material for using desferrioxamine infusions and intravenous 'port' devices.

2 An educational network is in place, with excellent informational materials for schools, outreach by specialist nurses into schools, and close liaison between hospital teachers and the children's own school teachers.

3 Many of the guidelines are of very high quality, especially the chelation folder, and several include criteria for discharge from hospital which is highly appropriate, but unusual.

4 The ambulance service ‘notification form’, whereby the local service holds details of the child and family, easing communication when the family needs to access hospital urgently, is an excellent innovation.

5 The check made by one of the CNS staff each day to A&E for attendances since the previous day is a good system to ensure appropriate input to care during the admission or contact at home if the child did not need admission.

6 The pro-active ‘trawling’ of wards by the critical care outreach team is ideal, so that acutely ill and deteriorating children are identified without relying solely on ward staff recognising and escalating any warning signs.

7 The availability of a psychologist offering sufficient sessions to see all children systematically, for input and neuro-cognitive assessments, is ideal.

8 There is a comprehensive nurse teaching programme.

9 A focus on the comfort and wellbeing of parents of attending and in-patient children was noticed.

10 The community services are outstanding in a number of ways. The offer of two home visits before the first hospital contact for the families of affected newborn is more than often undertaken, and the team described the way in which the second visit adds value. The first clinic visit takes place in the community centre, with the lead clinician coming there to meet the families, reducing over-medicalisation at this time when the baby is well. The quality of the data held at the Centre is exemplary. The community service is commissioned in an unusually detailed and specific manner, and the quality of the team’s work is closely monitored. Effective team working between the community and acute service teams is also noted.

APPENDIX 1: MEMBERSHIP OF THE REVIEW TEAM

Dr Marie Donohue	Consultant Haematologist	Nottingham University Hospitals NHS trust
Natalie Lawson	Senior Sister	Birmingham Children's Hospital NHS Foundation Trust
Elaine Miller	User Representative	UK Thalassaemia Society
Marie Claire Ngeumshe	User Representative	
Lee McPhail	Assistant Director of Operations	North Middlesex University Hospital NHS 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	*But no information in other languages, not travel advice for those with thalassaemia and no information intended for use by children at different ages.*

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 for GP / primary health care team was not seen*
3	Information should be available on transition to adult care. This information should cover all aspects of the transition (QR43).	N	*This is work in progress*
4	The SHT and its linked LHTs should have agreed a patient-held record for recording at least: a Information about the patient's condition b Current management plan c Regular medication d Named contact for queries and advice e Alternative contact for times when key contact is away.	N	*Not now in use*
5	The locally agreed patient-held record (QR4) should be in regular use within the LHT / SHT.	N	*As above*
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 are outstanding*
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	
16	The following services should be available a Paediatric high dependency care b Paediatric intensive care There should be agreed criteria for admission to each level of care.	Y	
17	The following support services should be available: a Interpreters b Social work c Play specialist d Hospital teacher (in-patient care only) e Child psychologist f Dietician	Y	
18	A service agreement for support from community services should be in place. This service agreement should cover, at least: a Guidelines for involvement of community paediatric services in the care of patients with haemoglobinopathies b Role of community services c Exchange of information between hospital and community services and vice versa d Arrangements for liaison with schools.	Y	
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	*Up to date training records for nursing staff and senior medical staff were seen.*
CLINICAL and REFERRAL GUIDELINES			
21	Guidelines should be in use covering a How to establish and confirm diagnosis b Parent and sibling testing	Y	
22	Clinical guidelines should be in use covering: a Recommended immunisations b Immunisations, other prophylaxis and travel advice prior to travel abroad. c Penicillin prophylaxis while awaiting clarification of diagnosis (sickle cell disease only)	Y	

Ref	Quality Requirement	Met?	Comment
23	*Clinical guidelines should be in use covering possible acute presentations including, at least:* **For patients with sickle cell disease:** a Fever and infection including major sepsis b Acute pain c Acute anaemia d Stroke and other acute ischaemic events e Acute chest syndrome f Acute splenic sequestration g Abdominal pain / jaundice h Priapism i Changes in vision, including urgent referral for an ophthalmologic opinion j Indications for 'top-up' and for exchange transfusion and practical protocol for undertaking exchange transfusion **For patients with thalassaemia:** a Fever and infection including major sepsis b Unexpected cardiac, hepatic, endocrine decompensation. These guidelines should ensure that patients are transferred to the paediatric ward for assessment as quickly as possible	*N*	*Mostly in place but no guidelines for management of children with thalassaemia presenting with acute problems*
24	Clinical guidelines should be in use covering routine out-patient monitoring and management between formal annual progress review visits, including: **All patients:** a General assessment of well-being including school attendance b Any difficulties adhering to treatment or other difficulties with care c Monitoring growth and development d Checking for palpable spleen **Patients with sickle cell disease:** a Discussion to ensure analgesics available for home use understood and effective b Informal assessment of cognitive function, learning and behavioural difficulties, including referral if needed c Checking for history of priapism in boys d Checking for and management of nocturnal enuresis e Checking all necessary immunisations up to date f Penicillin therapy and alternative therapy for children who are allergic to penicillin g Monitoring of oxygen saturation by pulse oximeter h demonstrating to parents / carers how to check for splenic enlargement i Monitoring of Hb levels, renal function tests, liver function tests j Indications for early referral to specialist haemoglobinopathy team (LHT only) **Patients with thalassaemia and regularly transfused sickle cell:** a Checking adherence specifically to chelation therapy and any difficulties b Monitoring Hb levels, iron levels, liver function and other biochemistry c Indications for early referral to specialist haemoglobinopathy team	Y	

Ref	Quality Requirement	Met?	Comment
25	Clinical guidelines should be in use covering **annual specialist review** visits including, at least: **All patients:** a Regularity of school attendance and reasons for absence b Any difficulties adhering to treatment or other difficulties with care c Monitoring growth and development, including review of centile charts d Health promotion and healthy lifestyle advice and support e Contraception and sexual health in relevant age group f Travel advice g Review Hb levels, renal and liver assessment, other biochemistry h Hepatitis B vaccination i Monitoring of hepatitis B antibodies and action to be taken should levels fall j Monitoring for hepatitis C in transfused patients k Discussion and preparation of child for any planned surgery l Indications for consideration of splenectomy m Preparations for splenectomy including recommended immunisations n Treatment of complications of splenectomy, including persistent thrombocytosis. **Patients with sickle cell disease:** a As QR 24 plus b Monitoring and screening for neurological complications, including transcranial Doppler ultrasonography c Indications for imaging to assess the extent of cerebrovascular disease d Indications for overnight oxygen saturation monitoring (sleep study) e Indications for echocardiography including possibility of pulmonary hypertension **Patients with thalassaemia:** a As QRs 31, and 32 where applicable [thalassaemia intermedia] plus b Review annual red cell consumption c Review adequacy and appropriateness of iron chelation regimen d Consideration of options for helping children, young people and their families to adhere to chelation therapy e Audiometry and ophthalmology check for those on desferrioxamine, from age 10 f Cardiological assessment (from age 10) g Testing for endocrine abnormalities (from age 10) h Bone mineral density assessment (from age 10).	Y	*There was no policy or guideline but reviews were evident in the medical records. The sickle annual review sheet was felt to be brief and did not cover all aspects of this QR, and no annual review sheet was seen for thalassaemia, but the team indicated in discussion that reviews took place, and there is a system of index cards [complex to outside visitors!] which apparently indicates which patients have had / are due annual review, and records the findings. The team may wish to develop a more accessible guideline / proforma to support this QR.*

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	*No specific reference to possibility of self referral but as all children meet the psychologist anyway not really required.*
27	Guidelines for referral for consideration of bone marrow transplantation should be in use.	Y	
28	Clinical guidelines should be in use covering: a Indications for regular transfusions b Investigations and vaccinations prior to first transfusion c Monitoring of haemoglobin levels	Y	
29	Clinical guidelines should be in use covering review by a specialist nurse or doctor prior to transfusion to ensure that each transfusion is appropriate.	Y	
30	A Transfusion Policy should be in use covering: a Area/s where transfusions will usually be given b Procedures for checking blood c Staff allowed to undertake cannulation d Recommended number of cannulation attempts e Maximum rate and volume of transfusion according to size of child f Monitoring the transfusions g Management of transfusion reactions h Arrangements for medical cover during transfusion i Arrangements for ensuring that a nurse with appropriate competences is in attendance throughout the transfusion.	Y	
31	Clinical guidelines for chelation therapy and monitoring iron load should be in use including: a Indications for starting chelation b Choice of regime c Dosage and dosage adjustment d Clinical assessment of tissue damage e Monitoring of serum ferritin f Use of non-invasive estimation of organ-specific iron loading heart and liver by T2*/R2 g Management of side effects of chelators.	Y	
32	Clinical guidelines for the management of thalassaemia intermedia should be in use including: a Indications for transfusion b Monitoring iron loading c Indications for splenectomy.	Y	
33	Clinical guidelines should be in use covering: a Indications for exchange transfusion b Arrangements for carrying out an exchange transfusion.	Y	
SERVICE ORGANISATION and LIAISON WITH OTHER SERVICES			
34	Clinical guidelines for acute and out-patient monitoring and management (QR23 and QR24) should be available and in use in appropriate areas including A&E, clinic and ward areas.	Y	

Ref	Quality Requirement	Met?	Comment
35	A protocol should be in use covering the initial clinic visit for patients with haemoglobinopathies covering, at least: a Giving each patient information relevant to their condition (QR1) b Giving each patient their patient-held record (QR4) c Allocation of a named contact for queries and advice to each patient. d Discussion of arrangements for future treatment and care e Sending the GP information relevant to their patient's condition (QR2)	Y	*Although no PHR in use.*
36	A protocol should be in use covering the initial clinic visit for patients previously treated outside the UK, including: a Full medical history and examination b Investigations c Referral to other specialist services (QR15) d All aspects of QR28	Y	
37	A protocol should be in use covering arrangements for care between SHT and LHT for ongoing care. This protocol should ensure that, to facilitate effective shared care: a All patients have an up to date patient held record and details of their care plan. b The LHT and the patient's GP have received details of the patient's care plan.	N	*No protocol was seen*
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	*And copy letters to patients were not consistently seen, in case records.*
39	The SHT and its linked LHTs should have agreed a policy on the communication of clinical information, management plans and important decisions regarding treatment between clinical teams. This protocol should cover information from specialist clinic visits as well as contacts with SHT / LHTs. The protocol should be specific about frequency / indications for communication with the patient's GP	N	*No policy seen though evidence of communication between Centre and local hospitals [clinic letters] was seen*
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	*This material was excellent.*
41	Patients and families should have choice of attending for blood tests, clinic appointments and blood transfusions 'out of hours' to minimise disruption to normal life	N	*Although some flexibility for transfusion times on weekday evenings is possible, and children can attend for phlebotomy until 5.30. No out of hours clinic appointments 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	
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	
45	The SHT should run a programme of training and awareness of the management of patients with haemoglobinopathy for its main referring LHTs.	N	*Some nurse teaching is in place to which staff from other units are invited, but no systematic programme of teaching for medical staff.*
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	*No evidence of such meetings being held*
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	*Weekly meetings in place, this is fully covered, with evidence of careful monitoring.*

DATA COLLECTION and AUDIT			
Ref	**Quality Requirement**	**Met?**	**Comment**
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 were seen regarding neutrophil counts compared with clinical severity for sickle cell children, time to first analgesia [March 09], and a first year audit of the genetic diagnostic service. None of the audits recommended here have been undertaken.*
51	Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry.	N	*No patients entered to date, but LREC application approved and funding now identified: to start shortly.*

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	

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	*Carefully maintained database gives assurance that all the newborn screening standards are 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)	Y	*[the first part of this standard overlaps with P3, and has been ignored]*
P5	Timely confirmation of diagnosis for infants with a positive screening result**	Y	
S1i	Failsafe to ensure ongoing care	Y	
S1ii	Up-to-date registers maintained of babies (cases) for which units are responsible	Y	*As P3*

** 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.