UK Forum on
Haemoglobin
Disorders

WM QRS West Midlands
Quality Review Service

Services for Children with Sickle Cell Disease or Thalassaemia at Birmingham Children's Hospital

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Quality Review Visit Report
Visit date: March 17th 2010

Report finalised: June 10th 2010.

CONTENTS

INTRODUCTION

This report presents the findings of the peer review visit to services for patients with sickle cell disease or thalassaemia at Birmingham Children's Hospital NHS Trust which took place on March 17th 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 Birmingham Children's 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 BIRMINGHAM CHILDREN'S HOSPITAL NHS TRUST

Service (as at March 2010)	Patient Numbers		Patients on long term red cell transfusions
	Sickle cell disease	Thalassaemia	
BCH	~ 300	80 Plus ~ 50 post BMT	~ 90
Wolverhampton	30	5	
Coventry	50	<5	
Stoke	<5	8	
Sandwell	25	<5	
Northampton	45	<5	
Kettering	35	-	

Background

Birmingham Children's Hospital is a large Foundation Trust set in the centre of the City. The Haematology Unit includes a large haemophilia and thrombosis service, paediatric oncology, managing approximately 60 new children with leukaemia annually, and a bone marrow transplant unit undertaking approximately 30 allografts a year. There are 5 consultant paediatric haematologists, of whom two provide the haemoglobinopathy service; for each this represents about 0.2 of their job plan. Thus the service is operating on approximately 0.4 wte consultant time. There are specialist registrars in paediatric haematology but their time is mostly taken up with care of the oncology children. They sometimes come to work in the red cell clinics (but not consistently). Other than the consultants, the service is led by the senior Day Unit Sister, and supported by a Haemoglobinopathy Liaison Sister and a community based nurse counsellor. There is an additional short-term funded nurse who is focussing on transition of children to adult services.

There is one weekly clinic for children with red cell disorders, in which the two consultants see 50 – 60 children each week. The clinic usually over-runs, starting at 9 am and finishing at 2 – 3 pm, although the last appointment is usually 12.30. On alternate weeks, when there is a parallel TCD clinic, there are insufficient clinic rooms and one consultant has to see children in a non-clinical area. An SpR sometimes comes to work in the clinic, but space restriction, and the staffing requirements of a parallel haemophilia clinic, make this inconsistent. There is an MDT after each clinic, which the community nurse specialist also attends.

Accident and Emergency

Children needing urgent assessment and care come to the A&E Department. It is not currently possible to offer direct access to a specialist assessment ward area. Children register and then wait to be called for formal 'triage'. Apparently a system of 'informal triage' has recently started whereby a nurse comes out from A&E periodically and checks the waiting children for any that appear to need more urgent input. The time from registration to triage / admission to the department is not formally recorded. This causes difficulty in assessing time from arrival to, for example, first analgesia (a standard service measure). Acutely sick children are brought straight through to resuscitation cubicles. Clinical guidelines are available to staff in A&E, both hard copy – given to every new trainee doctor on arrival – and electronically. According to the pain score, analgesia might be paracetamol / diclofenac, or oramorph, and those not responding within 30 minutes can be offered iv morphine. These children are admitted. PCA facilities are offered in-hours.

DAY CASE AND IN-PATIENT FACILITIES

The Day Unit offers transfusions to children on 4 days per week. Up to 25 children are transfused each week in a small open area with reclining chairs and beds. No out of hours transfusion service is available although children and families currently being treated have indicated that few of them would use this service at present; this issue could be revisited from time to time. The senior Day Unit sister reviews the children before transfusion, and offers cannulation. An 0.5 band 6 and two band 5 nurses are being trained to undertake this procedure. There are plans to convert a junior doctor post to an ANP post in order to support the Day Unit activity. It had been intended that this facility might offer an assessment / day care service for children with sickle cell pain but this has not been possible because of space and the unit is also difficult to access easily. Plans to re-site the Day Unit are being discussed. This is not yet finally decided, nor is the new siting of the Day Unit yet established. However, it is hoped that it will be more spacious and appropriate to the clinical activity it supports. Extending opening (eg 8 am – 10 pm) is being considered, and it may then become possible to offer the 'drop in' facility for sickle cell which the team would like to provide.

Although Ward 15 is the official 'home ward' for children with sickle cell / thalassaemia who need hospital admission, in practice it is often not possible to admit and manage them there. They may be managed on any of the in-patient wards, and on different wards on successive admission, so that while nursing staff on the different wards are familiar with the treatment protocols, continuity of nurse team input and development of a specialist in-patient team is not possible. The Day Unit Sister or Liaison Sister try to visit children and young people who are in-patients and support the ward nurses in their care. Bed shortages are reported to be common, and transfer out of BCH A&E Dept for lack of an in-patient bed is not infrequent – perhaps two children per month from this specialty. There is a new ward facility gifted by the Teenage Cancer Trust and, space allowing, older children with red cell disorders can in principle be admitted there, but it is expected that bed availability will limit this. Once children are 16, they will often be admitted to an adult ward, even if they have not yet been through formal transition to adult services.

LINKED HOSPITALS

There are no formal commissioning arrangements for the shared services with surrounding hospitals - which stretch from Stoke to Northampton and Kettering - and no formalised governance arrangements. There is, however, a clinically functioning network, with one of the specialist consultants offering at least twice yearly outreach clinics at each of the sites. This is worthy of

comment considering the relatively small proportion of his job plan which is committed to haemoglobinopathy work. The linked hospital consultant paediatrician who spoke with the review team described appropriate and readily available clinical support and advice regarding children causing concern, and also the use of local clinical guidelines which had been approved by the specialist consultant from the centre. TCDs are currently done in Birmingham and children travel in, but there are plans to make TCD part of the outreach service instead. There was reported to be a problem with getting a bed for a child needing urgent transfer in to BCH, although children needing PICU could usually be accommodated. It was felt by the BCH management team that delays in getting children transferred in had previously caused difficulties, but there was evidence of recent improvement.

Review Visit Findings

Achievements

1. This is a most impressive and dedicated medical and nursing team, with many long-serving members, offering services of a very high standard to a large number of patients. This is especially commendable given the constraints of facilities and staffing numbers under which they are working. Co-operation and close team working make this possible and team members appear to enjoy their work. Flexibility between the different nursing roles is noteworthy. The solidly nurse-led running of the Day Unit was felt to be an excellent model.

2. The positive contribution of the transition nurse is manifest. There are excellent materials and innovative ways of involving the older child in their care as they prepare to transfer to adult services. This is especially important in a unit such as this where adult facilities are at a different hospital.

3. The services and staff are highly valued by the user children and families.

4. The level of data collection and entry of children onto the National Haemoglobinopathy Registry is high especially as there is currently no data manager support.

5. The achievement of 100% of affected newborn being seen in clinic by the age of 3 months across the West Midlands [against a minimum standard of 90% and developmental standard of 95%] is outstanding.

IMMEDIATE RISKS

There were no immediate risks.

CONCERNS

1. There are aspects of the service which appear to be substantially under-resourced. The overall provision of approximately 0.4 wte consultant time for a service of this size, complexity and profile is markedly low compared with other units, and the provision of a single nurse lead, liaison nurse and community based nurse specialist, also responsible for antenatal counselling, is likewise thin. The Day Unit facility is cramped and clinic space is insufficient so that on alternate weeks one consultant sees children in a non-clinical area - the staff room - without examination facilities. This is inappropriate.

2. It is apparent that some of these constraints are not unique to this part of the hospital, and the review team was unclear about the level to which these concerns had been conveyed to senior management. There was, however, a sense that recently senior managers had been more open to considering the need for change and the team were optimistic that improvements might result.

3. The move of the haemoglobinopathy unit is soon to be decided. There is a possibility that facilities may be larger and more appropriate but it was thought that there might be temporary accommodation in the first instance and the Trust will want to ensure that any temporary or longer term siting does not further constrain or compromise this service.

4. The short-term funded 'transition nurse' is providing invaluable support in an important area of the service. If funding is not extended there will be an important loss of opportunity for continuing and extending this work, which is recognised to be key for the successful ongoing care, adherence to medication, and social integration of these young adults, in turn affecting clinical outcomes.

5. There are few reported audits of practice, although this – and the incompleteness of data held for the network – is not surprising given the lack of data manager time.

6. The difficulty in practice in admitting children to Ward 15, the official 'home ward' for inpatient care, is a concern given the desirability of developing a specialist and experienced nursing team for managing acute problems in children with these conditions. An additional concern is the

recurring, if not frequent, need for children to be transferred outside BCH to be managed by non-specialist medical teams, on occasions when no in-patient bed is available.

7. Communication with the BCH specialist team by teams caring for these children in other Birmingham hospitals is inconsistent. For example, a child may go for surgery at another hospital without the BCH team being informed or in a position to advise about peri-operative management. This could constitute clinical risk and lead to adverse incidents.

FURTHER CONSIDERATION

1. Some of the 'protocols' are currently in the form of a sheet describing current practice [for example those relating to out-patient monitoring, initial clinic visits, communication policy]. These could quite easily be combined to give a more cohesive and formal set of protocols which could then be shared with network hospitals as well as used at BCH. Other guidelines take the form of copies of the relevant UK clinical standards chapter. For some, for example iron chelation, this is bulky and not easy to use for immediate guidance. It might usefully be abbreviated and included into the general set of guidelines. A couple of guidelines have the logo of other hospitals, many need review dates and, in general, a robust document control system appeared to be lacking for most of the policies and protocols reviewed by the visiting team.

2. Play therapy input is available for children having problems with procedures, but there is no provision for a play specialist to be present at other than these specifically requested times.

3. The time of arrival in A&E for a child with sickle cell pain crisis should be recorded, and used to assess time to first analgesia, rather than the triage time which can be a variable time after arrival.

4. There is need for an overview of staffing and a consideration of adding data management support. If the outreach services were subject to formal SLAs, and if service line reporting and management becomes embedded, this service is likely to generate sufficient income to allow for more appropriate staffing levels and facilities.

GOOD PRACTICE

1. The aim of grouping children attending the day unit by age is commendable, undoubtedly making a positive contribution to the user experience. The value of a strong peer group as these children grow up is considerable.

2. There is active seeking of user opinion and use of patient feedback, with user comments being displayed on the ward. This appears to be a strong feature across the whole Trust.

3. The 'early years and young persons' protocol, developed by the community nurse specialist, is unusual and excellent.

4. The practice of copying every clinic letter to the community nurse specialist is good, and together with participation by that nurse in the weekly MDT, helps ensure proper communication between acute and community care teams.

5. Liaison with the newborn screening service is increasingly robust. Systems to ensure the failsafe tracking of all at risk pregnancies through to the newborn result and clinic attendance if needed are coming into place.

6. Brain MRI examinations in children with sickle cell disease are undertaken according to a minimum series recommended by the lead consultant, ensuring that adequate information is obtained and saving the need for repeat examination.

7. The use of stickers, alerting other hospital teams to inform the haemoglobinopathy team if the child is admitted, help towards ensuring that a child coming under the care of another team is seen also by the red cell team who can advise about specialist aspects of care.

8. Interpreters are readily available and in many cases have worked with the same families for many years, thus becoming real 'link workers'.

APPENDIX 1: MEMBERSHIP OF THE REVIEW TEAM

Dr Baba Inusa	Consultant Paediatric Haematologist	St Thomas' Hospital
Katherine Stevenson	Haemoglobinopathy Specialist Nurse	Manchester Royal Infirmary
Vicky Vidler	Nurse Consultant (Paediatric Haematology)	Sheffield Children's NHS Trust
Jessica Tudor-Williams	General Manager, Haematology	University College London Hospitals NHS Foundation Trust
Elaine Miller	Co-ordinator	UK Thalassaemia Society
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 some minor omissions – thalassaemia information was more limited, for example 1a for thalassaemia included only drug side effects. Part 2c was missing, as was 2f (though this is because there is no usual ward). 3c and e were not in the main patient information but are covered in 'transition' material. A good range of sources about how to access further information is included.*

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	*Specific information given to GP practices, apart from out-patient letters, was not found.*
3	Information should be available on transition to adult care. This information should cover all aspects of the transition (QR43).	Y	*A very good range of material, felt to be exemplary.*
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	*None for sickle cell affected children; UK Thalassaemia Society patient held filofax presented but it was unclear the extent to which this is in regular 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	*But facilities on the Day Unit are decidedly cramped and facilities are geared towards 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	*All thoroughly covered*
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	*The senior, band 7, nurse runs the Day Unit, and clearly functions at a very high level, but some aspects of the role of lead nurse – including responsibility for protocols, training, audit, liaison with local teams in linked hospitals – were not explicitly identified as being covered by this post.*
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	

Ref	Quality Requirement	Met?	Comment
14	There should be agreed cover arrangements for the outreach nurse specialist / counsellor.	N	*Flexibility of working between the liaison nurse and the community nurse allows this cover to work effectively for some patients although as the community nurse is employed by Heart of Birmingham PCT, she does not see patients outside that area.*
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	*Details are provided for all these services except Dental – subsequently informed takes place at the Dental Hospital - and contraception services – covered in transition material. Communication with some of the other specialist hospitals highlighted as problematic [see report]*
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	*Referral forms for each are presented, but in practice the psychology service is reported to be thinly stretched and access is not assured at all times needed.*
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	*There is no formal SLA, although there is an excellent protocol and evidence of good inter-team working*
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	*This is in place when the Day Unit Sister is present, but not when she is on leave; at those times cannulation is undertaken by one of the Oncology junior doctor. The 3 other day unit nurses are currently being trained to cannulate, which will then allow compliance with this QR.*

Ref	Quality Requirement	Met?	Comment
20	All staff involved in the care of children and young people with haemoglobinopathies should undertake regular child protection training.	Y	*Staff list provided, but some need repeat training [eg last attended training 2007, with 1 or 2 yearly training recommended]*
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	*But bears Barts and Royal London Hospital logo*
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	*Some are copies of relevant chapters of UK guidelines; thus complete but bulky and not easy to use for quick reference.*

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 currently written as a sheet describing current practice, rather than more formal protocol [see report]*

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	*Evidence of use in practice, on completed proforma in case notes*
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	

Ref	Quality Requirement	Met?	Comment
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	*Day unit nursing staff currently undertake, and ANP shortly to be in place to provide further support*
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	*Guidance for manual and automated exchange are presented*
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	*But as for 24*
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	*The brief chapter from UK standards for care is copied and highlighted for ease of use.*
37	A protocol should be in use covering arrangements for care between SHT and LHT for ongoing care. This protocol should ensure that, to facilitate effective shared care: a All patients have an up to date patient held record and details of their care plan. b The LHT and the patient's GP have received details of the patient's care plan.	N	*But outreach clinics are in place with all linked hospitals, and there are informal agreements about the division of care between BCH and these hospitals. BCH team are involved in the care of all long term transfused children. There are plans to establish more formal arrangements for shared care, also teaching and education for linked hospital teams.*
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 hand held record in use for sickle cell patients*
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	*But being developed, as for 37*
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	*And this is very comprehensive and patient focussed.*

Ref	Quality Requirement	Met?	Comment
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	*Transfusions currently 8 am – 4 pm 4 weekdays / week, but with plans to extend when re-locate. Plans for adolescent clinic 5 – 8 pm.*
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	*But as for 24 – sheet describing practice presented, rather than a protocol in regular use for reference.*
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	*This is very well covered in excellent material*
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	*Well evidence with feedback leaflets on the wards, 3monthly summary of feedback and actions resulting. Community champion for sickle cell and thalassaemia gained a Heart of Birmingham award 2009*
45	The SHT should run a programme of training and awareness of the management of patients with haemoglobinopathy for its main referring LHTs.	N	*But date set for first day meeting in October, room booked etc.*
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	*Minutes of meeting 19.11.09, well attended, and planned next meeting 4.10.10*
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	*Regular meetings now re-established, and minuted; liaison with screening is strong.*

Ref	Quality Requirement	Met?	Comment
	DATA COLLECTION and AUDIT		
50	The LHT / SHT should have audited compliance with key standards including: **Patients with sickle cell disease:** a Proportion of patients taking regular penicillin b Proportion of patients fully immunised against pneumococcus c Proportion of patients (HbSS and HbSβ^0) who have had Transcranial Doppler ultrasonography undertaken within the last year d Proportion of patients who have had their annual multi-disciplinary review within the last year e Effectiveness of action to contact families who have not attended for follow up appointments. f Review of the care of any patients who have died. **Patients with thalassaemia:** a Proportion of patients on chelation therapy b Proportion of patients who have had their annual multi-disciplinary review within the last year c Adequacy of recording of: ▪ Pre-transfusion Hb levels ▪ Regular monitoring of iron level ▪ Complications of iron overload ▪ Height / weight progression ▪ Spleen size ▪ Support for home chelation programme (QR40) d Effectiveness of action to contact families who have not attended for follow up appointments. e Review of the care of any patients who have died.	N	*Much data recorded and held on different data-sets, but none of the listed audits has yet been undertaken / reported.*
51	Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry.	Y	*Entry onto NHR is progressing well, with ~ 220 children so far entered.*

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	*No submission from Commissioners; discussion with specialist commissioner indicated that services currently commissioned by Heart of Birmingham as host PCT.*

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	N	*But close – 85% by 5 weeks, and longest 6/52*
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	*[first part of this standard ignored, duplicates P3 with different target]* *100% attend clinic by 3 months = excellent performance, across whole W Midlands*
P5	Timely confirmation of diagnosis for infants with a positive screening result**	N	*But there are plans to feedback to screening lead once confirmatory result is obtained; can then be monitored, and likely achieved.*
S1i	Failsafe to ensure ongoing care	Y	
S1ii	Up-to-date registers maintained of babies (cases) for which units are responsible	Y	*Database started from Jan 2010*

** 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.