UK Forum on
Haemoglobin
Disorders

WM QRS West Midlands Quality Review Service

Services for Children with Sickle Cell Disease or Thalassaemia
at
North Middlesex University Hospital NHS Trust

Quality Review Visit Report
Visit date: May 18th 2010

Report finalised: March 21st 2011

CONTENTS

INTRODUCTION

This report presents the findings of the peer review visit to services for children with sickle cell disease or thalassaemia at North Middlesex University Hospital NHS Trust, which took place on 18th May 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 North Middlesex University 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 NORTH MIDDLESEX UNIVERSITY HOSPITAL NHS TRUST

INTRODUCTION

The North Middlesex University Hospital NHS Trust serves a multi-cultural area of North London with a population of 500,000. The area has a high deprivation index with many single parent families, refugees and travelling families. Thirty per cent of the local population do not have English as their first language.

The Trust can provide almost all aspects of care for children with red cell disorders with in-patient and out-patient facilities on site. Community paediatric services were provided by two separate Trusts, Enfield Community Care Trust and Haringey Primary Care Trust. Junior doctors from Haringey contributed to the middle grade on-call rota.

A wide range of the hospital medical and surgical specialities were provided on site haematology, which focuses mainly on haemoglobinopathies, cancer services, renal dialysis, cardiology and HIV/AIDS. Patients were referred to Great Ormond Street Hospital for surgical procedures.

The North Middlesex University Hospital (NMUH) Paediatric Department was managed from 2004 by Great Ormond Street Hospital (GOSH) as part of a partnership arrangement. From May 2010 this

arrangement changed with managerial responsibility returning to North Middlesex University Hospital NHS Trust. The close links built up over the last five years have remained, however, with GOSH supporting the Department through education and training.

The Trust Chief Executive had an excellent knowledge of haemoglobinopathy services and was keen to support further service development.

Service (as at 2010)	Patient numbers sickle cell disease	Patient numbers thalassaemia	Patients on long term red cell transfusions
NMUH and Barnet General Hospital	219	<5	6
QE II Hospital, Welwyn Garden City	17		
Addenbrooke's Hospital, Cambridge	24	<5	
Princess Alexandra Hospital, Harlow	13		

Accident and Emergency

The hospital had a busy Accident and Emergency Department with approximately 120,000 new referrals per year, 25 to 30% of them children.

Outpatient Clinics

Keats – Outpatient Clinic

A dedicated Paediatric Outpatient Department was located in the tower block. There were weekly paediatric haematology clinics run jointly by the lead Paediatrician, a Paediatric Haematologist (joint post with GOSH) and a Haematologist. An after-school adolescent clinic was held bi-monthly and a family clinic was run quarterly, where all parents and children with haemoglobinopathies were seen together to save multiple clinic visits. Twice a month, the paediatric haematologist undertook a dedicated Transcranial Doppler (TCD) stroke screening clinic list which ran alongside the paediatric haematology clinic. Children and their parents were seen by nurse specialist for advice, education and support. A housing advice worker, provided by Haringey Social Services, attended the clinic on alternate weeks. Children requiring blood sampling before transfusion could use the Day Unit or the main hospital phlebotomy area, which was open until 6:45 pm two evenings a week.

Young people aged 14 years and over, requiring frequent hospital attendance due to regular blood transfusions or monitoring of hydroxyurea therapy, could attend the Tuesday afternoon or evening

haemoglobinopathy clinic, thus attending after school/college/work. Once a month this clinic was run by the paediatric haematologist and was specifically aimed at adolescents and young adults so continuing the transition process.

DAY CARE

Paediatric Day Assessment Unit (PDAU)

The Paediatric day assessment unit was nurse-led, with experienced staff undertaking pre-transfusion assessment and cannulation. It was open daily, Monday to Friday from 8.30 to 5pm, for review and treatment including blood transfusion. Children of school age requiring chronic blood transfusions could start after school and complete transfusions on the in-patient ward.

IN-PATIENT FACILITIES

Children with sickle cell or thalassaemia had direct access to Rainbow, the paediatric in-patient ward, at all times. At their initial clinic visit the families were given a 'pink passport' and invited to phone the ward at any time to discuss any concerns. Nursing staff also provided advice by phone or could advise attendance at the ward for assessment. This 'fast track' service was one of the first of its kind[1].

The inpatient ward, Rainbow, was a 25 bedded paediatric ward, including two high dependency beds. There was adequate cubicle accommodation, with two cubicles dedicated for oncology patients, and one bay designated for older children / adolescents. There were facilities for resident parents, school and play area.

Clinical cover was provided on a weekly rotational basis by paediatric consultants. The 'admitting consultant' for the week took clinical responsibility for all acute admissions, liaising as necessary with colleagues if specialty patients were admitted. The paediatrician with an interest in haematology and the haematology team were informed of all admissions in this patient group. The haematology registrars and paediatric consultants conducted a daily ward round on the children's ward, and children were seen on the twice-weekly haematology consultant ward round. High dependency facilities were available for the management of sick children, with clear admission criteria. There were links with the Children's Acute Transport Services team for patients requiring Paediatric Intensive Care.

[1] [ref 'Evaluation of fast track admission policy for children with sickle cell crises: questionnaire survey of parents' preferences'. Fertleman CR, Gallagher A, Rossiter MA. BMJ. 1997 Sep 13;315(7109):650].

COMMUNITY SERVICES

The George Marsh Centre for Sickle Cell and Thalassaemia located in Tottenham, an area where many of NMUH patients live, was opened in 1989. The Centre is managed by NMUH and the staff are NMUH employees. A team of 4.5 WTE staff provided antenatal counselling and prenatal diagnosis referrals. They were responsible for receiving the newborn screening results for Barnet, Enfield and Haringey, visiting the family of an affected newborn at home to give the diagnosis and early education, and providing support to affected families in the early weeks. The nurse usually accompanied the family to the first clinic visit. Languages spoken by the team include English, French, Portuguese, French, Lingala and Twi. The team also helped to set up the parents support groups. There was access to housing, social services and benefits advice for Haringey residents and it also houses educational resources for patients, carers and the community with a comprehensive library and frequent teaching days for health professionals and non health professionals. The team's main other role was to provide an outreach home pain assessment and treatment service for adult sickle cell patients, and to staff the adult red cell day unit.

LINKED HOSPITALS

A Paediatric Haematologist ran the TCD screening service for the whole North Central London sector, including University College Hospital and Whittington Hospitals, with outreach clinics held at each. Specialist team support was offered to the local teams at Barnet Hospital, Welwyn Garden City Hospital, Lister Stevenage, and Addenbrooke's Cambridge; consultant paediatricians from the first two of these attended to meet the review team. From May 2009 NMUH was identified as the specialist team for newborn babies with red cell disorders in Harlow. Children from these hospitals were seen at NMUH mostly on an annual basis to undertake annual specialist review. Some children attended more frequently, as necessary. Children in Cambridge were seen on an outreach basis, with TCD being offered at the same time. It was intended to offer outreach clinics at Harlow and Welwyn Garden City. As a specialist centre, one network meeting had been held to provide education and training to the local teams, describing the service, encouraging common protocols and practice. A second annual meeting was planned for September 2010 to continue to improve the functioning of the network.

COMMISSIONING ARRANGEMENTS

Discussions took place with a well informed local commissioner who clearly viewed haemoglobin services as an important local issue and understood the issues relating to providing services for

children with red cell haemoglobinopathies. There was commitment to improving community care with the aim of reducing admissions.

REVIEW VISIT FINDINGS

We would like to thank all the staff at NMUH for their work in preparing for this visit and for their support and help during the visit itself. It was particularly helpful to have the documentation so clearly laid out.

The overall findings from the visit were very positive. This is an excellent patient centred service with committed and enthusiastic staff. There was very strong, genuine, user involvement which ran throughout the service.

ACHIEVEMENTS

1 The level of information available was, in general, comprehensive and of a high standard. Protocols were readily available and up to date. Several excellent audits, including in screening, suggesting that information was getting out to families. There were strong links with the neonatal screening system which were working very well, although the meetings were, perhaps inevitably, more focussed on antenatal rather than neonatal screening. The only issues related to informing families of an affected newborn by four weeks of age; for some children this was related to the timeliness of getting data from the screening laboratory.

2 There were excellent transition arrangements with a genuinely 'seamless' transition to adult care and regular joint paediatric/adult clinics.

3 There was excellent patient involvement in the service through a support group. The users were very positive about the service but, crucially, also felt that when issues were identified they were dealt with promptly. This User Group already had links with the excellent George Marsh community centre. Both these service user groups should continue to be supported and encouraged to continue their input into service development.

4 Good educational and review meetings were accessible to both local and specialist teams. Network arrangements with Cambridge and other outlying Trusts had made real progress. The linked hospitals were now seen as part of the network. Outreach clinics existed, with plans for more, if feasible. Future development was largely dependent on resources being available.

Immediate Risks

There were no immediate risks identified.

Concerns

1 The Clinical Nurse Specialist was part-time and had no cover for absences. This was of particular concern because of the absence of specialist nurse support in some of the linked hospitals.

2 Links with linked hospitals, although developing well, were not yet sufficiently robust to ensure that all children and young people with red cell disorders within the network were receiving appropriate care, including annual review.

Further Consideration

1 The TCD Doppler screening programme was organised and undertaken by one Paediatric Haematologist. The paediatric nurse specialist had also undertaken training, however given her current workload, would find it difficult to support the service. So whilst the service seems comprehensive, it may be worth considering adding some support, in the form of an Ultrasonographer, to improve sustainability.

2 Some excellent nurse-led services were provided by the Clinical Nurse Specialists. This post was originally a home care, outreach post, designed to support families in managing pain episodes at home. More recently the demands of the hospital service had meant that the outreach function had diminished. The visiting team felt the service should be looking for additional nursing input in future years to ensure appropriate home care support is available to give families confidence in independent care, and for input to linked hospitals without specialist nursing staff.

3 PICU transfer: Transfer services were provided on a London-wide basis to available PIC beds. There were concerns about access to PICU beds and the speed of response of the system to a rapidly deteriorating patient with sickle cell disease. No specific instances of high risk incidents were cited.

4 There was similar concern about access to specialist urology services for the emergency treatment of priapism, and specialist neurology services for evaluation of symptomatic children or those with conditional or abnormal trans-cranial Doppler velocities. Pathways need

to be clear and robust. Many London centres are affected by these issues and by difficulties in access to PICU.

5 Resources available to the psychology service may benefit from review as the service appeared small and the resources modest for the number of patients.

6 The very small number of patients with thalassaemia get excellent care but may benefit from contact with others with the disorder.

7 If expansion of support to outlying hospitals is to move forward, with regular planned outreach clinics as planned, additional medical, nursing and data handing resources, in particular, will be needed.

8 The sustainability of the fast-track admissions policy may be affected by forthcoming reductions in junior doctors' hours and should, therefore, be kept under review.

9 Whilst Haringey Social Services provide social worker, housing and benefits advice, there was no similar provision for residents of other PCT's seen at NMUH, namely Enfield, Barnet and Edgware.

GOOD PRACTICE

1 The regular and routine practice of sending letters to families following consultations was excellent.

2 There was extensive community support available through the George Marsh Centre, including benefit and housing help. Liaison between this Centre and the hospital Trust was excellent. The Centre was very much an integral part of the service.

3 The availability of an after-school and evening clinic was excellent.

4 The fast track admissions policy enabled rapid assessment and ward admission. This works well and has been well evaluated, including high user satisfaction.

APPENDIX 1: MEMBERSHIP OF THE REVIEW TEAM

Elaine Miller	User Representative	UK Thalassaemia Society
Dr Asa'ah Nkohkwo	User Representative	Sickle Cell Society UK
Dr Baba Inusa	Paediatric Haematologist	Guy's and St Thomas' NHS Foundation Trust
Dr Jo Howard	Consultant Haematologist	Guy's and St Thomas' NHS Foundation Trust
Sally Riley	Deputy General Manager Women & Children	Whittington Hospital NHS Trust
Dr Phil Darbyshire	Consultant Paediatric Haematologist	Birmingham Children's Hospital NHS Foundation Trust

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	*Excellent comprehensive, evidence letters were routinely sent to parents.*

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	*This had recently started.*
5	The locally agreed patient-held record (QR4) should be in regular use within the LHT / SHT.	Y	
6	Services should be provided in a child friendly environment, including toys and books / magazines for children and young people of all ages.	Y	*Good facilities were available in all areas. The clinic was particularly impressive.*
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.	N	*Cover arrangements were not yet in place.*

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	*Off site facilities were detailed with access arrangements.*
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	*PICU access was via London transfer. See comments in main report.*
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	
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	
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	

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	

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	

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	*A new post holder was awaited.*
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	
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.	Y	
38	A protocol should be in use covering: a Updating patient-held records b Offering patients a permanent record of consultations at which changes to their care plan are discussed. c Recording changes of key contact d Giving further information (QR1) as patients' and families' needs change	Y	
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	Y	
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	Y	*There was good evidence of flexibility in arrangements*

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.	Y	*Education was in place and available to LHT s.*
47	The SHT should meet at least annually with its referring LHT teams to: a Identify any changes needed to network-wide policies, procedures and guidelines b Review results of audits undertaken c Review any critical incidents including those involving liaison between teams d Consider the content of future training and awareness programmes (QR45)	Y	
49	The SHT should meet at least annually with representatives of the neonatal screening programme to review progress, identify issues of mutual concern and agree action.	Y	

Ref	Quality Requirement	Met?	Comment
DATA COLLECTION and AUDIT			
50	The LHT / SHT should have audited compliance with key standards including: **Patients with sickle cell disease:** a Proportion of patients taking regular penicillin b Proportion of patients fully immunised against pneumococcus c Proportion of patients (HbSS and HbSβ^0) who have had Transcranial Doppler ultrasonography undertaken within the last year d Proportion of patients who have had their annual multi-disciplinary review within the last year e Effectiveness of action to contact families who have not attended for follow up appointments. f Review of the care of any patients who have died. **Patients with thalassaemia:** a Proportion of patients on chelation therapy b Proportion of patients who have had their annual multi-disciplinary review within the last year c Adequacy of recording of: ▪ Pre-transfusion Hb levels ▪ Regular monitoring of iron level ▪ Complications of iron overload ▪ Height / weight progression ▪ Spleen size ▪ Support for home chelation programme (QR40) d Effectiveness of action to contact families who have not attended for follow up appointments. e Review of the care of any patients who have died.	Y	
51	Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry.	Y	

Commissioners of Services

Ref	Quality requirement	Met?	Comment
52	Each Specialist Commissioning Group should have agreed the location of services for its population: a Specialist Haemoglobinopathy Team/s for children and young people b Local Haemoglobinopathy Team/s for children and young people c The expected referral patterns to each SHT and LHT. d The type of patients (sickle cell and / or thalassaemia) who will be treated by each team.	N	*Commissioners had a good understanding of expected referral patterns but these had not yet been formally agreed.*
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	*This work had not yet taken place.*

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	*Figures for 33 infants born between Jan 09 and April 10 were provided. These showed some delays in communication.*
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]. 86% babies were first seen in clinic by age 3 months over the whole period. Over more recent months 91% had been achieved.*
P5	Timely confirmation of diagnosis for infants with a positive screening result**	Y	*From notes audit of the cohort of 33 babies.*
S1i	Failsafe to ensure ongoing care	Y	*DNA policy*
S1ii	Up-to-date registers maintained of babies (cases) for which units are responsible	Y	

** 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.