UK Forum on Haemoglobin Disorders

WM QRS West Midlands Quality Review Service

Services for Children with Sickle Cell Disease or Thalassaemia at University Hospital Bristol NHS Foundation Trust: Bristol Royal Hospital for Children and the South West Region

Quality Review Visit Report

Visit date: April 28th 2010

Report finalised: December 2nd 2010

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Abbreviations in use in the report:

BMT	Bone Marrow Transplant
BRHC	Bristol Royal Hospital for Children
CNS	Clinical Nurse Specialist
OSCAR	Organisation for sickle cell anaemia research – a voluntary organisation
TCD	Trans-Cranial Doppler [scan to assess stroke risk in children with sickle cell disease
WTE	Whole Time Equivalent

INTRODUCTION

This report presents the findings of the peer review visit to services for patients with sickle cell disease or thalassaemia at University Hospital Bristol NHS Foundation Trust which took place on April 28th, 2010. The purpose of the visit was to review compliance with the '*Quality Requirements for Health Services Caring for Children and Young People with Haemoglobinopathies', 2009*. The visit was organised by the West Midlands Quality Review Service.

ACKNOWLEDGEMENTS

We would like to thank the staff of University Hospital Bristol NHS Foundation Trust: Bristol Royal Hospital for Children [BRHC] 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 and children 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 BRHC AND SOUTH WEST REGION

Service (as at April 2010)	Patient numbers sickle cell disease	Patient numbers thalassaemia	Patients on long term red cell transfusions
BRHC	27 [16 SS / $S\beta^o$] [11 SC / $S\beta^+$]	<5	<5
Gloucester	5 [SS]		
Exeter	<5		
Bath		<5	

BACKGROUND

Bristol Royal Hospital for Children, part of the University of University Hospital Bristol NHS Foundation Trust, is a large specialist hospital offering regional and supra-regional services for a number of paediatric specialties, including haematology, particularly haematological and other malignancies, haemophilia, and bone marrow transplantation. Red cell disorders constitute a relatively small part of the department's workload but patient numbers are growing. The patient

cohort is young. Fourteen children have been diagnosed since 2006, the majority through the newborn screening programme. There is a small and relatively stable number of patients with major thalassaemia syndromes.

Plans to provide services across the South West Region, mirroring those for paediatric malignancy, are in place and to some extent are already functioning in practice. At present, four of eight hospitals in the network do not have any patients with sickle cell disease or thalassaemia, three have a small number of patients and are developing links with the BRHC team. Derriford Hospital, Plymouth, shares care for children with paediatric malignancy, does not link functionally with BRHC for children with red cell disorders.

There are three paediatric haematology consultants, the third appointed early in 2010, and from this time the current lead consultant for these conditions took on this role. A staff grade doctor works exclusively on the Day Unit. There are now 1.5 specialist nurses for 'benign haematology' [red cell disorders and haemophilia]. The expansion from 1 wte was achieved in February 2010. There is 1 wte 'community clinical co-ordinator for sickle cell and thalassaemia', a post funded by Bristol PCT.

The department, as well as the range of general paediatric haematology presentations, cares for approximately 100 registered children with haemophilia [of whom 10 are severe] and about 80 new paediatric malignancy patients per year. About 40 allografts are undertaken annually.

Most required support services are in place and felt by the team to be satisfactory or good, the exception being access to a clinical psychology service.

Accident and Emergency

If children need urgent assessment and/or admission they present to A&E, where they are triaged usually as 'category 2' and should reach clinical assessment within 10 minutes. Analgesia for acute pain can be oral morphine, parenteral morphine, and sometimes intranasal diamorphine. What each child receives for first analgesia is detailed in a set of easily accessible individual 'care plans' kept in the same [hard copy] folder as the sickle cell and thalassaemia protocols. A full set of recently updated protocols is also available on the intranet.

Families are encouraged to phone ahead to alert staff that they are coming to A&E. Assessment is usually by the A&E team and the on-call paediatric haematology or oncology SpR. After 11 pm there is a 'hospital at night' paediatric team, including a paediatric ST3+. There is 24 hour cover by the paediatric haematologists in rota with paediatric oncologists. An e-mail is sent to the specialist

clinical team if a child is admitted out of hours, as a back up to clinical handover, so that their care will be picked up as appropriate early the next working day.

Time to first analgesia for children with all conditions presenting to A&E with pain was reported to be in line with the Royal College standard of over 90% within 20 minutes. Some families interviewed during the visit suggested that this standard was not always reached for children presenting with sickle cell crisis.

Transfer from A&E to ward takes place when the child is stable and a bed is ready. If there are any delays the child can be transferred to a small ward area adjacent to A&E, still under the care of the A&E team, until transfer.

OUTPATIENT CARE

Until recently, 'benign haematology' clinics for a range of disorders were held weekly, and the children with red cell disorders were seen here. However, in the recent past, transcranial Doppler scanning has been introduced in the clinic to assess stroke risk for the children with sickle cell disease and in order to reduce the number of clinics the sonographer attends, there is now a monthly red cell clinic into which these children are cohorted. This clinic is run by the lead paediatric haematologist, and SpR, one of the acute CNS nurses and the community co-ordinator. The medical records for children with red cell disorders, as well as those with malignancy, are held in this clinical area, and can be accessed by the site team 24/7 if a child is admitted through A&E.

DAY CARE

Day treatment including transfusions for children with red cell disorders takes place on a day unit shared with paediatric oncology and adult BMT services, which is adjacent to the out-patient clinic area. There are four cubicles, and two three bedded areas, one intended more for use by younger children and the other for older children / adults.

The unit is open 8am -6pm Monday to Friday, although if transfusions are ongoing after 6 pm the staff will stay until they finish, rather than moving the child elsewhere. They have been able to offer Saturday transfusions where it suited individual children. The CNS will often go to the child's home or school to take pre-transfusion blood samples to save a hospital visit for the family. Regularly transfused children attend clinic every two – three months, and are pre-assessed for each transfusion, and cannulated, by the dedicated day service staff grade doctor, covered in his absence

by one of the paediatric haematology specialist registrars. Nursing staff will establish venous access for the relatively few children with 'port' devices.

Children mostly lie on beds for their transfusions but there are suitable armchairs if they prefer. There is a treatment room where procedures are carried out, and where parents can be taught on a 'dummy' to access a port device. Play facilities are available in the Day Unit and in Clinics, and play specialists offer normalising as well as specialist / procedure-related play input.

IN-PATIENT FACILITIES

There is an in-patient ward, (34), designated for the care of children with paediatric haematology / oncology diagnoses up to the age of 10 to 12 years. An adjacent ward (35) cares for older children, usually from 12 up to 17 or 18, including all specialties. The standard of ward facilities for children and parents is high.

Those of the appropriate age with sickle cell acute presentations can usually be accommodated on ward 35, but for the younger children bed pressures often mean that they are cared for on one of the other medical wards. In-patient admissions for these children are infrequent, reflecting the relatively small patient numbers, around one admission every one or two months. On-going analgesia often takes the form of patient controlled analgesia [PCA] and the input of the Pain Team, lead by a consultant anaesthetist and nurse, is valued.

COMMUNITY SERVICES

The hospital-based CNSs do some outreach work in the community, visiting schools to take pre-transfusion samples from children, for example, but the majority of the community services are offered by a single-handed 'clinical co-ordinator', who works across children's and adult services, and comes to the children's outpatient appointments at BRHC. Although PCT-funded, she is not constrained by specific geographical boundaries and offers a consultation service for the whole region. She receives the results of the newborn screening programme, in parallel with the GP, the BRHC lead paediatrician, and the health visitor, and it is her responsibility to visit the home of affected newborn children to give the diagnosis and early education and support. She takes on a range of other activities, some which might be undertaken by an effective local voluntary organisation. There is a branch of OSCAR in Bristol, but it was not clear to the team exactly what its two part-time staff, who are PCT funded, offer to the service. It is reported not to cover children's services, and there is no 'support group' for children and families, which was identified by service users as well as providers to be an important gap.

COMMISSIONING ARRANGEMENTS

A member of the SW specialist commissioning team and two members of the local management team came to meet a member of the review team. On discussion, it was clear that there is understanding about the configuration of services for this patient group, and the intended network arrangements which, to a large extent, mirror those in place for oncology. Red cell disorders are not on the specialist commissioning team's work programme, which is not surprising considering the relatively low prevalence and other competing priority areas. However, there was a good level of interest and commitment to supporting and continuing to develop these services.

LINKED HOSPITALS

Numbers of patients attending linked hospitals outside Bristol are small. The intention is to include those at Gloucester, Exeter and Bath in the consultant outreach clinics already taking place at these hospitals for children with malignant disorders, on a quarterly basis. Any children presenting to or born in or near the other linked sites could be included as necessary. TCDs are likely to require the children to attend BRHC.

None of the local teams attended the visit, although Gloucester Royal Hospital had submitted some brief written evidence and a member of the review team had a telephone conversation with the lead paediatrician there. Referral pathways have historically been to Birmingham, but recently identified newborn have been linked instead for specialist care to the BRHC team. The intention is for this to continue for any additional new patients. Although the local team have not been using the BRHC protocols and guidelines, they have recently received these and are intending to use them. Clinical advice for the local team about individual patients, between outreach clinics, was reported to be easy to access from both the Birmingham and Bristol clinical teams. Transfer of a patient had not been necessary in the paediatrician's time in post in Gloucester.

The fact that a small number of patients are managed in Derriford Hospital, Plymouth, without any apparent link to a specialist service or team elsewhere, was felt to be of some concern.

REVIEW VISIT FINDINGS

ACHIEVEMENTS

1 The clinical and management team at BRHC are to be commended on how much they have put in place for this relatively small, although growing, patient group. The services are developing in a very positive way, with new appointments recently being made to the medical and nursing teams, and a trans-cranial Doppler screening service for stroke risk assessment having recently started. It is a team with many obviously committed and enthusiastic staff, and the community co-ordinator nurse role, and her contribution to the service generally, was felt to be very strong. There is good team working and flexibility especially between nursing roles. There are also strong working links with support departments, for example the A&E team, and the efforts of the core team in building these links is to be acknowledged.

2 Services in general are much appreciated by users, and the efforts of the nursing team generally are highly valued.

3 The Day Care service is impressive in its flexibility of working for the needs of this relatively small patient cohort. The efforts made to offer best services across age groups, and patients with different conditions, are impressive.

4 The facilities in general are of a high standard, with excellent provision for older children and adolescents.

5 Many of the written protocols and guidelines now in place are outstandingly strong, clear and comprehensive. These include transfusion, including that for exchange transfusion, annual review for thalassaemia, referral guidelines for bone marrow transplantation and thalassaemia intermedia.

IMMEDIATE RISKS

1 There were no immediate risks identified.

CONCERNS

1 There is no written 'did not attend' [DNA] policy, although core team members could describe what happens in practice. Given the importance of ensuring children are not lost to follow up, and thus not accessing appropriate preventative and screening measures, the process should be formalised and recorded.

2 There are no audits of practice against the key care standards. It is not possible to be certain, therefore, what percentage of children are receiving or are compliant with, the main aspects of care, for example regular penicillin prophylaxis, all recommended immunisations.

3 Psychology input for these children and families is not available. Although a dedicated psychologist is unlikely to be justified by the size of the service, it should be possible for this team to refer to a psychologist covering another part of children's services.

4 No support group is available for the families of children with red cell disorders. There is some confusion in the minds of the user families as to the roles of the community co-ordinator compared with the voluntary organisation OSCAR. In practice, the latter appears to offer little support for affected children or their families, and the community co-ordinator was, to some extent, filling this gap. These comments apply to the sickle cell children and families, not to those affected by thalassaemia who reported feeling well supported by the UK Thalassaemia Society, remotely from London, and by each other.

5 The lack of cover arrangements for the community co-ordinator nurse is of concern. For example if she is on leave, there may be delays in newborn screening results being communicated to families, who then lack the early education and support she offers. The community provision overall appears to be thinly stretched and families do not always feel that school visits occur as they should.

6 For out of hours periods no paediatric urologist is available to help manage acute fulminant priapism three nights and weekends in five. The arrangements for who should be called instead if required at these times are not clear. Given the potential damage which results from delay in treating priapism, these arrangements should be explicit.

FURTHER CONSIDERATION

1 Work with commissioners will help to formalise and further develop the network, to ensure that services are covering the whole population. Links with other hospitals are developing

but have further to go, for example, formalising use of shared protocols and guidelines. Although only a few children are managed in Plymouth, the lack of arrangements for them to link with any specialist team should be addressed. While recognising the challenge for commissioners of services spanning the border into Wales, there might be benefit in some joint working with the team in Cardiff.

2 Although dedicated clinical and educational meetings with linked hospitals would not be justified by the size of the outlying patient numbers, the red cell services and issues could usefully be considered at a session during the educational and other network meetings in place for paediatric oncology services.

3 As Bristol patient numbers are relatively few, some clinical activities, for example management of some of the acute complications in hospital, are necessarily low volume. There may be education and training benefit in team members, especially junior doctors and nursing staff, spending time attached to another larger Centre to extend their experience.

4 The process for smooth transition from children's to adult services is not yet established but there are plans in place to address this. One parent who met the team commented that she would like more parental involvement in this process.

5 The host team did not hold information which would allow them to check compliance with the 'screening standards' relating to management of newborn: the time by which the family are informed, are referred to clinic, are first seen in clinic, and are prescribed penicillin. The screening laboratory at Southmead Hospital apparently requests and monitors this information. The BRHC team might usefully ask to share these results.

6 The profile of these services in all the clinical areas is low, with no visibly displayed patient information leaflets or posters. Some of the materials available for this patient group might helpfully be visible in these areas.

7 There were some comments raised by service users regarding care in A&E. In particular, service users felt that time to first analgesia could frequently exceed the 20 minutes the A&E staff feel they are achieving and is more usually about 45 minutes. They also felt that sometimes staff in A&E are not experienced in accessing central venous port devices but try to do so, which can cause distress.

GOOD PRACTICE

Many of the observed good practices are encompassed in the overall 'achievements' earlier in the report. However, some additional specific aspects of practice were felt to be noteworthy:

1 Play input to the services was very strong. In particular, there was sufficient provision to offer 'normalising play' as well as specifically targeted or procedure-related input to support children with difficult aspects of their management.

2 The e-mail generated by A&E staff regarding children admitted out of hours to ensure early pick up by the specialist team is an excellent insurance against incomplete clinical handover.

3 The CNS's outreaching into the community and visiting schools to take routine blood samples is an unusual provision. This is helpful in minimising children's time out of school and reducing hospital visits for families.

4 The team remarked on the fact that refreshments were freely available for children and families from small 'trolleys' in the clinical areas, and the idea of 'treasure boxes' from which children could choose a small reward item after having a difficult or uncomfortable procedure. Both are novel ideas, felt likely to help improve the experience of care.

5 Pre-printed 'stickers' for prescribing of drugs for children of different body weights were noted on the in-patient ward. These are in use across the hospital and are an innovative and excellent way of reducing dose prescribing errors.

6 A recent small user satisfaction survey was well displayed and useful, and the 'follow up' actions taken were exemplary. Findings had been mailed to service users and a meeting was held to discuss the findings and ways of responding constructively to them. The enthusiasm with which user families had engaged with this process, and had wanted to continue the group discussion, highlights their need for a support group which, hopefully, can be established after discussion with the voluntary organisation OSCAR and Bristol PCT.

APPENDIX 1: MEMBERSHIP OF THE REVIEW TEAM

Dr Jo Howard	Consultant Haematologist	Guy's and St Thomas' NHS Foundation Trust
Natalie Lawson	Senior Sister	Birmingham Children's Hospital NHS Foundation Trust
Teresa Warr	Head of Service Development	South Central Specialised Services Commissioning Group
Asaah Nkohkwo	Chief Executive	Sickle Cell Society UK
S Kay Oliver	User Representative	
Dr Anne Yardumian	Clinical Lead for Peer Review Programme	

APPENDIX 2: COMPLIANCE WITH QUALITY REQUIREMENTS

Specialist Haemoglobinopathy Teams

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

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	*Clinic letters to GP's cover key clinical information but do not cover all the sections of QR1.*
3	Information should be available on transition to adult care. This information should cover all aspects of the transition (QR43).	N	*But clear plans are in place and a draft pathway was presented*.
4	The SHT and its linked LHTs should have agreed a patient-held record for recording at least: a Information about the patient's condition b Current management plan c Regular medication d Named contact for queries and advice e Alternative contact for times when key contact is away.	N	*No hand held records are in current use but copies of clinic letters sent to parents allows for some hand held clinical updates.*
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 good for children of all ages.*
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	*Name provided, and CME record including general haematology conferences / updates which include some red cell sessions.*
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 post-holder was new in post at the time of the visit, and had not yet undertaken specific training course although it was planned, and there was cover and support from a colleague with much experience and relevant training.*
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.	N	*As above*
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	*Details given in presentation at start of the visit.*
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	*Although criteria for admission were not found as a separate guideline, they were included in the individual sub-sections of clinical guidance [chest syndrome, deteriorating patient, etc] so QR is met.*
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	N	*Some support services are very strong eg play specialist input, but no access to psychology for these children is a gap in service.*
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	*No SLA in place but there is evidence in practice of good flexible working between the acute and community based specialist nurses. NOTE: although not a specific requirement of this QR, the lack of cover for community nurse when on leave is an issue.*
19	A nurse with competency in cannulation, starting and supervising a transfusion should be available at all times at which children are attending for transfusion.	Y	*Although nurses on the day unit do not cannulate, there is a 'resident' staff grade doctor who undertakes this role, so in practice the requirement is met.*
20	All staff involved in the care of children and young people with haemoglobinopathies should undertake regular child protection training.	Y	*No child protection records were seen at the visit, but were provided subsequently.*
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	*Very good guidelines overall.* *How to manage fulminant priapism in the absence of paediatric urologist should be clarified.*

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	*Good guidelines.* *Only lacking guidance about penicillin alternatives for children who are allergic to penicillin*.

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	*Good guidelines.* *Only lacking management of complications of splenectomy.*
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	*Gap in service; highlighted by team and at variance with current good practice guidance.*

Ref	Quality Requirement	Met?	Comment
27	Guidelines for referral for consideration of bone marrow transplantation should be in use.	Y	*Excellent guideline.*
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	*Lacking only guidance that patients should not have to wait for more than 1 hour for transfusion to be started, after arrival.*
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	*And useful monitoring sheets were presented, seen to be in use in medical records.*
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	*These were felt to be excellent.*
33	Clinical guidelines should be in use covering: a Indications for exchange transfusion b Arrangements for carrying out an exchange transfusion.	Y	*These are also very clear and comprehensive.*
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.	N	*As no patient held record, also no protocol was seen for care arrangements between centre and linked hospitals. Clinic letters are appropriately copied to linked hospital.*
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	*As above, although b is met by parents being sent copies of clinic letters.*
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	*This is met in practice, for the small number of patients who attend the centre and a linked hospital [medical record review].*
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	*Good, practically useful material was presented.*
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	*This was felt to be met in practice by the flexibility of opening of the day unit for transfusions, and the facility for pre-transfusion blood sampling to be taken by outreach nurse visiting school.*

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	N	*No written protocol was in place, although the team could describe the process in practice.*
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.	N	*Not yet in practice, although plans to introduce this are well advanced.*
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	*Although part c is an issue recognised as a challenge at present.*
45	The SHT should run a programme of training and awareness of the management of patients with haemoglobinopathy for its main referring LHTs.	N	*Although might be made a part of the network meetings currently in place for malignant haematology services.*
47	The SHT should meet at least annually with its referring LHT teams to: a Identify any changes needed to network-wide policies, procedures and guidelines b Review results of audits undertaken c Review any critical incidents including those involving liaison between teams d Consider the content of future training and awareness programmes (QR45)	N	*As 45*
49	The SHT should meet at least annually with representatives of the neonatal screening programme to review progress, identify issues of mutual concern and agree action.	N	*And might be useful as the screening service operates from a different Trust.*

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	*No audits of compliance with core standards seen, team confirm none undertaken at present.*
51	Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry.	N	*There is a local electronic list of patients seen at BRHC.*

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	N	*Although quite close in most cases recently.*
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) by 8 weeks and first attendance by 3 months of age	Y	*Figures sent after the visit show these standards were met for the small number of new babies seen in the preceding 18 months.*
P5	Timely confirmation of diagnosis for infants with a positive screening result**	N	*No evidence seen*
S1i	Failsafe to ensure ongoing care	N	
S1ii	Up-to-date registers maintained of babies (cases) for which units are responsible	Y	*But covers BRHC babies only.*

** 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.