UK Forum on Haemoglobin Disorders

WM QRS West Midlands Quality Review Service

Services for Children with Sickle Cell Disease or Thalassaemia At Sheffield Children's NHS Foundation Trust

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Quality Review Visit Report
Visit date: March 10th 2010

Report finalised: June 10th 2010.

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INTRODUCTION

This report presents the findings of the peer review visit to services for patients with sickle cell disease or thalassaemia at Sheffield Children's NHS Foundation Trust which took place on March 10th 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 Sheffield Children's NHS Foundation 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 SHEFFIELD CHILDREN'S NHS FOUNDATION TRUST

Service (as at October 2009)	Patient numbers Sheffield	Patient numbers Scunthorpe / Grimsby	Patient numbers Doncaster	Patients on long term red cell transfusions
Children with sickle cell disease	50	10	5	< 5 in Grimsby
Children with thalassaemia	10			10
Children with other haemoglobinopathies	<5	<5		<5

BACKGROUND

Sheffield Children's NHS Foundation Trust [SCH] provides a service for children with these conditions who live in Sheffield itself, and those in Rotherham and Barnsley. These are each approximately 30 minutes away by road and children from these towns have all their care provided at SCH. A small number of children with sickle cell disease / HbH in Scunthorpe / Grimsby and Doncaster receive care locally but the local teams network with the specialist team at SCH and almost all children, except a

very small number whose families are unwilling to travel, are reviewed centrally on an annual basis. There is a lead consultant [7.8 PA] with cover by two colleagues, one Haematology Consultant Nurse and one part-time Clinical Nurse Specialist, currently on a fixed term contract which it is very much hoped will be renewed. There is a haemoglobinopathy specialist nurse working in the community, employed by Sheffield PCT who undertakes antenatal counselling, and who handles the neonatal screen results for Sheffield. She also provides some continuing support to Sheffield families in the community.

Accident and Emergency

Children and families have open access to M3 ward, and so mostly bypass A&E. Children brought by ambulance are taken to A&E, nurse triaged and seen / assessed by an A&E doctor. The haematology team is always informed before the patient is discharged. There are comprehensive directories regarding possible presentations kept in A&E. It is intended to 'flag' the children with these conditions on the hospital's computer system so that if they attend for any reason, and their having sickle cell / thalassaemia is not reported, the attending team will be alerted. Service users reported that there is no provision for even short term parking for parents bringing children to hospital with acute emergencies.

Day Case and In-patient Facilities

In-patient and day care beds are grouped on M3 ward, used flexibly. There is no specific 'day care' area. Weekend transfusions are offered, but many families do not use that facility. There is a well used teaching room adjacent to the ward. All the facilities are of high quality and child-friendly.

Review Visit Findings

Overall the visiting team felt the quality of the services provided at SCH for these children and families was very high.

Achievements

1. This is clearly a very committed team, with strong clinical leadership from the lead consultant, the consultant nurse and the clinical nurse specialist. The team is much appreciated by the service users, who note how comfortable they are in accessing the team, and gratefully acknowledge all the support they receive.

2. The trans-cranial Doppler service is now well established and meticulous records of children's scan dates and repeats are held.

3. The appointment of the hospital based clinical nurse specialist is obviously making a difference, in terms of education for hospital staff. It has raised the profile of this service in the ward and across the hospital - as well as the individual care she delivers.

4. Holding a dedicated non-malignant haematology MDT every 3 weeks even with relatively small patient numbers demonstrates the high priority given to the children using this service.

5. There is a growing and strengthening relationship with the local teams more distant from Sheffield to the east, who reported that they were happy with the service and with the support the specialist team at SCH offers to them and their patients.

IMMEDIATE RISKS

There were no immediate risks identified.

CONCERNS

1. In the areas out of Sheffield, communication of a diagnosis of a major haemoglobin disorder to parents of a baby identified through the neonatal screening programme can currently be by telephone, sometimes followed up by a home visit by the GP. Arrangements should be put in place so that this news can be conveyed face – to – face by an appropriately experienced health care professional who can address some of the family's immediate concerns.

2. There was no evidence of any audit of practice or outcomes for the children with thalassaemia.

3. There is a lack of clarity in regard to the arrangements for community care. There is no service level agreement; the work undertaken by the PCT funded 'SCAT' is not clear and working between sectors does not appear to work effectively for the affected children and families. There is no formal cover arrangement for the community nurse specialist.

4. There is potentially a real loss to the service if the hospital nurse specialist's current fixed term contract is not extended.

FURTHER CONSIDERATION

1. The clinical guidelines overall are excellent and comprehensive, though the guideline relating to acute pain management is rather brief and non-specific considering how relatively frequent a presentation this is. For example, there is no indication of expected maximum time to first

analgesia, no reference to use of pain scores to guide level of analgesia and no guide as to frequency of re-evaluation and escalating analgesia.

2. Overall, in the information given to patients and in the facilities on the ward and day care areas, the focus appears to be directed more at younger children, and provision for older children / adolescents is less in evidence. Consideration might be given to alternatives to formal 'hospital beds' for use of older children attending for day transfusions.

3. Most regularly transfused children have indwelling 'port' devices which nurses are trained to access; in the small number of children for whom venous cannulation is required, this is undertaken by junior doctors rather than nurses. This is reported by users to engender delays, and – depending on the doctor - multiple attempts to cannulate may be necessary. Experienced nurses usually develop finer skills in this regard, and consideration might be given to training some additional nurses to undertake cannulation. It was noted by the review team that a higher percentage of regularly transfused children here used in-dwelling central venous 'port' access than is usual elsewhere.

4. The information letter sent to families of babies with newly diagnosed thalassaemia is comprehensive, but initially includes some quite complex genetic information which it was felt may be difficult to comprehend and off-putting. However, the letter is otherwise really helpful. Perhaps the genetic information could be put later on, in an 'appendix'?

5. The linked hospital teams do not meet formally regarding network functioning, although educational meetings are attended by specialist and local team members and it might be possible to add on a brief ' business meeting' , using the opportunity when key members are already together.

6. In one clinical guideline, it is recommended that pre-operative haemoglobinopathy screening be undertaken on children from 'African or African-Caribbean origin'; this might be broadened to include all children of other than northern European origin.

7. Although it is clear that this hospital service is working autonomously, with no clinical reference to the larger centre in Leeds, consideration might be given to some links (for example for review of TCDs or joint review of chelation practice). Such a link might provide useful audit, educational and training opportunities, given the relatively small number of patients treated in Sheffield and might be complementary, perhaps on an annual basis, to the local network meeting suggested in point 5.

8. Children are now being entered onto the National Haemoglobinopathy Registry but numbers were not given and there does not appear to be a formal way of identifying which have already been, and which need to be, entered, in order to facilitate working towards complete inclusion of all who give consent.

GOOD PRACTICE

Many examples of good practice were noted during the visit.

1. The move towards a 'flagging' system on the computer system alerting the treating team to the diagnosis if a child with a haemoglobin disorder came into A&E / any part of the hospital was felt likely to be very helpful

2. The red 'alert' sticker on the front of the case notes, with explanatory page at the front of the notes, is an excellent idea.

3. The letter written by the lead consultant to parents of newly diagnosed sickle cell disease following the first hospital visit, which runs through the information discussed at the visit, was felt to be especially helpful, supplementing the less personal written informational materials well.

4. Among the information to patients, 'what to do in an emergency' and the leaflet about starting Exjade, were outstanding.

5. The clinical guidelines overall are very comprehensive and clear.

6. The recent move to copy parents/ carers into all clinical correspondence is likely to help communication and understanding of care substantially.

APPENDIX 1: MEMBERSHIP OF THE VISITING TEAM

Dr Phil Darbyshire	Consultant Paediatric Haematologist	Birmingham Children's Hospital NHS Foundation Trust
Susan Smith	Sickle Cell & Thalassaemia Nurse Specialist	Bradford Teaching Hospitals NHS Foundation Trust
Lee McPhail	Assistant Director of Operations	North Middlesex University Hospital NHS Trust
Elaine Miller	Co-ordinator	UK Thalassaemia Society
Dr Anne Yardumian	Clinical Lead for Peer Review Programme	

APPENDIX 2: COMPLIANCE WITH QUALITY REQUIREMENTS

Specialist Haemoglobinopathy Teams

Ref	Quality Requirement	Met?	Comment
INFORMATION AND SUPPORT FOR PATIENTS AND THEIR CARERS			
1	Written information should be offered to patients and their families covering at least: 1 A simple explanation and description of the condition, how it might affect the individual, possible complications and treatment. For sickle cell disease this information should include: a Problems, symptoms and signs for which emergency advice should be sought b How to manage pain at home, including how to avoid pain, and non-pharmacological interventions c Importance of adequate fluid intake d Use of antipyretics for fever e Importance of regular antibiotics and full immunisation f How to feel the spleen and its significance. For thalassaemia this information should include: a Problems, symptoms and signs for which emergency advice should be sought b The importance of maintaining good haemoglobin levels by transfusion c Potential problems of iron load and how it can be managed. 2 Details of the services available locally including: a Clinic times and how to change an appointment b Key contact name and number c Alternative contact if key contact away d Who to contact for advice out of hours e How to use emergency services f Ward usually admitted to and its visiting times g Community services and their contact numbers h Details of support groups available. 3 Health promotional material including a Inheritance and implications for other family members b The importance of a good diet and regular exercise c Implications for travel d Age appropriate information on avoiding smoking and excess alcohol consumption e Age appropriate information on contraception and sexual health f Where to go for further information, including useful websites and national voluntary organisations.	Y	*Much excellent information/material was presented, but some of the subsections in this requirement were lacking, especially in 'health promotion' section [eg contraception / sexual health]. No information relating to acute emergencies in thalassaemia was found.*

Ref	Quality Requirement	Met?	Comment
2	Written information for the patient's primary health care team should be available covering at least: a All aspects of QR1 b The need for regular prescriptions, penicillin and analgesia (SC)	Y	
3	Information should be available on transition to adult care. This information should cover all aspects of the transition (QR43).	N	*The guidance given was non-specific to this group of disorders and no age appropriate information was shown.*
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	*This standards will, in effect, be achieved by the proposal in the near future to send all clinic letters and discharge summaries to parents / carers.*
5	The locally agreed patient-held record (QR4) should be in regular use within the LHT / SHT.	N	*As above*
6	Services should be provided in a child friendly environment, including toys and books / magazines for children and young people of all ages.	Y	*But the main focus appears to be particularly towards the younger child; facilities geared to adolescents / young people are not evident.*
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	*And impressive CME activity is recorded, considering this is not her only sub-specialty area.*
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	*Named cover consultants are given, but there was no evidence of specific CME activity relating to haemoglobinopathies.*
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	*And much experience / evidence training presented for this lead nurse.*
13	The SHT should have a nurse specialist or counsellor who provides outreach support for patients in the community. This nurse specialist / counsellor should have specific training in the care of patients with haemoglobinopathies.	Y	
14	There should be agreed cover arrangements for the outreach nurse specialist / counsellor.	N	*Currently if the community specialist nurse is on leave, there is no outreach cover, although if the 2nd nurse specialist post is continued, there is a stated intention for her to cover.*

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	*Although no specific names were given, the review team were confident all were in place and accessible.*
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: c Interpreters d Social work e Play specialist f Hospital teacher (in-patient care only) g Child psychologist h Dietician	Y	*Details of interpreting service and dietician support were not found by the reviewers of this standard, but references were found elsewhere and the hospital team reassured us the services were in place.*
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 service level agreement is in place*
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 do not undertake iv cannulation, a majority of children have 'Portacath's' and many nurses are trained / competent to access; list provided.*
20	All staff involved in the care of children and young people with haemoglobinopathies should undertake regular child protection training.	Y	*Details were received on request after the visit.*
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	*} All guidelines are in place and*
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	*} assessed as being very comprehensive*

Ref	Quality Requirement	Met?	Comment
24	Clinical guidelines should be in use covering routine out-patient monitoring and management between formal annual progress review visits, including: **All patients:** a General assessment of well-being including school attendance b Any difficulties adhering to treatment or other difficulties with care c Monitoring growth and development d Checking for palpable spleen **Patients with sickle cell disease:** a Discussion to ensure analgesics available for home use understood and effective b Informal assessment of cognitive function, learning and behavioural difficulties, including referral if needed c Checking for history of priapism in boys d Checking for and management of nocturnal enuresis e Checking all necessary immunisations up to date f Penicillin therapy and alternative therapy for children who are allergic to penicillin g Monitoring of oxygen saturation by pulse oximeter h demonstrating to parents / carers how to check for splenic enlargement i Monitoring of Hb levels, renal function tests, liver function tests j Indications for early referral to specialist haemoglobinopathy team (LHT only) **Patients with thalassaemia and regularly transfused sickle cell:** a Checking adherence specifically to chelation therapy and any difficulties b Monitoring Hb levels, iron levels, liver function and other biochemistry c Indications for early referral to specialist haemoglobinopathy team	Y	

Ref	Quality Requirement	Met?	Comment
25	Clinical guidelines should be in use covering **annual specialist review** visits including, at least: **All patients:** a Regularity of school attendance and reasons for absence b Any difficulties adhering to treatment or other difficulties with care c Monitoring growth and development, including review of centile charts d Health promotion and healthy lifestyle advice and support e Contraception and sexual health in relevant age group f Travel advice g Review Hb levels, renal and liver assessment, other biochemistry h Hepatitis B vaccination i Monitoring of hepatitis B antibodies and action to be taken should levels fall j Monitoring for hepatitis C in transfused patients k Discussion and preparation of child for any planned surgery l Indications for consideration of splenectomy m Preparations for splenectomy including recommended immunisations n Treatment of complications of splenectomy, including persistent thrombocytosis. **Patients with sickle cell disease:** a As QR 24 plus b Monitoring and screening for neurological complications, including transcranial Doppler ultrasonography c Indications for imaging to assess the extent of cerebrovascular disease d Indications for overnight oxygen saturation monitoring (sleep study) e Indications for echocardiography including possibility of pulmonary hypertension **Patients with thalassaemia:** a As QRs 31, and 32 where applicable [thalassaemia intermedia] plus b Review annual red cell consumption c Review adequacy and appropriateness of iron chelation regimen d Consideration of options for helping children, young people and their families to adhere to chelation therapy e Audiometry and ophthalmology check for those on desferrioxamine, from age 10 f Cardiological assessment (from age 10) g Testing for endocrine abnormalities (from age 10) h Bone mineral density assessment (from age 10).	Y	

Ref	Quality Requirement	Met?	Comment
26	Guidelines should be in use for referral of patients and their families to a clinical psychologist with experience in the care of patients with haemoglobinopathies (including the option for self-referral)	Y	
27	Guidelines for referral for consideration of bone marrow transplantation should be in use.	Y	
28	Clinical guidelines should be in use covering: a Indications for regular transfusions b Investigations and vaccinations prior to first transfusion c Monitoring of haemoglobin levels	Y	
29	Clinical guidelines should be in use covering review by a specialist nurse or doctor prior to transfusion to ensure that each transfusion is appropriate.	Y	
30	A Transfusion Policy should be in use covering: a Area/s where transfusions will usually be given b Procedures for checking blood c Staff allowed to undertake cannulation d Recommended number of cannulation attempts e Maximum rate and volume of transfusion according to size of child f Monitoring the transfusions g Management of transfusion reactions h Arrangements for medical cover during transfusion i Arrangements for ensuring that a nurse with appropriate competences is in attendance throughout the transfusion.	Y	
31	Clinical guidelines for chelation therapy and monitoring iron load should be in use including: a Indications for starting chelation b Choice of regime c Dosage and dosage adjustment d Clinical assessment of tissue damage e Monitoring of serum ferritin f Use of non-invasive estimation of organ-specific iron loading heart and liver by T2*/R2 g Management of side effects of chelators.	Y	
32	Clinical guidelines for the management of thalassaemia intermedia should be in use including: a Indications for transfusion b Monitoring iron loading c Indications for splenectomy.	Y	
33	Clinical guidelines should be in use covering: a Indications for exchange transfusion b Arrangements for carrying out an exchange transfusion.	Y	
SERVICE ORGANISATION and LIAISON WITH OTHER SERVICES			
34	Clinical guidelines for acute and out-patient monitoring and management (QR23 and QR24) should be available and in use in appropriate areas including A&E, clinic and ward areas.	Y	

Ref	Quality Requirement	Met?	Comment
35	A protocol should be in use covering the initial clinic visit for patients with haemoglobinopathies covering, at least: a Giving each patient information relevant to their condition (QR1) b Giving each patient their patient-held record (QR4) c Allocation of a named contact for queries and advice to each patient. d Discussion of arrangements for future treatment and care e Sending the GP information relevant to their patient's condition (QR2)	N	*This standard is nearly met. There is no patient held record and no mention of arrangements for future care with Local team.*
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	*Shared care clinical guidelines were demonstrated, but no protocol regarding what was expected regarding care at Local centre etc. There is no patient held record in regular use*
38	A protocol should be in use covering: a Updating patient-held records b Offering patients a permanent record of consultations at which changes to their care plan are discussed. c Recording changes of key contact d Giving further information (QR1) as patients' and families' needs change	N	*No protocol seen*
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	*No policy included*
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	*No policy, although because of use of alternative chelation regimens for most children this now relates only to small patient numbers*
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	*Although out of hours / weekend transfusion is offered if families choose, there is no out of hours provision for clinic or for phlebotomy.*

Ref	Quality Requirement	Met?	Comment
42	A protocol should be in use covering: a Follow up of children who do not attend b Communication and follow up of children who move to another area	Y	
43	A protocol should be in use covering transition to adult care. This should ensure: a Age guidelines for timing of the transfer. b Involvement of the young person in the decision about transfer. c Involvement of primary health care, social care and adult services in planning the transfer. d Allocation of a named coordinator for the transfer of care. e A preparation period and education programme relating to transfer to adult care. f Communication of clinical information to the adult services. g Arrangements for monitoring during the time immediately after transfer to adult care.	Y	*But due update [in progress]*
44	The team should have in place: h Mechanisms for receiving feedback from patients and carers about the treatment and care they receive. i Mechanisms for involving patients and carers in decisions about the organisation of the services. j Mechanisms for encouraging the development of local support groups.	N	*No formal mechanism was demonstrated for patient / carer feedback and involvement*
45	The SHT should run a programme of training and awareness of the management of patients with haemoglobinopathy for its main referring LHTs.	Y	*This is in place, as part of the broader haematology education and training with linked hospitals*
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	
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	
DATA COLLECTION and AUDIT			

Ref	Quality Requirement	Met?	Comment
50	The LHT / SHT should have audited compliance with key standards including: **Patients with sickle cell disease:** f Proportion of patients taking regular penicillin g Proportion of patients fully immunised against pneumococcus h Proportion of patients (HbSS and HbSβ^0) who have had Transcranial Doppler ultrasonography undertaken within the last year i Proportion of patients who have had their annual multi-disciplinary review within the last year j Effectiveness of action to contact families who have not attended for follow up appointments. k 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	*Audit of vaccination / penicillin might usefully be re-visited [2007, and 27 patients cf now 50 cared for].* *TCD records are detailed and complete.* *There appears to be no audit yet of completeness of annual review coverage.* *Although examples of letters regarding failed attendance were included, there was no record of how effective the actions had been in getting children back to clinic.* *No audits were presented relating to thalassaemia patients.*
51	Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry.	N	*No evidence was given presented about how many patients entered onto NHR, although this is in progress.*

Commissioners of Services

Ref	Quality requirement	Met?	Comment
52	Each Specialist Commissioning Group[1] should have agreed the location of services for its population: a Specialist Haemoglobinopathy Team/s for children and young people b Local Haemoglobinopathy Team/s for children and young people c The expected referral patterns to each SHT and LHT. d The type of patients (sickle cell and / or thalassaemia) who will be treated by each team.	N	*} No submission relating to*

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	*} these standards*

Additional Requirement – Screening Services

Ref	Quality requirement	Met?	Comment
P3	Timely communication of positive screening results (sickle cell disorder) – including a review of parental results	N	*But this standard is very nearly met, and all children are reported to be seen by 6 weeks*
P4	Effective follow-up of infants with positive screening results (sickle cell disorder) – all babies to be registered with a local clinic/centre (or clinic working as part of clinical network)	Y	*[the first part of this standard overlaps with P3, and has been ignored]* *2nd and 3rd parts are met.*
P5	Timely confirmation of diagnosis for infants with a positive screening result**	Y	
S1i	Failsafe to ensure ongoing care	N	*This is in place for Sheffield – born infants [the lead consultant and screening laboratory check lists 3/12ly, and are considering more frequent cross checking] but is not clearly in place at linked sites.*
S1ii	Up-to-date registers maintained of babies (cases) for which units are responsible	Y	*And the list usefully includes first language and post code*

** 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.