UK Forum on Haemoglobin Disorders

WM QRS West Midlands Quality Review Service

Services for Children with Sickle Cell Disease or Thalassaemia
At
Nottingham Children's Hospital, Nottingham University Hospitals NHS Trust

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Quality Review Visit Report
Visit date: January 26th 2011

Report finalised: 21st May 2011

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INTRODUCTION

This report presents the findings of the peer review visit to services for children with sickle cell disease or thalassaemia at NHS Trust, which took place on January 26th, 2011. 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 Nottingham University Hospitals NHS Trust [NUH], 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 NOTTINGHAM UNIVERSITY HOSPITALS NHS TRUST

Service (as at Jan 2011)	Patient numbers sickle cell disease	Patient numbers thalassaemias	Patients on long term red cell transfusions
Attending NUH	33	9 including HbH, BTI and transplanted BTM	7
Derby	13	<5	<5
Lincoln	<5		

Nottingham Children's Hospital has a separate identity within the large University Hospitals complex on the Queen's Medical Centre site. It forms part of the Family Health Directorate. Previously, children's services were split between this site and the City Hospital site, but in recent years children's services have been brought together here. There are 111 children's in-patient beds in total.

The haemoglobinopathy service is led by a paediatric haematologist, with a recently appointed second paediatric haematologist contributing largely to the service. Adult haematology services remain at the City Hospital site, but the adult haematologist with an interest in red cell disorders takes an active part in the paediatric service, with clinical input and cover also from other paediatric colleagues. A part time clinical nurse specialist post for the children's haemoglobinopathy service, for which funding was identified some time ago, has not yet been appointed to. There is currently no specific non-clinical manager responsible for this service although in preparation for the visit the help of a manager, whose remit is the integrated children's cancer services between Nottingham and Leicester, was very helpful.

Accident and Emergency

There is a separate children's Emergency Department, and children requiring acute care present there directly. A generic pain protocol was available. There did not appear to be a specific triage pathway for these children, and user families reported that there can be substantial delays before children receive analgesia and are admitted to the ward area. Average waits were reported to be an hour or more. No audits were reported giving a record of the range of times children waited for treatment.

Outpatient Clinics

Children with sickle cell disease or thalassaemia are seen in a morning non-malignant haematology clinic which takes place monthly. Outpatient facilities are good with a separate waiting area for teenagers, and there is a play specialist available during all clinics. Clinic nursing staff take children's blood samples, according to a pre-clinic discussion about need. Clinic is run by the paediatric haematologists, the adult haematologist, and attended also by a community nurse specialist and psychology assistant. Trans-cranial Doppler stroke risk assessment scans are undertaken by a Neuroradiologist in the clinic where possible, and there are plans to establish a separate monthly haemoglobinopathy clinic partly so that this service can be focussed on a session when most sickle cell children attend. There are currently no formal out-of-hours clinic sessions, but children can be seen at request on Day Care after clinic hours.

Transition to adult services for outpatient, day care and in-patient managements takes place flexibly, usually at 18 years. The transition process had not received much focus to date but is included in the remit of a psychology project [see below].

DAY CARE

The day care area, shared with paediatric oncology and other haematology services, is adjacent to the in patient ward, and nursing staff move flexibly between the two. The clinical area is pleasant and open. It is open 8 a.m to 8 p.m, Monday to Friday, allowing for after school transfusions. There are clinic rooms attached to the clinical area for private patient review. Notes for children with haemoglobin disorders, and others frequently attending, are held here. Some children have in-dwelling port devices for venous access, and the nursing staff can access these. Other children have to wait for a paediatric doctor to come to site intra-venous access, and this can engender delays. When an acute nurse specialist is appointed, this will be one of his / her duties.

Automated red cell exchange for planned or urgent transfusions was not available, as there was no machine on site to undertake this although some personnel from a different specialty would be able to offer the service if capital could be found.

IN-PATIENT FACILITIES

Children with red cell disorders are currently managed on a designated children's ward, shared with oncology, and bed availability is such that it is unusual for children to have to use other wards. The nursing team, some of whom have been in post for many years, know frequently attending children well. Paediatric haematologists and oncologists operate a weekly rotation for care of all in-patients, undertaking ward rounds every day including weekends, and twice a week all the consultants meet to review in-patients. Frequently the paediatric haematologists will visit any of their in-patients additionally when they are not on duty. Day to day care is provided by middle grade staff in paediatrics and paediatric haematology.

Efforts are already made to cohort older children into one ward area, and a ward expansion including a Teenage Cancer Trust unit is planned, adjacent to the current day care area. It has been agreed that whenever there is space all suitably aged children, not just those with malignant disease, can be cared for there.

A Pain Team has input into the management of children with sickle cell pain crisis, frequently using nurse controlled analgesia pumps. Although it was acknowledged that the requirements of these children can differ from those who are using strong analgesia for the first time, for example after an operation, there was no specific written guidance for analgesic doses titrated against pain scores, or for dose reduction, for these children.

Support Services

Psychology input to the team, previously limited, is now receiving considerable focus, with an assistant psychologist, working under supervision, undertaking a 12 month project to look at service needs and tasked with recommending what continuing requirement there would be. She is also offering some clinical services in parallel, including some behavioural approaches to pain management and some neuro-cognitive assessments although these are not systematic at present.

Specific social work support is not currently available for this service.

There is an excellent school facility on site, Ofsted rated 'outstanding' with a large pleasant out-door play area attached to it.

A 'complementary treatment' centre is based close to the ward, run by a children's nurse, and is freely available to children and parents.

Community Services

These are based at a multi-professional community centre, and are very active. They are run by almost 3 full time equivalent nurses and a clerical assistant, offering adult as well as paediatric services. As well as managing newborn screening results and visiting families in the early weeks, the team attend clinic at NUH routinely, and follow up children after discharge from hospital. Regular meetings with antenatal and neonatal screening laboratory are not currently in place, to ensure no identified affected baby has been 'missed', and although screening standard data were given for Nottingham and Leicester combined, there were no specific data relating to the Nottingham service. The community team are highly valued by service users and have been energetic in their efforts to gain feedback on services, having held a number of well attended events for user groups. A regular support group was not currently running because of poor attendance. The community services were reported to be likely to come under NUH management in the near future.

User Feedback

The user families who met the visiting team were generally appreciative of the services. One parent commented that he had come to the meeting particularly to express his gratitude. Acute and community services were highly valued, except that there were significant concerns expressed about the timeliness of treatment in the Accident and Emergency Department. For children attending with sickle cell pain crisis, waiting time was reported to be an hour or more.

LINKED HOSPITALS

There are some affected children living in Derby, who are served by a 6 monthly outreach clinic run by the Nottingham team. Currently Derby children with sickle cell disease come to Nottingham for their TCD screening service, but outreach with a mobile scanner is planned. The Derby consultant paediatrician reported that she was happy with the level of access and support she receives. A small number of children living in Lincoln inreach to the service at NUH. There are also operational links with the haemoglobinopathy service at the Leicester Centre, as part of a formal network established by the East Midlands Specialised Commissioning Team in 2009.

COMMISSIONING ARRANGEMENTS

The East Midlands Specialist Commissioning Group has been very effective in bringing focus on these services in the region, hosting network meetings, and providing pump prime funding for key posts. This was initially in response to concerns expressed by local clinicians at the failure to offer some basic services, such as trans-cranial Doppler stroke risk assessments for children with sickle cell disease. The Group continues to work with providers on network development, and clear monitored plans with timelines are in place. Some staff, for example the data manager, work across the East Midlands network although the visiting team gathered that the focus has been particularly on the Leicester services, and more time offered to the Nottingham team would be valuable. There was some frustration that a part-time hospital based nurse specialist at NUH had not yet been appointed to the post despite funding having been offered some time previously. Meeting held every two months, at different sites in the region, are chaired by the adult haematologist at Nottingham, and were intended to facilitate sharing of clinical expertise, and allow for case review and educational discussions as well as address operational issues, although the opportunity for clinical liaison had not been greatly utilised. The intention had been for the network to share guidelines but the two centres had not agreed a set they both found appropriate.

REVIEW VISIT FINDINGS

ACHIEVEMENTS

1 This hospital team is made up of some experienced long-serving members, together with some energetic new members, and hopefully one or more additional members to be appointed in the near future. It is committed and enthusiastic, and well integrated with community services, and it is offering good standards of care. The service is well respected by colleagues and appreciated by its users. Despite its relatively modest size, it has a high profile among hospital staff. It is well supported by a range of colleagues, for example in psychology and neuroradiology

2 The community nursing team is highly committed and effective, and has been helping to plug a gap in acute specialist nursing provision. The team has pulled together a lot of strong informational material for users and their carers, and communicates fully with parents and primary care colleagues about clinical matters. Efforts in canvassing user feedback to improving services accordingly have been outstanding.

3 There is an unusual degree of support from the East Midlands Specialised Commissioning team, taking a wider regional view as well as providing specific funding for key posts, and network development is progressing steadily.

IMMEDIATE RISKS

No immediate risks were identified.

CONCERNS

1 The management of acute pain in children with sickle cell disease needs some attention. There is no specific written pain management guidance. Children are treated, with input from a 'pain team' according to generic guidelines although it was acknowledged that their needs can be different. Specific written guidance about timing, type and doses of analgesia in Accident and Emergency and then on the ward, with renewed training of A&E and ward staff in this regard, is likely to lead to more consistent care.

2 User families expressed dissatisfaction and sometimes frustration at the delay from time of arrival in Accident and Emergency to first analgesia for their children presenting with acute sickle cell pain. Once specific written guidance is in place, an audit against this will provide assurance that waiting times are appropriate.

3 The failure to appoint a clinical nurse specialist, despite funding having been made available specifically for the part-time post, is concerning. The review team were informed that this was now 'signed off' and it is hoped that the process for appointment will move swiftly.

FURTHER CONSIDERATION

1 No local figures were available to assess compliance with the newborn screening programme standards.

2 There did not appear to be a system of assurance that all babies identified by the newborn screening programme were safely conveyed into the clinical services. It was thought that the newborn screening laboratory checked that this was occurring, but the community team did not have specific knowledge of this.

3 The service would benefit from more clarity about local management responsibility, which appears currently to be lacking.

4 The facility for offering automated red cell exchanges would be beneficial in terms of the speed of undertaking these procedures and in reducing iron load, and this would need capital investment.

5 Some aspects of network functioning could usefully be addressed, for example increasing the proportion of time the network data manager spends with the Nottingham team, and increasing the time spent on clinical discussions and shared case reviews at the network meetings.

6 Some potentially key posts are currently fixed term funded, including the acute nurse specialist and assistant psychology posts, and the team will need to focus on outcomes from the work undertaken by these post-holders, to support application for longer term funding.

7 Some committed time from a social worker would be very beneficial.

GOOD PRACTICE

1 Some specific patient and family information material was outstanding, including: a 'red amber green' alert sheet to guide families what symptoms they might safely manage at home and what should lead them to seek urgent advice, a thermometer card to keep in a baby's cot which had full contact details for the community centre team, information leaflets for parents, teachers and

carers for sickle cell and for thalassaemia, a ‘survival pack for teenagers’ and other general health promotional materials.

2 The flexible age at which patients could transfer to the adult services, up to 18+ years, offers individual choice, and although the process for transition is still not fully established, the plans and draft material to support the move are excellent.

3 There is a sense across the children’s hospital that efforts are made to focus on the needs of older children as well as younger children. For example there is a separate out-patient waiting area for those aged 11+, a dedicated ‘youth room’ for use of in-patient teenagers, and a monthly support meeting held for this age group. In-patient staff already try to cohort teenage children in one part of the ward, and this facility will be enhanced by the development of a Teenage Cancer Trust unit which children with other disorders will also be able to access.

4 The adjacency of the clinical day care unit with the in-patient ward, with a nursing team working flexibly across the two, is helpful in terms of continuity of care by familiar and experienced staff.

5 The haemoglobinopathy service ‘Operational Policy’ is excellent and comprehensive, although noted to be quite new, and how effective it is in supporting services will depend on how it is now used in practice. It should be available in all the clinical areas, alongside the clinical guidelines.

6 The out-patient facility is strong, in pleasant surroundings which show attention to children’s preferences. Out-patient nursing staff take blood samples on site, and there is good play provision. There is a facility to hold out-patient review with children at the time they attend for day care, which can avoid duplicate visits.

7 The system of a named ‘blood link nurse’ in each clinical area allows cascade training and competency assessment for nurses undertaking blood transfusions without the need for the hospital transfusion practitioners to visit all wards. Detailed records of training and assessment were seen in this clinical area.

8 Some additional facilities were worthy of note, including the excellent hospital school, and complementary treatment centre for children and families.

APPENDIX 1: MEMBERSHIP OF THE REVIEW TEAM

Dr Phil Darbyshire	Consultant Paediatric Haematologist	Birmingham Children's Hospital NHS Foundation Trust
Sandra Oliver	User/Carer	Glasso Afoofa Publishing Ltd
Ravinder Raj	User Representative	Sickle Cell & Thalassaemia Support Project, Wolverhampton
Vicky Vidler	Nurse Consultant (Paediatric Haematology)	Sheffield Children's Hospital NHS Foundation Trust
Neil Westerdale	Advanced Nurse Practitioner Haemoglobinopathies	Guy and St Thomas' NHS Foundation Trust
Dr Adrian Williams	Consultant Haematologist	Bradford Royal Infirmary
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	*There was a wide range of very good material.*

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	*And it was noted that an unusual amount of useful communication took place between the community nurses and GP's.*
3	Information should be available on transition to adult care. This information should cover all aspects of the transition (QR43).	N	*In development, and draft material seen was good.*
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	*A Leicester record was in the process of being adapted for local use.*
5	The locally agreed patient-held record (QR4) should be in regular use within the LHT / SHT.	N	*As for 4*
6	Services should be provided in a child friendly environment, including toys and books / magazines for children and young people of all ages.	Y	
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.	N	*There is a lead paediatrician and deputy, who work effectively in these roles, but no CPD records were made available.*
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.	N	*See comment above.*
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	*Although there was funding available for this post, it had not yet been advertised or appointed to.*
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	*Mostly provided by the community specialist nurses working across the acute and community boundary.*
14	There should be agreed cover arrangements for the outreach nurse specialist / counsellor.	Y	*As for 13*

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	*Full details and named contacts included in Operational Policy.*
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	*But no specific social work input was available for this 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.	Y	*This was not seen but was reported by the community team to be in place. The team is due shortly to transfer from PCT to NUH management.*
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	*And this can lead to delays in starting transfusion on the Day Care unit. Once appointed, the acute nurse specialist is expected to undertake this role.*
20	All staff involved in the care of children and young people with haemoglobinopathies should undertake regular child protection training.	N	*No records of doctors' child protection training were available.*
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	Guidelines should be in use covering possible acute presentations including, at least: **For patients with sickle cell disease:** a Fever and infection including major sepsis b Acute pain c Acute anaemia d Stroke and other acute ischaemic events e Acute chest syndrome f Acute splenic sequestration g Abdominal pain / jaundice h Priapism i Changes in vision, including urgent referral for an ophthalmologic opinion j Indications for 'top-up' and for exchange transfusion and practical protocol for undertaking exchange transfusion **For patients with thalassaemia:** a Fever and infection including major sepsis b Unexpected cardiac, hepatic, endocrine decompensation. These guidelines should ensure that patients are transferred to the paediatric ward for assessment as quickly as possible	N	*Although many guidelines are in place, some were judged to be rather sparse, and there is not sufficient guidance as to pain management for children with sickle cell pain crisis.* *Thalassaemia guidelines were appropriate.*

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	*Lacking - only reference to recording oxygen saturation in clinic.*

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	*And proformas for completion at the review visits were seen for sickle cell disease and thalassaemia.*

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)	N	*This is expected to be an output from the recently started psychology project.*
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	*Including good practical guidance for undertaking manual red cell exchange.*
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	
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	*Work in progress.*
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	*Work in progress.*
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	N	*Although no patients currently receiving desferrioxamine, it would be useful to have guidance for use if / when a new child requires this.*
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	*Although there are no formal out of hours clinic appointments, children can attend and be seen later on Day Care unit. Out of hours phlebotomy and transfusions are available.*

Ref	Quality Requirement	Met?	Comment
42	A protocol should be in use covering: a Follow up of children who do not attend b Communication and follow up of children who move to another area	Y	*There is a formal hospital DNA policy. The service's DNA handling, and guidance for communication about children who move out of area, is detailed in the new Operational Policy.*
43	A protocol should be in use covering transition to adult care. This should ensure: a Age guidelines for timing of the transfer. b Involvement of the young person in the decision about transfer. c Involvement of primary health care, social care and adult services in planning the transfer. d Allocation of a named coordinator for the transfer of care. e A preparation period and education programme relating to transfer to adult care. f Communication of clinical information to the adult services. g Arrangements for monitoring during the time immediately after transfer to adult care.	N	*But there are plans for progressing this, and draft written materials are good.*
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	*There is evidence of extensive involvement of users and carers in highlighting issues about care, very strong use of 'patient stories'.*
45	The SHT should run a programme of training and awareness of the management of patients with haemoglobinopathy for its main referring LHTs.	N	*Minutes of meetings were seen but did not evidence training aspects of discussions.*
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	*Not all elements of this QR were evidenced in the minutes presented.*
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	*Self assessment indicates yes, and the meeting frequency meets requirements, but the community team did not seem aware of these meetings.*

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	*Not all complete although key clinical sickle cell audits are in place for the Region in conjunction with the newborn screening laboratory. Local compliances not presented. None for thalassaemia.*
51	Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry.	Y	*This has now started.*

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.	Y	*Input of local specialised commissioning group, and understanding of local services is unusually high level.*
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.	Y	*As for 52*

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	*Regional data presented showing good compliance but not possible to extract local data from this; requested after visit but no figures submitted. Compliance for this centre therefore uncertain.*
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)	N	*[the first part of this standard overlaps with P3, and has been ignored]* *As for P3*
P5	Timely confirmation of diagnosis for infants with a positive screening result**	N	*As for P3*
S1i	Failsafe to ensure ongoing care	Y	
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.