UK Forum on
Haemoglobin
Disorders

WM QRS West Midlands Quality Review Service

# Services for Children with Sickle Cell Disease or Thalassaemia
# At
# Alder Hey Children's
# NHS Foundation Trust

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## Quality Review Visit Report
## Visit date: June 24th 2010

Report finalised: December 29th 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 Alder Hey Children's NHS Foundation Trust 2010, which took place on June 24th 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 Alder Hey 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 ALDER HEY NHS FOUNDATION TRUST

| Service (as at 2010) | Patient numbers sickle cell disease | Patient numbers thalassaemia | Patients on long term red cell transfusions |
|---|---|---|---|
| Alder Hey | 39 | <5 | 6 |
| Linked hospitals total [4 sites] | 6 | | |

Sickle cell and thalassaemia services at Alder Hey Hospital [AHH] are provided within the Paediatric Haematology Department. There are three paediatric haematologists, one is the named lead for this service and the other two cover in his absence. They both also take a part in the routine clinical work of the service and so are familiar with the patients. One trainee haematology doctor contributes to the service, and specialist nursing staff are key to its running. One CNS is the lead nurse, with 0.5 WTE for this role but provides an additional 0.4 WTE at band 5. Two additional CNS's provide cover.

## Accident and Emergency

Children needing urgent assessment can come to A&E, where they are triaged urgently. Those with sickle cell disease and acute pain are managed according to a standard protocol. During working hours, most families prefer to come to a 'Haematology Treatment Centre' where they are assessed and managed by the core team. Although controlled drugs are not kept in the Haematology Treatment Centre, a prescription can be written and dispensed by arrangement with a nearby pharmacy within 20 minutes. The lead nurse has a prescribing qualification. An average of four children per month present with an acute problem. Users reported that the direct access to the team at the Treatment Centre works well for them.

## Outpatient Clinics

Children with red cell disorders are seen in a weekly general haematology clinic. This is run by the lead clinician with the other two paediatric haematologists. The lead nurse, and sometimes the community nurse specialist, also come to the clinic. The children have transcranial Doppler appointments arranged in advance for the same day as a clinic attendance. Hard copy of the clinical guidelines are brought to the clinic for this session. For the first visit, the lead clinician tends to meet the family in the less formal setting of the Treatment Centre sitting room. 'Did not attend' rates at the clinic are very low at 4.5% for 2009, with telephone follow up of any children who miss appointments.

Transition to adult services has not been the focus of much attention to date. A meeting was organised at the community centre for the day after the visit, and the haematologist who takes over the care of these patients at the Royal Infirmary was considered to be engaged and interested so the team and users were optimistic that the transition process would improve.

## Day Care

The Day Unit is open 8 a.m – 6 p.m on weekdays. Children for transfusion can have their pre-transfusion samples taken at home by an outreach nursing team. The hospital CNS's cannulate the children on arrival. The nurses themselves have training in play and distraction techniques. Weekend transfusions can be arranged on the in-patient ward.

## IN-PATIENT FACILITIES

Children with these conditions are usually managed on ward C3. Occasionally it is necessary for them to be managed on a different ward but transfer out of Alder Hey due to bed pressures is very rare. There are up to two in-patients at any time. Oramorph is used initially, parenteral opiates as necessary, and patient controlled analgesia can be set up at any time. There are daily consultant ward rounds, including at weekends, the paediatric haematologists working a 1:3 rota. Users reported that pain management on the ward had not always been good, but noted recent improvements. They suggested that it can be difficult to get a doctor to visit the ward at night if they request to see one.

Red cell exchange, if necessary, is undertaken by a Blood Transfusion Service team coming to the hospital, which can be arranged at very short notice including weekends. There is an assessment proforma on the ward for nurses to use if families phone for advice, which gives guidance on the need for hospital assessment or admission.

## COMMUNITY SERVICES

A community specialist nurse and two screening link health visitors are based at a community centre about 4 miles from the hospital. Their responsibilities include antenatal counselling, referral for prenatal diagnosis if necessary, and informing the parents of carrier infants identified through the screening programme. The community nurse also runs the monthly sickle cell support group. She helps families with social and benefit issues, and works also with the adult patients [around 30 in total]. The hospital lead nurse visits the family of affected newborn at home. The community team are PCT funded and restricted in the area they cover. The team aim for the family to be informed of the diagnosis and the baby started on penicillin by two weeks of age, and this is nearly always achieved. The hospital nurse also visits schools, sometimes with the community nurse, if health education and support there is needed.

## LINKED HOSPITALS

Eight hospitals are linked with Alder Hey for haemoglobin disorder management. Six children are managed over four sites, and the other four hospitals currently see no children. Nonetheless, the lead clinician has been running a 6 monthly meeting for staff from all these sites, to discuss educational and organisational aspects of care. Guidelines are shared across this network. All the children are seen at Alder Hey for annual review.

### COMMISSIONING ARRANGEMENTS

The specialist commissioning lead locally is involved with services for children with red cell disorders, and has recently set up the pan-Regional group described above. This group has met twice so far, aiming to ensure appropriate access to investigation and management for all children and adults with red cell disorders in the NW Region and Merseyside. The group is currently developing clinical guidelines to be used across the region. There is no benchmarking of staffing or resource, but activity is monitored against an agreed plan. Commissioners would welcome some joint working between Alder Hey and the Manchester Children's Hospital, whose service manages a larger number of children, for combined educational meetings and discussion of difficult or complex cases.

## REVIEW VISIT FINDINGS

### ACHIEVEMENTS

1 The Alder Hey team is offering a very good service, with particularly strong nurse leadership. The range of roles undertaken by the lead hospital CNS is very wide, ranging from prescribing of medication, to home and school visits. The fact that she is currently undertaking an e-learning MSc in haemoglobinopathies while doing a demanding, full time job is commendable. The effort made by the team to streamline appointments for children needing multiple attendances is noted. There is obvious enthusiasm for the service, and commitment to it, from all the team members.

2 Users report positively on all aspects of the service and feel well supported and comfortable with members of the professional team. They note the improved access for urgent assessment and care since being able to use the Treatment Centre, and they very much appreciate this.

3 Close working links with the newborn screening laboratory allows for early detection of affected newborn, and screening standards relating to time to informing the family, and starting the baby on prophylactic antibiotics, are exceeded.

## IMMEDIATE RISKS

No immediate risks were identified.

## CONCERN

1 Some attention to the status of the acute clinical guidelines is needed. The version on the intranet is old, and a new draft set is in preparation. The visiting team felt that some areas of the draft might be revisited before finalising and distributing. The guideline for management of acute chest syndrome in children with sickle cell disease was not entirely clear, and there were no specific indications for the use of 'top up' and exchange transfusion. Document control in general needs to be tightened. In some cases there were two or three different versions of forms and guidance in the folders, which could lead to confusion.

## FURTHER CONSIDERATIONS

1 The hospital CNS is undertaking roles which elsewhere are more usually undertaken by community based colleagues. If there is not going to be an increase in hospital CNS sessions for this service, the team may need to review aspects of her work in the community. Some of the work currently undertaken by the community nurse might be offered by other agencies, for example social services or the voluntary sector.

2 Although children can be referred for psychology input, response time is slow and there is insufficient provision to allow the sort of systematic neuro-cognitive assessments that are recommended for children with sickle cell disease. Users also reported difficulty in accessing social work support.

3 The team might wish to consider the extent to which some links with the team at Manchester Children's Hospital might be beneficial, for example in holding joint educational meetings, and for shared review of complex cases including those with neurological problems or on iron chelation.

4 While having a consultant- and nurse-delivered service has obvious quality benefits, given how much out of hours assessment and treatment is inevitably delivered by junior medical staff, more involvement of paediatric trainees in the regular delivery of care should be considered. A less immediate consideration is the need, here and elsewhere, for junior

doctors to receive sufficient training to be the next generation of consultants in this growing sub-specialty.

5 Some sharing of information might be revisited. For example there is no written service description giving contact details, and little information is given to primary care teams about the conditions. Giving of written updates to parents in the form of copy clinic letters is, in practice, inconsistent.

6 The transition process is not yet fully developed but joint working with the team at the Liverpool Royal Infirmary and the community nurse specialist is planned. This work will need some continued attention to see it through to a successful pathway for transfer of care.

## GOOD PRACTICE

1 Much of the patient and family information is excellent, including some age-appropriate leaflets for young children in hospital and undergoing procedures. Information about thalassaemia, and clinical guidelines, are very good, especially considering the small patient numbers.

2 Some of the other clinical guidelines were also noteworthy. The outpatient guidance is comprehensive. The colour-coded proformas used for various clinic visits are helpful, and the sickle cell annual review proforma is very full.

3 There is a full set of clinical audits against key care standards.

4 Some of the support services are obviously particularly engaged and supportive, especially radiology, with an established transcranial Doppler service provided by a radiologist who is actively involved in training and quality issues.

5 The fact that the other two paediatric haematology consultants, in addition to the lead clinician, are involved in outpatient care of these children, and attend the MDT, is helpful as it allows for familiarity when they are managing in-patients.

6 Patient liaison is good, with evidence of user feedback and actions taken in response to it.

7 The longstanding twice yearly meeting held by the lead clinician for linked hospital staff is commendable, especially given the small number of outlying children in the network.

8 The sharing of protocols and guidelines across the network is good practice. Many other networks aspire to this but have not yet achieved it in practice.

9 The liaison between the hospital team and schools is valued. Users reporting that the attitude of school staff to the children noticeably improved after nurse visits.

10 Some specific details of the service were noteworthy, including the checklist and guidance for assessment of children over the telephone when parents call the ward, the fact that children can have pre-transfusion samples taken at home by the outreach nursing team, and the fact that nurses undertaking procedures themselves have had training in play and distraction techniques to use with worried or distressed children.

## APPENDIX 1: MEMBERSHIP OF THE REVIEW TEAM

| | | |
|---|---|---|
| Dr Phil Darbyshire | Consultant Paediatric Haematologist | Birmingham Children's Hospital NHS Foundation Trust |
| Elaine Miller | User Representative | UK Thalassaemia Society |
| Marie-Claire Nguemshe | User Representative | |
| Christine Williams | Service Director | Sickle Cell and Thalassaemia Centre, Hackney |
| Dr Christine Wright | Consultant Haematologist | Sickle Cell and Thalassaemia Centre, Sandwell and West Birmingham Hospital NHS Trust |
| 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:<br><br>1 A simple explanation and description of the condition, how it might affect the individual, possible complications and treatment.<br><br>For sickle cell disease this information should include:<br>a Problems, symptoms and signs for which emergency advice should be sought<br>b How to manage pain at home, including how to avoid pain, and non-pharmacological interventions<br>c Importance of adequate fluid intake<br>d Use of antipyretics for fever<br>e Importance of regular antibiotics and full immunisation<br>f How to feel the spleen and its significance.<br><br>For thalassaemia this information should include:<br>a Problems, symptoms and signs for which emergency advice should be sought<br>b The importance of maintaining good haemoglobin levels by transfusion<br>c Potential problems of iron load and how it can be managed.<br><br>2 Details of the services available locally including:<br>a Clinic times and how to change an appointment<br>b Key contact name and number<br>c Alternative contact if key contact away<br>d Who to contact for advice out of hours<br>e How to use emergency services<br>f Ward usually admitted to and its visiting times<br>g Community services and their contact numbers<br>h Details of support groups available.<br><br>3 Health promotional material including<br>a Inheritance and implications for other family members<br>b The importance of a good diet and regular exercise<br>c Implications for travel<br>d Age appropriate information on avoiding smoking and excess alcohol consumption<br>e Age appropriate information on contraception and sexual health<br>f Where to go for further information, including useful websites and national voluntary organisations. | Y | *And much of this material is very good and comprehensive.* |

| Ref | Quality Requirement | Met? | Comment |
|---|---|---|---|
| 2 | Written information for the patient's primary health care team should be available covering at least:<br>a All aspects of QR1<br>b The need for regular prescriptions, penicillin and analgesia (SC) | N | *Self assessment indicates that primary care information is provided in the form of a clinic letter. Those seen in medical records were insufficiently detailed to meet the requirement.* |
| 3 | Information should be available on transition to adult care. This information should cover all aspects of the transition (QR43). | N | *But transition processes and information are to be worked on in the near future.* |
| 4 | The SHT and its linked LHTs should have agreed a patient-held record for recording at least:<br>a Information about the patient's condition<br>b Current management plan<br>c Regular medication<br>d Named contact for queries and advice<br>e Alternative contact for times when key contact is away. | N | *But a full sickle cell patient held record is in development.* |
| 5 | The locally agreed patient-held record (QR4) should be in regular use within the LHT / SHT. | N | |
| 6 | Services should be provided in a child friendly environment, including toys and books / magazines for children and young people of all ages. | Y | *The facilities are all good.* |
| ***STAFFING and SUPPORT SERVICES*** | | | |
| 9 | The SHT should have a nominated lead paediatrician / paediatric haematologist consultant with an interest in the care of patients with haemoglobinopathies who should have responsibility for guidelines, protocols, training, audit relating to haemoglobinopathies and overall responsibility for liaison with referring LHTs. The lead consultant should undertake CME activity of relevance to care of children and young people with haemoglobinopathies. | Y | |
| 10 | The lead consultant (QR9) should have a named deputy who will provide cover for absences. The named deputy in the SHT should undertake CME activity of relevance to care of children and young people with haemoglobinopathies. | Y | |
| 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 ongoing CPD activities are noted.* |
| 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 | |

| Ref | Quality Requirement | Met? | Comment |
|---|---|---|---|
| 15 | Access to the following staff and services should be available:<br>a MRI and CT scanning<br>b Transcranial Doppler ultrasonography (SC)<br>c Hospital dental services<br>d Genetics services<br>e Bone marrow transplantation services<br>f Contraception and sexual health services<br>g Consultant cardiologist<br>h Consultant endocrinologist<br>i Consultant hepatologist<br>j Consultant neurologist<br>k Consultant ophthalmologist<br>l Consultant orthopaedic surgeon<br>m Consultant obstetrician<br>n Child and adolescent mental health services | Y | |
| 16 | The following services should be available<br>a Paediatric high dependency care<br>b Paediatric intensive care<br>There should be agreed criteria for admission to each level of care. | Y | *A Trust wide 'early warning system' is in use.* |
| 17 | The following support services should be available:<br>a Interpreters<br>b Social work<br>c Play specialist<br>d Hospital teacher (in-patient care only)<br>e Child psychologist<br>f Dietician | Y | |
| 18 | A service agreement for support from community services should be in place. This service agreement should cover, at least:<br>a Guidelines for involvement of community paediatric services in the care of patients with haemoglobinopathies<br>b Role of community services<br>c Exchange of information between hospital and community services and vice versa<br>d Arrangements for liaison with schools. | N | *And the roles of the community and the hospital CNS might need to be revisited as the service grows.* |
| 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 | *No written list of staff names, rotas, competency training was seen but the visiting team were assured by hospital CNS that this is in place.* |
| 20 | All staff involved in the care of children and young people with haemoglobinopathies should undertake regular child protection training. | Y | *List of key staff members by date and level of training was received after visit, although it is noted that the lead Consultant training to level 2 is not dated, and that staff in this service working predominantly with children and young people are recommended to be trained to level 3.* |

| Ref | Quality Requirement | Met? | Comment |
|---|---|---|---|
| ***CLINICAL and REFERRAL GUIDELINES*** | | | |
| 21 | Guidelines should be in use covering<br>a How to establish and confirm diagnosis<br>b Parent and sibling testing | Y | |
| 22 | Clinical guidelines should be in use covering:<br>a Recommended immunisations<br>b Immunisations, other prophylaxis and travel advice prior to travel abroad.<br>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:<br>**For patients with sickle cell disease:**<br>a Fever and infection including major sepsis<br>b Acute pain<br>c Acute anaemia<br>d Stroke and other acute ischaemic events<br>e Acute chest syndrome<br>f Acute splenic sequestration<br>g Abdominal pain / jaundice<br>h Priapism<br>i Changes in vision, including urgent referral for an ophthalmologic opinion<br>j Indications for 'top-up' and for exchange transfusion and practical protocol for undertaking exchange transfusion<br><br>**For patients with thalassaemia:**<br>a Fever and infection including major sepsis<br>b Unexpected cardiac, hepatic, endocrine decompensation.<br>These guidelines should ensure that patients are transferred to the paediatric ward for assessment as quickly as possible | N | *Some new draft guidelines are in preparation; the visiting team felt that some aspects might need revising before they are finalised including clearer guidance for managing acute chest syndrome, reviewing the time from start of priapism when a family is advised to present, and addition of clear indications for top up and exchange transfusion. Acute visual disturbance is not currently included.*<br><br>*The version currently on the intranet is over 10 years old, and the new draft should replace it as soon as possible.*<br><br>*No guideline for acute presentation in thalassaemia was seen. Patient numbers are small but a line indicating that a senior paediatric haematologist should be alerted if there is such a presentation would be helpful.* |

| 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:<br>**All patients:**<br>a General assessment of well-being including school attendance<br>b Any difficulties adhering to treatment or other difficulties with care<br>c Monitoring growth and development<br>d Checking for palpable spleen<br>**Patients with sickle cell disease:**<br>a Discussion to ensure analgesics available for home use understood and effective<br>b Informal assessment of cognitive function, learning and behavioural difficulties, including referral if needed<br>c Checking for history of priapism in boys<br>d Checking for and management of nocturnal enuresis<br>e Checking all necessary immunisations up to date<br>f Penicillin therapy and alternative therapy for children who are allergic to penicillin<br>g Monitoring of oxygen saturation by pulse oximeter<br>h demonstrating to parents / carers how to check for splenic enlargement<br>i Monitoring of Hb levels, renal function tests, liver function tests<br>j Indications for early referral to specialist haemoglobinopathy team (LHT only)<br>**Patients with thalassaemia and regularly transfused sickle cell:**<br>a Checking adherence specifically to chelation therapy and any difficulties<br>b Monitoring Hb levels, iron levels, liver function and other biochemistry<br>c Indications for early referral to specialist haemoglobinopathy team | Y | *These guidelines are good and comprehensive, including those for thalassaemia.* |

| Ref | Quality Requirement | Met? | Comment |
|---|---|---|---|
| 25 | Clinical guidelines should be in use covering **annual specialist review** visits including, at least:<br><br>**All patients:**<br>a Regularity of school attendance and reasons for absence<br>b Any difficulties adhering to treatment or other difficulties with care<br>c Monitoring growth and development, including review of centile charts<br>d Health promotion and healthy lifestyle advice and support<br>e Contraception and sexual health in relevant age group<br>f Travel advice<br>g Review Hb levels, renal and liver assessment, other biochemistry<br>h Hepatitis B vaccination<br>i Monitoring of hepatitis B antibodies and action to be taken should levels fall<br>j Monitoring for hepatitis C in transfused patients<br>k Discussion and preparation of child for any planned surgery<br>l Indications for consideration of splenectomy<br>m Preparations for splenectomy including recommended immunisations<br>n Treatment of complications of splenectomy, including persistent thrombocytosis.<br>**Patients with sickle cell disease:**<br>a As QR 24 plus<br>b Monitoring and screening for neurological complications, including transcranial Doppler ultrasonography<br>c Indications for imaging to assess the extent of cerebrovascular disease<br>d Indications for overnight oxygen saturation monitoring (sleep study)<br>e Indications for echocardiography including possibility of pulmonary hypertension<br>**Patients with thalassaemia:**<br>a As QRs 31, and 32 where applicable [thalassaemia intermedia] plus<br>b Review annual red cell consumption<br>c Review adequacy and appropriateness of iron chelation regimen<br>d Consideration of options for helping children, young people and their families to adhere to chelation therapy<br>e Audiometry and ophthalmology check for those on desferrioxamine, from age 10<br>f Cardiological assessment (from age 10)<br>g Testing for endocrine abnormalities (from age 10)<br>h Bone mineral density assessment (from age 10). | Y | *Although no protocol was seen, the process of annual review is clearly in place, evidenced by reference to medical records and audit data.* |
| 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 | |
| 27 | Guidelines for referral for consideration of bone marrow transplantation should be in use. | N | *No explicit indications for consideration of BMT were included.* |

| Ref | Quality Requirement | Met? | Comment |
|---|---|---|---|
| 28 | Clinical guidelines should be in use covering:<br>a Indications for regular transfusions<br>b Investigations and vaccinations prior to first transfusion<br>c Monitoring of haemoglobin levels | N | *The self assessment response indicates that these are currently in development, for sickle cell disease [and should include reference to general hospital transfusion policy].*<br><br>*They are in place for thalassaemia.* |
| 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:<br>a Area/s where transfusions will usually be given<br>b Procedures for checking blood<br>c Staff allowed to undertake cannulation<br>d Recommended number of cannulation attempts<br>e Maximum rate and volume of transfusion according to size of child<br>f Monitoring the transfusions<br>g Management of transfusion reactions<br>h Arrangements for medical cover during transfusion<br>i Arrangements for ensuring that a nurse with appropriate competences is in attendance throughout the transfusion. | Y | *But – as in 28 – might usefully cross refer to sickle cell transfusion guidelines.* |
| 31 | Clinical guidelines for chelation therapy and monitoring iron load should be in use including:<br>a Indications for starting chelation<br>b Choice of regime<br>c Dosage and dosage adjustment<br>d Clinical assessment of tissue damage<br>e Monitoring of serum ferritin<br>f Use of non-invasive estimation of organ-specific iron loading heart and liver by T2*/R2<br>g Management of side effects of chelators. | N | *These are not complete, as per self assessment.* |
| 32 | Clinical guidelines for the management of thalassaemia intermedia should be in use including:<br>a Indications for transfusion<br>b Monitoring iron loading<br>c Indications for splenectomy. | Y | *Self assessed as 'no' but inclusion of UK Standards guidance is sufficient.* |
| 33 | Clinical guidelines should be in use covering:<br>a Indications for exchange transfusion<br>b Arrangements for carrying out an exchange transfusion. | N | |
| ***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:<br>a Giving each patient information relevant to their condition (QR1)<br>b Giving each patient their patient-held record (QR4)<br>c Allocation of a named contact for queries and advice to each patient.<br>d Discussion of arrangements for future treatment and care<br>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:<br>a Full medical history and examination<br>b Investigations<br>c Referral to other specialist services (QR15)<br>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:<br>a All patients have an up to date patient held record and details of their care plan.<br>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:<br>a Updating patient-held records<br>b Offering patients a permanent record of consultations at which changes to their care plan are discussed.<br>c Recording changes of key contact<br>d Giving further information (QR1) as patients' and families' needs change | N | |
| 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 was seen, and as medical records for children also attending other local hospital were not seen it was not possible to assess this.* |
| 40 | An operational policy should be in use covering:<br>a Teaching children, young people and their patients how to set up an administer subcutaneous desferrioxamine infusions<br>b Encouraging children to participate in setting up and administering their own infusion<br>c Regular assessment and updating administration techniques<br>d Recording of assessments of administration techniques | N | |
| 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 | *There is some flexibility for phlebotomy and transfusion. Out of hours clinic appointments are being considered as part of transition work, but not currently available.* |

| Ref | Quality Requirement | Met? | Comment |
|---|---|---|---|
| 42 | A protocol should be in use covering:<br>a Follow up of children who do not attend<br>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:<br>a Age guidelines for timing of the transfer.<br>b Involvement of the young person in the decision about transfer.<br>c Involvement of primary health care, social care and adult services in planning the transfer.<br>d Allocation of a named coordinator for the transfer of care.<br>e A preparation period and education programme relating to transfer to adult care.<br>f Communication of clinical information to the adult services.<br>g Arrangements for monitoring during the time immediately after transfer to adult care. | N | *This is work in progress.* |
| 44 | The team should have in place:<br>a Mechanisms for receiving feedback from patients and carers about the treatment and care they receive.<br>b Mechanisms for involving patients and carers in decisions about the organisation of the services.<br>c Mechanisms for encouraging the development of local support groups. | Y | |
| 45 | The SHT should run a programme of training and awareness of the management of patients with haemoglobinopathy for its main referring LHTs. | Y | *And these meetings have been held regularly every 6 months since 2005.* |
| 47 | The SHT should meet at least annually with its referring LHT teams to:<br>a Identify any changes needed to network-wide policies, procedures and guidelines<br>b Review results of audits undertaken<br>c Review any critical incidents including those involving liaison between teams<br>d Consider the content of future training and awareness programmes (QR45) | Y | *As for 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 | *These meetings take place every 6 months, and are minuted.* |

| Ref | Quality Requirement | Met? | Comment |
|---|---|---|---|
| ***DATA COLLECTION and AUDIT*** | | | |
| 50 | The LHT / SHT should have audited compliance with key standards including:<br>**Patients with sickle cell disease:**<br>a Proportion of patients taking regular penicillin<br>b Proportion of patients fully immunised against pneumococcus<br>c Proportion of patients (HbSS and HbS$\beta^0$) who have had Transcranial Doppler ultrasonography undertaken within the last year<br>d Proportion of patients who have had their annual multi-disciplinary review within the last year<br>e Effectiveness of action to contact families who have not attended for follow up appointments.<br>f Review of the care of any patients who have died.<br>**Patients with thalassaemia:**<br>a Proportion of patients on chelation therapy<br>b Proportion of patients who have had their annual multi-disciplinary review within the last year<br>c Adequacy of recording of:<br>▪ Pre-transfusion Hb levels<br>▪ Regular monitoring of iron level<br>▪ Complications of iron overload<br>▪ Height / weight progression<br>▪ Spleen size<br>▪ Support for home chelation programme (QR40)<br>d Effectiveness of action to contact families who have not attended for follow up appointments.<br>e Review of the care of any patients who have died. | Y | *All relevant key audits are in place, some ongoing.* |
| 51 | Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry. | N | *No children yet entered onto NHR. Consent is being sought.* |

## Commissioners of Services

| Ref | Quality requirement | Met? | Comment |
|---|---|---|---|
| 52 | Each Specialist Commissioning Group[1] should have agreed the location of services for its population:<br>a Specialist Haemoglobinopathy Team/s for children and young people<br>b Local Haemoglobinopathy Team/s for children and young people<br>c The expected referral patterns to each SHT and LHT.<br>d The type of patients (sickle cell and / or thalassaemia) who will be treated by each team. | N | |

| Ref | Quality requirement | Met? | Comment |
|---|---|---|---|
| 53 | Each Specialist Commissioning Group should have:<br>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.<br>b Agreed a plan for the development of SHTs and LHTs located within its area.<br>c Monitored achievement of the agreed plan at least annually.<br>The agreed development plan should ensure that QRs 7 to 19 are met within 2 to 5 years. | N | |

**Additional Requirement – Screening Services**

| Ref | Quality requirement | Met? | Comment |
|---|---|---|---|
| P3 | Timely communication of positive screening results (sickle cell disorder) – including a review of parental results | Y | *Small patient numbers, but target times regularly exceeded* |
| 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]*<br><br>*As for P3.* |
| P5 | Timely confirmation of diagnosis for infants with a positive screening result** | Y | |
| S1i | Failsafe to ensure ongoing care | Y | *Robust process described by hospital and community specialist nurses.* |
| 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 ($\beta^{+}$, $\beta^{0}$, 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.