UK Forum on Haemoglobin Disorders

WM QRS West Midlands Quality Review Service

Services for Children with Sickle Cell Disease or Thalassaemia at St George's Healthcare NHS Trust, London

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Quality Review Visit Report
Visit date: April 22nd 2010

Report finalised: 30th July 2010

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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 St George's Healthcare NHS Trust which took place on April 22nd 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 St George's Healthcare 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 ST GEORGE'S HEALTHCARE NHS TRUST

Service (as at 2010)	Patient numbers sickle cell disease	Patient numbers thalassaemia	Patients on long term red cell transfusions
SGH	210	5	6 [sickle] 5 [thal]
Queen Mary @ St Helier	34		<5
Kingston Hospital	10	<5	<5

St George's Hospital, now St George's Healthcare NHS Trust [SGH,] has a long established reputation for the treatment of red cell disorders in children. There is now a developing network and linked hospitals in the South West London Network for these disorders are Queen Mary at St Helier and Kingston Hospital. Some patients from Croydon who attend Mayday Hospital also attend SGH. As Mayday is having a separate peer review visit, the shared SGH / Mayday services were not reviewed at this visit and a subsidiary report may follow. Some senior medical leads left SGH in recent years

and there have been a series of locum consultants in post, both in paediatrics and adult haematology. Approximately one year ago, two substantive paediatric consultants were appointed who have energetically assumed the roles of clinical lead and deputy for sickle cell and thalassaemia services. There is strong nursing leadership, with a very experienced and involved Day Unit sister, consultant nurse, ward sister and a CNS in paediatric haematology/oncology who offers support. There is no dedicated red cell haematology CNS at present. Community services in Wandsworth are led by a very senior and experienced lead nurse counsellor, and she and her team work effectively across acute and community boundaries.

There is a full range of specialty services available on site, including PICU. Children with red cell disorders are referred to off-site specialty teams only for cardiac assessment for iron overload, nephrology and hepatology.

Accident and Emergency

Children do not usually access services via A&E as there is direct access to the in-patient ward at all times. Occasionally a patient will present to A&E, and children brought by ambulance will be taken there. A full set of clinical guidelines is available in A&E, and staff the team spoke to at the visit indicated that if a child arrived there not needing resuscitation, they would be directed straight to the ward. In any event they would contact the paediatric team immediately for advice / assessment.

Outpatient, Day Case and In-Patient Facilities

A morning clinic on alternate weeks for children with sickle cell or thalassaemia takes place in a general paediatric outpatient area. About 20 children attend and the clinic is run by two consultants. Regularly transfused children attend clinic approximately three monthly. A monthly afternoon clinic has been running for transition-age children, of whom there are many [83 children aged 13 to 18]. From early summer, once a ‘backlog’ is cleared, it is intended to reduce these to quarterly sessions run jointly by paediatric and adult teams, at which individual care plans will be agreed. This clinic will be followed by a tour of the adult facilities, then a ‘workshop’ including some education and entertainment. It is hoped that input from a charitable organisation offering complementary treatments can be included.

Children attending clinic can have blood samples taken at the clinic, or in the general phlebotomy area which is less suitable. There are plans to appoint a full time phlebotomist for children in the

clinic. This would reduce pressure on the Day Unit, to which hospital children also sometimes go for blood tests.

Jungle Day Unit is a 12 bedded ward area with a play area, small sitting room, small kitchen area and procedures room. It is open from 7 a.m to 8 p.m. Monday to Friday. It caters for medical and surgical day cases, mostly elective, accommodating up to 60 children per day. It is managed by an experienced senior sister, and she and many of the other staff cannulate, as well as pre-assessing children for transfusion. Although the 12 stations have hospital beds, younger children often have their transfusion in the play area, and older children in the sitting room area.

Children requiring admission are treated on Freddie Hewitt ward, with direct access at all times, bypassing A&E. It is unusual for bed pressures not to allow for this. It is staffed by committed nurses who are experienced in the management of sickle cell children. The facilities are good. They are mostly geared towards younger children although there are bays into which the team try to cohort older children and adolescents. Refurbishment of the floor is being planned, but not finally agreed. If this is possible it will improve the facilities especially for these older children.

LINKED HOSPITALS

Kingston Hospital submitted some written material relating to their service, which was assessed against the Quality Requirements for Local Haemoglobinopathy Teams [see Appendix 2]. None of the Kingston team attended to meet the review team. The team from Queen Mary's at St Helier also submitted written material for review and the consultant paediatrician, consultant haematologist, and CNS came to SGH for a discussion with some members of the review team.

Services at St Helier are well established, and there is considerable experience in the management of these conditions, with teams led by experienced consultants and with sufficient patients [approximately 90 in all, approximately half of whom were children] to maintain clinical expertise. To a large extent the services run autonomously. Annual review visits are undertaken 'in house' at the clinic visit nearest the child's birthday. The St Helier team can make direct referrals to the SGH trans-cranial Doppler service [TCD] for stroke risk assessment, with results returned directly to them. Some children have not yet accessed this service but recently there has been more systematic referral and it is intended that all will be screened in the near future. Responsibility for receiving newborn screening results, communicating results to families, accompanying families to first clinic visits, and ongoing support of families rests with one community CNS [Sutton and Merton] who also works through transition and into the adult red cell service.

Now that services at SGH are becoming re-established, it is intended to firm up network links, with some joint meetings and sharing of protocols which currently are different at each of the three sites reviewed at this visit. One joint meeting has already taken place. For children with specific problems such as difficult iron overload problems, referral has previously been to another centre such as UCLH. Referral pathways are being re-considered now that the team is re-established at SGH.

COMMISSIONING ARRANGEMENTS

Commissioning for the services at SGH is relatively well understood and is under the remit of the South West London Acute Commissioning Unit. One individual – who attended the visit to meet reviewers – has responsibility for all acute services at SGH. There is interest in red cell services, especially following reports of some user issues in the adult services. There is an intention for sickle cell disease, and specifically quality of in-patient care, to be one of the ' CQUIN' quality indicators used in 2010/2011. Continued dialogue between the commissioner and local manager is likely to be helpful in progressing improvements in these services.

REVIEW VISIT FINDINGS

ACHIEVEMENTS

1 This is a very well-established service, although the departure of two key medical leads from the hospital service was followed by a period of uncertain medical leadership. A strong new clinical and management team has significantly developed the service in only one year. The efforts and energies are the new team are commended, and there is clear support from local and Trust management for continuing improvements.

2 User feedback about the services was very positive. In particular, the role of the community based nurse specialists was greatly appreciated. Their support for children and families was reported as being 'invaluable'.

3 There is evidence of good team working between the paediatric and adult teams, who communicate well.

4 The tremendous work of Day Unit team in offering an accessible, flexible, very high quality nurse-led service is noteworthy. The senior nursing staff involved in the service are all highly committed to providing excellent services for the children using these services.

5 The constructive way in which the teams had responded to the process of preparation for the visit was striking. Many individuals commented that they felt the work they had put in had led to tangible improvements and better communication between clinicians, managers and commissioners which they felt would make for continued progress.

6 The visiting team formed a very favourable impression of the services at Queen Mary at St Helier, on the acute and community sites, and much excellent patient information material had been written by the CNS for Sutton and Merton.

IMMEDIATE RISKS

No immediate risks were identified.

CONCERNS

1 Although direct access to the in-patient ward makes access for children with sickle cell pain crisis easy and quick, the time from arrival to first analgesia was reported by users sometimes to be long. The stated aim of less than 60 minutes is longer than usually recommended [< 30 minutes] and families reported that waits of over an hour are frequent. An audit to identify where delays are occurring might usefully allow this issue to be addressed.

2 The lack of a hospital based CNS for this large service was felt to be a gap. Appointment to this post would be likely to lead improvement in the quality of services available and would probably also reduce admissions and reduce length of stay.

3 There was a lack of cover for absences of some key individuals. This applied to the community CNS's and the Day Unit sister at SGH (whose most senior and experienced 'second' has recently left the service). It is hoped that a suitably banded 'junior sister' will be appointed.

4 Although psychology input is available, no psychology time is specifically allocated to this service. Children can reach the service at times of crisis but, there is no provision for regular input, including neurocognitive assessments, which now form part of regular care. The St Helier team do not have adequate access to psychology services and would additionally refer to a dedicated service at SGH.

FURTHER CONSIDERATION

1 The extent to which the South West London Trusts are functioning as a network is not quite clear. There is not yet sharing of guidelines / protocols, although there would be clear advantages of combining those in use at the different sites, which are strong and comprehensive in different sections. Amalgamation would lead to very robust and useful clinical guidance. The linked hospitals are not, at least yet, referring to the specialist team at SGH for specialist annual review, although this may change now that the medical team at SGH is re-established. There are plans for at least annual network meetings. The visiting team considered there may also be value in jointly reviewing difficult cases, joint protocol development and, possibly, sharing of posts across the network, to ensure cover is available for absences.

2 Some Croydon patients who mainly attend Mayday Hospital also use the services at SGH although this had not been identified as a linked hospital in the network by the SGH team. Mayday-based children also access King's College Hospital. Choice for families as to which specialist hospital they wish to access is appropriate, as long as there is clarity about which services they are using on which site, and appropriate continuity of care is achieved.

3 Phlebotomy is currently offered for the children using this service in three places: out-patients, the Day Unit, and the main hospital phlebotomy area. The latter is less suitable for children, and the extension to a full time phlebotomy service in children's out-patients, which the team hope will be possible, would improve this situation for users as well as taking some pressure off the very busy Day Unit staff.

4 The development of the 5th floor ward facility to include dedicated bays for use by older children / adolescents would enhance the service for this age group.

5 There is an opportunity for the hospital team to work with commissioners for the service, who already have an interest in and focus on sickle cell disease / thalassaemia, in order to progress some of the planned improvements and to work jointly on the quality of in-patient stays as part of the CQUINS programme.

6 There is no systematic way of recording which children have had their 'annual review' undertaken.

7 Social work provision for children using the service is felt to be patchy, and this presents real difficulty for some families and their health professionals.

8 No data were presented to show age of infant at the time the family were visited and given the diagnosis, nor the time of the first clinic visit. The community team were confident that these were timely, but an audit should be undertaken to be certain that these standards are being systematically met.

GOOD PRACTICE

1 Direct access to the specialist in-patient ward was available at all times.

2 Individual care plans are kept on the ward and updated after each clinic visit by copy letter. This allows staff to be aware of current problems at any time a child is admitted.

3 There is a strong training focus on appropriate monitoring and safety of opiate use for children being managed for pain crisis.

4 There is 24 hour access to automated red cell exchange, which is unusual, and in some clinical situations likely to be very beneficial.

5 The 'packs', produced by the Sutton and Merton community nurse, for use by the family of a newly diagnosed infant, and for schools, are particularly good. The pack for transition age children is also excellent.

6 Various individual documents in use are noteworthy:

a. The 'checklist' for use by the nurse counsellor visiting the home to inform the family of a newly identified infant is an excellent way of ensuring and recording that all important aspects have been covered in the discussion.

b. The DNA letter having, on its reverse, some reminder guidance of what to do if the child becomes ill before the next appointment.

c. The 'paediatric cannulation and venepuncture assessment' document was very comprehensive.

7 The offer of complementary therapies by a charity working within the hospital ['Full Circle'] is likely to lead to improved patient experience and, possibly, shorter length of stay.

8 The full set of updated clinical guidelines are held in hard copy in all the relevant clinical areas at SGH, easily identified by staff who are familiar with them, and also easily accessible on the 'intranet'.

APPENDIX 1: MEMBERSHIP OF THE REVIEW TEAM

Karen Madgwick	Transfusion Practitioner	North Middlesex University Hospital NHS Trust
Dr Gail Miflin	Consultant Haematologist	University College London Hospitals NHS Foundation Trust
Lee McPhail	Assistant Director of Operations	North Middlesex University Hospital NHS Trust
Ravinder Raj	User Representative	
Dr Kate Ryan	Consultant Haematologist	Manchester University Hospitals NHS Foundation Trust
Rajpal Singh	User Representative	
Dr Anne Yardumian	Clinical Lead for Peer Review Programme	

APPENDIX 2: COMPLIANCE WITH QUALITY REQUIREMENTS

The majority of written evidence supplied for the review team was from SGH, but the linked SWL hospitals Queen Mary @ St Helier, and Kingston Hospital, also submitted written evidence against the QR's which apply to LHT's. The overall compliance rating [Y/N] relates to SGH; comments about Queen Mary @ St Helier [QMSH], and Kingston Hospital [KH] are included where relevant in the final column.

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	Y	*Overall the information was present and was appropriate, but some was not found - including how to feel for the spleen and its significance [1f] although this was included in the checklist used by CNS visiting the family at home to give diagnosis and initial information. For thalassaemia, parts b and c were not explicit. An old UK Thalassaemia leaflet was included in the evidence presented.* *Information about local services was complete.* *Health promotional material was included but was sparse in regard to diet / exercise, smoking and alcohol avoidance.* *QMSH information was stronger in some respects, including some aspects of health promotion.* *KH indicated use of same material as SGH.*

Ref	Quality Requirement	Met?	Comment
	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.		
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	*But it was not clear what information the linked hospitals offer to GP's.*
3	Information should be available on transition to adult care. This information should cover all aspects of the transition (QR43).	Y	*This was a little 'thin' on the SGH site but strong materials are offered @ QMSH.* *KH folder indicated this should be discussed with the team at the visit but no team members attended.*
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	*Although a sickle cell patient-held record was included in the evidence folders, none of the attending families knew of it or used it, and no patient- held record was offered for thalassaemia.*
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	*At SGH site although more geared towards younger children in the present facilities.*
STAFFING and SUPPORT SERVICES			
7 LHT	The LHT should have a nominated lead paediatrician / paediatric haematologist with responsibility for guidelines, protocols, training, audit relating to haemoglobinopathies, liaison with community services and overall responsibility for liaison with the SHT. The lead consultant should undertake CME activity of relevance to care of children and young people with haemoglobinopathies.	Y	*In place for QMSH, and KH.*
8 LHT	There should be agreed arrangements for cover for absences of the lead paediatrician.	Y	
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	Y	

Ref	Quality Requirement	Met?	Comment
	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.		
10	The lead consultant (QR9) should have a named deputy who will provide cover for absences. The named deputy in the SHT should undertake CME activity of relevance to care of children and young people with haemoglobinopathies.	Y	
12	The SHT should have a lead haemoglobinopathy nurse with responsibility, with the lead consultant (QR9), for guidelines, protocols, training, audit relating to haemoglobinopathies and liaison with referring LHTs. The lead nurse should have specific training in the care of patients with haemoglobinopathies.	Y	*The community nurse specialist and consultant nurse at SGH provide most of the functions although the lack of a dedicated acute service CNS was noted, and plans are in place to address this.*
13	The SHT should have a nurse specialist or counsellor who provides outreach support for patients in the community. This nurse specialist / counsellor should have specific training in the care of patients with haemoglobinopathies.	Y	
14	There should be agreed cover arrangements for the outreach nurse specialist / counsellor.	Y	
15	Access to the following staff and services should be available: a MRI and CT scanning b Transcranial Doppler ultrasonography (SC) c Hospital dental services d Genetics services e Bone marrow transplantation services f Contraception and sexual health services g Consultant cardiologist h Consultant endocrinologist i Consultant hepatologist j Consultant neurologist k Consultant ophthalmologist l Consultant orthopaedic surgeon m Consultant obstetrician n Child and adolescent mental health services	Y	
16	The following services should be available a Paediatric high dependency care b Paediatric intensive care There should be agreed criteria for admission to each level of care.	Y	*No criteria for referral to PICU were presented in the evidence folders but were found on ward / PICU visits.*
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	*Psychology services were reported to be inadequate, with input only at times of crisis rather than systematic psychology sessions. QMSH have difficulty accessing psychology services and would use that at SGH if available for them.*
18	A service agreement for support from community services should be in place. This service agreement should cover, at least:	N	*The integration of acute and community services in 2010/2011 will help further to improve links*

Ref	Quality Requirement	Met?	Comment
	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.		*between acute and community personnel .*
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	*At SGH many of day unit nursing team can cannulate. At QMSH, one certificate of competence was included, but it was not clear if nurse cannulation was available at all times children attend for transfusion.*
20	All staff involved in the care of children and young people with haemoglobinopathies should undertake regular child protection training.	Y	*Also evidenced for QMSH.*
CLINICAL and REFERRAL GUIDELINES			
21	Guidelines should be in use covering a How to establish and confirm diagnosis b Parent and sibling testing	Y	*There was limited information on establishing full diagnosis.*
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*	*These guidelines were not together in an easily accessible form. Some elements of the guidance needing clarification to be practically useful – eg for chest syndrome the suggestion 'consider exchange transfusion' could be expanded to include circumstances under which this should / might not need to be undertaken. Specific guidance for acute anaemia, and referral for urgent eye assessment, were not included.* *For QMSH, different clinical guidelines were presented; these were very strong in areas, sometimes more comprehensive than the SGH equivalents. KH team*

Ref	Quality Requirement	Met?	Comment
			presented a third set, again with some areas of particular focus and detail.
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	*These aspects of outpatient care were covered in the guideline 'Routine healthcare maintenance guideline for sickle cell patients'*
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,	N	*This is in place for thalassaemia, but not for sickle cell except the regularly transfused children with sickle cell disease.*

Ref	Quality Requirement	Met?	Comment
	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).		
26	Guidelines should be in use for referral of patients and their families to a clinical psychologist with experience in the care of patients with haemoglobinopathies (including the option for self-referral)	Y	*Good and comprehensive guidance*
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	Y	*And this guidance was considered to be very strong and comprehensive, including*

Ref	Quality Requirement	Met?	Comment
	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.		*governance aspects.*
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.	N	*Guidance about chelation was found to be deficient in certain areas, including clear indications for starting, choice of regimen, dose adjustment, and only a few of the potential side effects and their management were included. The guidance indicates Exjade treatment start from age 5, although 6 is the licensing cut off.*
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 guidelines are very good, and some of the content could usefully be transported into the thalassaemia major clinical guidelines.*
33	Clinical guidelines should be in use covering: a Indications for exchange transfusion b Arrangements for carrying out an exchange transfusion.	Y	
SERVICE ORGANISATION and LIAISON WITH OTHER SERVICES			
34	Clinical guidelines for acute and out-patient monitoring and management (QR23 and QR24) should be available and in use in appropriate areas including A&E, clinic and ward areas.	Y	
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)	N	*Although a patient held record was presented, none of the 5 user families who attended the visit knew of it or used it.* *Direct contact with the ward was known to all and was valued and works in practice.*
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	*Included in the guideline 'Evaluation and Management of Children with Sickle Cell Anaemia Presenting after the Neonatal Period'*

Ref	Quality Requirement	Met?	Comment
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	*A draft document relating to sickle cell disease [not thalassaemia] was seen but is not in use in practice.*
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 no patient held record in routine use, this cannot be compliant although other aspects of the QR are met.*
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	*A patient letter which included copies to relevant parties was included as evidence, but dated 2006, and no general policy or guidance was seen.*
40	An operational policy should be in use covering: a Teaching children, young people and their patients how to set up an administer subcutaneous desferrioxamine infusions b Encouraging children to participate in setting up and administering their own infusion c Regular assessment and updating administration techniques d Recording of assessments of administration techniques	Y	*Self assessed as N/A but relevant guidance 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	N	*Phlebotomy out of hours is offered on the day unit, but children cannot arrive at day unit after school for transfusion, if booked for afternoon have to take time off, and no out of hours clinic availability.* *No out of hours transfusion facility available at QMSH or KH.*
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 the protocol is not specific for these conditions, and the decision to discharge after DNA being at the clinician's discretion might need to be re-visited for these conditions.*
43	A protocol should be in use covering transition to adult care. This should ensure: a Age guidelines for timing of the transfer.	Y	*The material in use at QMSH particularly strong in this regard. At SGH the transition process is –*

Ref	Quality Requirement	Met?	Comment
	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.		*as per team presentation – not yet fully in place.*
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	*A 'PET tracker' is in use on ward.*
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 there are plans in place to progress this.*
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 for QR 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.	Y	*Minutes of meeting were presented.*
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	Y	*But it was noted that the denominator of most of sickle cell audits was 166, as compared with the 193 total patients the SGH team indicated were under their care. It was understood from discussion that as the new clinical team has come in place they are now systematically recording patients being managed in a manner which may have previously lapsed.* *Detailed completed results proformas were included for all patients, which would make*

Ref	Quality Requirement	Met?	Comment
	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	*pulling together audits very easy.* *Audits for QMSH were included and were complete, with good results. None seen for KH.*
51	Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry.	N	*Consent is being sought and the new data manager will hopefully undertake this.*

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	*Although no data were supplied, the commissioner who met the team had a good sense of what the service offered, and was interested to continue working with the provider team to improve quality of services.*
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	*As 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	Y	*Although only a small sample [5 babies over 2 years] was provided, the community nurses who attended the visit assured the review team that this was met.*
P4	Effective follow-up of infants with positive screening results (sickle cell disorder) – all babies to be registered with a local clinic/centre (or clinic working as part of clinical network)	Y	*As for P3*
P5	Timely confirmation of diagnosis for infants with a positive screening result**	N	*No data were provided, and the one set of case notes of an infant ~ 6 months old did not include a laboratory confirmatory result.*
S1i	Failsafe to ensure ongoing care	Y	*Detailed description of efforts undertaken were given to the review team in interview.*
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.